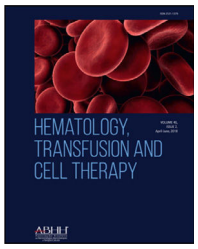
HEMATOLOGY, TRANSFUSION AND CELL THERAPY

www.htct.com.br



POSTER PRESENTATIONS

ADULT HEMATOLOGY ABSTRACT CATEGORIES

ACUTE LEUKEMIAS

PP 01

ASPARAGINASE INDUCED SINUS VEIN THROMBOSIS IN AN ADULT YOUNG ALL PATIENT

Beyza OLUK, Özden ÖZLÜK, Murat ÖZBALAK, Simge ERDEM, Sevgi KALAYOĞLU BEŞIŞIK

İstanbul University, Istanbul Medical Faculty, Department of Internal Medicine, Division of Hematology

Objective: Asparaginase has a very important role in ALL treatment among increasing of remission rate and duration. Multiple side effects prevent its regulatory use. Asparaginase reduce antithrombin 3, heparin cofactor II, protein C and plasminogen synthesis. By the way, P-selectin, PAI 1, tissue factor activity, and vWF antigen levels increase. Hypofibrinogenemia may be a marker of of hemostasis disturbances and decreased protein synthesis. **Case report:** A 25-year-old male patient with acute T-cell lymphoblastic leukemia diagnosis was initiated on induction chemotherapy according to augmented BFM protocol. After 4 weeks, remission was confirmed. During consolidation with the third dose of standard L-asparaginase of 10000 units, headache, nausea and vomiting started and confusion developed. Biochemical investigations, PT, aPTT were within normal limits. Fibrinogen was 92 mg/dL and D-dimer was high.Contrast-enhanced MRI showed a thrombus occluding. **Methodology:** the superior sagittal sinus. He was intubated and followed up in the intensive care unit due to a rapid decline in the Glasgow score and epileptic seizures. Correction of fibrinogen with cryoprecipitate anticoagulation treatment contributed to symptoms improvement. unfortunately, asparaginase was removed from the protocol. During maintenance therapy, he has had severe COVID-19 pneumonia and during this time he did not experience any thrombosis related complications. **Results:** Asparaginase therapy is associated with low antithrombin and fibrinogen levels and 7% of thrombosis rate. Previous studies showed that, in ALL patients, thrombosis occurred far more frequently during cycles that contained asparaginase than those that did not. **Conclusion:** Therefore, studies have investigated the role of fresh frozen plasma or cryoprecipitate supplementation to reduce the thrombohemorrhagic risk of therapy. Retrospective studies suggest antithrombin concentrates may have a beneficial effect on the outcomes of adults treated with asparaginase for ALL.

https://doi.org/10.1016/j.htct.2021.10.1011

PP 02

VENETOCLAX AZACITIDINE COMBINATION THERAPY IN FIRST-LINE TREATMENT OF ACUTE MYELOID LEUKEMIA PATIENTS: A SINGLE CENTER EXPERIENCE

Melda Cömert, Tuba Bulduk, Bilge Uğur, Murat Yıldırım, Selim Sayın, Alparslan Merdin, Ebru Kılıç Güneş, Elifcan Aladağ Karakulak, Meltem Aylı

Gülhane Education and Training Hospital, Department of Hematology

Objective: The response of elderly patients with acute myeloid leukemia (AML) to standard induction therapy is quite poor due to higher frequency of adverse genomic features, increased resistance to treatments, comorbidities and performance status. BCL-2 overexpression is implicated in survival of AML cells and treatment resistance. Preclinical data demonstrated the anti-leukemic effect of venetoclax, a selective BCL-2 overexpression is implicated in survival of AML cells and treatment resistance. **Methodology:** Venetoclax has received FDA approval for the treatment of AML patients >75 years of age and in combination with hypomethylating agents/low-dose cytarabine in patients not eligible for intensive therapy. Six newly diagnosed AML patients who were followed up in Gülhane Education and Training Hospital, Hematology clinic and treated with azacitidine+venetoclax were evaluated retrospectively. **Results:** F/M:1/5,the mean age was 77.3 (63-87).Two patients were secondary AML.All

patients had normal karyotype.Venetoclax+azacytidine treatment was started in all patients as first-line treatment after obtaining off-label consent.The average number of courses of venetoclax + azacitidine administered 3.5 (1-8).Patients received 200 mg/day venetoclax because of fluconazole usage concomittantly.One patient died with a FEN attack at the end of the second cycle, and 5 patients are still being followed up. **Conclusion:** Azacitidine or decitabine monotherapy yields low response rates (10%-50%, including hematologic improvement), require 3.5 to 4.3 months to achieve best response, and are not curative, with a median OS of less than 1 year.Targeted therapies capable of rapidly inducing a high rate of clinical response, with better tolerability and durable responses for elderly patients with AML. The novel combination of venetoclax with decitabine or azacitidine was effective and well tolerated in elderly patients.

https://doi.org/10.1016/j.htct.2021.10.1012

PP 03

A REGISTRY-BASED, OBSERVATIONAL SAFETY STUDY OF INOTUZUMAB OZOGAMICIN (INO) IN PATIENTS WITH B-CELL PRECURSOR ACUTE LYMPHOBLASTIC LEUKEMIA (ALL) PROCEEDING TO HEMATOPOIETIC STEM CELL TRANSPLANT (HSCT)

David MARKS [1], Marcos DE LIMA [2], Partow KEBRIAEI [3], Francesco LANZA [4], Christina CHO [5], Sergio GIRALT [5], Gizelle POPRADI [6], Michael HEMMER [7], Xin ZHANG [8], Richa SHAH [8], Verna WELCH [8], Erik VANDENDRIES [8], Matthias STELLJES [9], Wael SABER [7]

[1] *University Hospitals Bristol*
[2] *UH Cleveland Medical Center*
[3] *MD Anderson Cancer Center*
[4] *Ospedale di Ravenna*
[5] *Memorial Sloan Kettering Cancer Center*
[6] *McGill University Health Centre*
[7] *CIBMTR, Medical College of Wisconsin*
[8] *Pfizer Inc*
[9] *Universitätsklinikum Münster*

Objective: InO is a CD22-directed antibody-drug conjugate indicated for treatment of relapsed/refractory (R/R) ALL. InO has been associated with hepatotoxicity and hepatic veno-occlusive disease/sinusoidal obstruction syndrome (VOD/SOS), particularly post-HSCT. Registry data from the Center for International Blood and Marrow Transplant Research (CIBMTR) was analyzed to assess toxicity in patients (pts) with ALL who received InO prior to HSCT. **Methodology:** CIBMTR patient data are being collected from 2017-2022 after US approval of InO. Data accrued from 2017–2020 from 131 US adult pts (median age 40 y) treated with InO who proceeded to allogeneic HSCT were included. Using interim data at 3 y, we evaluated post-HSCT outcomes, including clinical status, overall survival (OS), non-relapse mortality (NRM), relapse, death after relapse, and investigator-defined adverse events, including hepatic VOD/SOS. All statistical analyses are descriptive. **Results:** Before HSCT, 36% of pts received 1 InO cycle, 46% had 2 cycles, 17% had ≥3 cycles. Median time from last InO dose to HSCT was 2.0 mos (range: 0.4–26.2). At data lock (Nov 2020, n=131), VOD/SOS incidence within 100 d post-HSCT was 13% (18% of R/R ALL pts, n=91). Post-HSCT 12 mo OS was 55%; post-HSCT 12 mo NRM was 21%; post-HSCT 12 mo relapse was 36%; non-HSCT-related 12 mo mortality was 25%. Most pts (89%) who underwent HSCT during complete remission (CR) experienced continued CR post-HSCT. **Conclusion:** Incidence of VOD/SOS after first HSCT in InO-treated pts with R/R ALL in this study was similar to the 18-19% reported in pooled analyses of 2 clinical trials among InO-treated pts with R/R ALL and in the INO-VATE study. The NRM at 1 y of 21% (23% R/R ALL) is lower than the NRM at 1 y of 38% reported in the pooled analyses of R/R ALL InO recipients.

https://doi.org/10.1016/j.htct.2021.10.1013

PP 04

ANTI-CD52 TREATMENT EXPERIENCE IN A T-CELL PROLYMPHOCYTIC LEUKEMIA PATIENT:CASE REPORT

Senem MARAL, Murat ALBAYRAK, Hacer Berna OZTURK, MerihREIS ARAS, Fatma YILMAZ, Pınar TIGLIOGLU, Mesut TIGLIOGLU, Buğra SAGLAM

Dışkapı Research and Training Hospital, Department of Hematology

Objective: T-cell prolymphocytic leukemia (T-PLL) is a rare and highly aggressive T cell neoplasm with rapidly progressing clinical course. T-PLL accounts for 2% of mature lymphocytic leukemia in adults. Median overall survival with modern therapy is reported one to three years. Here we report a T-PLL patient with peritoneum involvement and progressive ascite despite anti-CD52 treatment. **Case report:** A 65-year-old man with diabetes mellitus was admitted to hospital due to fatigue for a few weeks. Laboratory workup revealed that white blood cell count $469 \times 103/\mu l$ (90% lymphocytes), haemoglobin of 11.4 g/dl, platelets of $104 \times 103/\mu l$. Medium sized atypical lymphoid cells with partial chromatin condensation and a visible nucleolus were observed on blood smear. **Methodology:** On physical examination, palpable inguinal lymph nodes, splenomegaly 3 cm below the rib margin and a palpable lesion on the helix of left ear were noticed. Punch biyopsy of skin lesion was reported as a mature and immature T cell infiltration which are CD3 and CD10 positive and Tdt, CD34, CD20, CD99 negative. Flow cytometric study of peripheral blood sample was revealed that T-Chronic Lymphocytic Leukemia (T-CLL). **Results:** FMC protocol (fludarabine, mitoxantrone, and cyclophosphamide) was initiated and followed by intravenous alemtuzumab at a dose of 3 mg on day 1, 10 mg on day 2 and 30 mg on day 3. However after two months of

anti-CD52 treatment, his general situation dete-riorated and pleural effusion and abdominal distension developed due to massive ascites. Small, mature lymphocytic cell infiltration was shown in ascites fluid on cytological examination. He died after six months of diagnosis. **Conclusion:** T-PLL is a very aggressive disease with a median survival of less than 1 year. Not all patients diagnosed with T-PLL require treatment immediately.Currently, IV alemtuzumab (anti-CD52) is the accepted best avaliable treatment with very high response rates when given as first-line treatment. However, treatment is notcurative and a minority of T-PLL patients experience long-term disease-free survival.

https://doi.org/10.1016/j.htct.2021.10.1014

PP 05

RETROSPECTIVE EVALUATION OF PATIENTS WITH ACUTE MYELOID LEUKEMIA RECEIVING VENATOCLAX-BASED TREATMENT

EMINE DURAK, MEHMET CAN UGUR, BETUL KOYUNCU, SINEM NAMDAROĞLU, OKTAY BILGIR

Health Sciences University, Bozyaka Training and Research Hospital, Department of Hematology

Objective: Acute myeloid leukemia (AML) is the disease of elderly patients. Therefore, a significant number of patients are not suitable for intensive induction chemotherapy. In this study, it was aimed to retrospectively evaluate patients with AML who were treated Venatoclax-based regimens in our center. **Methodology:** The data of the patients who were treated Venatoclax-based regimens with the diagnosis of AML in the Bozyaka Training and Research Hospital Department of Hematology were scanned retrospectively from their files. **Results:** Data of 11 patients in total were reached. The mean age of the patients was 73.9. 8 of 11 patients were follow-up with diagnosis of AML , 3 patients with MDS RAEB II. Average follow-up time was 13.6 months. 5 patients died during follow-up. HMA +venatoclax was given to 6 patients as first-line, 4 patients second-line and 1 patient third-line therapy. Complete response was found in 3 patients, partial response in 1 patient, stable disease in 1 patient, and refractory disease in 1 patient. **Conclusion:** Venatoclax is a promising treatment option because it is an oral agent that can be tolerated by elderly patients and improves response rates and survival.

https://doi.org/10.1016/j.htct.2021.10.1015

PP 06

BONE MARROW NECROSIS IN ACUTE LYMPHOBLASTIC LEUKEMIA: A CASE REPORT

Melike Nur Ünal [1], Demet Kiper Ünal [2], Tuğba Çetintepe [2], Betül Bolat Küçükzeybek [3], Şerife Solmaz [2], Bahriye Payzın [2]

[1] *İZMİR KATİP ÇELEBİ UNIVERSITY, ATATÜRK TRAINING HOSPITAL, DEPARTMENT OF INTERNAL MEDICINE*
[2] *İZMİR KATİP ÇELEBİ UNIVERSITY, ATATÜRK TRAINING HOSPITAL, DEPARTMENT OF IINTERNAL MEDICINE, HEMATOLOGY DEVISION*
[3] *İZMİR KATİP ÇELEBİ UNIVERSITY, ATATÜRK TRAINING HOSPITAL, DEPARTMENT OF PATHOLOGY*

Objective: Bone marrow necrosis (BMN) is an entity that necrotic cells are seen on amorphous eosinophilic ground with medullary infarctus but withouth cortical bone involvement. BMN is a postmortem diagnosis in most of the reports. Bone marrow biopsy and aspiration is essential for the diagnosis. The case we report here is a patient who is diagnosed BMN and ALL at the same time with the first bone marrow biopsy, which is showed extensive necrosis. **Case report:** A 42-year-old man applied to our E.R. with lumbal pain. The initial blood count showed leukocyte:5.13 × 109/l, neutrophil:2.68 × 109/l, Hgb:10.7 g/dl, Hct: %31.5, thrombocyte:127 × 109/l, LDH:539 u/l (N:0-250), ALP:185 u/l (N:40-150), Total Bilirubin:0.81 mg/dl, CRP:337mg/l (N<5)), ESH: 94mm/h, folic acide: 2.8ng/ml (N>5.4), Vitamin B12:398 pg/ml (N:210-900), ferritin: 5607 ug/l (N:22-320), fibrinogen:1304 mg/dl (N:200-400), D-Dimer:646 ug/l (N<243) and a normal range for PZ, aPTZ, INR. **Methodology:** Peripheral smear showed %38 PMN, %56 lymphocyte, %6 monocyte, normoblasts, rare tear drop cells and rare thrombocytes. Pathological evaluation revealed hypercellular bone marrow (%95), extensive necrosis, CD3(-) CD5(-) CD20(+), CD38(-), CD10 diffuse(+), BCL2(+) MPO(-) CD117(-), CD34(+) CD79a, Pax5 and TdT suboptimal (+). Flow cytometry showed no significant result because of the deficiency of material. PCR revealed no BCR-ABL transcript. **Results:** The patient diagnosed B precursor ALL. With the BFM IA protocol complete remission obtained. At the control BMB CD3, CD20, CD79a, Pax5, TdT, MPO, CD34 was applied but there was no neoplastic involvement. After the BFM IB protocol, complete remission has been pursued. The patient is currently receiving the BFM IC protocol. **Conclusion:** BMN is an uncommon pathology with poor prognosis. Primary etiology is malignancies, especially hematologic malignancies, at %90 of the cases. As we see at this case, while the clinical and laboratory findings are insignificant; when a patient shows fever with unknown origin, bone pain, newly developed cytopenias, we must keep in mind the diagnosis of BMN and if a patient is diagnosed BMN, necessary scaning must be done immediately for malignancies as the primary cause.

https://doi.org/10.1016/j.htct.2021.10.1016

PP 07

NEXT GENERATION SEQUENCING PRACTICES IN HEMATOLOGY: A RECENT EXPERIENCE OF A SINGLE CENTER

Muruvvet Seda AYDIN, Funda CERAN, Simten DAGDAS, Gulsum OZET

Ankara City Hospital, Department of Hematology

Objective: Next-generation sequencing (NGS)-based technologies are novel methodologies for the diagnosis, prognostic assessment and decision of individualized treatment strategy in hematological neoplasia. NGS led to a more comprehensive understanding of the mutational landscape, especially in the myeloid neoplasms. Herein, we present the results of the patients who underwent NGS with the suspicion of myeloid neoplasia. **Methodology:** Retrospective data from a total of 13 patients were analyzed who were diagnosed between 01.10.2018 and 01.06.2021. There were four myeloid panels in the NGS. Panel 1 consists of ASXL1, CALR, CBL, CEBPA, CSF3R, and DNMT3A mutations. Panel 2 consists of EZH2, FLT3, IDH1, IDH2, JAK2, KIT, KRAS, and MPL mutations. Panel 3 consists of NPM1, NRAS, RUNX1, SETBP1, and SF3B1 mutations. Panel 4 consists of SH2B3/LNK, SRSF2, TET2, TP53, U2AF1, ZRSR2 mutations. **Results:** Median age was 48. Diagnoses were AML (n=7), AA (n=1), MDS (n=2), DLBCL (n=1), MM (n=1), and Evans syndrome (n=1). Seven cases with malignant diagnoses were eligible for intensive therapy. There were no mutations detected by NGS in MM, AA, DLBCL, and Evans syndrome cases. Biallelic CEBPA mutation accompanied FLT3 mutation in 1 case. IDH1 and NPM mutation were detected in 1 APL case. MPL, SRSF2, ASXL1, CBL, U2AF1, SF2B1, and TET2 were mutations detected in cases with dysplasia. **Conclusion:** In our cohort, NGS did not add any significant information in the lymphoid malignancies and benign hematological cases. NGS helped to define the allelic ratio of FLT3+ mutations and helped to accurately define the ELN risk of AML. Mutations that were detected in the cases with dysplastic bone marrow findings were concordant that were reported in the literature. Larger case series are needed in order to define the therapeutic and prognostic implications.

https://doi.org/10.1016/j.htct.2021.10.1017

PP 08

INAPPROPRIATE ADH SYNDROME OCCURING DURING B-ALL TREATMENT

Merih Reis Aras [1], Senem Maral [1], Hacer Berna Afacan Ozturk [1], Pinar Tiglioglu [1], Mesut Tiglioglu [1], Bugra Saglam [1], Fatma Yilmaz [1], Mustafa Onder [2], Murat Albayrak [1]

[1] *University of Health Sciences Ankara Diskapi Yildirim Beyazit Training and Research Hospital, Hematology Department*
[2] *University of Health Sciences Ankara Diskapi Yildirim Beyazit Training and Research Hospital, Family Medicine Department*

Case report: Inappropriate ADH syndrome is a cause of hyponatremia with increased ADH secretion despite normal plasma osmolality and euvolemic state.There are many related drugs in its etiology.Inappropriate ADH syndrome occured in two B-ALL diagnosed patient during cyclophosphamide and vincristine treatment regimen.The detection of inappropriate ADH syndrome in both patients with euvolemic hyponatremia shows the importance of reviewing the drugs used by the patient in the etiology of hyponatremia.

https://doi.org/10.1016/j.htct.2021.10.1018

PP 09

T-ACUTE MYELOID LEUKEMIA CASE THOUGHT TO BE ASSOCIATED WITH RADIOIODINE (I^{131}) TREATMENT

Merih Reis Aras [1], Pinar Tiglioglu [1], Mesut Tiglioglu [1], Bugra Saglam [1], Fatma Yilmaz [1], Neslihan Olgun [2], Senem Maral [1], Hacer Berna Afacan Ozturk [1], Murat Albayrak [1]

[1] *University of Health Sciences Ankara Diskapi Yildirim Beyazit Training and Research Hospital, Hematology Department*
[2] *University of Health Sciences Ankara Diskapi Yildirim Beyazit Training and Research Hospital, Family Medicine Department*

Case report: Ionizing radiation and chemotherapeutic agents can cause carcinogenic effects by causing DNA damage.A 40-year-old female patient diagnosed with thyroid papillary carcinoma in 2016 and subsequently administered 150 mCi radioiodine (I^{131}). Leukocytosis was detected in the examinations performed due to urinary system infection. She was diagnosed t(9:22) p210 positive AML M1-2. The patient had a history of Stargardt Syndrome.Development of t-AML after radioiodine treatment is very rare.

https://doi.org/10.1016/j.htct.2021.10.1019

PP 10

FLT3-ITD POSITIVITY IN AML; CASE SERIES

Kemal FİDAN

Erciyes University, Faculty Of Medicine, Department Of Hematology

Objective: Diagnosis of AML requires additional procedures, including pathological examination, immunophenotyping, cytogenetic examination, and molecular diagnosis. The determination of the specific cytogenetic abnormality is important for the selection of appropriate treatment and prognostic analysis.The 2 most common mutations of the FLT3 gene are FLT3-ITD and FLT3-D835. Here, we will present FLT3-ITD positive AML cases admitted to our clinic between 2019-2021. **Case report:** We have 5 cases of AML FLT3-ITD heterozygous. In all our cases, Midastaurin was given with 7+3 chemotherapy (CT) in the initial treatment. While 1 of our cases went into remission, the other 4 relapsed. All of the patients who relapsed were given FLAG CT, no remission was achieved and they were switched to ADE CT. Remission was achieved in 2 of 4 patients, 2 of them were refractory. One patient was given gilteritinib. HSCT was performed in 2 patients. While 2 of our

patients are dead, 2 of them are in remission and 1 of our patients is still under treatment. **Results:** FLT3 gene mutations are strongly associated with leukocytosis and poor prognosis in patients with AML(1-3). Patients with any of these mutations have a higher risk of recurrence and a lower survival rate[3]. All of our patients had leukocytosis at the time of diagnosis. Four of our patients relapsed and all of these patients were refractory to chemotherapy. Our patients who were refractory to treatment had a high mortality rate (50%) and a lower survival time (8-12 months). **Conclusion:** FLT3 signaling inhibitors have been used both alone and in combination to improve clinical outcomes in patients with AML with FLT3 mutations. While inhibitor monotherapy provides clinical response, its efficacy is usually temporary and resistance develops rapidly. Various combination therapies are used to enhance efficacy and prevent or overcome resistance. More studies are needed to evaluate its efficacy and explain the development of resistance.

https://doi.org/10.1016/j.htct.2021.10.1020

PP 11

EXPERIENCE OF GLASDEGIB IN PATIENTS WITH ELDERLY ACUTE MYELOID LEUKEMIA

Nanişe Gizem FENER, Oktay BİLGİR

SBÜ Bozyaka Training and Research Hospital Department of Hematology

Aim: Acute myeloid leukemia (AML) is characterized by the increase of high levels of myeloid cells in the bone marrow. In general, AML is a disease of older adults. Many adults with AML are unable to receive intensive chemotherapy because of its toxicity.In this study, it was aimed to retrospectively evaluate patients with AML who were treated Glasdegib-based regimens in our center. **Case 1:** A 75-year-old male patient was diagnosed with congestive heart failure + AML, and idarubicin and cytarabine chemotherapy protocol was started. The patient started to receive covid treatment due to covid lung involvement. The patient, whose treatment was interrupted for 1 month, received the second course as LDAC + glasdegib. In the bone marrow biopsy performed after the 5th cycle, it is observed in remission. **Case 2:** An 82-year-old male patient was diagnosed with chronic renal failure. With a diagnosis of AML intermediate risk LDAC+glasdegib treatment was given to the patient who did not go into remission after 4 cycles of decitabine treatment. The patient, who was followed in remission after 4 cycles, died due to pneumonia. **Case 3:** A 76-year-old male patient has a diagnosis of cerebrovascular disease. The patient, who was followed up in remission after 2 courses of azacitidine with a diagnosis of medium risk AML, was approved for glasdegib in the 3rd course of treatment, and he is being followed up with the 3rd course of LDAC+glasdegib therapy. **Material and method:** The data of the patients who were treated Glasdegib-based regimens with the diagnosis of AML in the Bozyaka Training and Research Hospital Department of Hematology were scanned retrospectively from their files. **Results:** In our 3 patients, 1 person died due to pneumonia, but the patient was being followed in remission. Our other 2 patients are still being actively followed up with LDAC + glasdegib treatments. **Discussion:** In 2018, the FDA approved glasdegib, a Hedgehog signaling pathway inhibitor, in adults 75 years of age or older with a diagnosis of AML or those with comorbidities that preclude the use of low-dose cytarabine plus intensive induction chemotherapy. Approval is based on interim results from the phase 2 BRIGHT 1003 study evaluating glasdegib in combination with LDAC or LDAC alone. The median OS was 4.9 months for LDAC versus 8.8 months in patients treated with glasdegib+LDAC. This difference represented an approximately 50% reduction in mortality in patients treated with glasdegib+LDAC. The final result of the BRIGHT 1003 study confirmed that glasdegib LDAC significantly improved OS compared to LDAC alone (hazard ratio, 0.495 [95% CI, 0.325-0.752]; P =0.0004). Also, the addition of glasdegib to LDAC did not cause a significant increase in adverse events.

https://doi.org/10.1016/j.htct.2021.10.1021

CHRONIC LEUKEMIAS

PP 12

SYNTHETIC BIOLOGY MEETS PRECISION MEDICINE: DRUG REPURPOSING FOR BLOOD CANCER PRECISION MEDICINE

Deepak Balaji Thimiri Govindaraj Thimiri Govindaraj

Council for Scientific and Industrial Research - Biosciences CSIR

Objective: Optimizing Drug discovery and Translation is one of the key tracks in Global Challenges Annual meeting 2019 and is the critical factor in achieving UN Sustainable Development Goals 3 Good Health and Well Being. WHO reports Cancer is the second leading cause of death globally. The aim of the proposal is to establish robust drug screening platform which can identify drugs and drug combinations that are effective in precision medicine for relapsed individual South African Leukemia patients. **Methodology:** Here, cancer cell will be either directly analyzed using high-throughput drug screening or single cell drug screening will be performed using flow cytometry /microfluidics to provide datasets on drug sensitivity for individual patientsI. WP1: Establishing the culture setting for CLL/MM and high-throughput drug screening on CLL/MM.II. WP2: “Signaling pathway-only” limited drug screening for CLL/MMIII. WP3: Establishing setting for Precision Microfluidics-driven single cell drug screening; **Results:** Expected Results are using full-library drug screening results, we will identify effective drugs and drug combinations that inhibit cancer cell proliferation either through cytostatic or cytotoxic effects. These results will provide the basis for identifying effective drug combinations using our predictive analytics, which will be packaged as preclinical information for a precision clinical trial. Thus, we would establish cutting-edge platform that can handle blood cancer and also solid tumor. **Conclusion:** Using this pipeline, we aim to identify drug combinations that can overcome relapse stage and ultimately provide tailored-specific therapy options for

Cancer patients. Our intention is to develop technologies that provide clinically relevant drug combinations information to oncologists within a timeframe of 7 days. The development and validation of the screening pipeline will incorporate the first CSIR platforms for cancer translational research with respect to identifying effective cancer drugs.

https://doi.org/10.1016/j.htct.2021.10.1022

PP 13

AVASCULAR NECROSIS IN CHRONIC MYELOID LEUKEMIA: A REVIEW OF PATHOPHYSIOLOGY, PATIENTS' CHARACTERISTICS, AND CLINICAL OUTCOMES

Abdulrahman Al-Mashdali [1], Husam Al-Dubai [1], Mohamed Yassin [2]

[1] *Department of Internal Medicine, Hamad Medical Corporation*
[2] *National Center for Cancer Care and Research, Department of Oncology, Hematology and BMT Section, Hamad Medical Corporation*

Objective: The exact incidence of avascular necrosis (AVN) in chronic myeloid leukemia (CML) is still unknown as the number of cases is limited. AVN was reported as an initial presenting manifestation in few CML patients. On the other hand, AVN was linked to medications used in CML treatment, specifically interferon-alpha (IFN-α) therapy and tyrosine kinase inhibitors (TKI). Our review aims to describe the pathophysiology, patients' clinical characteristics, and outcomes of AVN in CML. **Methodology:** We searched PubMed and Google Scholar for the case reports and series of patients with CML who developed AVN from inception to July 2021. We found 21 cases of AVN in CML patients,17 cases with AVNFH, and four cases with ONJ. Articles in the grey literature and non-English language publications were excluded. Patient characteristics, hematological parameters, management, and outcomes of AVN were extracted from those articles. **Results:** The median age was 39 years with an almost equal distribution between males and females. WBC counts were strikingly elevated in patients who initially presented with AVNFH (above 10,0000 in most cases). AVN related to CML management has been linked to TKIs and standard IFN-α therapies. Only 6 (out of 17) patients who developed AVNFH eventually required a hip replacement, and one (out of 17) developed a recurrent episode of AVNFH. All the reported cases of CML with ONJ were associated with TKIs **Conclusion:** Given the lack of data, we could not conclude whether AVN has an adverse prognostic effect on CML. However, the overall prognosis is comparable with AVN associated with other conditions. Clinicians should consider AVN in CML patients with either hip or jaw pain because early detection and management are essential to decrease morbidity and long-term disability in such patients. A further prospective study with a larger sample size is needed to clarify the different aspects of AVN in CML patients.

https://doi.org/10.1016/j.htct.2021.10.1023

PP 14

SECONDARY CHRONIC MYELOID LEUKEMIA FOLLOWING RADIOACTIVE IODINE (I131)

Yousef Hailan, Mohamed Yassin

Hamad Medical Corporation

Objective: Radioactive iodine (RAI) with I131 has an established role in managing differentiated thyroid carcinoma, namely papillary thyroid carcinoma (PTC) and follicular thyroid carcinoma. However, concerns have been raised about its possible carcinogenic effects. Papers of t-CML following I131 are increasingly reported, and thus this review is dedicated to highlighting it. **Methodology:** All reports from the 1960s to date related to CML following RAI therapy were searched on Google Scholar and PubMed. Different search terms with Boolean function to search for the relevant articles. **Results:** We identified ten articles reporting 12 cases, as presented in table 1. We found that most of the reports were for men (8/12) under the age of 60 years (10/12), and the primary tumor was of PTC characteristics (5/12 were PTC, and 3/12 were mixed papillary-follicular carcinoma). The dose of I131 ranged between 30 millicuries (mCi) to 850 mCi; the mean dose was 331 mCi. Also, t-CML developed within the first ten years (9/12), mainly between 4-7 years post-exposure. **Conclusion:** A few reports found a statistically significant increased risk of leukemia following RAI therapy; some suggested a relative risk of 2.5 for I131 vs. no I131. Observed findings from these studies include a linear relationship between the cumulative dose of I131 and the risk of leukemia, doses higher than 100 mCi were associated with a greater risk of developing secondary leukemia, and most of the leukemias developed within the initial ten years of exposure.

https://doi.org/10.1016/j.htct.2021.10.1024

CHRONIC MYELOPROLIFERATIVE DISEASES

PP 15

CONCOMITANT LATENT POLYCYTHEMIA VERA AND MGUS.

Fidan KHALILOVA, Azer KERIMOV, Zohra ALIMIRZAYEVA

National Center of Hematology and Blood Transfusion

Objective: With the introduction of changes in the diagnostic criteria for polycythemia vera (PV) in the 2016 WHO classification, it became necessary to revise the diagnosis in some patients. Cases with latent (masked) polycythemia vera (LPV) were identified. Bone marrow trepanobiopsy takes one of the most important place in the differential diagnosis of LPV with other myeloproliferative diseases.We describe a case with coexistence of LPV and MGUS in a patient at the onset of the disease. **Case report:** Patient F.I., aged 62, was admitted with complaints of burning sensation in both feet, pain in the left lower extremity, back pain, nocturia 2-3 times per night and

weight loss of 6-7 kg.Lumbar and whole spinal MRI revealed changes in the intensity of the medullary signal, mild decrease in the height of L2-L3 and T10 . EMG revealed polyneuropathy.PET showed a moderate uptake of FDG in the localization of the bone marrow. The spleen was enlarged-size-157 mm. **Methodology:** Laboratory findings: hemogram-WBC-11.15 × 103/μL, Hgb-15g / dL, HCT-48%, PLT-604 × 103/μL. Bone marrow biopsy, imprint, aspiration revealed moderately hipercellular bone marrow with increasing in all 3 series, groupings in megakaryocytes, containing limited (3-4%) kappa monoclonal plasma cell population; moderately increasing reticulin fibers (grade 1 according to WHO). Karyotype 46, XY; multiple myeloma FISH panel: translocation 4; 14 and translocation 11; 14 (+). JAK2V617F-50.48% (+). **Results:** The key point in the diagnosis was trilineage hyperplasia of the bone marrow,because the reticulin fibrosis may occurs in 20% of PV cases. Thus, the patient was diagnosed with LPV. Due to the detection of plasma cells in the bone marrow (3-4%), kappa light chains, with the diagnosis of LPV, the diagnosis of MGUS was established.The patient was prescribed ASS 100 mg per os, Hydrea at a dose of 500 mg every other day. For MGUS, the "wait and watch" tactic was chosen. **Conclusion:** In the diagnosis of LPV, along with molecular genetic research, trepanobiopsy of the bone marrow plays a leading role. The possibility of a combination of myeloproliferative and other diseases should not be ruled out.

https://doi.org/10.1016/j.htct.2021.10.1025

PP 16

THE OUTCOME OF FATHERHOOD IN PATIENTS WITH PHILADELPHIA NEGATIVE MYELOPROLIFERATIVE NEOPLASMS, A SINGLE INSTITUTION EXPERIENCE

Elrazi Ali, Mohammad Abu-Tineh,
Anas Babiker, Yousef Hailan, Bashir Ali,
Qusai Maharmah, Zakaria Maat,
Abdulatif Waggad, Mohamed Yassin

Hamad Medical Corporation

Objective: The aim of this retrospective study is to evaluate fertility in the Philadelphia-negative MPN male patients and the effect of treatment received on male fertility and the outcome. **Methodology:** This is a single-center, mixed-design study (retrospective + phone interviews) conducted within the National Center for Cancer Care and Research. **Results:** 120 patients were interviewed, only 19 patients (15.7%) had met the inclusion criteria. The majority of patients had lost follow-up or cannot be contacted, and 29.1% of patients had their families completed by the time of diagnosis. The treatment received includes hydroxyurea, interferon, and ruxolitinib. The mode of delivery was normal vaginal delivery in 68% of the pregnancies. The total number of conceptions was 27; three stillbirths were reported. **Conclusion:** The data showed that most MPN male patients on treatment had their offspring born normally with no delivery complications, no reported congenital anomaly or growth retardation, and no report of MPN-related cancers. Though, further studies with a larger sample size are required to fully understand the effect of medications on the outcome of fatherhood in Philadelphia negative MPN patients.

https://doi.org/10.1016/j.htct.2021.10.1026

PP 17

CONCOMITANT JAK2 AND BCR-ABL1 IN PATIENTS WITH CHRONIC MYELOID LEUKEMIA CLINICAL IMPACT AND RESPONSE TO THERAPY: A SYSTEMATIC REVIEW

Elrazi Ali, Elabbass Abdelmahmuod,
Mohamed Yassin, Ashraf Ahmed, Maab Faisal

Hamad Medical Corporation

Objective: The aim of this review is to assess patients with chronic myeloid leukemia with concomitant JA2 positive for - their characteristics - response to treatment **Methodology:** We searched the English literature (Google Scholar, PubMed, and SCOPUS) for studies, reviews, case series, and case reports of patients with chronic myeloid leukemia who had JAK2 mutation. Inclusion criteria: were the presence of JAK2 mutation in CML patients with BCR-ABL1 rearrangement and, secondly, age ≥18yrs. The search included all articles published up to 20th April 2021. **Results:** A total of 25 patients met our criteria of the search. Twenty-two patients were diagnosed in the chronic phase, 2 patients in the accelerated phase, and one patient transformed to the blast phase. More females n=16 and 10 males. The mean age at the time of diagnosis was 61.3 years. 9 patients had to be switch to second-line therapy. Age and gender distribution and the presence of splenomegaly or organomegaly are almost the same. Males were slightly more than females. **Conclusion:** It is difficult to conclude that multi-kinase inhibitors are superior to imatinib in treating CML with concomitant JAK2 mutation. But the result of the reported cases showed that multi-kinase inhibitors are more likely to be successful in achieving remission and loss of JAK2 mutation. However, it is difficult to generalize the result without further studies due to the few numbers of patients and the unusual association.

https://doi.org/10.1016/j.htct.2021.10.1027

COAGULATION DISEASES

PP 18

DOUBLE HETEROZYGOTIC FV DEFECT WITH HETEROZYGOTIC FV LEIDEN MUTATION AND FV DEFICIENCY IN THROMBOSIS

İSMAİL ALTUĞ DEMİR,
HATİCE DEMET KİPER ÜNAL, ŞERİFE SOLMAZ,
BAHRİYE PAYZIN, TUĞBA ÇETİNTEPE

İZMİR KATİP ÇELEBİ UNIVERSITY, ATATÜRK TRAINING HOSPITAL, DEPARTMENT OF INTERNAL MEDICINE

Objective: FV Leiden mutation causes activated protein C (APC) resistance and causes an increase in thrombin level.

Although moderate bleeding is seen in severe factor V deficiency, less than 1% of patients experience bleeding. Cases in which thrombosis is prominent in the presence FV Leiden mutation and FV deficiency have been reported. Here, we present a patient with FV deficiency with FV Leiden heterozygous mutation in the etiology of recurrent abortion. **Case report:** A 41-year-old female patient who applied to her primary care physician with bilateral lumbar pain upon finding INR: 1.43 (0.8-1.2) and APTT: 37.6 seconds (25-36.5), the patient was recommended to apply to our out patient clinic. The patient who described two spontaneous abortions (at the age of 25, the first in the 2nd trimester and the other in the 3rd trimester), also had a history of ecchymosis in the extremities caused by minor trauma at intervals. **Methodology:** PT, INR and APTT returned to normal with the mixing test performed on the patient (12.1 sEC, 1.03 and 28.6 sec, respectively).Afterwards, FV, which is one of the factors in the common pathway of coagulation, was found low in the examination repeated twice (12.3% and 10.2%) (N: 62-139%). The APCR studied twice in screening for thrombophilia was 1.4 and 2.4 (N: 2.61-3.32) Protein C, protein S, antithrombin III levels were within normal limits, LAC and APA were negative. **Results:** According to this result, FV Leiden heterozygous mutation was detected in the genetic thrombophilia panel. Also the patient had FV deficiency . **Conclusion:** Authors termed the coexistence of heterozygous FV Leiden mutation and type1 FV deficiency as pseudohomozygous FV Leiden mutation. In our and other studies, we concluded that thrombosis was clinically significant, where as bleeding was rare and mild. We think that prolonged PT and APTT results in patients with a history of thrombosis with FV Leidenmutation are also stimulating in evaluating FV activity.

https://doi.org/10.1016/j.htct.2021.10.1028

PP 19

BLEEDING MANAGEMENT DURING DELIVERY AND POSTPARTUM PERIOD IN GLANZMANN THROMBASTHENIA: EXPERIENCES FROM TWO CASES

Özden ÖZLÜK [1], Mustafa Murat ÖZBALAK [1], Tarık Onur TİRYAKİ [1], Sevgi KALAYOĞLU BEŞIŞIK [1], Tuba SARAÇ SİVRİKOZ [2]

[1] *İstanbul Medical Faculty, Hematology*
[2] *İstanbul Medical Faculty, Gynecology and Obstetrics*

Objective: Glanzmann thrombasthenia (GT) is a hereditary bleeding disorder. The platelets lack αIIbβ3integrin and fail to aggregate. Pregnancy can also lead to isoantibody formation when fetal cells with β3integrins pass into the circulation of a mother lacking them; a consequence is neonatal thrombocytopenia and a high risk of mortality. We here present our experience with two GT patients, in which rFVIIa was our choice for bleeding prophylaxis and/or control during delivery and postpartum period. **Case report:** Case 1: A 28-year-old woman with GT was hospitalized. She was on 38th gestational week (GW). Vaginal delivery was completed with rFVIIa prophylaxis. Postpartum 5th day rFVIIa stopped. The patient discharged with a minimal vaginal bleeding. Postpartum10th day, she developed severe bleeding. GT seemed to be the only related factor. rFVIIa restarted with tranexamic acid and misoprostol. Two apheresis units of platelets were transfused. That time, rFVIIa continued 7 days. **Methodology:** Case 2: A 26-year-old woman with GT developed hematuria on 30th GW. No urinary system pathology was found. With. rFVIIa treatment, hematuria was ceased. On 39th GW, during labor she developed massive bleeding. As urgent management, 8 random units of platelet and 5 units of packed red blood cell were transfused with local vaginal compress. rFVIIa treatment was initiated. On 10th days of rFIIa with minimal vaginal bleeding the patient was discharged from the hospital. **Results:** In both of the patients, no major neonatal bleeding problem was experienced. **Conclusion:** GT patients are at risk for heavy bleeding during labor, deliver or postpartum. Platelet transfusion is simple and easy option for bleeding control. In alloimmunized patients pooled platelet should be used. The use of rFVIIa appears to be safe and relatively effective.

https://doi.org/10.1016/j.htct.2021.10.1029

PP 20

CASE REPORT OF MARGINAL ZONE LYMPHOMA DETECTED WHILE INVESTIGATING ETIOLOGY FOR HEMOSTASIS DISORDER

Mesut Tığlıoğlu, Pınar Tığlıoğlu, Merih Reis Aras, Buğra Sağlam, Fatma Yılmaz, Senem Maral, Hacer Berna Afacan Öztürk, Murat Albayrak

Diskapi Yildirim Beyazit Training and Research Hospital

Case report: In this article, we wanted to present our case in which we detected SMZL during examining for defects in coagulation tests and correlated the PT and aPTT elevation with the development of inhibitors against coagulation factors related to this disease. The PT and aPTT values of the patient diagnosed with MZL did not improve in the mixing test, and no other etiology was found. With the second course of chemotherapy, the patient's values improved.

https://doi.org/10.1016/j.htct.2021.10.1030

PP 21

ACQUIRED FACTOR XIII DEFICIENCY WITH RUNX1 MUTATION, A REPORT OF TWO CASES TREATED WITH FACTOR XIII CONCENTRATE

Alfadil Haroon, Ali Alahmari, Nadiah Alobaidi, Ahmed Syed Osman, Hazzaa Alzahrani

Oncology Centre, King Faisal Specialist Hospital and Research Centre, Riyadh, KSA

Acquired FXIII deficiency has been described in association with malignancies or autoimmune disorders. We report two cases of acquired FXIII deficiency associated with hematologic malignancies. The first patient is a 60-year-old male with CMML who presented 4 weeks after confirming his diagnosis with non-traumatic anterior abdominal wall hematoma. Workup revealed FXIII deficiency. He was treated with FXIII replacement and other supportive measures. The hematoma resolved and patient was maintained on factor replacement. Unfortunately, his disease transformed to AML and he succumbed to death after starting AML therapy despite achieving complete remission. The second patient is a 24-year-old male patient post haploidentical transplant for intermediate risk AML. He developed hemorrhagic cystitis day 36 post-transplant and non-traumatic subdural hematoma on day 60 post-transplant. Workup revealed FXIII deficiency. He was treated with factor replacement and the subdural hematoma resolved with improvement of the hemorrhagic cystitis. Both patients had RUNX1 mutation which regulates expression of F13A1 in megakaryocyte this can decreased platelet expression of F13A1 in patient with RUNX1 haplodeficiency which lead to platelet dysfunction. FXIII deficiency should be considered for patient with unexplained bleeding with normal routine workup.

https://doi.org/10.1016/j.htct.2021.10.1031

LYMPHOMA

PP 22

A CASE OF MULTI REGIONAL PRIMARY MUSCLE LYMPHOMA

Beyza OLUK, Metban MASTANZADE,
Nur Seda IBILI, Sevgi KALAYOĞLU BEŞIŞIK

İstanbul University, Istanbul Medical Faculty, Department of Internal Medicine, Division of Hematology

Objective: Primary extranodal non-Hodgkin's lymphoma (eNHL) usually presents at an early stage, as an extranodal organ involvement along with draining lymph nodes only or the predominant site is extranodal. As an eNHL, primary skeletal muscle lymphoma is very rare. The usual clinical picture is local swelling and pain with or without systemic symptoms. MRI features are distinctive and FDG-PET/CT may help to evaluate the stage and monitor the response to the treatment. **Case report:** A 56-year-old male, presented with a one-month history of swelling and pain on his left ankle. There was no history of trauma or any physical strain. A mass lesion was palpated on the calcaneus bone. MRI showed diffuse muscle involvement. The clinical picture was not consistent with infection or hematoma. The blood cell count and biochemical investigations were within normal limits. Serology for hepatitis B, C and HIV were negative. Biopsy was decided. **Methodology:** Histological examination revealed CD19, CD20, bcl-2 and bcl-6 positive B-cell lymphoma with a Ki67 proliferation index of 95%. Myc, bcl-2, and bcl-6 gene rearrangements were not detected. Diffuse large B cell lymphoma was diagnosed. FDG-PET/CT showed lesions in multiple regions only limited to skeletal muscles but no other organ involvement. He had no adverse risk factors but bulky lesion (11cm sized lesion). After 6 courses of R-CHOP protocol, he had complete anatomic and metabolic response. **Conclusion:** Healthy skeletal muscles do not have lymphatic system. Lymphomatous involvement of muscles occurs by 3 pathways as dissemination via the haematogenous or lymphatic pathway, extension from adjacent organs, such as the bones or lymph nodes, and de novo primary extranodal disease. Most of the histology primary skeletal lymphomas have the aggressive B-cell immunophenoty. In general, treatment is similar to nodal lymphomas. In conclusion, we aimed to contribute in experience with this rare eNHL type.

https://doi.org/10.1016/j.htct.2021.10.1032

PP 23

A RARE CASE: POSTTRANSPLANT NK/T CELL LYMPHOMA

Pınar Tığlıoğlu, Mesut Tığlıoğlu,
Merih Reis Aras, Buğra Sağlam, Fatma Yılmaz,
Senem Maral, Hacer Berna Afacan Öztürk,
Murat Albayrak

Diskapi Yildirim Beyazit Training and Research Hospital

Case report: We wanted to present our patient who was diagnosed with NK/T cell type PTLD after kidney transplantation, to contribute to the literature. Posttransplant lymphoma in NK cell phenotype (EBV unrelated) was detected in biopsy taken from the lesions that developed in mouth 11 years after kidney transplantation. It was detected as stage 1E with the examinations. As a result, early recognition of such rare cases and start treatment and reducing immunosuppressive agents are important

https://doi.org/10.1016/j.htct.2021.10.1033

PP 24

A VERY RARE CAUSE OF DIARRHEA IN A CHEMOTHERAPY-INDUCED NEUTROPENIC PATIENT: PELLAGRA

Fatma YILMAZ, Murat ALBAYRAK,
Senem MARAL, Hacer Berna AFACAN ÖZTÜRK,
Merih REİS ARAS, Pınar TIĞLIOĞLU,
Mesut TIĞLIOĞLU, Buğra SAĞLAM

Health Sciences University Diskapı Yıldırım Beyazit Training and Research Hospital, Hematology Clinic

Case report: Pellagra is a systemic disease caused by a deficiency of vitamin B3 .A 19-year-old male patient, who was diagnosed with Burkitt's lymphoma was admitted to the hematology clinic for the second cycle of R-CODOX-M chemotherapy treatment. The patient at risk of malnutrition developed dermatit,diare and demans during treatment. The

cause of diarrhea in the neutropenic patient is mostly in the form of infective diarrhea. Diarrhea due to vitamin deficiency should be kept in patients with malnutrition .

https://doi.org/10.1016/j.htct.2021.10.1034

PP 25

CAVITARY PRIMARY PULMONARY LYMPHOPLASMOCYTIC LYMPHOMA COMPLICATING HENOCH–SCHÖNLEIN PURPURA

Burak GULTEKIN [1], Ege Sinan TORUN [2], Ahmet GUL [2], Sevgi KALAYOGLU-BESISIK [3]

[1] *Istanbul University, Istanbul Faculty of Medicine, Department of Internal Medicine*
[2] *Istanbul University, Istanbul Faculty of Medicine, Department of Internal Medicine, Division of Rheumatology*
[3] *Istanbul University, Istanbul Faculty of Medicine, Department of Internal Medicine, Division of Hematology, Istanbul, Turkey*

Introduction: Non-Hodgkin lymphoma (NHL) may occur in the chest, often as secondary involvement but occasionally as primary disease. Low-grade pulmonary B-cell lymphoma is the most frequent form. The diagnosis based on histological examination of surgical samples. Henoch–Schönlein purpura (HSP) as a systemic vasculitis typically less commonly affects adults. Triggers including infections, medications and malignancy for HSP have been recognized. **Case report:** We report a patient presenting with HSP who had primary pulmonary lymphoplasmocytic lymphoma (PPLL) as an underlying malignancy. Case: 57-year-old male patient developed chest pain with a hemoglobin level 5.9g/dL. Symptoms resolved after erythrocyte transfusions. He has been diagnosed as having type 2 myocardial infarction. The detailed investigation contributed to warm autoimmune hemolytic anemia (AIHA) diagnosis. Steroid was started. He had high eryhtrocyte sedimentation rate. Further workup revealed bilateral multiple hilar lymphadenopathies and nodular cavitary pulmonary lesions on torax CT. The clinical picture and laboratory evaluation were not consistent with invasive fungal infection and tuberculosis. Purified protein derivative (PPD) skin test was negative. Bronchoalveolar lavage did not reveal any atypical cell and culture positivity. Thoracoscopic lymph node excision was performed. Histologic investigation showed plasma cells in the paracortical area with a slight increase in kappa to lambda ratio (3:1). A fine needle aspiration biopsy of lung tissue revealed lymphoplasmocytosis. PET-CT documented cavitary nodular lesions and hilar lympadenomegalies but no other suspicious lesion. Biopsy sample from one lesion sized 18×12 mm with SUVmax 5 revealed plasma cell infiltration with an IgG kappa phenotype. PPLL was diagnosed. Meanwhile AIHA responded to steroid but recurred during dose tapering. PPLL treatment with bortezomib and rituximab based regimen was decided. AIHA went in remission but relapsed after one year with HSP associated clinical picture. He had severe abdominal pain with intestinal wall thickness. Biopsy samples from kidney showed IgA vasculitis and from skin granular type of IgA and C3 deposition in the walls of small diameter vessels in the papillary dermis. Pulse steroid followed by cyclophosphamide controlled the clinical picture. **Conclusion:** We wished to highlight that in adults presenting with HSP may be a sign of underlying malignancy relapse.

https://doi.org/10.1016/j.htct.2021.10.1035

PP 26

ANTICARDIOLIPINIC ANTIBODIES IN NON-HODGKIN LYMPHOMA

Sanda Buruiana, Minodora Mazur, Maria Robu, Victor Tomacinschii

SUMPh "Nicolae Testemiţanu"

Objective: Identification of hemostasis changes in patients with non-Hodgkin's lymphoma (NHL) and anticardiolipin antibodies (aCL). **Methodology:** The study included 83 patients (men-34, women-49) with a mean age of 63.2 years, with NHL, investigated complex, by research of lupus anticoagulant (LA) by Turbidimetry; antiβ2glycoprotein I IgG, IgM and aCL antibodies, by ELISA method. Hemostasis disorders were evaluated according to the type of NHL, stage, tumor size. **Results:** aCL were detected in 10 (12%) patients: 6 patients with aggressive type lymphoma and 4 patients with indolent type lymphoma, with advanced stage B cell NHL in 60%, mean age 52.8 years. LA was present in 80% of cases, unlike aCL IgG antibodies (10%) and antiβ2glycoprotein I IgG (10%). Hemostasis disorders were found in 6 (60%) patients: thrombotic events-at 4 (40%) patients with Mantle cell lymphoma (1 patient), Small lymphocytic lymphoma (1 patient), lymphoblastic lymphoma (2 patients). Local stage (I and II) of the lymphoma was in 75%, but with a large size of the tumor (> 11 cm), and hemorrhage at 2 (20%) patients with stage IV Small lymphocytic lymphoma, in which immune thrombocytopenia developed. **Conclusion:** The presence of antiphospholipid antibodies, in particular of lupus anticoagulant, advanced age, generalized stage, and large tumor size are risk factors for the development of hemostasis diseases in NHL patients, especially thrombosis.

https://doi.org/10.1016/j.htct.2021.10.1036

PP 27

A CASE OF STAGE 1E DIFFUSE LARGE B-CELL LYMPHOMA PRESENTED WITH KNEE INVOLVEMENT

Mesut Tığlıoğlu, Murat Albayrak, Pınar Tığlıoğlu, Merih Reis Aras, Buğra Sağlam, Fatma Yılmaz, Senem Maral, Hacer Berna Afacan Öztürk

Diskapi Yildirim Beyazit Training and Research Hospital

Case report: Bone involvement is rare in DLBCL. 70-yearold patient, applied to the orthopedics clinic dueto knee pain. Kneeprosthesis was planned. During operation suspicious nontumoral lesion with unclear borders was observed. Bone biopsy was taken from the intraoperatively detected lesion and a knee prosthesis was placed. According to PETCT and bonemarrow biopsy results, patient was diagnosed as stage 1E. Awareness of DLBCL with atypical presentation are of great importance in terms of early diagnosis

https://doi.org/10.1016/j.htct.2021.10.1037

PP 28

THE EFFECT OF COMORBIDITY AND BODY MASS INDEX ON SURVIVAL IN PATIENTS WITH MARGINAL ZONE LYMPHOMA

Esra TURAN ERKEK, Tuba TAHTALI

Kartal Dr. L.Kırdar City Hospital

Objective: Marginal zone lymphoma is a biologically and clinically heterogeneous group of indolent lymphoproliferative diseases, constituting 5-15% of all NHLs (Non-Hodgkin Lymphoma) [1]. By the World Health Organization; subgroups as extranodal marginal zone lymphoma (ENMZL, MALT lymphoma, Maltoma), nodal marginal zone lymphoma (NMZL), splenic marginal zone lymphoma (SMZL) constitute 70%, 20%, 10% of MZL (Marginal Zone Lymphoma) cases, respectively. **Methodology:** A total of 50 patients with a diagnosis of MZL who applied to our hospital between 2013 and 2021 were included in this retrospective study. All analyzes were performed on SPSS v21. The Kolmogorov-Smirnov test was used for normality control. Data are given as mean ± standard deviation for continuous variables and frequency for categorical variables. Survival times were calculated using the Kaplan Meier method. Cox regression analysis (enter method) was performed to identify important prognostic factors. p<0.05 values were accepted as statistically significant results. **Results:** The mean age of 50 people in the study group was 62.88 ± 11.50 years and ranged from 34 to 84 years. 50% of the participants were male and 50% were female . The mean follow-up period of the patients was 51.80 ± 27.47 months. It was observed that none of the parameters measured in the study, such as age, gender, body mass index, diabetes, heart disease, thyroid diseases, non-hematological malignancies, chemotherapy, and radiotherapy intake, had an effect on survival. **Conclusion:** Age at diagnosis should be considered in risk assessment of patients with marginal zone lymphoma. It is thought that the fact that the patients are predominantly in the advanced stage MZL group, and the relatively short follow-up period compared to the indolen lymphoma group, has an effect on the absence of a determining effect of comorbid diseases on mortality. Prognostic markers determined by multicenter and detailed studies are needed to provide a better prediction.

https://doi.org/10.1016/j.htct.2021.10.1038

PP 29

CASE REPORT: FOLLICULAR LYMPHOMA PRESENTED WITH CHYLOTHORAX

Senem MARAL, Murat ALBAYRAK, Berna AFACAN ÖZTÜRK, Merih REİS ARAS, Fatma YILMAZ, Pınar Tığlıoğlu, Mesut TIĞLIOĞLU, Buğra SAĞLAM

Dışkapı Research and Training Hospital, Department of Hematology

Objective: Chylothorax is the leakage of chylous contents into the pleural space as a result of damage to the thoracic duct. Chylous effusion is seen often unilateral but may be bilateral rarely. Etiology includes non-traumatic and traumatic causes. While sarcoidosis, amyloidosis, superior vena cava thrombosis and congenital anomalies are non-traumatic causes, non-Hodgkin lymphomas are the most common causes.Herein, we present a follicular lymphoma patient who was presented chylothorax at diagnosis. **Case report:** A 31-year-old male patient presented with fatigue, and dyspnea. On physical examination, inguinal and axillary multiple palpable lymphadenopathies (LAP) were observed, and respiratory sounds were significantly decreased on the left side.Computed tomography imaging revealed prevascular, paratracheal, subcarinal LAPs and 5 cm thick pleural effusion in the deepest part and compression atelectasison the left. Excisional LAP biopsy revealed follicular lymphoma **Methodology:** When thoracentesis was performed and milky effusion was classified as an exudative. The high triglyceride level was consistent with a chylous effusion. After 6 cycles of R-CHOP treatment, the patient had a significant regression in the initial LAPs, while the chylous effusion persisted. When cytological examination of thoracentesis did not reveal lymphoma, the patient was followed-up. **Conclusion:** Chylothorax is associated with significant morbidity and mortality if left untreated. Control of the underlying malignancy is still the mainstay of treatment and reported as the most effective. In the literature, successful results were reported with the treatment of the underlying lymphoma. owever, it is known, chylothorax may recur and patients should be follow-up closely.

https://doi.org/10.1016/j.htct.2021.10.1039

MYELOMA

PP 30

LENALIDOMIDE ASSOCIATED İMMUNE THROMBOCYTOPENIA: A CASE REPORT

İbrahim Halil Acar, Birol Guvenc

Çukurova University Faculty of Medicine

Objective: Autoimmune cytopenia is observed in many hematological malignancies, whereas immune thrombocytopenia is rarely observed in plasma cell dyscrasias, such as multiple myeloma. On the other hand, cytopenias secondary to myelosuppression due to lenalidomide use are frequently observed,

whereas immune thrombocytopenia is a rarer complication. **Case report:** A 63-year-old female patient without any known disease was performed bone marrow biopsy in January 2019 due to anemia and high sedimentation rate. She was diagnosed with IgG-kappa type multiple myeloma and administered four cycles of bortezomib-cyclophosphamide-dexamethasone treatment. She went into remission after this treament and was then performed autologous stem cell transplantation followed by a consolidation therapy comprising 2 cycles of bortezomib-lenalidomide-dexamethasone treatment. Subsequently, she was administered lenalidomide maintenance therapy with regular follow-up visits. Isolated thrombocytopenia was observed in the patient in her last follow-up visit and was therefore hospitalized for further examination. No schistocyte was observed in the peripheral smear as well as no rouleaux formation. It was determined that her LDH (lactate dehydrogenase) levels were normal and that she did not have organomegaly. The results of the Coombs test, in addition to the results of hepatitis B, hepatitis C, HIV (Human Immunodeficiency Virus), EBV (Ebstein-Barr Virus), and ANA (antinuclear antibody) tests, which were run in order to determine whether she had any viral diseases, came out as negative. Post-transfusion purpura was ruled out in the patient as she had no history of transfusion in the last three months. She was then performed bone marrow biopsy, since her platelet count did not increase after discontinuation of lenalidomide treatment despite the fact that she was given platelet suspension transfusion. Subsequently, it was was determined that her megakaryocyte count increased, whereas her plasma cell ratio was less than 5%. In view of the foregoing, she was pre-diagnosed with lenalidomide-related immune thrombocytopenia, and was thus given 1 gr of methylprednisolone for 3 days followed by the administration of methylprednisolone at a daily dose of 1 mg/kg for 5 days. However, a sufficient increase in her platelet count could not be achieved with the said treatment. Therefore, she was administered eltrombopag therapy instead, since she was refractory to other treatments that could have been administered as a replacement treatment, such as IVIG (Intravenous Immunoglobulin), rituximab or cyclophosphamide. The patient, whose platelet count increased after the administration of eltrombopag therapy, was then discharged with full recovery. **Conclusion:** The aim of this case report is to demonstrate that lenalidomide-associated immune thrombocytopenia should also be considered when there is isolated thrombocytopenia in patients with multiple myeloma without a decrease in other cell lines.

https://doi.org/10.1016/j.htct.2021.10.1040

PP 31

CLINICAL PARAMETERS OF MULTIPLE MYELOMA PROGRESSION IN RESIDENTS OF THE GOMEL REGION OF BELARUS

Zhanna KOZICH [1], Victor MARTINKOV [1], Janna PUGACHEVA [1], Ludmila SMIRNOVA [2], Svetlana MIHNO [1], Anna DOMANTSEVICH [1]

[1] *State Institution "Republican Research Center for Radiation Medicine and Human Ecology*

[2] *Education Institution "Belarusian Medical Academy for Postgraduate Education", Minsk*

Objective: To study clinical parameters of multiple myeloma progression in residents of the Gomel region of Belarus **Methodology:** The study included 159 MM patients who were examined at the State Institution "Republican Research Center for Radiation Medicine and Human Ecology", Gomel from 2018 to 2021. The average age was 62. Female patients prevailed and amounted 57.1%. MM was diagnosed according to international criteria. The criteria for progression were determined when new foci of destruction or extramedullary lesions appeared, and at an increase in the number of plasma cells in the bone marrow> 10%. **Results:** Progression was in 10.7%(17). No differences in the immunological variant of MM. CD20 expression>20% was found 6.18 more often in progressed patients (p=0.0001). CD56>20% was 2.37 more common at progression (p=0.006). CD117>20% was 2.34 more often at progression, (p=0.116). M-protein>15 g/l was 6.22 more often at progression (p = 0.0001). Abnormal κ/λ was in 81.3% at progression (p=0.027). LDH was different (p=0.023). Kidney damage and destructive syndrome did not affect progression (p=0.797). **Conclusion:** Identification of markers of progression at the initial examination, such as excess expression of CD20> 20%, CD56> 20%, excess of M-protein> 15 g/l, abnormal κ/λ ratio can predetermine the outcome of the disease. Our findings are consistent with the literature data, but much remains unclear, for instance, cases with normal LDH values in patients with progression. This gives rise to future research.

https://doi.org/10.1016/j.htct.2021.10.1041

PP 32

THE EFFECT OF BISPHOSPHONATE USE ON TREATMENT RESPONSE AND OVERALL SURVIVAL IN MULTIPLE MYELOMA PATIENTS

Ayşe Uysal [1], Mustafa Merter [1], Serhat Göçüncü [2]

[1] *Fırat University School Of Medicine Hematology Department*

[2] *Fırat University Scool of Medicine Internal Medicine*

Objective: Bisphosphonates are pyrophosphate analogs with a high affinity for calcium crystals. Due to the affinity of bisphosphonates for calcium, they bind rapidly to calcium-containing hydroxyapatite crystals, especially in the resorption zone. In this way, they prevent bone resorption. In this study, we aimed to investigate the effect of bisphosphonate use on treatment response and overall survival in patients with MM. **Methodology:** All patients with MM who followed by the Hematology department of Fırat University Hospital in the last 10 years were included in this retrospective observational study. Age, gender, end-organ involvement, ISS staging, LDH level, IG subtype in diagnosis, bisphosphonate use (duration and dose), treatments, response status and survival was investigated. **Results:** Ninety-one patients, of whom 53 were

male and 38 females, were included in this study. At the time of diagnosis,14 patients with high calcium, 77 patients had normal calcium. There was no significant difference in survival between bisphosphonate intake status and IG subtypes (p>0,05). There was no significant difference in progression-free survival between the ISS category, bisphosphonate intake status, creatinine category, and IG subtypes (p>0,05). **Conclusion:** In this study, OS, and PFS in MM patients were not affected by bisphosphonate use. however, LDH level influenced both OS and PFS, the increase in LDH level negatively affected the survival.

https://doi.org/10.1016/j.htct.2021.10.1042

PP 33

INSIGHTS INTO DIAGNOSIS AND MANAGEMENT OF ADVANCED MULTIPLE MYELOMA

Vasile Musteata [1], Doina Ranga [2], Larisa Musteata [3], Cristina Dudnic [3], Nina Sghibneva-Bobeico [1]

[1] *Institute of Oncology, State University of Medicine and Pharmacy*
[2] *State University of Medicine and Pharmacy*
[3] *Institute of Oncology, State University of Medicine and Pharmacy*

Objective: The advanced stages of multiple myeloma (MM) commonly manifest a recurrent evolution, unfavorable prognosis and negative socio-economic impact. The increased rates of morbidity and DALYs, frequent complications and relapses, unfavorable socio-economic impact characterize MM as an actual issue of hematology and public health. The objective of the study was the identification of diagnostic patterns and the evaluation of short- and long-term results of treatment of the advanced stages of MM. **Methodology:** The study is a cross-sectional descriptive analysis of a cohort of 50 newly diagnosed patients with advanced stages of MM, who have been treated and followed-up at the Hematology Dept. of the Oncology Institute from Moldova during 2016-2020. The diagnosis was assessed by cytological, immunohistochemical examinations of the bone tissue and bone marrow, and ELISA immunological test of the peripheral blood. The stage was asserted in each case according to the Revised International Staging System. **Results:** The patients age ranged between 28-75 years (median - 57.7 years). MM developed mainly in persons aged 60-69 (52%) years and in rare cases under 39 years (6%). Females were 29 (58%), and males - 21 (42%). 31 (62%) patients were diagnosed in stage III, 14 (28%) - in stage II and 5 (10%) - in stage I. Immunoglobulin (Ig) G isotype was detected in 28 (56%) cases, IgA - in 12 (24%), light chains (Bence Jones MM) - in 10 (20%). Very good partial responses were achieved in 25 (50%) of patients. **Conclusion:** MM was diagnosed mostly in patients of 60-69 years, females and stage III disease. Bone marrow myeloma cells ranged between 30-67% (median - 46%). Concerning the Ig isotype distribution in MM, IgG accounted the majority of cases. Refractory chronic renal failure was the most common complication (50% of cases) in advanced MM. Targeted chemotherapy proved to be efficient in the advanced stages of MM regardless of the gender, age and disease span. Very good partial responses lasted 12-24 months.

https://doi.org/10.1016/j.htct.2021.10.1043

PP 34

CASE REPORT: COEXISTENCE OF CELIAC DISEASE AND MULTIPLE MYELOMA

Filiz YAVASOGLU [1], Ciğdem OZDEMIR [2]

[1] *Afyonkarahisar Health Sciences University Hospital, Hematology department*
[2] *Afyonkarahisar Health Sciences University Hospital, Pathology department*

Objective: Celiac disease is a systemic disease in which the natural and adaptive immune system is affected by the effect of gluten exposure and environmental factors in individuals with genetic predisposition. Multiple myeloma; is characterized by an increase in clonal plasma cells. It is the most common hematological malignancy after lymphomas.We aimed to present a case siagnosed with celiac disease and multipl myeloma **Case report:** A 56-year-old female patient with a diagnosis of asthma and celiac disease for 1 year was referred to the Hematology department because her refractory anemia. Serum IgA level of the patient was 4490 mg/dl without renal failure and hypercalcemia.bone marrow biopsy compatible with myeloma. The patient received 6 cycles of bortezomib, cyclophosphamide, and dexamethasone and 3 cycles lenalidomid dexametazon chemotherapy.After chemotherapy, Autologous stem cell transplantation was performed. **Conclusion:** Celiac disease is an autoimmune disease, characterized by inflammation and villus atrophy in the small intestine mucosa as a result of sensitivity to gluten, resulting in malabsorption. The incidence of lymphoma and gastrointestinal system malignancy is increased in individuals with celiac disease. Multiple myeloma may also be accompanied by autoimmune diseases such as ankylosing spondylitis, scleroderma, and sjögren's syndrome. Coexistence of multiple myeloma and celiac disease is rare.

https://doi.org/10.1016/j.htct.2021.10.1044

PP 35

A RARE AND COMPLEX CAUSE OF IMPOTENCE POEMS SYNDROME

Buğra Sağlam [1], Murat Albayrak [1], Mustafa Önder [2], Hacer Berna Afacan Öztürk [1], Merih Reis Aras [1], Pınar Tığlıoğlu [1], Mesut Tığlıoğlu [1], Fatma Yılmaz [1], Senem Maral [1]

[1] *University of Health Sciences Ankara Dışkapı Yıldırım Beyazıt Training and Research Hospital, Department of Hematology, Ankara, TURKEY*

[2] *University of Health Sciences Ankara Dışkapı Yıldırım Beyazıt Training and Research Hospital, Department of Family Medicine, Ankara, TURKEY*

Objective: Although plasma cell neoplasms occupy a large place in hematology practice, POEMS syndrome is very rare. Serum lambda light chain elevation and polyneuropathy, together with organomegaly, endocrinopathy, and skin lesions are the main components of the syndrome. We share our case, which we diagnosed in our clinic, with the belief that it will contribute to the literature. **Case report:** A 51-year-old male patient, who had no history of co-morbidity, drug use, or exposure to toxic substances, was started on supportive treatment in February 2021, who first developed the complaint of impotence. Later, he applied to the neurology outpatient clinic with complaints of weakness and weakness in the feet. After detecting polyneuropathy in his evaluation, IgG Lambda monoclonal gammopathy was detected in serum immune electrophoresis in his evaluation for etiology. **Methodology:** Thereupon, it started to be investigated in terms of plasma cell neoplasms. In the examinations performed, immunoglobulin levels, serum-urine kappa and lambda light chain levels, plasma increase in the bone marrow biopsies and a solitary 3.3 cm sclerotic lesion in the sacral region were detected in the PET-CT of the patient, whose ethology could not be diagnosed. **Results:** A tru-cut biopsy was taken from the sclerotic lesion of the patient, who was thought to be a plasmacytoma and a 20% monoclonal IgG lambda plasma increase was detected. In his physical examination, it was seen that he had increased lesions (Figure-1) and acrocyanosis (Figure-2) on the skin for the last 3-4 months. The patient's current complaints and laboratory results were evaluated with a preliminary diagnosis of POEMS syndrome (Table-1). **Conclusion:** POEMS syndrome is a rare disease and its exact incidence is unknown. It is frequently seen in 5-6 decades, with a median age of 51 years, and 63% of cases are male patients [1]. Chronic and excessive production of proinflammatory and other cytokines (IL-1β, TNFα, IL-6, vascular endothelial growth factor (VEGF) etc.), microangiopathy, edema, effusions, increase in vascular permeability, increase in neovascularization are important in the pathophysiology of the disease.

https://doi.org/10.1016/j.htct.2021.10.1045

PLATELET DISEASES

PP 36

IMMUNE THROMBOCYTOPENIA RELAPSE POST COVID-19 VACCINE IN YOUNG MALE PATIENT

Hana Qasim

Hamad Medical Corporation

Case report: We report a 28-year-old Asian male patient, known for ITP and in partial remission for eighteen months, who presented to emergency department with ITP relapse (platelets count of 1 × 10^3 /uL), four days after receiving the second dose of Pfizer SARS-CoV-2 vaccine, which required treatment with intravenous immunoglobulins and dexamethasone, we discuss as well the likely underlying pathophysiology and the suggested approach in patients known for ITP who are willing to receive mRNA COVID vaccines.

https://doi.org/10.1016/j.htct.2021.10.1046

PP 37

INTEGRATED EFFICACY RESULTS FROM THE PHASE 2 AND PHASE 3 STUDIES WITH CAPLACIZUMAB IN PATIENTS WITH ACQUIRED THROMBOTIC THROMBOCYTOPENIC PURPURA

Flora Peyvandi [1], Spero Cataland [2], Marie Scully [3], Paul Coppo [4], Paul Knoebl [5], Johanna A. Kremer Hovinga [6], Ara Metjian [7], Javier de la Rubia [8], Katerina Pavenski [9], Jessica Minkue Mi Edou [10], Filip Callewaert [11], Hilde De Winter [12]

[1] *Angelo Bianchi Bonomi Hemophilia and Thrombosis Center, Fondazione IRCCS Ca' Granda Ospedale Maggiore Policlinico, and Department of Pathophysiology and Transplantation, Università degli Studi di Milano, Milan, Italy*
[2] *Division of Hematology, Department of Internal Medicine, The Ohio State University, Columbus, OH, USA*
[3] *Department of Haematology, University College London Hospitals NHS Trust, London, UK*
[4] *Department of Hematology, Reference Center for Thrombotic Microangiopathies (CNR-MAT), Saint-Antoine University Hospital, AP-HP, Paris, France*
[5] *Department of Medicine 1, Division of Hematology and Hemostasis, Medical University of Vienna, Austria*
[6] *Department of Hematology and Central Hematology Laboratory, Inselspital, Bern University Hospital, University of Bern, Bern, Switzerland*
[7] *Division of Hematology, Department of Medicine, University of Colorado–Anschutz Medical Center, Denver, CO, USA*
[8] *Hematology Department, Internal Medicine, School of Medicine and Dentistry, Catholic University of Valencia and Hospital Doctor Peset, Valencia, Spain*
[9] *Departments of Medicine and Laboratory Medicine, St. Michael's Hospital and University of Toronto, Toronto, ON, Canada*
[10] *Clinical Development, Ablynx, a Sanofi company, Ghent, Belgium*
[11] *Medical Affairs, Sanofi, Diegem, Belgium*
[12] *Formerly Clinical Development, Ablynx, a Sanofi company, Ghent, Belgium*

Objective: An integrated analysis based on the Phase 2 TITAN (NCT01151423) and Phase 3 HERCULES (NCT02553317) studies with caplacizumab (CPLZ) in acquired thrombotic thrombocytopenic purpura (aTTP) was performed to assess treatment

differences on efficacy and safety outcomes that may have been undetected in the individual trials. **Methodology:** In both trials, patients with an acute episode of aTTP were randomized to receive CPLZ or placebo (PBO) in addition to therapeutic plasma exchange (TPE) and immunosuppression. All randomized patients from both studies were included in the integrated efficacy analyses (CPLZ: n=108; PBO: n=112), and those who received at least 1 dose of the study drug were included in the safety analyses (CPLZ: n=106; PBO: n=110). **Results:** CPLZ significantly reduced mortality (0 vs 4 deaths; P<0.05) and refractory TTP (0 vs 8 events; P<0.05) versus PBO and improved time to platelet count response (hazard ratio, 1.65; P<0.001). CPLZ also reduced the composite endpoint of TTP-related death, exacerbation, or any treatment-emergent major thromboembolic event during the treatment period (13.0% vs 47.3%; P<0.001) and median number of TPE days (5.0 vs 7.5 days) versus PBO. Mild mucocutaneous bleeding was the main safety finding for CPLZ. **Conclusion:** This integrated analysis provided new evidence that CPLZ prevents mortality and refractory disease in aTTP and reinforced the individual trial efficacy and safety findings. No new safety signals were identified for

https://doi.org/10.1016/j.htct.2021.10.1047

PP 38

EPIDEMIOLOGY, TREATMENT PATTERNS, AND CLINICAL OUTCOMES AMONG PATIENTS WITH ACQUIRED THROMBOTIC THROMBOCYTOPENIC PURPURA (ATTP) IN THE UNITED STATES: AN ELECTRONIC HEALTH RECORDS ANALYSIS

Ayoade Adeyemi [1], Filip Callewaert [1], Francesca Razakariasa [2], Rui de Passos Sousa [1]

[1] *Sanofi*
[2] *Quinten France*

Objective: Acquired thrombotic thrombocytopenic purpura (aTTP) is an ultra-rare, potentially life-threatening thrombotic microangiopathy (TMA). Data on epidemiology, disease management, and clinical outcomes are scarce and often heterogeneous. The aim of this study was to assess the epidemiology, disease management, and clinical outcomes in patients with aTTP in the United States. **Methodology:** This longitudinal retrospective observational study of the Optum-Humedica database included patients with aTTP diagnosis from October 2015 to December 2019 if they had ≥1 documented ADAMTS13 activity <10% or ≥1 aTTP episode (≥1 inpatient stay with TMA diagnosis and ≥1 therapeutic plasma exchange [TPE] during the same stay); patients with conditions that mimic aTTP were excluded. Patients were followed until loss to follow-up, end of study period, or death. All analyses were descriptive. **Results:** Among 666 patients with aTTP diagnosis, 302 (45%) had ≥1 aTTP episode. Annual incidence of ≥1 aTTP episode was 1.81/million (based on data from 2016–2019). Patients with ≥1 aTTP episode received a mean of 16.7 TPE sessions; 59% used rituximab. Among patients with ≥1 aTTP episode, exacerbations occurred in 17% (52/302); relapse occurred in 11% (34/302). Mortality rate was 25% (167/666) among all patients with aTTP diagnosis and 14% (41/302) among patients with ≥1 aTTP episode. **Conclusion:** Despite treatment with TPE and immunosuppressants, the high mortality and morbidity observed in this patient population demonstrates the need for more effective therapies to improve clinical outcomes.

https://doi.org/10.1016/j.htct.2021.10.1048

STEM CELL TRANSPLANT

PP 39

THE ROLE OF SUBPOPULATIONS OF MOBILIZED PERIPHERAL HEMATOPOIETIC STEM CELLS IN THE RESTORATION OF HEMATOPOIESIS DURING HIGH-DOSE CHEMOTHERAPY IN CANCER PATIENTS

Liudmila GRIVTSOVA [1], Nikolai TUPITSYN [2]

[1] *A.F. Tsyb Medical Radiological Research Center – branch of the National Medical Research Radiological Center, Ministry of Health of Russia*
[2] *FSBI NMITs oncology named after N.N. Blokhin "of the Ministry of Health of Russia*

Objective: Mobilized peripheral hematopoietic stem cells are transplanted to cancer patients as support for high-dose chemotherapy. It is believed that the effectiveness of restoring all hematopoietic sprouts during HSC transplantation depends on the total dose of CD34+ cells. At the same time, CD34+ stem cells are a heterogeneous cell pool, including progenitor cells of different levels of differentiation and different ability to proliferate. Accordingly, it can be expected that the subpopulation composit. **Methodology:** We have studied of HSC subsets in 569 specimens of hemopoietic tissue (blood cells and LP cells) from 167 adult cancer patients and on 557 specimens of hemopoietic tissue from 263 pediatric cancer patients. Also, 61 samples of LP from 50 healthy HSC donors were studied. All patients were managed at bone marrow transplantation units of hematology malignancy and oncology department of N.N. Blokhin Cancer Research Center from 1996 to 2014. **Results:** Peripheral hemopoietic stem cells (HSC) that are transplanted to cancer patients to reduce critical pancytopenia vary in subset composition and include early polypotent precursors (CD38- and/or HLA-DR-, CD90+, CD45-negative), lymphoid precursors (CD10+, CD7+, CD2+, CD19+, CD56+), megakaryocyte- (CD61+) and myeloid-committed precursors (CD117+, CD13+, CD33+). These subsets of early and committed HSC are found in different proportions in cancer patients and normal donors. **Conclusion:** So, the pool of mobilized HSC is heterogeneous and represented by pluripotent precursors and committed HSC in different proportions that are in variable, rather sophisticated interrelations. Mobilization effect of SC individual subsets is related with disease type. To achieve fast recovery of granulocyte lineages after HSC autologous or allogeneic transplantation one should not

focus only on proportion of committed myeloid HSC: optimal HSC content to be transplanted should be in a certain balance.

https://doi.org/10.1016/j.htct.2021.10.1049

PP 40

HEMATOLOGICAL FINDINGS IN COVID-19 AND INSIGHTS TO STEM CELL THERAPY

Ghada ELGohary

King Khaled University Hospitals

Objective: As the COVID-19 was spreading to all countries, its manifestations were identifying gradually, which were related to several organs. COVID-19 is associated with distinct hematological changes, increased serum inflammatory markers, and coagulopathy. **Methodology:** Most of the known COVID 19 complications are related to the patients' prognosis and mortality, particularly in those with severe disease, the issue which attract the scientists and the medical physicians all over the world to find the proper treatment for such monter, we discussed the associations between COVID-19 clinical features and complications, and secondly, its hematological findings and coagulopathy are investigated. **Conclusions:** Such associations not only may shed light on our prognostic view of patients with COVID-19 but also will entail significant therapeutic implications. One of its key implications is to utilize the mesenchymal stem cells (MSCs) to treat patients with COVID-19. Herein, this kind of novel therapy has been discussed, as well with its cons and pros points

https://doi.org/10.1016/j.htct.2021.10.1050

PP 41

OUTCOMES OF ALLOGENEIC SC TRANSPLANT IN HEMATOLOGICAL MALIGNANCY PATIENTS USING BUSULFAN 3 (9.6 MG/KG) BASED CONDITIONING REGIMEN

Majed Altareb, Yazeed Bajuaifer, Walid Rasheed

King Faisal Specialest Hostpital

Objective: To study the outcomes of allo-HCT in patients with hematological malignancy who received BU3 (9.6 mg/kg) based conditioning from matched related or unrelated donors. **Methodology:** A retrospective analysis of KFSHRC-BMT Database, we identified 65 patients who received Allo-HCT between October 2005 and December 2019 at King Faisal Specialist Hospital & Research Center. The patients received SCT from full HLA matched related or unrelated donors. We excluded Mismatched MUD, Cord & Haplo-identical Stem Cell sources. **Results:** We identified 47 AML (72.3%), 8 MDS (12.3%), 8 Myelofibrosis (12.3%) & 2 CML (3.1%) patients. Acute GvHD grade II-IV and III-IV occurred in 29% and 14% respectively. Chronic GvHD occurred in 55% and was extensive in 24% of patients. With Median follow-up 60.5 months, 2 years and 5 years OS were 58.5 % and 44.1% respectively. The 2 years and 5 years DFS were 52.9% and 44.5% respectively. Cumulative incidence of relapse and NRM at 2- years were 29.5% and 17.4% respectively. Day +100 TRM were 10.7% **Conclusion:** Allogeneic SCT using BU3 based regimen appears feasible to use in patients who are not suitable for fully myeloablative (BU4) regimen. TRM, DFS & OS rate were comparable to reports from studies using BU4 based regimen, warranting prospective studies in these patients.

https://doi.org/10.1016/j.htct.2021.10.1051

PP 42

A CASE OF REFRACTORY IMMUNE THROMBOCYTOPENIA APPLIED WITH AUTOLOGOUS HEMATOPOETIC STEM CELL TRANSPLANTATION

Mine KARADENIZ [1], Mete UCDAL [2], Hakan GOKER [1], Umit Yavuz MALKAN [1]

[1] *Hacettepe University Faculty of Medicine, Department of Hematology, Ankara, Turkey*
[2] *Hacettepe University Faculty of Medicine, Department of Internal Medicine, Ankara, Turkey*

Case report: A 61-year-old male patient who had previously been splenectomized for immune thrombocytopenia, hospitalized with mucosal bleeding. Upon failure to respond to steroid, intravenous immunoglobulin, rituximab, danazol, azathioprine and eltrombopag treatment, autologous hematopoietic stem cell transplantation was performed to the patient. At the end of the first month, he had normal platelet count.

https://doi.org/10.1016/j.htct.2021.10.1052

PP 43

INVESTIGATION OF DRUG-DRUG INTERACTIONS INVOLVING ANTI-INFECTIVE DRUGS IN PATIENTS UNDERWENT HEMATOPOIETIC STEM CELL TRANSPLANTATION

Ayse Gunay [1], Eren Demirpolat [1], Ali Unal [2]

[1] *Erciyes University, Faculty of Pharmacy, Clinical Pharmacy Department*
[2] *Erciyes University, Faculty of Medicine, Hematology Department*

Objective: Drug-drug interactions are an important cause of adverse drug events. The preventable or manageable nature of drug-drug interactions puts them at the center of interventions. Since hematopoietic stem cell transplantation is a challenging and multi-drug process, drug-drug interactions are frequently encountered. **Methodology:** In our study, the drugs used by a total of 100 patients with 50 autologous and 50

allogeneic bone marrow transplants for 10 days before transplantation, on the day of transplantation and for 10 days after transplantation were examined retrospectively in terms of interaction. Two paid softwares and two free softwares were used to examine interactions. The obtained data were analyzed with Microsoft Excel program. **Results:** A total of 3805 interactions were observed in the 21-day period in 50 patients who underwent allogeneic stem cell transplantation, and these interactions occurred with the repetition of 1017 interactions in different patients. For the same period in 50 autologous stem cell transplant patients, 2906 interactions were detected, and this number occurred with 725 different interactions seen in different patients. It has been understood that anti-infectives cause serious interaction load. **Conclusion:** Hematopoietic stem cell transplantation is a period in which prophylactic or anti-infective treatment for the detected microorganism is applied intensively. Interactions of anti-infectives with each other and with other drugs in the treatment regimen are frequently encountered during the transport process. Interactions should be identified and their clinical significance should be demonstrated. It should be handled with the partnership of physician-clinical pharmacist.

https://doi.org/10.1016/j.htct.2021.10.1053

PP 44

EVALUATION OF COMMON INTERACTIONS INCLUDING ANTI-INFECTIVE DRUGS IN PATIENTS UNDERWENT AUTOLOGOUS AND ALLOGENEIC STEM CELL TRANSPLANTATION

Ayse Gunay [1], Ali Unal [2], Eren Demirpolat [1]

[1] *Erciyes University, Faculty of Pharmacy, Clinical Pharmacy Department*
[2] *Erciyes University, Faculty of Medicine, Hematology Department*

Objective: Hematopoietic stem cell transplantation is a challenging process involving polypharmacy. Drug-drug interactions are common due to the large number of drugs used in patients, and antiinfectives are frequently involved in interactions due to their widespread use. **Methodology:** In our study, the drugs used by a total of 100 patients with 50 autologous and 50 allogeneic bone marrow transplants for 10 days before transplantation, on the day of transplantation and for 10 days after transplantation were examined retrospectively in terms of interaction. Two paid softwares and two free softwares were used to examine interactions. The obtained data were analyzed with Microsoft Excel program. **Results:** 1017 different interactions were detected in patients with allogeneic bone marrow transplantation and 725 different interactions in patients with autologous bone marrow transplantation. It was observed that 342 interactions were common in the two transplant types. Interactions involving anti-infectives have been studied and the data showed antifungals, antibacterials and antivirals cause significant interaction load. Some interactions were found to be dependent on the transplant process. **Conclusion:** Allogeneic bone marrow transplantation and autologous bone marrow transplantation are challenging processes in which intensive drug therapy is applied. Knowing the interactions that are common to both types of transplantation and the interactions involving anti-infectives specific to a certain period of the transplantation process allows the process to be managed effectively. It is important to manage interactions in physician-clinical pharmacist collaboration

https://doi.org/10.1016/j.htct.2021.10.1054

TRANSFUSION MEDICINE AND APHERESIS

PP 45

THERAPEUTIC PLASMA EXCHANGE IN PATIENTS WITH NEUROLOGICAL DISEASES: A 9-YEAR, SINGLE-CENTER EXPERIENCE

Merih Reis Aras [1], Mehlika Panpalli Ates [2], Murat Albayrak [1], Hacer Berna Afacan Ozturk [1], Abdulkerim Yildiz [3], Fatma Yilmaz [1], Bugra Saglam [1], Selim Selcuk Comoglu [2]

[1] *University of Health Sciences Ankara Diskapi Yildirim Beyazit Training and Research Hospital, Hematology Department*
[2] *University of Health Sciences Ankara Diskapi Yildirim Beyazit Training and Research Hospital, Neurology Department*
[3] *Ministry of Health Hitit University Erol Olcok Training and Research Hospital, Hematology Department*

Objective: Therapeutic plasma exchange (TPE), is based on the removal of pathogenic substrates from plasma with replacement fluid. TPE is being used in the treatment of many neurological diseases, especially Myasthenia Gravis (MG) and Guillain Barre Syndrome (GBS). The aim of this study is to analyse the efficay and safety of TPE experience in neurological disorders. **Methodology:** We reviewed the medical records of all 59 patients who received a total of 267 therapeutic cycles between 2012 and 2021 in our tertiary care university hospital. Respond assesment was evaluated with Medical Research Council (MRC) scoring system. Neutrophil count, lymphocyte count and neutrophil/lymphocyte ratio was recorded before any treatment and 7 days after the last plasmapheresis cycle. **Results:** Of the 59 patients, 30 (50.8%) were male and 29 (49.2%) were female. Of these patients 44.1% were diagnosed with MG, 27.3% with GBS, %8.5 with Multiple Sclerosis (MS). The median number of TPE sessions per patient was 5 [1-7]. 33.9 % of patients had at least one complication that hypotension was the most seen (%22). Overall response rate was %76.3. MRC score was significantly higher in the group with response than the group without symptom regression (p <0.05). **Conclusion:** TPE is a safe and an effective treatment option in neurological diseases. TPE related side effects/complications were generally mild to moderate and manageable. Performing the TPE response evaluation with the MRC scoring system was beneficial for the reliability of the efficacy as a concrete finding.

https://doi.org/10.1016/j.htct.2021.10.1055

OTHER DISEASES

PP 46

DOES BLOOD TYPE HAVE AN EFFECT ON THE COURSE OF COVID-19?

Fatma YILMAZ [1], Murat ALBAYRAK [1],
Abdülkerim YILDIZ [2],
Hacer Berna AFACAN ÖZTÜRK [1],
Senem MARAL [1], Pınar AKYOL [1],
Merih REİS ARAS [1], Buğra SAĞLAM [1],
Mesut TIĞLIOĞLU [1]

[1] *University of Health Sciences, Diskapi Yildirim Beyazit Training and Research Hospital, Department of Hematology*
[2] *University of Hitit,Erol Olçok Training and Research Hospital, Department of Hematology*

Objective: Predictive parameters that can affect the course of this infection have been the main topic of research since the beginning of the COVID-19 (Coronavirus disease 2019) pandemic. Since the discovery of blood groups, the effect of these on infectious diseases has always been of interest **Methodology:** To analyze the effect of ABO blood group on mortality, hospitalization duration and hematological and cytokine storm parameters in patients with COVID-19. This retrospective study was conducted on 140 patients diagnosed with COVID-19. Demographic characteristics, laboratory parameters including ABO blood group, complete blood count (CBC) parameters, biochemical tests, cytokine storm parameters, duration of hospitalization, and final status (discharge or death) were recorded. **Results:** The 140 patients included in the analysis comprised 72 (51.4%) males and 68 (48.6%) females with a mean age of 66.3±14.0 years. . Age and gender, hospitalization duration and mortality rates were similar in all blood group types. Only D-dimer values were found to be higher in blood group A compared with other blood groups. **Conclusion:** Although no difference in mortality was determined between groups, the D-dimer level was statistically significantly higher in COVID-19 patients with A blood group. Larger studies are needed to reflect D-dimer levels on the clinical course of infection, and thus on daily practice

https://doi.org/10.1016/j.htct.2021.10.1056

PP 47

RECURRENT AUTOIMMUNE HEMOLYTIC ANEMIA AFTER MRNA COVID-19 VACCINE (PFIZER-BIONTECH)

Fatma YILMAZ, Murat ALBAYRAK,
Merih REİS ARAS, Senem MARAL,
Hacer Berna AFACAN ÖZTÜRK,
Pınar TIĞLIOĞLU, Mesut TIĞLIOĞLU,
Buğra SAĞLAM

Health Sciences University Diskapı Yıldırım Beyazit Training and Research Hospital, Hematology Clinic

Case report: One of the causes of autoimmune hemolytic anemia is drugs. Vaccination is the most important step in the management of the COVID-19 pandemic. After receiving the m-RNA COVID-19 (Pfizer-BioNTech) vaccine, the patient admitted with weakness and jaundice for the last three days. Laboratory results are consistent with AIHA recurrence. Splenectomy was performed after the patient stabilized with rituximab therapy. Especially newly developed therapeutic agents have a potential risk of new side effects.

https://doi.org/10.1016/j.htct.2021.10.1057

PP 48

EXTRAMEDULLARY HEMATOPOIESIS IN PATIENTS WITH TRANSFUSION DEPENDENT β-THALASSEMIA (TDT): A SYSTEMATIC REVIEW

Eihab Subahi [1], Fateen Ata [1], Hassan Choudry [2],
Phool Iqbal [1], Mousa AlHiyari [1],
Ashraf Soliman [3], Vincenzo De Sanctis [4],
Mohamed Yassin [1]

[1] *Hamad Medical Corporation*
[2] *Faisalabad Medical University*
[3] *University of Alexandria*
[4] *Quisisana Hospital*

Objective: Thalassemia is one of the most common hemoglobinopathies, with around 5% of the world's population expected to have some degree and type of thalassemia. Beta thalassemia (BT) occurs due to a deficient production of the beta-globin chain of hemoglobin. Extramedullary hematopoiesis (EMH) is one of the complications of BT, mainly observed in minor/intermedia subtypes. EMH is the production of blood cells outside the marrow as a compensatory response to long-standing hypoxia. Due to chronic transfusions, it is not expected in patients with beta-thalassemia major (BTM). However, there are increasingly reported cases of EMH in BTM. The incidence of EMH in BTM is thought to be <1%. However, it seems that the true incidence is much higher than expected. This review aims to pool the available data and provide cumulative evidence on the occurrence of EMH in BTM patients. **Methodology:** We aim to conduct a systematic review via searching multiple electronic databases (PubMed, Scopus, Google Scholar) to identify eligible articles from any date up to December 2020. Eligible studies should report extramedullary hematopoiesis in beta-thalassemia major. Case reports, case series, observational studies with cross-sectional or prospective research design, case-control studies, and experimental studies will be included if found relevant. Two reviewers (FA and ES) will individually analyze the study quality using the statistical methodology and categories guided by the Cochrane Collaboration Handbook, PRISMA guidelines, and Joanna Briggs Institute checklist for case reports and series. **Results:** Data from 253 cases of EMH in BTM patients were extracted with mean age of 35.3 +/- 0.5 years. Mean hemoglobin at presentation with EMH was 8.2 +/- 2.1 mg/dL. Lower limb weakness was the most common presenting feature (N=23) (paraspinal EMH). Magnetic

resonance imaging (MRI) was the most widely used diagnostic modality (N=226). Overall, blood transfusion was the commonest reported treatment (N=30), followed by radiotherapy (N=20), surgery (N=15), hydroxyurea (N=12), steroids (N=6), and exchange transfusion (N=2). An outcome was reported in 20% of patients, all recovered, with no reported mortality. **Conclusion:** EMH is considered a rare phenomenon in BTM patients. It can occur in any organ system with varied clinical features. MRI can effectively diagnose EMH, and conservative management has similar results compared to invasive treatments. Larger studies, focusing on outcomes may enhance guidelines on preventive and therapeutic strategies for managing EMH in BTM.

https://doi.org/10.1016/j.htct.2021.10.1058

PP 49

LATE DIAGNOSIS OF CONGENITAL METHEMOGLOBINEMIA IN A 33-YEAR-OLD CYANOTIC PATIENT, CASE REPORT AND REVIEW OF LITERATURE

MAYA ALDEEB, MOHAMED YASSIN

Hamad Medical Corporation

Objective: Due to mild symptoms, congenital methemoglobinemia is rarely diagnosed and reported as a cause of the cyanosis, especially in adults. Despite the benign nature of congenital methemoglobinemia, it is crucial to keep it in the differential diagnosis list when assessing cyanotic patients, mainly if he has a normal PaO2. Patients are usually asymptomatic and are treated for cosmetic purposes, but they might suffer from severe complications if exposed to oxidative agents. **Case report:** A 33-year-old lady presented to ED with difficulty breathing and bluish discoloration gradually increased over days, without fever or cough. she mentioned having recurrent similar episodes since childhood. family history positive for similar episodes in the sister. physical examination positive for central and peripheral cyanosis, with o2sat of 88% and RR of 20, the rest of examination within normal limits. laboratory tests normal except for ht 48.9%, PO2 160 on ABGs. **Methodology:** The patient's clinical picture of cyanosis with no evidence of cardiovascular or pulmonary diseases and the discrepancy between PaO2 and O2 saturation on oximeter required thinking of methemoglobinemia as a possible diagnosis despite the patient's age and the absence of any exposures. Methemoglobin level 20.9% (0-1.5%). Hemoglobin electrophoresis did not detect any abnormal hemoglobin. The activity of NADH cytochrome b5 reductase or the level were not done. **Results:** We searched PubMed and Google Scholar, we found 22 articles with a total of 30 patients with congenital methemoglobinemia. The mean age of the patients was 25 years (range 2 days-61 years); most of them were previously healthy. Out of 30 patients, 16 were treated with ascorbic acid or methylene blue or both with improvement, 14 either were not treated or treatment not mentioned in the report. Our patient received ascorbic acid 500 mg BID orally and improved clinically and laboratory. **Conclusion:** Due to mild symptoms, congenital methemoglobinemia is rarely diagnosed and reported as a cause of the cyanosis, especially in adults. Despite the benign nature of congenital methemoglobinemia, it is crucial to keep it in the differential diagnosis list when assessing cyanotic patients, mainly if he has a normal PaO2. Patients are usually asymptomatic and are treated for cosmetic purposes, but they might suffer from severe complications if exposed to oxidative agents.

https://doi.org/10.1016/j.htct.2021.10.1059

PP 50

A RARE ASSOCIATION IN A CASE WITH HEREDITARY SPHEROCYTOSIS: SPECTRIN BETA (SPTB) AND JAK-2 MUTATION

Fatma YILMAZ, Murat ALBAYRAK, Merih REİS ARAS, Senem MARAL, Hacer Berna AFACAN ÖZTÜRK, Pınar TIĞLIOĞLU, Mesut TIĞLIOĞLU, Buğra SAĞLAM

Health Sciences University Diskapı Yıldırım Beyazit Training and Research Hospital, Hematology Clinic

Case report: Five types of gene variants are seen in hereditary spherocytosis (ANK, SPTB, SPTA1, SLC4A1, EPB42). JAK2 V617F mutation; is most common seen in bcr-abl negative chronic myeloproliferative diseases. As a result of NGS performed before splenectomy, SPTB c.4973+2T> C and JAK-2 c.1849G>T p.(Val617Phe) mutations were detected. Co-occurrence of these two mutations requires special attention in terms of the management of thrombocytosis and side effects that may occur after splenectomy.

https://doi.org/10.1016/j.htct.2021.10.1060

PP 51

RETROSPECTIVE EVALUATION OF PATIENTS WITH LANGERHANS CELL HISTIOCYTOSIS FOLLOWED IN OUR CLINIC

Mehmet Can Ugur [1], Cansu Atmaca Mutlu [2], Sinem Namdaroglu [1], Füsun Gediz [3], Oktay Bilgir [1]

[1] *Health Sciences University, Bozyaka Training and Research Hospital, Department of Hematology*
[2] *Izmir Democrasy University, Buca Seyfi Demirsoy Training and Research Hospital, Hematology*
[3] *Izmir Economy University, Medicalpark Hospital, Deparment of Hematology*

Objective: Langerhans cell histiocytosis (LCH) is a rare histiocytic disorder that can be especially seen in children and young adults. The clinical presentation of patients with LCH varies according to the sites of involvement. In about half of patients, the disease is limited to a single organ system and bone involvement is very common. In this study, it was aimed

to retrospectively evaluation the patients diagnosed with LCH who were followed up and treated in our clinic. **Methodology:** The data of patients over the age of 18 years who were followed up and treated in Bozyaka Training and Research Hospital Hematology Clinic between 2015-2021 were scanned retrospectively from the hospital system. **Results:** Data of 6 patients were obtained. The mean age of the patients was 33.6. There was no difference between the genders. Pain was the reason for admission in 4 patients and was the most common symptom. While the most frequently involved system was the skeletal system with 5 patients, lung involvement was seen in 2 patients. Vinblastine and prednisolone combination therapy was given to 1 patient, who developed steroid-related avascular necrosis. One patient who was planned for combination treatment **Conclusion:** LCH is a rare disease especially seen in children and young adults. It can involve the skeletal system, lungs, and other organs. The prognosis is often good with excision of the lesion or systemic treatment.

https://doi.org/10.1016/j.htct.2021.10.1061

PP 52

PILOMATRIX CARCINOMA WAS BEYOND ANY EXPECTATIONS: A CASE REPORT OF CARCINOMA CLINICIAN SHOULD BE AWARE OF IT

Abdalraouf Omar [1], Suad Elnnami [2], Ibtihaj Hassan [3]

[1] *Tripoli University Hospital and Albadry policlinic*
[2] *Tripoli University Hospital and University of Tripoli*
[3] *Tripoli University Hospital*

Introduction: Pilomatrix carcinoma is a rare cutaneous tumor derived from follicular matrix with about 150 reported cases and it is considered as locally aggressive tumor with a tendency to recur. In addition to lymph node metastases and poor prognosis, it is non-chemotherapy responding in systemic metastasis. **Method and result:** A 37-year-old Libyan man presented with two large, coalesced nodules in the face measuring about 3*2 cm and 3*3 cm, treated as a case of lipoma by surgical excision based on clinical diagnosis, reappearing of larger nodule 9*6*4 cm involving almost all the left check, surgical excision was done with histopathological features of pilomatrix carcinoma infiltrating to the subcutaneous adipose tissue and deep striated muscle with no clear margins. Conventional radiotherapy by electron beam was started using the linear accelerating machine, with total radiotherapy dose 60 gray (Gy) in 30 fraction for six weeks. No local recurrences, nor lymph node or systemic metastasis since June 2020. **Conclusion:** Pilomatrix carcinoma must always be considered in the differential diagnosis of nodular tumors of the head-and-neck due to high recurrence rate after simple excision. Furthermore, local radiotherapy post incomplete surgical excision is the best adjuvant therapy.

https://doi.org/10.1016/j.htct.2021.10.1062

PP 53

WHAT HAVE WE EXPERIENCED WITH COVID-19 IN PATIENTS WITH HEMATOLOGICAL DISORDERS?

Simge ERDEM [1], Metban MASTANZADE [1], Mustafa Murat OZBALAK [1], Sevda SERT EKINCI [2], Dilek Ozden OZLUK [1], Tarık Onur TIRYAKI [1], Beyza OLUK [1], Ipek YONAL HINDILERDEN [1], Mustafa Nuri YENEREL [1], Meliha NALCACI [1], Tufan TUKEK [2], Sevgi KALAYOGLU BESISIK [1]

[1] *Istanbul University Istanbul Faculty of Medicine, Internal Medicine Department/Hematology Division*
[2] *Istanbul University Istanbul Faculty of Medicine, Internal Medicine Department*

Objective: Patients with cancer are considered highly vulnerable to the COVID-19 disease. However, there are still few data in hematologic patients. Some small studies have shown a high mortality on patients with hematologic malignancies and COVID-19. In this study we aim to report a single center experience of the hematologic patient population with COVID-19 disease. **Methodology:** This single centre, retrospective, cohort study included a total of 111 adult patients (aged ≥18 years) with diagnosis of neoplastic and non-neoplastic hematologic diseases between March 2020 and August 2021. Ethics committee approval was obtained from the Istanbul University Istanbul Faculty of Medicine Clinical Research Ethics Committee. Categorical variables were compared using Pearson's Chi-square test. STATA16-MP was used for the statistical analysis. **Results:** A total of 111 patients (median age:55) with hematologic disease were diagnosed with COVID19. Ninety patients had neoplastic hematologic disorder. Fourty-five patients were receiving anti-neoplastic treatment at the time of COVID19 diagnosis. A total of 21 patients (overall mortality rate:19%) died and 19 of them had neoplastic disorder. The malignancy mortality rate was estimated to be 21%. 45 of 90 cases were receiving chemotherapy. Ten of these 45 patients (22%) died due to COVID19 disease. **Conclusion:** In our study the majority of patients who died due to COVID-19 had hematological malignancies. The cytokine storm which affects the clinical outcome in COVID-19 may contribute to dismal prognosis in hematologic malignancies in which cytokine increase is a part of process. Most of the succumbed patients were relapsed refractory multiple line treated which may reflect the immune insufficiency. It seemed COVID-19 progress is mostly poor in hematologic malignancies compared otherwise healthy people.

https://doi.org/10.1016/j.htct.2021.10.1063

PP 54

A RARE CAUSE OF ANEMIA IN ADULTHOOD CONGENITAL DYSERYTHROPETIC ANEMIA

Buğra Sağlam, Murat Albayrak, Hacer Berna Afacan Öztürk, Merih Reis Aras,

Pınar Tığlıoğlu, Mesut Tığlıoğlu, Fatma Yılmaz, Senem Maral

University of Health Sciences Ankara Dışkapı Yıldırım Beyazıt Training and Research Hospital, Department of Hematology, Ankara, TURKEY

Objective: Congenital dyserythropoietic anemia is a group of diseases characterized by ineffective erythropoiesis and multinuclear erythroblasts, mostly diagnosed in childhood. Although there are 3 main types, type II is the most common. We present our patient with congenital dyserythropoietic anemia, who was not diagnosed until the age of 49, to contribute to the literature. **Case report:** A 49-year-old male patient was admitted to our hospital with abdominal pain, weakness and yellowing of the eyes. His examinations revealed splenomegaly, cholelithiasis, anemia and hyperbilirubinemia. In the patient's anamnesis, he stated that he had jaundice and weakness since childhood, and that he knew that he had abdominal pain and spleen enlargement with advancing age. **Methodology:** Bone marrow biopsy was performed to the patient for a different diagnosis and cause. Binuclear erythroblasts were observed in the patient (fig. 1). As a result of the new generation sequencing performed on the patient who was evaluated as familial non-immune hemolytic anemia, c.1733T>C homozygous mutation in exon 15 of the SEC23B gene was detected and a diagnosis of congenital dyserythropetic anemia type II was made **Results:** Congenital dyserythropoietic anemias (CDA) represent a large group of diseases that mainly result in ineffective erythropoiesis. Morphological changes observed in the bone marrow over a long period of time were its main diagnostic features. Together with 3 main subtypes, they are examined in a total of 5 subtypes. CDA type II is most common. Clinically normal or slightly increased reticulocyte count is characterized by a variable degree of normocytic anemia. **Conclusion:** Diagnosing CDA cases: It is closely related to the clinician's ability to remember and access genetic tests, especially in advanced ages. Considering that access to genetic tests will increase in the future, many undiagnosed cases may come up. Although our treatment possibilities are limited in the current situation, future treatment methods are promising. However, studies are still needed to understand this disease and its mechanisms.

https://doi.org/10.1016/j.htct.2021.10.1064

QUALITY IMPROVEMENT / PATIENT SAFETY

PP 55

HEMATOLOGIC REFERENCE VALUES FOR HEALTHY ADULT SAUDIS

Salwa Bakr

Faculty of Medicine, Fayoum

Background: Laboratory hematological tests are widely used in clinical practice to assess health and disease conditions. Although, the International Federation of Clinical Chemistry and Laboratory Medicine (IFCC) and the Clinical and Laboratory Standards Institute (CLSI) recommended that reference ranges should be established for each region , to the best of our knowledge, no study has described the reference values of routine hematological parameters in healthy Saudi adults. **Objectives:** To provide reference values of routine hematological parameters in Saudi adults according to age and gender. Material and Methods: A total of 827 adults potentially healthy Saudi participants with age ranging from 15 to 65 years were enrolled in this cross-sectional study from the central province of Saudi Arabia, Riyadh city. **Results:** The reference values of routine hematological parameters (full blood count, hemostatic profile, and biochemical tests of serum hematinic) according to gender were provided in detail (mean, SD, range, median, upper and lower limits) after exclusion of 157 due to various reasons. No difference in any hematological values were observed in relation to age. Current study hematological parameters' reference ranges were mostly different to the universal established ranges. **Conclusion:** This novel study provides the reference ranges of routine hematologic parameters for adult Saudi population for accurate assessments and appropriate management of routine clinical care, hence, to improve quality in health care.

https://doi.org/10.1016/j.htct.2021.10.1065

PEDIATRIC HEMATOLOGY ABSTRACT CATEGORIES

COAGULATION AND FIBRINOLYSIS DISORDERS

PP 56

A CASE REPORT WITH SEVERE CONGENITAL FACTOR XIII DEFICIENCY AND AN UNCOMPLICATED PREGNANCY AND BIRTH PROCESS

İbrahim Eker [1], Özge Vural [2], Mehmet Yılmazer [3], Nilgün Eroğlu [1], Yeter Düzenli Kar [1]

[1] *Afyonkarahisar Health Sciences University Department of Pediatric Hematology and Oncology*
[2] *Gazi University Medical School Department of Pediatric Hematology and Oncology*
[3] *Afyonkarahisar Health Sciences University Department of Obstetrics and Gynecology*

Introduction: Factor XIII deficiency is an extremely rare type among bleeding diathesis. In factor XIII deficiency, normal results of coagulation screening tests are expected. It usually does not cause spontaneous bleeding. Apart from bleeding diathesis, it may cause delayed wound healing and recurrent spontaneous abortions in women. Here, we present a 32-year-old case with severe congenital factor XIII deficiency who had an uncomplicated pregnancy and birth with regular replacement therapies. **Case report:** A 32-year-old patient with severe congenital factor XIII deficiency, who had a history of spontaneous abortion at the 11th week of her first pregnancy, applied to our center with a request for childbirth. It was learned that the factor XIII levels of the patient could not be measured, that she was using plasma-derived FXIII concentrate at a dose of 25 units/kg every time once a month, and in cases where this could not be obtained, 5 units/kg

cryoprecipitate was given instead. After the completion of the pre-pregnancy assessments, starting 3 months before the planned pregnancy and continuing for the whole pregnancy and for 3 months after birth, 25 units/kg plasma-derived concentrate at a dose of 25 units/kg was applied each time and every two weeks, and in cases where this could not be provided, the follow-up was continued by applying cryoprecipitate at a dose of 5 units / kg instead. During this whole process, FXIII levels ranged between 70% and 100%. The patient, who developed an abortion risk due to decidual bleeding in the first trimester, was hospitalized and an additional 25 units / kg plasma-derived FXIII concentrate was administered and a parenteral dose of 30 mg / kg tranexamic acid was applied until the signs of decidual bleeding disappeared. An additional 50 units/kg dose of plasma-derived FXIII concentrate was administered to the patient 30 minutes before birth who had a planned delivery by cesarean section at 38 weeks of gestation, and 30 mg/kg parenteral tranexamic acid was administered for 7 days following the delivery. FXIII level was detected 50% in the child of a healthy, 3500-g born boy. The patient and her baby, who are in the first year after birth, are followed up without any complications, and prophylactic plasma-derived FXIII concentrate or cryoprecipitate is administered to the patient once a month. **Discussion and Conclusion:** Inherited bleeding diathesis lead to an increased risk of bleeding and abortion in obstetric patients. Factor XIII deficiency is an extremely rare type among them. FXIII has a role in angiogenesis as well as hemostasis. Therefore, wound healing and tissue repair are impaired in Factor XIII deficiency. The risk of premature separation of the placenta, miscarriage especially in the first trimester, and postpartum uterine bleeding are increased in FXIII deficiency. Tranexamic acid can be used safely in obstetric patients with bleeding diathesis. It may be possible to ensure that patients with factor XIII deficiency have an uncomplicated pregnancy and delivery with regular follow-ups, regular prophylactic factor preparations, plasma replacements if they are not found, and in cases of bleeding, with additional doses of factor preparations or plasma replacement applications with tranexamic acid.

https://doi.org/10.1016/j.htct.2021.10.1066

PP 57

EVALUATION OF THE FREQUENCY OF ARTERIAL AND VENOUS THROMBOSIS AND PREDISPOSING FACTORS IN PATIENTS USING ELTROMBOPAG

Derya Deniz Kurekci, Melda Isevi, Engin Kelkitli, Mehmet Turgut

19 Mayıs University Department of Hematology

Objective: Eltrombopag is a small molecule thrombopoietin-receptor agonist used orally for the treatment.There is a high risk of thrombosis associated with the use of Eltrombopag. Our aim in this study is evaluating the incidence of arterial and venous thrombosis in patients using Eltrombopag and followed up in our center with the diagnosis of ITP, MDS and aplastic anemia, and contributing to the literature with the data of Central Black Sea by retrospectively evaluating the predisposing factors. **Methodology:** In this study, the data of 144 patients who were treated with Eltrombopag with the diagnosis of ITP, MDS and aplastic anemia at Ondokuz Mayıs University Faculty of Medicine Hematology Clinic between 2009-2019 were analyzed retrospectively. The data of the patients were obtained retrospectively from the hospital management information system. **Results:** The study included 144 patients who treated with Eltrombopag. 66 (45.8%) of the patients were male and 78 (54.2%) were female. The mean age of the patients was 54.12 ± 20.08 years. 102 (70.8%) of the patient were diagnosed with ITP, 31 (21.5%) with aplastic anemia and 12 (7.6%) with MDS. Thrombosis was observed in 7 (4.9%) of 144 patients who treated with Eltrombopag. Venous thrombosis was found in 6 (4.2%) of these patients and arterial thrombosis was found in one patient (0.7%). **Conclusion:** Eltrombopag treatment poses a risk for thromboembolic events. The treatment process should be followed closely especially in patients with known risk factors for thrombosis.

https://doi.org/10.1016/j.htct.2021.10.1067

PP 58

ETIOLOGY, TREATMENT AND FOLLOW-UP OF THROMBOSIS IN CHILDREN, ONE CENTER PROSPECTIVE TRIAL

Yunus Murat Akcabelen [1], Volkan Köse [1], Dilek Gürlek Gökçebay [1], Turan Bayhan [1], Neşe Yaralı [2], Namık Yaşar Özbek [1]

[1] *University of Health Sciences, Ankara City Hospital, Pediatric Hematology and Oncology*
[2] *Ankara Yıldırım Beyazıt University Ankara City Hospital, Pediatric Hematology and Oncology*

Objective: The aim of the study; To determine the frequency, etiological factors, treatment, long-term follow-up and recurrence rates of thrombosis in children. **Methodology:** Children with thrombosis in Ankara City Hospital between December 2018 and August 2021 were included. Patients were called or examined at 6–12-month intervals. **Results:** A total of 328 patients with a mean age of 6.9 were included. Catheter-related thrombosis was present in 52.7%.There were 14% arterial thrombosis and 59% venous thrombosis. Intracardiac thrombosis 16.2%, pulmonar thrombosis 2.4%, serebral thromboembolism %20 were detected.In the treatment, subcutaneous ondansetron (78.6%) was used mostly, but intravenous ondansetron was given in 6 patients and TPA was given 20 patients.In a mean follow-up of 15.8 months, 5 (1.5%) patients died due to thrombosis. **Conclusion:** Determining the etiological factors of patients with recurrence thrombosis is important for the duration of treatment in the follow-up.

https://doi.org/10.1016/j.htct.2021.10.1068

PP 59

THE EVALUATION OF HEALTHY CHILDREN WITH INCIDENTAL PROLONGATION OF PROTHROMBIN OR ACTIVATED PARTIAL THROMBOPLASTIN TESTS

Gökcen MERAL [1], Volkan KÖSE [1], Vildan ÇULHA [1], AYÇA KOCA YOZGATLI [1], NAMIK YAŞAR ÖZBEK [1], HÜSNİYE NEŞE YARALI [2]

[1] *MINISTRY OF HEALTH ANKARA CITY HOSPITAL, DEPARTMENT OF PEDIATRICS*
[2] *ANKARA YILDIRIM BEYAZIT UNIVERSITY FACULTY OF MEDICINE, DEPARTMENT OF PEDIATRICS*

Objective: This cross-sectional study aimed to reveal possible hemostatic disorders in patients referred to the Pediatric Hematology Department due to the prolongation of the prothrombin test (PT) or activated partial thromboplastin test (aPTT). **Methodology:** In this study, patients aged 0-18 years without known hematologic disease were referred to investigate the incidental prolonged PT and/or aPTT were evaluated. Mixing studies were performed in patients with continued PT/aPTT prolongation in the control examinations. Coagulation factor activities were analyzed in patients with improvement in mixing study. Antiphospholipid antibodies were studied in patients whose results did not improve with mixing studies. **Results:** Coagulopathy was found in 30% of 103 patients. Lupus anticoagulant positivity was found in two patients (1.9%). The most common factor (F) deficiencies were FVII deficiency (10.6%), FXI deficiency (7.8%), FXII deficiency (7.8%), FV deficiency (0.9%), FVIII deficiency (0.9%), fibrinogen and FVII deficiency (0.9%) and von Willebrand factor (vWF) deficiency (0.9%). Coagulopathy was more common in patients with bleeding disorders in their families, and this difference was statistically significant. **Conclusion:** In our study, mild factor deficiencies were more common than expected. Coagulation factor deficiencies can be seen in the patients without any finding of physical examination, personal and family histories. There is often no evidence of bleeding in mild factor deficiencies, and the clinical significance is unknown. We recommend using PT and aPTT as screening tests, especially before a major surgical intervention is performed.

https://doi.org/10.1016/j.htct.2021.10.1069

PLATELET DISORDERS / THROMBOSIS AND ANTITHROMBOTIC THERAPY

PP 60

CHILDHOOD IMMUNE THROMBOCYTOPENIA: A MULTICENTER QUESTIONNAIRE STUDY

Ayşegül Ünüvar [1], Melike Sezgin Evim [2], Serap Karaman [1], Arzu Akçay [3], İbrahim Eker [4], Funda Tayfun Küpesiz [5], Namık Özbek [6], Mehmet Ertem [7], Sultan Aydın [8], Zuhal Keskin [9], Yusuf Ziya Aral [10], Zülfükar Gördü [11], Murat Elli [12], Ayşe Özkan Karagenç [13], Burcu Belen Apak [14], Hülya Uzel [15], Murat Söker [15], Tuba Karapınar [16], Yeşim Oymak [16], Nihal Karadaş [17], Alper Özcan [18], Ersin Töret [19], Ülker Koçak [20], Sinan Akbayram [21], Şule Ünal Cangül [22], Aylin Canbolat Ayhan [23], Tiraje Celkan [24], Deniz Tuğcu [1], Bülent Zülfikar [25], Rejin Kebudi [25], Şadan Hacısalihoğlu [26], Erol Erduran [27], Sema Aylan Gelen [28], Nazan Sarper [28], Fatih Erbey [29], Emin Kürekçi [30], Hüseyin Gülen [31], Barış Yılmaz [32], Ömer Doğru [32], Ahmet Koç [32], Selma Ünal [33], Hüseyin Tokgöz [34], Canan Albayrak [35], Yılmaz Ay [36], Fatih Orhan [37], Davut Albayrak [38], Neslihan Karakurt [39], Betül Orhaner [40], Emine Türkkan [41], Yıldız Yıldırmak [42], Hadi Geylani [43], Begüm Koç [44], Ahmet Fayik Öner [45], Çetin Timur [46], Hale Ören [47]

[1] *Istanbul University, Istanbul School of Medicine, Division of Pediatric Hematology&Oncology*
[2] *Uludag University School of Medicine, Division of Pediatric Hematology*
[3] *Acibadem Altunizade Hospital, Pediatric Hematology&Oncology Unit*
[4] *Afyonkarahisar University of Health Sciences Medical Faculty, Division of Pediatric Hematology*
[5] *Akdeniz University School of Medicine, Division of Pediatric Hematology&Oncology*
[6] *Ankara City Hospital, Pediatric Hematology Unit*
[7] *Ankara University School of Medicine, Division of Pediatric Hematology*
[8] *Antalya Training and Research Hospital, Pediatric Hematology & Oncology Unit*
[9] *Ataturk University School of Medicine, Division of Pediatric Hematology&Oncology*
[10] *Aydin Adnan Menderes University School of Medicine, Division of Pediatric Hematology*
[11] *Aydin Maternity and Children's Hospital, Pediatric Hematology Unit*
[12] *Bagcilar Medipol Mega University Hospital, Division of Pediatric Hematology&Oncology*
[13] *Basaksehir Cam and Sakura City Hospital, Pediatric Hematology&Oncology Unit*
[14] *Baskent University School of Medicine, Division of Pediatric Hematology*
[15] *Dicle University School of Medicine, Division of Pediatric Hematology&Oncology*
[16] *Dr Behcet Uz Children's Hospital, Pediatric Hematology&Oncology Unit*
[17] *Ege University School of Medicine, Division of Pediatric Hematology*
[18] *Erciyes University School of Medicine, Division of Pediatric Hematology&Oncology*

[19] Eskisehir Osmangazi University School of Medicine, Division of Pediatric Hematology&Oncology
[20] Gazi University School of Medicine, Division of Pediatric Hematology
[21] Gaziantep University School of Medicine, Division of Pediatric Hematology
[22] Hacettepe University School of Medicine, Division of Pediatric Hematology
[23] Istanbul Medeniyet University School of Medicine, Prof. Dr. Suleyman Yalcin City Hospital, Pediatric Hematology&Oncology Unit
[24] Istanbul University-Cerrahpasa School of Medicine, Division of Pediatric Hematology&Oncology
[25] Istanbul University Oncology Institute, Division of Pediatric Hematology&Oncology
[26] Kanuni Sultan Suleyman Training and Research Hospital, Pediatric Hematology&Oncology Unit
[27] Karadeniz Technical University School of Medicine, Division of Pediatric Hematology
[28] Kocaeli University School of Medicine, Division of Pediatric Hematology
[29] Koc University School of Medicine, Division of Pediatric Hematology&Oncology
[30] Losante Child and Adult Hospital, Pediatric Hematology Unit
[31] Manisa Celal Bayar University School of Medicine, Division of Pediatric Hematology
[32] Marmara University, Istanbul Pendik Training and Research Hospital, Division of Pediatric Hematology&Oncology
[33] Mersin University School of Medicine, Division of Pediatric Hematology
[34] Necmettin Erbakan University, Meram School of Medicine, Division of Pediatric Hematology &Oncology
[35] On dokuz Mayis University School of Medicine, Division of Pediatric Hematology
[36] Pamukkale University School of Medicine, Division of Pediatric Hematology
[37] Sakarya University School of Medicine, Division of Pediatric Hematology&Oncology
[38] Samsun Medicalpark Hospital, Pediatric Hematology Unit
[39] Sancaktepe Training and Research Hospital, Pediatric Hematology Unit
[40] SBU Bursa Yuksek Ihtisas Training and Research Hospital, Pediatric Hematology&Oncology Unit
[41] SBU Okmeydanı Training and Research Hospital, Pediatric Hematology Unit
[42] SBU Sisli Hamidiye Etfal Training and Research Hospital, Pediatric Hematology Unit
[43] SBU Van Training and Research Hospital, Pediatric Hematology Unit
[44] Umraniye Training and Research Hospital, Pediatric Hematology&Oncology Unit
[45] Van Yuzuncu Yil University School of Medicine, Division of Pediatric Hematology&Oncology
[46] Yeditepe University School of Medicine, Division of Pediatric Hematology&Oncology
[47] Dokuz Eylul University School of Medicine, Division of Pediatric Hematology

Objective: A questionnaire form was prepared by the Turkish Pediatric Hematology Society- Subcommittee of Hemostasis, Thrombosis and Hemophilia to determine the current approaches in the diagnosis and treatment of childhood ITP in our country. Our aim was to share the results of this study, and to do new, national, multicenter prospective studies. **Methodology:** This form, which consists of twenty questions with multiple choices, but a brief explanation is requested when there is a different approach other than the options given, was sent to all pediatric hematologists via e-mail. **Results:** The response was obtained from 55 hematologists experienced in ITP from 47 centers in total. Due to space constraints, this summary could not present the survey questions and answers. **Conclusion:** In conclusion, the approaches for diagnosis and management of childhood ITP differ between centers.

https://doi.org/10.1016/j.htct.2021.10.1070

PP 61

THE CLINICAL PICTURE AND LABORATORY WORK-UP OF GLANZMANN THROMBASTHENIA

Joanna Zdziarska [1], Teresa Iwaniec [1], Ewa Wypasek [2], Tomasz Sacha [1]

[1] Department of Haematology, Jagiellonian University Medical College, Krakow, Poland
[2] Laboratory of Molecular Biology, John Paul II Hospital, 31-202 Krakow, Poland; Faculty of Medicine and Health Sciences, Andrzej Frycz Modrzewski Krakow University, 30-705 Krakow, Poland

Case report: We present the clinical picture and laboratory work-up of Glanzmann thrombasthenia, based on a group of 7 patients. Bleeding history was significant in all patients and included both mucosal and postsurgery bleeds. Laboratory analysis revealed decreased or absent platelet aggregation (< 10%) with all physiologic agonists (ADP, collagen, epinephrin, arachidonic acid) together with normal agglutination response to ristocetin. In three patients diagnosis was confirmed by flow cytometry.

https://doi.org/10.1016/j.htct.2021.10.1071

PP 62

THE USE OF ROMIPLOSTIM IN AN INFANT

Fatma Burçin KURTİPEK, Turan BAYHAN, Vildan ÇULHA, Neşe YARALI

Ankara City Hospital

Case report: Immune thrombocytopenia (ITP) is the most common platelet disorder in children, peaking between the

ages of 1-7.The first line therapy consists of intravenous immunoglobulin, anti-D immunoglobulin or corticosteroids. Second-line treatment options are immunosuppressive therapy, Rituximab.Thrombopoietin receptor agonists are used, which increase platelet production in the bone marrow. Our case report on a child with refractory chronic ITP, who failed the first and second line therapy.

https://doi.org/10.1016/j.htct.2021.10.1072

PP 63

ESSENTIAL THROMBOCYTOSIS IN CHILDREN

Canan ALBAYRAK [1], Davut ALBAYRAK [2]

[1] *Ondokuz Mayıs University Medical Faculty*
[2] *Samsun Medicalpark Hospital*

Objective: Sporadic essential thrombocytosis is a very rare disease in the childhood age group and its frequency has been reported as 1/1,000,000. WHO 2008 essential thrombocytosis diagnostic criteria; high platelet count for more than one year (>450 × 109/l), exclusion of reactive or secondary causes of thrombocytosis (iron deficiency, megaloblastic anemia, acute phase reactants, trauma, operation), no family history of myeloproliferative neoplasm and thrombocytosis, and WHO It can be summarized as the absence of myeloid neoplasm criteria. **Methodology:** In this study, seven cases diagnosed as sporadic essential thrombocytosis in our Pediatric Hematology clinic are presented. Six of the patients were girls and one was a boy. The median age at presentation was 13 years (the youngest 5 months, the oldest 15 years old). Application complaints: Headache, vertigo and tinnitus in adolescent children were not present in young children, they were detected incidentally. Thrombus was not detected in any patient. The median platelet count at diagnosis was 1442 × 109/l (range 963- 2438). **Results:** An increase in megakaryocytes was detected in bone marrow aspiration, no cytogenetic anomalies were found. Jak-2 (V617F) mutation was detected in one case and CALR mutation in two cases. No MPL (W515L) mutation was found in any case. In one case with a CALR mutation, a known type 2 mutation was detected, and in the other a new, previously unidentified mutation was detected. In the other four cases, no clonality was detected. Three cases with mutations and two cases with no mutations are being followed up with hydroxyurea therapy. The other two cases are using low-dose aspirin. Follow-up periods range from six months to nine years. No complications developed. **Conclusion:** Thrombocytosis is a common problem in childhood. Reactive and secondary causes are usually identified. Essential thrombocytosis is a diagnosis that should be considered after excluding other causes. Mutation studies should be performed in pediatric patients who meet the WHO 2008 criteria. While Jak-2 (V617F), CALR and MPL (W515L) mutations are seen in 90% of cases in adults, these three mutations are only seen in 25% of the childhood age group. The high number of cases with no mutations indicates that new candidate genes should be sought and studied.

https://doi.org/10.1016/j.htct.2021.10.1073

RED BLOOD CELL DISORDERS

PP 64

GARDNER DIAMOND SYNDROME: A CASE REPORT

Elif Güney [1], Yasemin Ardıçoğlu Akışın [2], Nejat Akar [1]

[1] *TOBB ETU Faculty of Medicine Department of Pediatrics*
[2] *TOBB ETU Faculty of Medicine, Biochemistry*

Case report: Gardner Diamond Syndrome (GDS) is a rare autoimmune disorder also known as autoerythrocyte sensitization syndrome represented with skin lesions. These lesions mostly occur after a triggering factor. The pathophysiology of the disease is not completely understood yet. In this case report, the characteristic features of GDS is presented; furthermore, our aim is to emphasize the effect of emotional stress during the disease.

https://doi.org/10.1016/j.htct.2021.10.1074

IMMUNODEFICIENCIES / NEUTROPHIL DISEASES

PP 65

MYELOPEROXIDASE DEFICIENCY: SINGLE CENTER EXPERIENCE

Özlem ARMAN BİLİR, Namık Yaşar ÖZBEK

Ankara City Hospital

Objective: Myeloperoxidase (MPO) is a hemoprotein expressed in azurophilic granules of neutrophils and lysosomes of monocytes. It is caused by mutations in the MPO gene on chromosome 17 and is estimated to affect 1:2000-4000 people. It is the most common inherited defect of phagocytes. Microbial killing is impaired in patients with MPO deficiency, but most patients are asymptomatic, except for diabetic patients. In this article, we aimed to present our patients diagnosed with primary MPO deficiency. **Methodology:** During the investigation for the etiology of neutropenia in the hematology department of our hospital, patients who were diagnosed with MPO deficiency were examined. In the evaluation of the patients, it was observed that the neutrophil count in the hemogram printout and the counted neutrophil count in the peripheral smear were inconsistent. We performed MPO staining with FCM from the peripheral blood samples of the patients and we found that the neutrophils were MPO negative. **Results:** A 1-day-old male patient has no additional disease (c.608A>C H mutation). C.578G>C mutation was detected in the follow-up due to ANA+ in a 6.5-year-old female patient. A c.2031-2A>C mutation was found in the 18-year-old patient who was being followed up with the diagnosis of Granulamatous Polyangiitis and his sister. A c.493del mutation was detected in an 11-year-old patient who was diagnosed with ITP 5 years ago. The noval mutation was detected in the patient followed up with the diagnosis of

retinoblastoma. **Conclusion:** MPO deficiency may occur primarily as well as secondary. A number of point germ line mutations cause primary MPO deficiency. Most patients asymptomatic without an increase in infection. Severe infectious complications were not observed in any of our patients. We wanted to emphasize that MPO deficiency should also be kept in mind in patients whose neutropenia etiology was investigated.

https://doi.org/10.1016/j.htct.2021.10.1075

LEUKEMIA

PP 66

CHARACTERISTICS AND OUTCOME OF T(8;21)-POSITIVE CHILDHOOD ACUTE MYELOID LEUKEMIA: A SINGLE INSTITUTION'S EXPERIENCE

Volkan KÖSE, Dilek KAÇAR,
Özlem ARMAN BİLİR, Ayça KOCA YOZGAT,
Hüsniye Neşe YARALI

Division of Pediatric Hematology and Oncology, Ankara City Hospital, University of Health Sciences

Objective: Compared with other cytogenetic acute myeloid leukemia (AML) groups, patients with core-binding factor AML (CBF-AML) are considered as a favorable AML risk group based on their high remission rate and survival probabilities. However, up to 30-40% of these patients can still relapse after standard intensive induction and consolidation chemotherapy. **Methodology:** From 2004 to 2020, 147 AML patients reviewed. Ten of 147 patients were followed up with t(8;21) chromosomal anomaly. The t(8;21)(q22;q22) was detected by reverse transcription polymerase chain reaction (RT-PCR) and/or floresan in situ hibridizasyon (FISH). We analyzed patients' demographic data: sex, white blood cell count at diagnosis, central nervous system status, additional cytogenetic anomaly and recurrence rates, stem cell transplant status and survival rates. **Results:** Two of 10 patients were female. The median age was 10 years (3-17 years). Median followup was 36 months (2-114 months). The mean white blood cell count of 10 patients was 21.5 ($\times$ 109/l) at diagnosis. One out of 10 patients had granulocytic sarcoma and 2 had central nervous system involvement. Additional cytogenetic anomalies were detected in 90% of the patients, of which 2 relapsed and 3 died. One patient received hematopoietic stem cell transplantation and died because of HSCT complications. **Conclusion:** Recent studies show that CBF-AML includes different groups with different clinical outcomes. We found that 50% of our patients achieved complete remission and 50% experienced relapsed disease or death. After we were able to monitor the t(8;21) level with RT-PCR, we diagnosed relapsed disease in 1 patient with additional cytogenetic anomaly. RT-PCR is essential for optimal handling of these patients to predict patients' relapse risk and to detect minimal residual disease.

https://doi.org/10.1016/j.htct.2021.10.1076

PP 67

BK-VIRUS ASSOCIATED HAEMORRHAGIC CYSTITIS CONCOMITANT WITH CHEMOTHERAPY IN AN ADOLESCENT GIRL WITH ACUTE LYMPHOBLASTIC LEUKEMIA

Aslı Turgutoğlu Yılmaz, Dilek Kacar,
H. Nese Yarali, Namik Yasar Ozbek

Department of Pediatric Hematology/Oncology, Ankara City Hospital Children's Hospital

Case report: Haemorrhagic cystitis (HC) is characterized by focal or diffuse haemorrhagic and inflammatory changes of the bladder mucosa. Polyoma BK virus (BKV) infection is an important underlying condition that provokes hematopoietic stem cell transplantation (HSCT)-related HC. Although commonly reported in transplant recipients, BKV associated HC, and tubulointerstitial nephritis rarely occurs in paediatric acute lymphoblastic leukemia (ALL) patients receiving chemotherapy. A 15-year-old girl diagnosed with T cell ALL, receiving high-risk chemotherapy protocol, complained about dysuria and lower abdominal pain with macroscopic haematuria. Her complaints started under meropenem, teicoplanin, amikacin, and caspofungin treatment due to neutropenic fever with severe mucositis. There wasn't any bacterial growth in the urine or blood culture. PCR analysis detected 2,2 $\times$ 109 copies/mL of BKV in urine. The antibiotics other than ciprofloxacin were discontinued. Her complaints are alleviated day by day. She did not experience any urinary symptoms or haematuria, and the BKV copy number declined to 3,3 $\times$ 107 copies/mL during follow-up.Contributing factors of BKV associated HC are highly relevant in HSCT recipients. However, patients receiving intensive chemotherapy may have similar conditions. A predisposing and potential manageable factor such as BKV should be searched in paediatric haematology practice.

https://doi.org/10.1016/j.htct.2021.10.1077

PP 68

A CASE OF METHOTREXATE-INDUCED PHOTOSENTIVITY REACTION

Aslı Turgutoğlu Yılmaz, Dilek Gurlek Gokcebay,
Ayca Koca Yozgat, H. Nese Yarali

Department of Pediatric Hematology/Oncology, Ankara City Hospital Children's Hospital

Case report: Methotrexate is an essential drug effectively used in acute lymphoblastic leukemia. Doses above 500 mg/m2 are defined as high-dose methotrexate (HDMTX). Since HDMTX is known to cause serious morbidity, it is given with a standard

rescue therapy to prevent toxicity. Besides myelosuppression and mucositis, other side effects of methotrexate are hepatotoxicity, erythema, desquamation, allergic reactions and neurotoxicity. Methotrexate is also associated with radiation recall and false photosensitivity. A 10-year-old girl with pre-B ALL underwent hematopoietic stem cell transplantation two times due to marrow and central nervous system (CNS) relapse. On the follow-up, 3 months later she had a bone marrow relapse. After remission obtained with high dose chemotherapy, maintenance treatment was given due to relapse/refractory disease. One year later she had isolated CNS relapse again and treated with intrathecal methotrexate, Ara-C and dexamethasone. The patient was started on relapse/refractory maintenance therapy, and 1 g/m2 methotrexate was given every 4 weeks. Immediately after intravenous methotrexate was given to the patient in the 13th week of her treatment, she complained of burning, pain and redness in the areas that had previously been desquamated due to sunburn. No additional treatment was given, except alkaline hydration and calcium folinate, when the findings were observed. The patient was started on antihistamine therapy. Methotrexate drug level reached 0.02 umol/L at the 54th hour, the i.v. hydration was stopped. The patient's red and itchy lesions healed within 2 days by benefiting from the antihistamine. She is being followed-up at our outpatient clinic weekly chemotherapy without any sign of relapse. This sunburn-like erythema after methotrexate administration might be associated with impaired mononuclear cell response in sun-exposed tissues. Our case stated that he went to the sea two weeks ago and that the bullae secondary to the sunburn that developed afterwards peeled off after they burst. In conclusion, patients with a history of recent generalized sunburn should have their methotrexate delayed to avoid this complication.

https://doi.org/10.1016/j.htct.2021.10.1078

PP 69

UNUSUAL METABOLIC COMPLICATIONS OF CHILDHOOD ACUTE LYMPHOBLASTIC LEUKEMIA: HYPERCALCEMIA, HYPERAMONEMIA, LACTIC ASIDOSIS

Dilek Kaçar, Aslı Turgutoğlu Yılmaz,
Ayça Koca Yozgat, Neşe Yaralı

Ankara City Hospital, Department of Pediatric Hematology and Oncology

Case report: We present three children with precursor B acute lymphoblastic leukemia (ALL). The first one had malignancy associated hypercalcemia at diagnosis. The second one experienced hyperamonemia during induction. Both of them had been treated successfully. The last one had refractory leukemia and died because of lactic acidosis due to extensive infiltration of the liver by tumor cells. The rare but potential fatal metabolic complications of ALL needs high clinical suspicion and prompt treatment.

https://doi.org/10.1016/j.htct.2021.10.1079

PP 70

CENTRAL HYPOTHROIDISM DUE TO ACUTE LYMPHOBLASTIC LEUKEMIA WITH CENTRAL NERVOUS SYSTEM INFILTRATION

Dilek Kaçar, Seda Şahin,
Fatma Burçin Kurtipek, Neşe Yaralı

Ankara City Hospital, Department of Pediatric Hematology and Oncology

Case report: We describe a five-year-old girl with high risk B precursor acute lymphoblastic leukemia with central nervous system involvement. Laboratory tests suggested the presence of central hypothyroidism (thyroid-stimulating hormone [TSH]: 0.30 mU/ml, normal range 0.64–6.27 mU/ml; serum free thyroxine [FT4]: 0.70 ng/dl, normal range 0.86–1.4 ng/dl). Magnetic resonance imaging detected heterogeneous contrast enhancement of pituitary gland in addition to cerebral and cerebellar atrophy.

https://doi.org/10.1016/j.htct.2021.10.1080

PP 71

BONE AS A SITE OF EXTRAMEDULLARY DISEASE IN ACUTE LYMPHOBLASTIC LEUKEMIA

Fatma Tuba YILDIRIM, Dilek KAÇAR,
Aslı TURGUTOĞLU YILMAZ, Burçin KURTİPEK,
Ayça KOCA YOZGAT,
Dilek GÜRLEK GÖKÇEBAY, Neşe YARALI

Ankara City Hospital

Case report: We describe 3 children with pre B acute lymphoblastic leukemia (ALL).The first two were evaluated in orthopedic clinics because of limping due to ischium involvement and bone fracture suspicion due to involvement of upper limb bones.As a result of normal hemograms in both cases, leukemia diagnosis delayed.The third patient experienced bone marrow and vertebral column relapse of ALL presenting with nuchal rigidity mimicking meningitis.Bone should be considered as a site of extramedullary disease.

https://doi.org/10.1016/j.htct.2021.10.1081

PP 72

A CHALLENGE IN PEDIATRIC ACUTE LEUKEMIA TREATMENT: UNEXPECTED, PROLONGED CYTOPENIA. IS IT BE CALLLED 'INCOMPLETE HLH'?

Ersin Toret, Sumeyye Emel Eren,
Zeynep Canan Özdemir, Özcan Bör

Eskisehir Osmangazi university

Objective: The diagnostic criteria set for HLH may look like symptoms of cancer or a severe bacterial infection common occurring when patients are immunosuppressed due to

ongoing chemotherapy. Features similar to immune dysregulation in HLH also occur during pediatric acute leukemias. This immune dysregulation results unexpected cytopenias, fever, and splenomegaly in children with acute leukemia. We aim to analysis the pediatric acute leukemia pateints who had unexpected, prolonged cytopenias, and did not full-fill the HLH-2004 criteria set and received pulse methylprednisolone therapy up to three days **Methodology:** Data was analyzed retrospectively. The diagnosis of HLH was defined according to the HLH-2004 criteria set but two criterias (NK cell activity and sCD25 level) of HLH diagnosis were not studied due to lack of necessary equipment. Treatment response was defined as increasing neutrophil count above 500/mm3 in patients within the first seven days. **Results:** 12 patients received steroid for unexpected, prolonged cytopenias. Five or six of six criteria was not found. Four criteria in four, three criteria in five and two criteria in three patients was determined. All patients had cytopenia at least two of three lineages in peripheral blood, one of which was neutropenia. Hemophagocytosis in bone marrow sample was detected in eight patients. Ten patients (87%) recovered within the first seven days. Seven of nine thrombocytopenic patients recovered. **Conclusion:** In this report, the efficiency of short-term steroid treatment was demostrated in patients with unusual cytopenias who did not full-fill HLH criteria.

https://doi.org/10.1016/j.htct.2021.10.1082

PP 73

EVALUATION OF VACCINATION RESPONSE IN CHILDREN AFTER TREATMENT FOR ACUTE LEUKEMIA

Elif KILIC KONTE [1], Ayca KOCA YOZGAT [2], Aysun KARA UZUN [2], Bahar CUHACI CAKIR [3], Hüsniye Nese YARALI [2]

[1] *Muş State Hospital*
[2] *Ankara City Hospital*
[3] *Gazi University Medical Faculty Hospital*

Objective: Our study aims to evaluate the patients' immunity regarding childhood vaccination after leukemia treatment and determine the vaccines that require additional doses. **Methodology:** Sixty-six patients who were followed up with the diagnosis of ALL and AML between 2013 and 2016 were included in our study. The patient's gender, age at diagnosis, leukemia type, leukemia risk groups, vaccination status before chemotherapy (CT) and serologies of hepatitis A, hepatitis B, varicella, measles, rubella, mumps at the end of CT were recorded. **Results:** At the end of the treatment, loss of protective antibody response against hepatitis A (47.4%), hepatitis B (68.2%), varicella (64.2%), measles (45.5%), rubella (43.9%), and mumps (50%) vaccines were shown. Loss of protective antibodies against hepatitis A (66.7%), hepatitis B (100%), varicella (100%), measles (100%), rubella (91.7%), and mumps (91.7%) in high-risk ALL patients was higher than patients in standard-intermediate risk ALL. **Conclusion:** Loss of humoral immunity against hepatitis A, hepatitis B, varicella, MMR was shown in patients with leukemia at the end of the treatment. Due to the significant decrease in hepatitis B and MMR protective antibodies in the high-risk group, we recommend patients with leukemia who have completed chemotherapy to be vaccinated with hepatitis B vaccine three months and MMR vaccine six months after the treatment.

https://doi.org/10.1016/j.htct.2021.10.1083

PP 74

PONATINIB EXPERIENCE IN A PEDIATRIC CHRONIC MYELOID LEUKEMIA PATIENT

Serap Karaman [1], Mustafa Bilici [1], Ayşegül Ünüvar [1], Deniz Tuğcu [1], Gülşah Tanyıldız [1], Rumeysa Tuna Deveci [1], Gülçin Yegen [2], Şifa Şahin [1], Zeynep Karakaş [1]

[1] *Department of Pediatric Hematology and Oncology, Istanbul University, Istanbul, Turkey*
[2] *Department of Pathology, Istanbul University, Istanbul, Turkey*

Objective: Chronic myeloid leukemia (CML) is rarely seen in children. The development of myelofibrosis in CML is not uncommon and is associated with a poor prognosis. In cases unresponsive to treatment, tyrosine kinase mutation should be checked for drug resistance, second generation tyrosine kinase inhibitor (TKI) drugs (dasatinib/nilotinib) should be switched to and a suitable donor for bone marrow transplantation should be sought. Third-choice TKI can be used in children who are unresponsive to treatment and do not have a suitable donor. **Materials and Methods:** Experience of third-choice TKI(ponatinib) in a child with CML diagnosis due to unresponsiveness to treatment. **Results:** A 5.5-year-old female patient with no known disease was referred to us because of hepatosplenomegaly (liver 5 cm, spleen 10 cm). There was no laboratory disorder except for anemia (hgb 8.9 g/dL) and high LDH (1104 U/L). WBC was $11.1 \times 10^3/\mu$L neu $6.92 \times 103/\ \mu$L plt 304000/μL. Peripheral smear showed leukoerythroblastosis. Bone marrow biopsy result was evaluated as compatible with myelofibrosis and an increase in blast rate from 8% to 18% in the bone marrow. The patient was diagnosed CML accelerated phase with cytogenetic (46,XX,t(9:22)(q34;q11))and translocation (t(9:22)- p210,BCR/ABL positive) results and. Imatinib treatment was started at 400 mg/m^2.The copy number of BCR-ABL p210 checked before treatment was 72% IS. However, the patient developed febrile neutropenia, and imatinib dose reduction (< 200 mg/m^2) and interruption were required in the follow-up.Under imatinib treatment, BCR-ABL copy number was 16%IS at 1 month, 11%IS at 3 months, and 95%IS at 5 months. Due to the increase in the BCR-ABL copy number, nilotinib was switched to as a second-choice TKI(230 g/m2/dose, in 2 doses).No mutation could be detected in the c-ABL gene, which was examined for tyrosine kinase resistance. HLA groups were sent from the family and compatible donors were not found. Due to severe neutropenia in the follow-up, nilotinib could be continued at 50% dose. Under nilotinib treatment, the BCR-ABL copy number was 13% IS at 1 month, 10% IS at 2 months, and 31% IS at 3 months. The patient was started on ponatinib (18 mg/m^2/day)

as a third choice TKI. However, due to the deep neutropenia of the patient, it was possible to continue with a dose of 10 mg/m^2 from the 2nd week. With this dose, the neutrophil is around 0.8-1 $\times$ 10^3/μL. Under ponatinib treatment, BCR-ABL copy number was 6.6% IS at 1 month, 0.8% IS at 3 months, 0.09% at 5 months, and 0.05% at 6 months. No significant side effects were observed except neutropenia. **Conclusion:** There is no approved treatment in pediatric CML cases where the second choice TKI fails and there is no donor for transplantation. FDA approval for ponatinib in adult patients was obtained in December 2020. Ponatinib is a natural or mutant pan-BCR-ABL mutation inhibitor. It also inhibits VEGFR, FGFR,PDGFR,EPH and SRC kinases as well as KIT,RET,TIE2 and FLT3. The use of ponatinib should be evaluated by monitoring side effects/tolerance in pediatric cases where there is no other treatment option, and there is a need for studies on this subject.

https://doi.org/10.1016/j.htct.2021.10.1084

INHERITED BONE MARROW FAILURE DISEASES

PP 75

INVESTIGATION OF SALIVARY miR-9, miR-34a ve miR-196a LEVELS IN FANCONI ANEMIA AND ORAL SQUAMOUS CELL CARCINOMA PATIENTS

Zisan Asal Kilic [1], Nevin Yalman [2], Tulin Tiraje Celkan [3], Bora Basaran [4], Haydar Murat Yener [5], Selcuk Dasdemir [1], Mehmet Güven Günver [6]

[1] *Department of Medical Biology, Istanbul University Istanbul Faculty of Medicine*
[2] *Department of Pediatric Hematology Oncology, Yeditepe University Hospital*
[3] *Department of Pediatric Hematology Oncology, Istinye University Faculty of Medicine, İstanbul, Turkey*
[4] *Department of Otorhinolaryngology, Istanbul University Istanbul Faculty of Medicine*
[5] *Department of Otorhinolaryngology, Istanbul Cerrahpasa University Faculty of Medicine*
[6] *Department of Bioistatistics, Istanbul University Istanbul Faculty of Medicine, İstanbul, Turkey*

Objective: Fanconi anemia (FA) is a rare bone marrow deficiency syndrome due to the DNA repair gene mutations, and Oral Squamous Cell Carcinoma (OSCC) is seen more frequently in FA patients than in the general population. The dysregulation of PI3K and Wnt signaling has been implicated in OSCC pathogenesis and abnormal expressions of miRNAs (a class of non-coding small regulatory RNAs) associated with these signaling pathways has been reported in OSHK patients. Salivary miRNAs are valuable biomarker candidates for OSCC development and prognosis. In this study, salivary levels of miR-9, miR-34a and miR-196a miRNAs related to PI3K and Wnt signaling pathways were examined in OSCC and FA patients and compared with the healthy control group. **Methodology:** Saliva samples were collected from 89 subjects including 25 OSCC patients, 24 FA patients and 40 healthy controls. Total RNA was isolated using Quick-RNA Miniprep Kit (Zymo Research) due to the kit instructions. cDNA was generated with miRCURY LNA miRNA PCR Assay (Qiagen, Hilden, Germany) and Quantitative real-time PCR was performed with miRCURY LNA SYBR Green PCR Kit (Qiagen, Hilden, Germany) according to the manufacturer's protocol. For the normalization of the expression levels of each miRNA, the mean expression U6 SnRNA was used as reference. The ΔΔCt value and the normalized miR-9, miR-34a and miR-196a salivary levels were calculated with Livak Method. **Results:** Our results showed that miR-9 and miR-34a levels in OSCC patients were significantly lower compared to healthy control groups (p= 0,01 and p= 0,012), and there was no significant difference in miR-196a levels (p> 0,05). In FA patients, miR-9 and miR-34 levels were lower than in control groups, likewise the OSCC patients (p =0,017 and p =0,014). There was no significant difference between miR-9, miR-34a, and miR-196a levels of FA patients and OSCC patients (p >0.05). **Conclusion:** According to our results, low levels of miR-9 and miR-34a in saliva are biomarker candidates that may be important for OSCC development. In FA patients, close follow-up of the levels of miR-9 and miR-34 would be appropriate considering OSCC development. Further studies are needed to confirm the potential of miR-9 and miR-34a as biomarkers for OSCC.

https://doi.org/10.1016/j.htct.2021.10.1085

PP 76

A NOVEL MISSENSE MUTATION OUTSIDE DNAJ DOMAIN OF DNAJC21 IS ASSOCIATED WITH SHWACHMAN-DIAMOND SYNDROME

Mohammad Bilal Alsavaf [1], Jeffrey M. Verboon [2], Muhammed E. Dogan [3], Zehra Busra Azizoglu [4], Fatma Zehra Okus [1], Alper Ozcan [1], Munis Dundar [3], Ahmet Eken [4], Hamiyet Donmez Altıuntas [5], Vijay G. Sankaran [2], Ekrem Unal [1]

[1] *Division of Pediatric Hematology, Oncology & HSCT Center, Department of Pediatrics, Erciyes University, Faculty of Medicine*
[2] *Division of Hematology/Oncology, Boston Children's Hospital, Harvard Medical School, Boston*
[3] *Department of Medical Genetic, Erciyes University*
[4] *Gevher Nesibe Genom and Stem Cell Institution, Betul Ziya Eren Genome and Stem Cell Center (GENKOK), Erciyes University, Faculty of Medicine*
[5] *Department of Medical Biology, Erciyes University, Faculty of Medicine*

Shwachman-Diamond Syndrome (SDS) and related bone marrow failure disorders are characterized by early onset pancytopenia with a hypocellular bone marrow, short stature, and pancreatic insufficiency, along with an increased risk for myeloid malignancies. Recently, several cases with an SDS-like syndrome have been reported to harbor mutations in the DNAJ domain of *DNAJC21*. Here, we report an intriguing case

of a 13.5 years-old female born to Turkish consanguineous parents with a novel missense mutation occurring outside the DNAJ domain of the *DNAJC21* gene. Whole-exome and Sanger sequencing confirmation revealed a homozygous missense mutation in *DNAJC21* gene c.463T>C, p.W155R which was considered as pathogenic in in silico analyses. Initially, this patient's vague and atypical symptoms led to uncertainty of the underlying diagnosis. Upon confirmation of the genetic mutation, a number of functional studies such as diepoxibutane test, proliferation test from peripheral blood mononuclear cells, and cytokinesis-block micronucleus cytome assay performed with the patient cells confirmed the likely diagnosis of an SDS-like syndrome attributable to DNAJC21 dysfunction. Through the analysis of this rare case, we illuminate the pleiotropic features of this unique bone marrow failure syndrome and emphasize the paramount role of genomic testing to discriminate a range of closely related bone marrow failure disorders.

https://doi.org/10.1016/j.htct.2021.10.1086

STEM CELL TRANSPLANTATION

PP 77

THE ROLE OF THERAPEUTIC DRUG MONITORING OF INTRAVENOUS BUSULFAN FOR PREVENTION OF SINUSOIDAL OBSTRUCTION SYNDROME IN CHILDREN

Dilek GURLEK GOKCEBAY [1], ÖzlemArman Bilir [1], Seda Şahin [2], İkbal Ok Bozkaya [1], Namık Yaşar Özbek [1]

[1] *University of Health Sciences Ankara City Hospital Department of Pediatric Hematology Oncology*
[2] *University of Health Sciences Ankara City Hospital Department of Pediatrics*

Objective: Busulfan is a widely used alkylating drug for conditioning of hematopoietic stem cell transplantation (HSCT). Higher exposure of Bu is associated with toxicity and (sinusoidal obstruction syndrome) SOS, whereas lower exposure is associated with graft failure or relapse risk. Therapeutic drug monitoring (TDM) has been recommended to overcome these issues. We aimed in this study to compare HSCT outcomes in children with and without TDM of Bu. **Methodology:** This retrospective study conducted at our Transplantation Unit between 2012 and 2021. Patients aged 0-18 y underwent HSCT who received Bu-based conditioning and completed post-transplant +100 days included in the study. Data were collected including demographic information, primary diagnoses, conditioning regimen, graft-related data, dose of Bu, time to neutrophil and platelet engraftment, presence of SOS, acute or chronic GvHD, and clinical outcomes. SPSS 18.0 was used for statistical analysis. **Results:** 172 patients (59 girls, 113 boys) with a median age of 4.70 years (IQR 2.41-10.01) were enrolled in the study. TDM of Bu was performed in 126 patients. 32 patients (19%) developed moderate or severe SOS. Incidence of SOS was significantly higher in the group without TDM. A multivariable analysis showed that presence of acute GVHD and 2 or more alkylating agents in conditioning regimen were associated were SOS. HSCT related outcomes, relapse, OS and EFS did not different between two groups. **Conclusion:** To improve treatment outcomes of Bu, TDM and dose adjustment, following the first dose, has highly recommended regardless of the dosing guideline was used. We also demonstrated the incidence of SOS decreased in patients with TDM, but other HSCT related outcomes were not influenced. Optimal cumulative Bu exposure canbalance between efficacy and toxicity of HSCT in children.

https://doi.org/10.1016/j.htct.2021.10.1088

PP 78

A CASE OF POLYCYTHEMIA DIAGNOSED AS HEMOGLOBIN ANDREW-MINNEAPOLIS

Mustafa Bilici [1], Serap Karaman [1], Aysegul Unuvar [1], Deniz Tugcu [1], Gülsah Tanyildiz [1], Ayca Dilruba Aslanger [2], Oya Uyguner [2], Rumeysa Tuna Deveci [1], Sifa Sahin [1], Zeynep Karakas [1]

[1] *Department of Pediatric Hematology and Oncology, Istanbul University, Istanbul, Turkey*
[2] *Department of Genetics, Istanbul University, Istanbul, Turkey*

Objective: Polycythemia is a rare condition in which an increase in erythrocyte mass is observed. It can be primary or secondary. Primary polycythemia occurs as a result of congenital or acquired mutations that regulate erythroid development. Although secondary polycythemia is mostly seen secondary to hypoxia due to cardiac/pulmonary reasons, it also develops as a result of congenital mutations. Globin gene mutations that increase the affinity of hemoglobin for oxygen are one of these rare causes. **Materials and Methods:** We present a male case who was referred to us for polycythemia. **Results:** A 15-year-old male patient with no known disease was referred to us after his school screening revealed high hemoglobin (18 g/dL). In complete blood count, other series were normal (wbc $5.8 \times 10^3/\mu L$ neu $3.3 \times 10^3/\mu L$ plt $174 \times 10^3/\mu L$), bilirubins and liver functions were within normal limits. On physical examination, conjunctiva and hands were pletoric, there was no hepatosplenomegaly, intermittent headaches were present, and neurological examination was normal. The patient was examined for the etiology of polycythemia. Hyperchromic erythrocytes were found in peripheral smear, no signs of hemolysis were observed. EPO level (8 mIU/ml) was in the normal range and JAK2 (V617F) mutation was negative. The patient's cardiac and pulmonary functions were within normal limits. Hemoglobin electrophoresis was sent from the patient. HbA was determined as 59.2, HbA2 2.8, Variant Hb 38. c.435G>T mutation was detected in the HBB genetic analysis, and this was considered to be compatible with Hemoglobin Andrew-Minneapolis. It was learned that the patient's mother and her cousins had similar findings, and some of them had undergone phlebotomy. Phlebotomy was planned in the presence of the patient's hemoglobin value > 18 g/dL and clinical findings. Phlebotomy was performed 3 times, aspirin was not started because there was no

history of thromboembolism. In our 1-year follow-up, the hemoglobin value was 17-17.5 g/dL. **Conclusion:** More than a hundred globin gene mutations associated with erythrocytosis have been described. Hemoglobin Andrew-Minneapolis mutation is one of them. Hemoglobin's affinity for oxygen has increased and EPO level is normal/increased. Due to the low number of cases, treatment recommendations were prepared based on polycythemia vera guidelines. Patients should be closely monitored in terms of hyperviscosity and thromboembolism, aspirin prophylaxis and phlebotomy are recommended according to symptoms. While investigating the etiology of polycythemia, hemoglobin electrophoresis is necessary, although it is very rare.

https://doi.org/10.1016/j.htct.2021.10.1089

LYMPHOMAS

PP 79

THE SMALLEST PRIMARY BONE LYMPHOMA

Fatma Burçin KURTİPEK, Volkan KÖSE,
Seda ŞAHİN, Derya ÖZYÖRÜK, Neriman SARI,
Sonay İNCESOY ÖZDEMİR, Arzu YAZAL ERDEM,
Meriç KAYMAK, İnci İLHAN ERGÜRHAN

Ankara City Hospital

Case report: Primary lymphoma of bone (PLB) is a rare malignant condition with lymphocytic infiltration of the bone; it accounts for 2–3% of all primary bone tumours in adults and children .Here we report a little girl with isolated PLB of B cell lineage focussing on diagnosis, evaluation and treatment strategy. Our case can help to get acquaintance with PBL,it should be taken into consideration as a different diagnosis for osteolytic lesions of bone.

https://doi.org/10.1016/j.htct.2021.10.1090

PP 80

MRI FINDINGS OF BONE MARROW AT THE BEGINNING OF LEUKEMIA

Seda Şahin, Neriman Sarı, Tülin Demirkan

Ankara Şehir Hastanesi

Case report: Pediatric ALL/lymphoma (LBL) is a clonal hematopoietic stem cell disorder which's highly aggressive. There is an overlap between ALL and LBL which shouldn't cause delay in the diagnosis of each other.We'll describe a patient who presented with leukemia symptoms such as fever,bone pain, who didn't have obvious atypical cells in his peripheral smear,BM aspirationand involvement in scintigraphy but had diffuse bone marrow(BM)involvement in the lower extremities in his MRI. BM biopsy showed ALL/LBL.

https://doi.org/10.1016/j.htct.2021.10.1091

PP 81

THREE CASES WITH BURKITT LYMPHOMA PRESENTING WITH CHOLESTASIS

Fatma Tuba YILDIRIM, Derya ÖZYÖRÜK,
Arzu YAZAL ERDEM, Selma ÇAKMAKCI,
Neriman SARI, Sonay İNCESOY, İnci İLHAN

Ankara City Hospital

Case report: Cholestasis secondary to neoplasm is rare in children. It is also rare in Burkitt lymphoma and may be cause to treatment delay. We report 3 cases diagnosed with Burkitt lymphoma with cholestasis. All patients had jaundice and high direct biluribin levels. They were given LMB chemotherapy protocol. After COP chemotherapy, cholestasis disappeared rapidly in all patients. In conclusion, cholestasis at initial resolves rapidly with chemotherapy despite high liver function tests in Burkitt lymphoma.

https://doi.org/10.1016/j.htct.2021.10.1092

BRAIN TUMOURS

PP 82

HIGH GRADE GLIOMA OF CENTRAL NERVOUS SYSTEM: SINGLE CENTER TREATMENT EXPERIENCE

Nur OLGUN [1], Deniz KIZMAZOGLU [1],
Batuhan OZDOGAR [2], Dilek INCE [1],
Emre CECEN [1], Ceren KIZMAZOGLU [3],
Şefika AKYOL [1], Ayşe DEMIRAL [4],
Rıza CETINGOZ [4], Ayşe ONAL [1], Handan UCAR [5],
Erdener OZER [6]

[1] *Dokuz Eylul University Institute of Oncology, Pediatric Oncology*
[2] *Dokuz Eylül University Faculty of Medicine, Department of Pediatrics*
[3] *Dokuz Eylül University Faculty of Medicine, Department of Neurosurgery*
[4] *Dokuz Eylül University Faculty of Medicine, Department of Radiation Oncology*
[5] *Dokuz Eylul University Faculty of Medicine, Department of Radiodiagnostic*
[6] *Dokuz Eylül University Faculty of Medicine, Department of Pathology*

Objective: To evaluate characteristics and treatment responses of patients with high grade gliomas (HGG) in our center. Medical files of patients with malignant CNS tumors between 1987-2020 were analyzed retrospectively. There were 44 patients with HGG. **Case report:** Diagnosis of patients as follows: 21 pons glioma, 2 anaplastic astrocytoma, 11 anaplastic ependimoma, 7 glioblastoma multiforme, 1 glioblastoma, 2 gliomatosis cerebri. The median age at diagnosis was 6,5 yrs (7 – 17 yrs), M/F:25/19. Age distribution: <5 yrs 12 patients, 5-10 yrs 18 patients, 10-18 yrs 14 patients. The most frequent complaints for pons gliomas: cranial nerve paralysis

(52%), visual impairment (48%), headache (38%), power loss (43%) and speech disorder (30%). **Methodology:** Surgery was performed to extrinsic component of mass in 3 patients of pons gliomas.For other HGG: 7 subtotal resection and 16 gross total resection had performed.7 patients died before RT. And other 37 patients received radiotherapy. RT total doses varied between 50-60 Gy.7 patients were not received chemotherapy, 3 of them died before chemo, and others received only RT. For other HGGs, platin based regimens used for the first line treatment. Temozolamide, bevacizumab, irinotecan as the other options. **Results:** Median progression free survival time was 6 mos (2weeks-25 mos) for pons gliomas, for other gliomas median progression free survival time was 14 mos(0 -74 mos). For pons gliomas: Event free survival rate for 6 mos was 75%, for one year 17%;one year, 18 mos, and two years overall survival rates were 84%,52% and 10%respectively.For other HGGs: Event free survival rate for one year and two years were 57% and 17% respectively. One year and two years overall survival rates were 73% and 36% respectively. **Conclusion:** High grade glioma is a group of tumors in which still the helplessness experienced in treatment. Despite radiotherapy and chemotherapy, prognosis is very poor. The progression free and overall survival rates of patients were similar to literature. With new developments in molecular pathology, as the use of molecular target therapies, the progression free survival rates newly will improve.

https://doi.org/10.1016/j.htct.2021.10.1093

PP 83

ONC 201 PRACTICE IN DIFFUSE MIDLINE GLIOMA (H3K27M MUTANT)

Bahattin Tanrıkulu [1], Cengiz Canpolat [2], Ahmet Harun Yaşar [3], Ayça Erşen Danyeli [4], M. Memet Özek [1]

[1] *AcibademUniversity School of Medicine Department of Neurosurgery Division of Pediatric Neurosurgery*
[2] *AcibademUniversity School of Medicine Department of Pediatrics Division of Pediatric Hematology and Oncology*
[3] *AcibademUniversity School of Medicine Department of Neurosurgery*
[4] *AcibademUniversity School of Medicine Department of Pathology*

Objective: Diffuse Midline Gliomas (DMG), H3 K27M-mutant have the poorest prognosis among all pediatric high-grade gliomas, with a median survival of 9-11 months. Although radiotherapy (RT) is standard treatment for these tumors, unfortunately there has been no approved and effective treatment which completely diminishes the tumor yet. In our clinic, we started an up-to-date approach to manage DMG, which is adjuvant fractionated external beam radiotherapy along with ONC 201 after tissue diagnosis. **Methodology:** Between January 2016 and June 2021, a total of 11 patients with H3 K27M-mutant diffuse midline glioma, diagnosis confirmed by Next-Generation Sequencing (NGS) were enrolled in study. All patients received ONC201 orally once a week following radiotherapy. Safety, and radiological evaluations were regularly assessed every 12 weeks. **Results:** Among the 11 patients, the median age of diagnosis was 5. Seven (63.6%) patients were male and 4 (36.4%) were female. Primary lesions were localized in the pons in 5 (45.5%) patients, unilateral thalamus (2 on the left, 1 on the right) in 3 (27.3%) patients, bilateral thalamus in 2 (18.2%) patients, and temporo-insular in 1 patient (9.1%). Median progression-free interval was 10 months and median overall survival was 16 months. **Conclusion:** Diffuse midline glioma has dismal prognosis. None of the treatment options made any dramatic changes in disease course during last 30 years. In our series, diffuse midline glioma patients who had ONC201 tend to have few months more progression free and overall survival (16 vs 11 months) in comparison to patients who had classical treatment in literature. As a neurooncology team, we strongly advocate to obtain tissue samples from diffuse tumors, to establish definite diagnosis and to perform NGS

https://doi.org/10.1016/j.htct.2021.10.1094

NEUROBLASTOMA

PP 84

PARATESTICULAR INVOLVEMENT IN NEUROBLASTOMA

Volkan KÖSE [1], Sabri DEMİR [2], Neriman SARI [1]

[1] *Division of Pediatric Hematology and Oncology, Ankara City Hospital, University of Health Sciences*
[2] *Division of Pediatric Surgery, Ankara City Hospital, University of Health Sciences*

Case report: Neuroblastoma is 7-10% of all pediatric cancer cases. Primary testicular and paratesticular neuroblastoma is very rare in the literature. We aimed to present our experience with a 4-year-old patient with an abdominal and right paratesticular mass. The patient's imaging revealed extensive lung and bone metastases. In the diagnostic biopsy, the primary tumour consistent with poorly differentiated neuroblastoma and the right paratesticular mass biopsy revealed neuroblastoma metastasis.

https://doi.org/10.1016/j.htct.2021.10.1095

BONE TUMOURS

PP 85

PRIMARY EWING'S SARCOMA OF SPHENOID BONE EXTENDING TO BRAINSTEM; AN ORDINARY TUMOUR AT AN EXTRAORDINARY LOCATION AND INVOLVEMENT

Seda ŞAHİN, İnci İlhan

Ankara Şehir Hastanesi

Case report: Ewing's sarcoma (ES) is the 2nd primary bone tumor of childhood, mostly located in the lower extremities. The incidence of primary cranial ES is <1%. Our patient is 9 years old female who has intracranial primary ES extending from the sphenoid bone corpus to the clivus border. This is a rare case of childhood that's originating from the sphenoid bone and spreading to such a very large intracranial area. Our aim is to provide data on the clinical and therapeutic course of a rare case.

https://doi.org/10.1016/j.htct.2021.10.1096

SUPPORTIVE CARE AND PALLIATIVE CARE

PP 86

ACTINOMYCES ODONTOLYTICUS: A RARE CAUSE OF PEDIATRIC FEBRILE NEUTROPENIA

Meric KAYMAK CIHAN [1], Sonay INCESOY OZDEMIR [1], Belgin GULHAN [2], Arzu YAZAL ERDEM [1], Derya OZYORUK [1], Neriman SARI [1], Inci ERGURHAN ILHAN [1]

[1] *ANKARA CITY HOSPITAL, DEPARTMENT OF PEDIATRIC ONCOLOGY*
[2] *ANKARA CITY HOSPITAL, DEPARTMENT OF PEDIATRIC INFECTION DISEASES*

Case report: Actinomyces spp. are gram-positive bacilli found in humans as a common flora of the oropharynx, gastrointestinal tract, and urogenital tract. We describe a case of Actinomyces odontolyticus bacteremia in an Ewing sarcoma and febrile neutropenic girl. This is the first time that bacteremia due to A. odontolyticus has been reported in a pediatric cancer patient. . This case suggests that A. odontolyticus should be regarded as a possible cause of bacteremia in neutropenic pediatric cancer patients.

https://doi.org/10.1016/j.htct.2021.10.1097

TUMOR BIOLOGY, IMMUNOLOGY AND IMMUNOTHERAPY

PP 87

WITHDRAWN: THE EFFECT OF NIVOLUMAB IN PEDIATRIC MALIGNANT TUMORS: A SINGLE CENTER EXPERIENCE WITH EIGHT PATIENTS

Veysel GOK [1], Firdevs AYDIN [1], Alper OZCAN [1], Ebru YILMAZ [1], Ekrem UNAL [1], Musa KARAKUKCU [1], Türkan PATIROGLU [1], Mehmet Akif OZDEMIR [1], Filiz KARAMAN [2], Orhan GORUKMEZ [3], Ozlem GORUKMEZ [3], Atil BISGIN [4]

[1] *Division of Pediatric Hematology and Oncology, Department of Pediatrics, Erciyes University*
[2] *Division of Pediatric Radiology, Department of Radiology, Erciyes University*
[3] *Department of Medical Genetics, Bursa Yüksek Ihtisas Training and Research Hospital*
[4] *Department of Medical Genetics, Medical Faculty, Çukurova University*

This abstract has been withdrawn: please see Elsevier Policy on Article Withdrawal (http://www.elsevier.com/locate/withdrawalpolicy).

This abstract has been withdrawn at the request of the author.

The publisher regrets that an error occurred which led to the premature publication of this paper. This error bears no reflection on the article or its authors.

https://doi.org/10.1016/j.htct.2021.10.1098

PP 88

A CASE DIAGNOSED WITH FOUR DIFFERENT TUMORS

Fatma Tuba YILDIRIM, Derya ÖZYÖRÜK, Arzu YAZAL ERDEM, Selma ÇAKMAKCI, Neriman SARI, Sonay İNCESOY, İnci İLHAN

Ankara City Hospital

Case report: Chromosomal breakage syndromes are characterized by cancer predisposition. Here we present a 27-month-old female with Fanconi Aplastic Anemia diagnosed with 4 tumors. Imaging showed brain mass causing the shift, liver mass and left kidney mass. She had diagnosed with high grade intracranial tm, wilms tm and hepatocellular ca. Because of refractory pancitopeni, she underwent HSCT. After 2months she developed intracranial embryonal tumor. The patient died with progression. Genetic tests revealed no mutation.

https://doi.org/10.1016/j.htct.2021.10.1099