

# HEMATOLOGY, TRANSFUSION AND CELL THERAPY



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## **POSTER PRESENTATIONS**

Adult Hematology Abstract Categories

Acute Leukemias PP 01

CYTARABINE-INDUCED NEUROTOXICITY WELL-RESPONDING TO METHYL PREDNISOLONE

Berrin Balik Aydin<sup>1</sup>

<sup>1</sup> Gazi Yasargil Education And Training Hospital

Objective: Neurotoxicity is a well-recognized complication of high-dose cytosine arabinoside (HIDAC). We describe a patient with AML who suffered cerebellar toxicity following high-dose cytarabine and showed excellent response to methylprednisolone. Methodology: A 34-year-old male with acute myeloid leukemia (AML) M1 presented with dysarthria in the inpatient clinic. He had been previously diagnosed with myeloblastic leukemia with maturation type AML, negative for t(8:21), t(9:22), CEBPA and FLT3-TKD mutations by PCR. Cytogenetics were 46, XY. Prior to her presentation, he was treated with induction chemotherapy, which consisted of cytarabine 200 mg/m<sup>2</sup> on days 1–7, idarubicin 12 mg/m<sup>2</sup> on days 1-3. After induction chemotherapy, he had complete morphologic and immunophenotypic remission of her leukemia on bone marrow biopsy, which was followed by one consolidation cycle of high-dose cytarabine (3 gm/m<sup>2</sup> on days 1, 3 and 5). After the first cycle of consolidation therapy, on day six he began to complain of dysarthria, dizziness, gait disorder and balance loss. His cumulative dose of cytarabine at that time was 37.400 g. On physical exam, he had not be able to walk in a straight line, he had dysarthria but he had not dysmetria and dysdiadochokinesia. Gait was ataxic and the rest of neurologic examination was generally normal. MRI and CT of the brain showed no acute pathologic findings. His neurologic symptoms were presumed to be secondary to cytarabine neurotoxicity. He was started on prednisone 80 mg daily over 7 days with rapid resolution of his symptoms within a few days of starting corticosteroids. The steroids were tapered by halving the dose each 3 days over the

following 2 weeks. The patient did not receive any additional consolidation treatments with cytarabine, though he remained on maintenance allogeneic hematopoietic cell transplantation (HCT), donor was his brother. He is currently doing well and remains in remission from his disease, without neurologic deficits. Results: An excellent response to methylprednisolone in our patient strongly suggests an immune mediated mechanism of neurotoxicity. The patient's improvement in symptoms may have been spontaneous or due to the steroid effect but suggests a possible treatment approach. Conclusion: There are no standardized treatments for cytarabine- induced neurotoxic effects, besides discontinuation of the drug. There are only a few cases in the literature.<sup>(1)</sup> We choice treatment with corticosteroids in this case. The patient presented with neurologic deficits soon after followed by rapid resolution of symptoms after initiation of corticosteroids. This case support the theory of an immunemediated mechanism and will hopefully serve as a potential treatment for those experiencing neurotoxicity with cytarabine in the future

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## Adult Hematology Abstract Categories

Chronic Leukemias PP 02

## REACTIVATION OF HEPATITIS B IN A PATIENT WITH UNTREATED CHRONIC LYMPHOCYTIC LEUKEMIA

Sinan Demircioğlu<sup>1</sup>, Atakan Tekinalp<sup>1</sup>, Ali Demir<sup>2</sup>

<sup>1</sup> Necmettin Erbakan University Meram Faculty of Medicine, Department of Hematology, Konya, Turkey

<sup>2</sup> Necmettin Erbakan University Meram Faculty of Medicine, Department of Gastroenterology, Konya, Turkey

Objective Introduction: The natural course of hepatitis B virus (HBV) infection is determined by the interaction between viral replication and the host's immune response. HBV persists in the body of all infected patients, even those with evidence of serological recovery. Therefore, individuals with a history of HBV infection receiving immunosuppressive therapy are at risk for HBV reactivation. HBV reactivation can result in increased serum aminotransferase levels, fulminant liver failure, and/or death. HBV reactivation has been described in patients receiving chemotherapy for various hematological and solid tumors. We present our patient with a diagnosis of chronic lymphocytic leukemia (CLL) who was spontaneously reactivated during follow-up without treatment. Case report: A female patient, who had been followed up with the diagnosis of chronic lymphocytic leukemia for 13 years without treatment, presented in August 2022 with complaints of loss of appetite, weight loss, and jaundice in the eye. It was learned in her history that she had not received chemotherapy or radiotherapy before, had no known disease, and did not use any medication. On physical examination, sclera icteric, multiple lymph nodes with a size of 1 cm in the cervical region and splenomegaly were detected. The patient's Rai stage 2, CLL-IPI score of 3, 17p(-), mutation in the IGHV gene was detected. Detection of leukocyte count 157,000/mm<sup>3</sup>, lymphocyte count 149.660/mm<sup>3</sup>, hemoglobin 13.5 g/dL, platelet count 117.000/mm<sup>3</sup>, alanine aminotransferase (ALT) 2045 U/L, aspartate aminotransferase (AST) 2252 U/L, total bilirubin 10.72 mg/dL, direct bilirubin 9.62 mg/dL, protrombin time 18.7 sec, active partial thromboplastin time 30 sec. While IgG was normal, IgA and M were low. HBsAg positive, anti-HBs negative, anti-HBc IgM negative, anti-HBcIgG positive, HBV-DNA 28,000,000 IU/mL were determined to explain liver dysfunction. One year ago, HBsAg was negative, anti-HBs negative, anti-HBc-IgM negative, while anti-HBc-IgG was positive. The patient was accepted as HBV reactivation and tenofovir disoproxil was started. In the first month of treatment, AST-ALT and bilirubin values returned to normal limits. In the 3rd month of the treatment, HBsAg and HBV-DNA became negative and anti-HBs became positive. The patient was followed up without treatment for CLL. Long-term use of tenofovir disoproxil was planned, despite the possibility of spontaneous HBV reactivation again. Methodology detected: Detection of leukocyte count 157,000/mm<sup>3</sup>, lymphocyte count 149.660/ mm<sup>3</sup>, hemoglobin 13.5 g/dL, platelet count 117.000/mm<sup>3</sup>, alanine aminotransferase (ALT) 2045 U/L, aspartate aminotransferase (AST) 2252 U/L, total bilirubin 10.72 mg/dL, direct bilirubin 9.62 mg/dL, protrombin time 18.7 sec, active partial thromboplastin time 30 sec. While IgG was normal, IgA and M were low. HBsAg positive, anti-HBs negative, anti-HBc IgM negative, anti-HBcIgG positive, HBV-DNA 28,000,000 IU/mL were determined to explain liver dysfunction. One year ago, HBsAg was negative, anti-HBs negative, anti-HBc-IgM negative, while anti-HBc-IgG was positive. The patient was accepted as HBV reactivation and tenofovir disoproxil was started. In the first month of treatment, AST-ALT and bilirubin values returned to normal limits. In the 3rd month of the treatment, HBsAg and HBV-DNA became negative and anti-HBs became positive. The patient was followed up without treatment for CLL. Long-term use of tenofovir disoproxil was planned, despite the possibility of spontaneous HBV reactivation again. Results: determined to explain liver dysfunction. One year ago, HBsAg was negative, anti-HBs negative, anti-HBc-IgM negative, while anti-HBc-IgG was positive. The patient was accepted as HBV reactivation and tenofovir disoproxil was started. In the first month of treatment, AST-ALT and bilirubin values returned to normal limits. In the 3rd month of the treatment, HBsAg and HBV-DNA became negative and anti-HBs became positive. The patient was followed up without treatment for CLL. Long-term use of tenofovir disoproxil was planned, despite the possibility of spontaneous HBV reactivation again Conclusion: tenofovir disoproxil was planned, despite the possibility of spontaneous HBV reactivation again. Discussion: HBsAg positive individuals are at greater risk for HBV reactivation compared to HBsAg negative individuals. It has been reported that HBV reactivation rate is up to 70% in HBsAg-positive individuals receiving standard chemotherapy.For those with cured infection (defined as HBsAg-negative, anti-HBc-positive, HBV DNA-negative), reactivation ranged from 0.3% to 9.0%.

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#### PP 03

## ESTABLISHMENT OF EX VIVO DRUG SENSITIVITY SCREENING PLATFORM FOR LEUKAEMIA AND MULTIPLE MYELOMA USING A SOUTH AFRICAN PATIENT COHORT

Vanelle Kenmogne L<sup>1</sup>, Austin Malise Thudzelani Takalani<sup>1</sup>, Ekene Emmanuel Nweke<sup>1</sup>, Mutsa M Takundwa<sup>1</sup>, June Fabian<sup>2</sup>, Heather Maher<sup>2</sup>, Justin Du Toit<sup>2</sup>, Vinitha Philip-Cherian<sup>3</sup>, Pascaline Fonteh Fru<sup>1</sup>, Deepak Balaji Thimiri Govindaraj<sup>4,\*\*</sup>

 <sup>1</sup> Department of Surgery, University of the Witwatersrand, Johannesburg, South Africa
<sup>2</sup> Synthetic Nanobiotechnology and Biomachines, Synthetic Biology and Precision Medicine Centre, NextGeneration Health Cluster, Council for Scientific and Industrial Research, Pretoria, South Africa
<sup>3</sup> Wits Donald Gordon Medical Centre, Johannesburg, South Africa
<sup>4</sup> Department of Haematology, Chris Hani Baragwanath Academic Hospital, Johannesburg South Africa

**Objective:** Our objective is to develop a functional precision medicine platform designed to directly identify tailored drug regimens that target individual patient cancer cells and give benefit to the same donors by supporting clinical decision-making. We demonstrate our *ex vivo* drug sensitivity screening platform for precision medicine using Leukaemia and Multiple Myeloma samples from a South African patient

cohort as proof of concept. Methodology: Through collaboration with Chris Hani Baragwanath and Donald Gordon Hospitals, Johannesburg, South Africa, we performed patient sample collections of n=80. Collected patient samples include Acute myeloid leukaemia (AML) (n=7), Chronic lymphocytic leukaemia (CLL) (n=4), Chronic myeloid leukaemia (CML) (n= 30), Multiple Myeloma (n=40) and health donor (n=5). For each patient sample, peripheral blood mononuclear cell (PBMC) isolation was performed and cryopreseved in liquid nitrogen. Results: Our preliminary demographic analysis results show that we can group patients based on diagnosis, staging, exclusion and inclusion criteria. From our demographic analysis, we have also identified highly frequent chemotherapy drugs used in the cohort. Further, we can identify the most frequent chemotherapy drugs given as medication to the patient cohort. We then selected 30 drugs that are relevant for leukemia and multiple myeloma for Ex vivo drug sensitivity screening test. Conclusion: Using our results we will then select effective drugs for monotherapy and also drug combinations. Selected drug combinations will then be validated on patient samples using our ex vivo drug sensitivity test. These results will be analyzed using our statistical capabilities and developed as a packaged product of preclinical information for precision clinical trials. Thus, we are progressing our cutting-edge translational platform from technology readiness level (TRL4) to TRL6 on blood cancer.

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#### PP 04

## IBRUTINIB RELATED NEUROPATHY: A CASE REPORT

Zeynep Tuğba Güven<sup>1</sup>, Nesibe Taşer Kanat<sup>1</sup>, Neslihan Mandacı Şanlı<sup>1</sup>, Ali Ünal<sup>1</sup>

<sup>1</sup> Erciyes University Faculty of Medicine, Department of Hematology, Kayseri, Türkiye

Case report Introduction: Chronic lymphocytic leukemia (CLL) is the most common leukemia seen in adulthood and mostly affects the older age group. The treatment of CLL has completely changed in recent years with the discovery of new agents. Today, ibrutinib, an oral inhibitor of the Bruton kinase signaling pathway, has become one of the commonly used agents in the treatment of CLL. Ibrutinib, a generally well tolerated agent, has manageable side effects. However, lifethreatening side effects such as major bleeding, AF, and infections can be seen. Here, we present a case of CLL who developed peripheral sensorimotor neuropathy during ibrutinib treatment. Case Report: A 62-year-old female patient who was diagnosed with CLL 5 years before her admission was followed up in remission after R-FC chemotherapy. The patient, who received his last chemotherapy about 2 years ago, applied to the polyclinic with complaints of weakness and pallor for 2 weeks. Hepatosplenomegaly and diffuse (cervical, axillary, inguinal) lymphadenopathies were found in the outpatient clinic examination. In his abdominal ultrasonography, the liver was 16 cm, and the spleen was 14 cm. There were paratracheal and mediastinal LAPs on thorax tomography. Bicytopenia was detected in whole blood examination. The patient was thought to have CLL recurrence and Ibrutinib treatment was started at a dose of 420 mg/day. The patient presented with the complaint of weakness in the legs that started after ibrutinib treatment and continued to increase 3 weeks later. There was no significant finding in the patient's lumbar MR imaging. EMG examination of the patient revealed motor sensory axonal neuropathy. Ibrutinib was discontinued due to neuropathy thought to be related to ibrutinib. Neuropathy symptoms regressed in the patient's followup. After about 6 weeks, the patient's neuropathic symptoms regressed. Venetoclax treatment was started in the patient with persistent lymph nodes and B symptoms. The patient, whose neuropathic symptoms regressed, continues to be followed up. Discussion: With the introduction of new agents in the treatment of CLL, the chance of treatment in relapsed refractory patients has increased. In the treatment of CLL, standard R-FC (Rituximab-Fludarabine, Cyclophosphamide), and R-Bendamustine regimens were previously used as firstline therapy. Today, these treatments have been replaced by BTK inhibitors (Ibrutinib, Acalabrutinib), PI3K protein inhibitors (Idelalisib), BCL-2 inhibitors (Venetoclax) and CD-20 antibodies (Obinituzumab, Ofatumumab). The reason for this drastic change in the CLL treatment algorithm is that the newly discovered agents have less side-effect profiles, ease of use, and positive effects on mortality. Although these new treatments have less side effect profile, each newly reported side effect is very important for the follow-up of patients after treatment. Ibrutinib is a Bruton Tyrosine kinase inhibitor and is the first-line therapy for CLL. Among the side effects of ibrutinib, diarrhea, cough, nausea, HT, AF, major bleeding can be counted. It is mentioned in the literature that ibrutinib may cause neuropathy. In our case, motor neuropathy also developed, and symptoms regressed after discontinuation of the drug. The side effect of motor neuropathy should also be considered in patients given ibrutinib, and if this side effect develops, the treatment plan should be reconsidered.

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## PP 05

## RİCHTER SYNDROME TRANSFORMATION UNDER VENETOCLAX TREATMENT: A CASE REPORT OF A 51-YEAR-OLD FEMALE WITH CLL

İbrahim Halil Açar<sup>1</sup>, Birol Güvenç<sup>2</sup>

<sup>1</sup> Department of Hematology, Osmaniye State
Hospital, Osmaniye, Turkey
<sup>2</sup> Department of Hematology, Cukurova University,

Adana, Turkey

**Background:** Richter syndrome (RS) is typified by the emergence of an aggressive lymphoma in individuals who have been previously or simultaneously diagnosed with chronic lymphocytic leukemia (CLL) or small lymphocytic lymphoma (SLL). Although it is relatively rare, appearing in 2% to 10% of CLL patients, RS often proves to be lethal due to its rapid progression and the scarcity of specific therapies. Venetoclax, a BCL2 inhibitor, has demonstrated efficacy in CLL but its role remains less explored in RS. Hence, there is a paucity of information regarding the direct employment of Venetoclax in the treatment regimen for RS. This study presents a case of Richter transformation being managed under treatment with Venetoclax. Case report: Case: A 51-year-old female patient, diagnosed with CLL with negative 17p deletion following investigations in 2015 due to autoimmune immune thrombocytopenia (ITP) and lymphocytosis, was given 6 cycles of FCR (fludarabine, cyclophosphamide, rituximab) due to steroid-resistant autoimmune thrombocytopenia, and complete response (CR) was achieved according to iwCLL criteria. After remission, the patient was monitored without treatment, and in 2020, full blood count, biochemical analysis, and peripheral smear were performed due to fatigue symptoms. The complete blood count showed leukocytes: 44600/mm3, lymphocytes: 39000/mm3, MCV: 86 fl, and hemoglobin: 9.5 g/dL. The patient, with no signs of hemolytic anemia, had no nutritional (Fe, B12, folate) deficiency, and normochromic normocytic anemia was detected. There were no mutations in the immunoglobulin heavy chain variable region (IGHV) genes. The patient, evaluated as relapsed stage 3 disease, was started on venetoclax-rituximab treatment. In the 11th month of the treatment, due to symptoms of fatigue, fever, night sweats, and weight loss, a bone marrow biopsy was performed after pancytopenia was observed, and a diagnosis of diffuse large Bcell lymphoma was made. Due to Richter transformation, DA-R-EPOCH (dose-adjusted rituximab, etoposide, prednisolone, vincristine, cyclophosphamide, doxorubicin) treatment was initiated. After 4 cycles of DA-R-EPOCH treatment, single-agent ibrutinib was started due to treatment-resistant disease and an ECOG performance score of 2. The patient, whose disease continued to progress under ibrutinib treatment, died from septic shock. Conclusions: This case underscores the complexities in treating Richter syndrome, particularly with venetoclax, and emphasizes the need for careful monitoring and understanding of potential transformations. The development of Richter transformation under venetoclax treatment highlights an area that requires further investigation and consideration in the management of CLL. Prospective studies and a comprehensive approach are vital to enhancing treatment strategies and improving outcomes for patients with this aggressive form of lymphoma.

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Adult Hematology Abstract Categories

Chronic Myeloproliferative Diseases PP 06

## BIOMEDICAL ANALYSIS OF RED BLOOD CELLS IN POLYCYTHEMIA VERA, APPLICATION OF RAMAN SPECTROSCOPY

Weronika Lebowa<sup>1</sup>, Jakub Dybaś<sup>2</sup>, Stefan Chłopicki<sup>2</sup>, Tomasz Sacha<sup>3</sup>

<sup>1</sup> Department of Hematology, University Hospital, Jagiellonian University Medical College, Krakow, Poland

<sup>2</sup> Jagiellonian University Medical College, Doctoral School of Medical and Health Sciences, Faculty of Medicine, Krakow, Poland

<sup>3</sup> Jagiellonian Centre for Experimental Therapeutics, Krakow, Poland

Objective: Polycythemia vera (PV) is a chronic myeloproliferative neoplasm characterized by increased red blood cell mass. Excess erythrocytosis leads to elevated hematocrit, resulting in increased blood viscosity, a condition that promotes thrombosis. For years, red blood cells (RBCs) in PV were considered to be morphologically and functionally normal. This analysis aimed to check whether there are biochemical alterations in RBCs in PV that may be associated with thrombotic complications. Methodology: We included 5 patients with PV and 5 healthy individuals in the preliminary analysis of the biochemical properties of isolated RBCs focused on different forms of hemoglobin and heme. The analysis was conducted using Raman spectroscopy. Results: The results of the Raman spectra obtained from isolated RBCs suggest a larger contribution of ferrous heme iron in the sample of a patient with PV compared to a control sample. In the PV sample, a greater contribution of the high-spin heme iron, a molecular state typical for deoxyhemoglobin, was observed, which stays in line with higher ferrous content. The effect may indicate a weaker linkage of the protein with oxygen. Conclusion: Our analysis suggests the occurrence of biochemical alterations in RBCs in PV, together with RBC overproduction. Changes in the structure of hem and hemoglobin affect oxygen affinity. Our future study will focus on determining if described alterations in RBCs may contribute to the pathogenesis of thrombotic complications in PV.

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## PP 07

## DISCONTINUATION OF TYROSINE KINASE INHIBITORS IN TUNISIAN CHRONIC MYELOID LEUKEMIA PATIENTS

Rim FRIKHA<sup>1</sup>, Moez ELLOUMI<sup>1</sup>, Hassen KAMOUN<sup>1</sup>

<sup>1</sup> UNIVERSITY HOSPITAL OF SFAX-TUNISIA

**Objective:** Some patients who achieve deep molecular remission (DMR) can successfully discontinue tyrosine kinase

inhibitors (TKI). TKI discontinuation in chronic phase CML is being implemented in the clinical routine. To investigate the outcome of the patients with chronic myeloid leukemia (CML) discontinued tyrosine kinase inhibitors (TKI) therapy Case report: TKI was prospectively discontinued in patients who were diagnosed with CML in the chronic phase treated with TKI for ≥5 years, and sustained molecular response 4.5 (MR4.5) for  $\geq$ 2 years. Molecular relapse was defined as a single loss of major molecular response (MMR) (BCR-ABL1<sup>IS</sup> >0.1%). Methodology: Standard qRT-PCR techniques were performed to evaluate minimal residual disease (MRD) Results: Twentyone patients with chronic-phase CML were enrolled. The median duration of TKI treatment before discontinuation was 117 months (49-177) months. The median follow-up time after TKI discontinuation was 20 months (range: 1-117 months). The estimated TFR rate was 62% and 47.6% at 12 and 24 months after discontinuation respectively. Five patients experienced loss of MMR within 7 months after TKI discontinuation. All relapsed patients promptly resumed TKI therapy and regained at least major molecular response. Conclusion: Our data on the Tunisian population may provide a basis for the safety and feasibility of TKI discontinuation particularly in CML patients who are in sustained deep molecular response with longer TKI treatment duration.

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#### Adult Hematology Abstract Categories

Coagulation Diseases PP 08

## AZERBAIJAN EXPERIENCE OF HAEMOPHILIA CARE

Gunel Alızada <sup>1</sup>, Mehpara Kazımova <sup>2</sup>, Elmira Gadımova <sup>1</sup>, Hikmet Ibrahımlı <sup>2</sup>

<sup>1</sup> Azerbaijan State Advanced Training Institute for Doctors named after Aziz Aliyev, Department of Haematology

<sup>2</sup> The State Agency on Mandatory Health Insurance

Objective: As the management of haemophilia is complex, it is essential that those with the disorder should have ready access to a range of services provided by a multidisciplinary team of specialists. There is a State Program aimed at solving this problem in Azerbaijan. The purpose of the study to learn complex epidemiological characteristics which are necessary for justification of strategy on treatment and prevention of haemophilia. Methodology: For planning of prophylactic treatment in Baku city, there was obtained the database of all patients (by sex, age, diagnosis, severity)registered in the city (625 persons). The main group consisting of 52 patients with severe and 40 patients with moderate haemophilia-A was formed.Different variant treatment of 162 patients was organized in HTC:chemical synovectomy with rifampicin (44); phonophoresis with refined naftalan oil (44); phonophoresis with hydrocortisone (28); electrophoresis with KJ (35). Results: 77.9% of patients observed in treatment and prophylaxis

facilities in Baku were men, 59% were diagnosed with haemophilia A, 18.8% with severe and 31.5% with moderate haemophilia.Prophylactic treatment reduces the average annual number of bleeding episodes by 2.2 times in severe haemophilia and 2.1 times in moderate haemophilia. The model of prophylactic treatment of hemophilia can be applied in the infusion model 2 or 3 times a week as far as possible. Conclusion: The role of physiotherapeutic methods of hemarthrosis treatment was assessed and positive results were obtained. Due to the prevalence of polymorbidity in patients with hemophilia the complexity of their observation and treatment and the participation of specialists from several specialties is necessary. As the duration of haemophilia is proportional to the frequency of its complications, starting the prophylactic treatment at the stage when patients are first diagnosed is recommended.

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#### PP 09

## CHARACTERIZATION AND MANAGEMENT OF PATIENTS WITH HEREDITARY FACTOR X DEFICIENCY: A RETROSPECTIVE SINGLE CENTER EXPERIENCE

Nigar Abdullayeva<sup>1</sup>, Fahri Sahin<sup>1</sup>, Zuhal Demirci<sup>1</sup>, Bahar Sevgili<sup>1</sup>

<sup>1</sup> Ege University Medical Faculty Hospital Adult Hemophilia and Thrombosis Center, Izmir, Turkey

Objective: Factor X deficiency (FXd) is a rare coagulation disorder that can be either hereditary or acquired. Case report: We characterized patients with FXd and evaluated their bleeding patterns and treatment strategies. Methodology: This retrospective review includes patients with FXd managed at Ege University Medical Faculty Hospital Ege Adult Hemophilia and Thrombosis Center. We analyzed demographic characteristics, laboratory results, bleeding scores, and treatments of five patients with FXd (Table). Patient 1 was admitted for further evaluation of menometrorrhagia and prolonged postpartum bleeding. She required treatment following birth, tooth extraction, and fractional curettage during follow-up. Coagulation tests were run as a part of in vitro fertilization in patient 2 and were abnormal. Family history was significant for a history of thrombosis in her mother. Blood tests were positive for Prothrombin 20210 G/A heterozygous mutation and lupus anticoagulants. The patient has never had any bleeding episodes in the follow-up. Patient 3 has a history of menometrorrhagia, gingival bleeding, and prolonged bleeding after an abortion. The sister of the patient has FXd. In followup, she was treated for subcutaneous hematoma, gingival, and post-cesarean bleeding. Patient 4 presented for evaluation of menometrorrhagia. She was treated for polypectomy, two cesarean sections, tooth extraction, intermittent recurrent ecchymosis, and epistaxis. Patient 5 was diagnosed at age one and was referred to us for further management of his condition. His initial presentation was consistent with subdural hematoma. In the follow-up, he was treated for epistaxis, hematuria, subcutaneous hematoma, and gastrointestinal and gingival bleeding. He continues to take Factor X concentrate prophylactically. All the patients are currently healthy and regularly follow up in our center. **Results Conclusion:** Since there is no FX concentrate in our country yet, FFP is used. Patients should be treated with the appropriate FX preparation and a prophylactic approach should be applied in necessary patients.

Table. Patient Characteristics and Diagnostic Laboratory Results

Patient No	Age at analysis	Gender	FX %	PT sec 10.9-14.7	PTT sec 22.5-31.3	Bleeding score*	Treatment
1. 2. 3. 4. 5.	41 25 18 34 1	F F F M	0.2 12.3 0.8 34.4 1	60.4 31.5 37 13.9 180	64.1 57.9 19.3 28.3 138	11 0 11 15 10	FFP, ES, PCC Not need FFP, ES, PCC FFP FFP, FXC, PCC

\*- International Society for Thrombosis and Hemostasis/Scientific and Standardization Committee Bleeding Assessment Tool (ISTH-BAT), FFP- fresh frozen plasma, ES- erythrocyte suspension, PCCprothrombin complex concentrate, FXC- Factor X concentrate, Ffemale, M-Male

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#### Adult Hematology Abstract Categories

Lymphoma PP 10

## REACTION OF THE CIRCULATING REGULATORY T CELLS AFTER CHEMORADIATION THERAPY OF HODGKIN LYMPHOMA

Tatiana Mushkarina <sup>1</sup>, Evgenija Kuzmina <sup>1</sup>, Tatiana Bogatyreva <sup>1</sup>, Ludmila Grivtsova <sup>1</sup>

<sup>1</sup> A. Tsyb Medical Radiological Research Centre MRRC

Objective: Purpose of the research is to determine the reaction of regulatory T cells after chemoradiation therapy of Hodgkin lymphoma. Methodology: 29 samples of peripheral blood of patients with Hodgkin lymphoma (before treatment - 10; after chemotherapy - 9; after consolidation radiotherapy -10). Chemotherapy was carried out according to the following schemes: ABVD, BEACOPP with the addition of 1-2 courses of CVPP or COPP. The subsequent consolidation of radiation therapy was accomplished to a dose of 20-24 Gy. Treg-cells were identified by phenotype CD45+CD4+CD25+CD127-. Control group consisted of 40 practically healthy people. The group data were compared using the Mann-Whitney U test. Results: At the onset of Hodgkin lymphoma the percentage and absolute count of regulatory T cells corresponded to normal values (5.19%/0.036\*109 cells/l - Hodgkin lymphoma vs 3.69%/0.031\*10<sup>9</sup> cells/l - control level, p>0.05). After chemotherapy the percentage of regulatory T cells increased to 9.09%, p<0.05; the absolute count remained at the same level (0.037\*10<sup>9</sup> cells/l, p>0.05). After consolidation of radiation therapy the percentage of regulatory T cells was determined

at the level of 9.19%, p>0.05. The decrease of absolute count of regulatory T cells was statistically significant difference and was near 0.019\*10<sup>9</sup> cells/l. **Conclusion**: There is a relative redistribution of cells within a subpopulation of activated CD4+CD25+T cells towards an increase in the level of regulatory T cells after chemotherapy of Hodgkin lymphoma. The subsequent radiotherapy consolidation at a dose of 20-24 Gy continued to increase the sensitivity of regulatory T cells to the radiation component of chemoradiation therapy.

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## PP 11

## CUTANEOUS RICHTER TRANSFORMATION IN THE 16TH YEAR OF FOLLICULAR LYMPHOMA DIAGNOSIS

Ulviyya Hasanzade<sup>1</sup>, Yunus Catma<sup>1</sup>, Nur Seda Ibili Cetinkaya<sup>1</sup>, Beyza Oluk<sup>1</sup>, Simge Erdem<sup>1</sup>, Cem Hacialioglu<sup>1</sup>, Ahmet Oguz Celik<sup>2</sup>, Musa Falay<sup>2</sup>, Sevgi Kalayoglu Besisik<sup>1</sup>

 <sup>1</sup> Istanbul University İstanbul Medical Faculty, Department Of Internal Medicine Division Of Hematology
<sup>2</sup> İstanbul University Istanbul Medical Faculty, Department Of İnternal Medicine

**Case report:** Richter transformation may develop in lymph nodes or rarely extranodally. A 70-year-old male with an exhausted appearance had a large malodorous wound progressing to necrosis on the left chest wall. He received two treatment lines 5 years apart for follicular lymphoma and was in remission. Histological evaluation showed triple hit diffuse large B cell lymphoma. PET-CT showed localized cutaneous and lymph node involvement. Two treatment lines did not control the disease. He passed on progression.

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#### PP 12

## AUTOLOGOUS HEMATOPOIETC CELL TRANSPLANTATION (HCT) FOR HODGKIN LYMPHOMA, REAL WORLD EXPERIENCE OF A SINGLE CENTER EXPERIENCE

Carmino De Souza<sup>1</sup>, Marcos Colella<sup>1</sup>, Eliana Miranda<sup>1</sup>, Lorena Bedotti<sup>1</sup>, Afonso Vigorito<sup>1,2</sup>

 <sup>1</sup> University of Campinas - UNICAMP, Hematology and Hemotherapy Center
<sup>2</sup> University of Campinas - UNICAMP, Bone Marrow Transplantation Unit, Hematology and Hemotherapy Center

**Objective:** Hodgkin's Lymphoma (HL) during the years became a high curable hematology malignant disease.

Despite high curable rates, up to 30% of patient will relapse or will be refractory to first line therapy (R/R). In this scenario, hematopoietic cell transplantation (HCT) is an important treatment modality to reverse the poor prognosis of these R/R HL patients. Hence, our goal was to evaluate the outcomes of R/R HL pts who underwent an autologous HCT. Methodology: Pts who underwent an autologous or allogeneic HCT for R/R HL at the University of Campinas, Bone Marrow Transplantation Unit of Clinical Hospital, from 1994 to 2023, had their charts revised, retrospectively. 144 procedures were performed, 121 autologous HCT, and 23 allogeneic HCT, It was analyzed 119 (95%) patients for the first autologous HCT. Descriptive analyses, Kaplan-Meier Method, Log-Rank test to compare groups and Cox Regression were applied by IBM-SPSS 24.0. Results: The median age was 27 years (9-72), 60% male. Nodular sclerosis (63%) was the most common histology. The time from diagnosis and HCT was 23 months (6-96); 44% pts had chemoresistant disease (CT\_R) and 56% chemosensitive (CT\_S); the OS and PFS pts with CT\_R were worse and Cox Regression analyzes confirmed as worst prognosis (OS: HR 2.29, 95%CI 1.29-4.07, p=0.004), besides that for PFS the time from diagnosis and HCT (PFS: HR 0.98, 95%CI: 0.97-0.99, p=0.007) was also another factor. Conclusion: Despite the small number of enrolled pts, our data can be compared to literature regarding OS and PSF. Chemosensitivity disease at HCT was associated with better outcome, and Autologous-HCT allows for long-term survival in R/R HL.

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## PP 13

SYNDROME OF INAPPROPRIATE ANTIDIURETIC HORMONE SECRETION AS CENTRAL NERVOUS SYSTEM LYMPHOMA RELAPSE SIGN OF NODAL DIFFUSE LARGE B-CELL LYMPHOMA

Nur Seda İbili Çetinkaya<sup>1</sup>, Ulviya Hasanzade<sup>1</sup>, Gülşah Alagöz<sup>2</sup>, Nur Rana Karakaya<sup>2</sup>, Nigar Ağzada<sup>2</sup>, Mehmet Babüroğlu<sup>3</sup>, Sevgi Beşışık<sup>1</sup>

 <sup>1</sup> Istanbul University, Istanbul Medical Faculty, Department of Internal Medicine, Division of Hematology
<sup>2</sup> Istanbul University, Istanbul Medical Faculty, Department of Neuroradiology

<sup>3</sup> Istanbul University, Istanbul Medical Faculty, Department of Internal Medicine

**Case report:** A woman (65) with nodal diffuse large B-cell lymphoma in remission developed confusion and communication loss before the 6th chemotherapy. She had no fever and no meningeal sign.Biochemistry revealed hyponatremia consistent with the secretion of inappropriate ADH.MRI showed contrast enhancement on the mesencephalic aqueductus cerebri and on 3rd ventricle.Cerebrospinal fluid had low glucose,

high protein, and lymphocytes. Central nervous system lymphoma with SIADH as a relapse sign was diagnosed.

#### https://doi.org/10.1016/j.htct.2023.09.063

#### Adult Hematology Abstract Categories

Myeloma PP 14

## INFECTION RATES ACROSS THE AUTOLOGOUS STEM CELL TRANSPLANTATION WITH REFLECTION OF MULTIPLE MYELOMA INDUCTION STORY IN TURKEY

Shirkhan Amikishiyev<sup>1</sup>, Sevgi Kalayoglu Besisik<sup>2</sup>, Ipek Yonal Hindilerden<sup>2</sup>, Mustafa Nuri Yenerel<sup>2</sup>, Arif Atahan Cagatay<sup>3</sup>, Simge Erdem<sup>2</sup>, Gulkan Ozkan<sup>4</sup>, Meliha Nalcaci<sup>2</sup>, Deniz Sargin<sup>2</sup>

 <sup>1</sup> Istanbul University, Istanbul Faculty of Medicine, Department of Internal Medicine, Istanbul, Turkey
<sup>2</sup> Istanbul University, Istanbul Faculty of Medicine, Department of Internal Medicine, Division of Hematology, Istanbul, Turkey
<sup>3</sup> Istanbul University, Istanbul Faculty of Medicine, Department of Infectious Diseases and Clinical Microbiology, Istanbul, Turkey
<sup>4</sup> Goztepe Medical Park Hospital, Istanbul, Turkey

Objective: This study aimed to investigate the frequency of infections after autologous hematopoietic stem cell transplantation (HSCT) in patients who were diagnosed with multiple myeloma (MM) in our tertiary center. Methodology: We conducted a single-center retrospective study between May 2007 and November 2016. All patients with MM diagnoses were screened on our institutional electronic database and European Society of Blood and Marrow Transplantation datacollecting forms. Results: Total 150 patients enrolled in the study. Nearly all patient developed fever. The median time from SCT to fever development was 7.4  $\pm$ 2.8 days. The most frequently encountered infection type was pneumonia and soft tissue infections. Other clinically documented infections were oropharyngeal candidiasis, herpetic stomatitis, skin and soft tissue infections, and neutropenic colitis. One patient developed CMV colitis. Blood and urine cultures were positive in 18.6% and 20%, respectively. Conclusion: The number of pre-transplant treatment regimens and antimicrobial lines was not statistically significant (p=0.34). No correlation was found between the timing of the SCT and the number of antimicrobial lines after transplantation (p=0.44). There was no statistical significance between febrile neutropenia and CD34 cell count (p=0.34). Early mortality rate was 0.6%. The early mortality rate covering the first 100 days was acceptable.

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## PP 15

## DEVELOPMENT OF GIANT PLASMACYTOMA IN A PATIENT WITH BONE MARROW RESPONSE DURING TREATMENT: A CASE REPORT

Rafiye Ciftciler<sup>1</sup>, Serhat Sayın<sup>1</sup>, Mehmet Daglı<sup>1</sup>

<sup>1</sup> Selcuk University Faculty Of Medicine Department Of Hematology

Objective: A plasmacytoma is a myelomatous mass that can develop into a widespread illness, be seen alone, or be combined with multiple myeloma (MM). Bone marrow does not always indicate MM, but over the course of 4-5 years, about 50% of cases advance to this disease. In this study, we aimed to present a patient who was diagnosed with multiple myeloma and developed giant plasmacytoma despite bone marrow response during follow-up. Case report: During the 4th cycle, a giant plasmacytoma developed at the patient's right arm proximal humerus level.Ultrasound imaging performed on the right upper extremity was reported as 'Diffuse skin-subcutaneous thickness, increased echogenicity and linear fluid areas were observed. A large  $5 \times 3$  cm hypoechoic nodular lesion with markedly increased blood flow was observed in the proximal medial neighborhood of the patient's incision line. Plasmacytoma continued to shrink with radiotherapy and chemotherapy Methodology: At the time of diagnosis, EPs are seen in around 7% of individuals with MM and are best identified by PET/CT scans; the presence of EP is linked to a worse prognosis. Later in the course of the disease, 6% more patients will get EP. Large, crimson-colored, subcutaneous masses can be a symptom of EP. The creases on the palms and/or soles may be affected by plane xanthomas, which may be a paraneoplastic condition. Rarely, cutaneous spicules made partially of the monoclonal (M) protein may form. Results Conclusion: We presented a case that developed a giant plasmacytoma based on multiple myeloma. This case is important because, after the diagnosis, a giant plasmacytoma developed during the 4th cycle of chemotherapy, although the patient's laboratory examinations and clinic responded to chemotherapy after 3 cycles of chemotherapy.





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#### Adult Hematology Abstract Categories

Platelet Diseases PP 16

A PHASE 3 STUDY TO EVALUATE THE EFFICACY AND SAFETY OF CAPLACIZUMAB WITHOUT FIRST-LINE THERAPEUTIC PLASMA EXCHANGE IN ADULTS WITH IMMUNE-MEDIATED THROMBOTIC THROMBOCYTOPENIC PURPURA

Sriya GUNAWARDENA, MD<sup>1</sup>, Angela HU, MD<sup>1</sup>, Laurel A. MENAPACE, MD<sup>1</sup>, Hikaru OKADA, MD, PhD<sup>2</sup>, Beverly ACCOMANDO, MS<sup>1</sup>, Julie LIN, MD<sup>1</sup>

<sup>1</sup> Sanofi, Cambridge, MA, USA <sup>2</sup> Sanofi, Tokyo, Japan

**Objective:** Caplacizumab (CPLZ) is indicated, in combination with therapeutic plasma exchange (TPE) and immunosuppressive therapy (IST), for the treatment of immune-mediated TTP (iTTP). TPE is a mainstay of iTTP treatment but is burdensome and associated with complications. Real-world data suggest efficacy of TPE-free CPLZ regimens in iTTP, but clinical trial data is unavailable. This trial evaluates the efficacy and safety of CPLZ with IST without first-line TPE in adults with iTTP. **Methodology:** MAYARI (NCT05468320) is a Phase 3 multicenter study. Adults with a clinical diagnosis of initial/recurrent iTTP are eligible pending ADAMTS13 activity level confirmation within 48 hours of enrollment. Participants will receive CPLZ and IST. CPLZ

treatment will be continued until ADAMTS13 activity level of  $\geq$ 50% at 2 consecutive visits after platelet count normalization or for up to 12 weeks, whichever occurs first; follow-up period is 12 weeks. TPE may be started after 24 hours if indicated. Results: The primary endpoint is the proportion of participants achieving remission without requiring TPE during the overall study period (Table). Revised outcomes definitions from the International Working Group for iTTP will be utilized (Cuker et al. Blood. 2021;137[14]:1855-1861). An adequate number of participants will be enrolled to ensure  $\geq$ 55 participants with ADAMTS13 activity levels <10% at baseline are available for primary endpoint analysis; around 61 participants are expected to be enrolled. Conclusion: The current standard of care in patients with iTTP includes a combination of TPE, IST, and CPLZ. This novel study will define the efficacy and safety of CPLZ and IST without first-line TPE in adults with iTTP. This regimen would avert the risks for substantial complications associated with TPE and represents a paradigm shift in the frontline management of iTTP. This content was first presented at ASH 2022 (abstract #1174).

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Adult Hematology Abstract Categories

Other Diseases PP 17

THE CLINICAL EFFICACY OF EPOETIN ALFA AND DARBEPOETIN ALFA IN PATIENTS WITH LOW-RISK OR INTERMEDIATE-1-RISK MYELODYSPLASTIC SYNDROME: RETROSPECTIVE MULTI-CENTER REAL-LIFE STUDY

Müzeyyen Aslaner Ak<sup>1</sup>, Birsen Sahip<sup>1</sup>, Ayfer Geduk<sup>2</sup>, Mehmet Ali Uçar<sup>3</sup>, Hacer Kale<sup>4</sup>, Tugba Hacibekiroglu<sup>5</sup>, Merve Gokcen Polat<sup>2</sup>, Yasin Kalpakcı<sup>5</sup>, Ali Zahit Bolaman<sup>3</sup>, Birol Guvenc<sup>3</sup>, Sehmus Ertop<sup>1</sup>

 <sup>1</sup> Department of Hematology, Zonguldak Bulent Ecevit University Faculty of Medicine
<sup>2</sup> Department of Hematology, Kocaeli University Faculty of Medicine
<sup>3</sup> Department of Hematology, Cukurova University Faculty of Medicine, Adana
<sup>4</sup> Department of Hematology, Adnan Menderes University Faculty of Medicine
<sup>5</sup> Department of Hematology, Sakarya Training and

Research Hospital

**Objective:** This study aimed to evaluate the clinical efficacy of epoetin alfa and darbepoetin alfa in patients with myelodysplastic syndromes (MDS) in the real-life setting. **Methodology:** A total of 204 patients with low-risk or intermediate-1-risk MDS who received epoetin alfa or darbepoetin alfa were included. Hemoglobin levels and transfusion need were recorded before and during 12-month treatment. **Results:** Hemoglobinlevelsweresignificantlyhigherateachfollowupvisitwhencomparedtobaseline levelsinbothepoetinalfaanddarbepoetinalfagroups.Transfusionneedsignificantly decreasedfrombaselineateachstudyvisi intheepoetinalfagroup.Hemoglobin levels or transfusionneedwassimilarbetween treatmentgroups. **Conclusion:** This reallife retrospective study revealed similar efficacy of epoetin alfa and darbepoetin alfa among low risk or intermediate-1 risk MDS patients with no difference in treatment response between treatment groups, whereas a likelihood of earlier treatment response in the epoetin alfa group(figure 1).

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PP 18

## RETROSPECTIVE EVALUATION OF BONE MARROW FINDINGS IN AUTOIMMUNE HEMOLYTIC ANEMIAS

Eren Arslan Davulcu<sup>1</sup>, Tarık Onur Tiryaki<sup>2</sup>, Elif Aksoy<sup>1</sup>, Emine Gültürk<sup>1</sup>, İpek Yönal Hindilerden<sup>3</sup>, Meliha Nalçacı<sup>3</sup>, Fehmi Hindilerden<sup>1</sup>

 <sup>1</sup> University of Health Sciences Bakırkoy Dr. Sadi Konuk Training and Research Hospital, Hematology Clinic, Istanbul, Turkey
<sup>2</sup> University of Health Sciences, Şişli Hamidiye Etfal Training and Research Hospital, Department of Internal Medicine, Division of Hematology, Istanbul, Turkey
<sup>3</sup> Istanbul University Faculty of Medicine,

Department of Internal Medicine, Division of Hematology, Istanbul, Turkey

Objective: Autoimmune hemolytic anemias (AIHA) are rare disorders where autoantibodies destroy self-red blood cells. AIHA includes warm AIHA (wAIHA), cold AIHA (cAIHA or cold agglutinin disease), mixed AIHA (mAIHA), paroxysmal cold hemoglobinuria (PCH), and atypical AIHA (aAIHA) based on direct antiglobulin test (DAT) results. We studied bone marrow features and their link to disease outcomes in AIHA cases with bone marrow trephine biopsies during the disease course. Methodology: AIHA patients, who had bone marrow aspiration and trephine biopsy between 2005-2023, were assessed retrospectively. Data included demographics, baseline/follow-up laboratory results (HB, hematocrit, reticulocyte count/percentage, corrected reticulocyte, lactate dehydrogenase, bilirubin, haptoglobin levels, DAT results), bone marrow features (cellularity, erythroid hyperplasia, dyserythropoiesis, marrow reticulin fibrosis, lymphoid infiltrates), treatment details, response, and outcomes. Results: A total of 43 AIHA patients were studied (32 females), with the median age at diagnosis of 55 years. Patients with grade≥1 MF received more treatment lines (p=0.012). Reticulocytosis was less frequent in ≥MF1 group (p=0.03). Grade 0-1 MF and grade≥2 MF had no difference in treatment response (p=0.089, p=0.055); grade  $\geq 2$ MF had less frequent reticulocytosis than grade 0-1 MF (p=0.024). Dyserythropoiesis had no impact on treatment or relapse (p=1, p=0.453).MF grade didn't affect relapse (p=0.503).

**Conclusion:** Our study provides valuable insights into the relationship between bone marrow characteristics and treatment response in AIHA patients. The findings indicate a significant correlation between the degree of MF and a decrease in bone marrow reticulocyte response. Additionally, as the degree of MF increased, the number of treatment lines also increased, suggesting a potential impact on disease progression and management.

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## PP 19

## LOCALIZED AL AMYLOIDOSIS OF THE URINARY BLADDER PRESENTING WITH PAINLESS MASSIVE HEMATURIA

Kıvanç Koruk <sup>1</sup>, Murat Özbalak <sup>2</sup>, Ali Altay <sup>3</sup>, Gülçin Yeğen <sup>3</sup>, Sevgi Beşışık Kalayoğlu <sup>1</sup>

 <sup>1</sup> Istanbul University Istanbul Medical Faculty, Department of Internal Medicine Division of Hematology, Istanbul Turkey
<sup>2</sup> Istanbul University Istanbul Medical Faculty, Department of Pathology Istanbul Turkey
<sup>3</sup> Başakşehir Çam ve Sakura City Hospital Department of Internal Medicine Division of Hematology,Istanbul,Turkey

Objective: Amyloid deposits can be localized as a wall thickness or mass lesion either as AA amyloidosis or AL amyloidosis and may develop nearly on all organs. It is generally a mild, non-lifethreatening entity with a good prognosis and rarely showed progression to systemic disease Methodology: We present two cases of urinary bladder localized AL amyloidosis that presents with painless hematuria and imaging studies mimic malignant tumors. Cystoscopic evaluation and biopsy were performed. Results: 63 years male presents with massive hematuria. Ultrasonography revealed a  $17 \times 14$ mm mass lesion on the bladder wall. Transurethral biopsy specimen histology showed lambda-type amyloid. The second patient was a 71-year-old male and evaluation for painless hematuria revealed a bladder wall mass lesion whose histology was consistent again with AL amyloidosis. Both patients did not have systemic amyloidosis signs and symptoms Conclusion: The literature did not include long-term outcomes. Usually, benign nature was depicted, and surgical removal is the preferred treatment. Since the contributing factors are not clear, we are concerned about the risk of recurrence and experienced the challenge of anti-plasma cell therapy giving or not.

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## PP 20

A Rare Cause Of Lymphadenopathy: Kikuchi Fujimoto

Gülcan Erbaş<sup>1</sup>

<sup>1</sup> İstanbul Faculty of Medicine

Kikuchi Fujimoto Disease (KFD) is known as NecrotizingHistiocytic Lymphadenitis. It is a self-limiting clinical situation that is seen especially in women younger than 30 years of age. It is caracterized by progresses with multiplecervical lymphadenopathy and high fever, and regresses in 1-4 months. Its etiology is still not fully elucidated. It is thought tobe a hyperimmune reaction triggered by variousmicroorganisms (Herpesviruses, especially Ebstein BarrVirus). This is a disease that should be kept in mind in the presence of fever and lymphadenopathy of unknown origin, and can be diagnosed by pathology after exclusion of otheretiological agents. Here, a case who applied to our hospitalwith swelling and pain in the neck is presented. Case: A previously healthy 13year-old female patientpresented with complaints of swelling and pain in the neck. Inher history, it was learned that her complaint had been for 20 days. It was learned that she applied to an external center andused antibiotics with the diagnosis of acute lymphadenitis, but her complaint did not regress. There were no B symptoms. Inher resume, it was learned that she was born at term and thatshe did not have the medication she used all the time. Adenoidectomy was performed six years ago. There was nofeature in her family history. Physical examination revealedpalpable lymphadenopathy of approximately 3 cm in the rightposterior cervical region. The patient's blood count wasnormal. Sedimentation was 36 mm/hr. Acute phase reactantswere negative; peripheral smear was normal. EBV, CMV, hepatitis, toxoplasma, brucella, bartonella, tuberculosis testswere negative. The pediatric infection unit was consulted forfurther investigations. There was no mediastinal width on chest X-ray. Immunoglobulin levels were normal. The doublenegative T cell rate was 6.6%. Biopsy of the lesion and simultaneous bone marrow was performed to the patient. As a result of the pathology, diffuse necrosis and apoptotic changeswere detected. The present findings were pathologicallycompatible with Kikuchi-Fujimoto. The patient is currentlybeing followed up with pediatric immunology. Conclusion: Clinical management of patients presenting withpalpable lymph node is very important. The diagnosis of lymphoma, which is one of the most common childhoodmalignancies, should definitely be kept in mind. Kikuchi-Fujimoto disease is extremely rare. It is very difficult toconsider them among the differential diagnoses. Our aim in presenting this case is to raise awareness about Kikuchi-Fujimoto disease in our daily clinical practice. KikuchiFujimoto disease should be among the differential diagnosesin patients with lymph node enlargement.

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## PP 21

# A RARE CAUSE OF CYANOSIS: HEMOGLOBIN KANSAS

Metban Mastanzade <sup>1</sup>, Alper Koç <sup>1</sup>, Mustafa Hakan Demirbaş <sup>2</sup>, Serkan Özen <sup>3</sup>

<sup>1</sup> Elazığ Fethi Sekin City Hospital, Department of Hematology <sup>2</sup> Elazığ Fethi Sekin City Hospital, Department of Genetics

<sup>3</sup> Elazığ Fethi Sekin City Hospital, Department of Intensive Care

Objective: Hemoglobin Kansas is a variant of hemoglobin with low oxygen affinity and decreased heme-heme interaction. Patients with this variant may have asymptomatic cyanosis and polycythemia. We herein report a Hb Kansas case from Elazığ/Turkey. Case report: A 25-year-old male patient was consulted from the intensive care unit because of low oxygen saturation and peripheral cyanosis. Primary cardiac and pulmonary diseases were excluded in the tests performed before the hematology evaluation. His SpO2 was 40% in room air. Complete blood count was unremarkable except mild polycythemia (Hemoglobin (Hb), 16.9 g/dL; hematocrit, 47.6%; mean red blood cell volume, 94.4 fL; white blood cell count, 9600/ mm3, and platelet count 207 × 109/L). Methodology: There was no evidence of hemolysis. An arterial blood gas analysis (under 8 L/min oxygen) showed that the arterial partial pressure of oxygen (PaO2) was 99.1 mmHg and the SaO2 was 61.4%. Both carboxyhemoglobin and methemoglobin levels were in normal range. Hb electrophoresis revealed an abnormal band between HbA and HBA2 in close proximity to the location of HbA (Figure A). Beta globin gene analysis was performed to determine the variant. Results: The HBB gene sequence analysis revealed a c.308A>C missense change resulting in substitution from asparagine to threonine at codon 103 (Hb Kansas). His daughter and father had the same clinic. Conclusion: Hb variants with low oxygen affinity could be considered in patients with unexplained cyanosis if there is dissociation between PaO2 and SaO2. Such patients do not require any special treatment and have a good prognosis. Considering the diagnosis will help prevent unnecessary investigations and treatments.



#### Haemoglobin Electrophoresis

Name	%	Normal Values %
НЬ А	55.3	
Z8 zone	41.2	
Hb E zone	0.6	
Hb A2	2.9	

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#### PP 22

## CAN RADIOTHERAPY INDUCE A CLINICAL RESPONSE WITH OCCASIONAL LONG-TERM REMISSION IN RECURRENT GRANULOSA CELL TUMORS OF THE OVARY?

Ebtihaj Hassan<sup>1</sup>, Suad Enaami<sup>1</sup>

<sup>1</sup> Radiotherapy Department, Tripoli University Hospital, Tripoli Libya

Objective: Our objective was to review the impact of adjuvant radiotherapy on recurrent granulosa cell tumor of the ovary. Case report: Adult-type Granulosa cell tumors are uncommon neoplasms arising from the ovary's sex-cord stromal cells and account for 2-4% of all ovarian cancer. The hormonal features of AGCT explain the clinical manifestations for early diagnosis and recurrence prediction. Surgery is crucial for both initial and recurrent treatments, whereas adjuvant radiotherapy or chemotherapy therapy can induce clinical response and reasonable prevention of recurrence. Methodology: A 47-year-old Libyan woman had history of stage I AGCT of ovary diagnosed in 2012 after ovarian cystectomy, recure in 2016 with bilateral adnexal complex masses, fertility-sparing surgery was done followed by six cycles of chemotherapy then she starts hormonal therapy. In June 2021accedintal Para aortic lesion was discovered, but lost F/U. In January 2022, scans showed a right lateral vaginal vault lesion and other six lesions in the pelvis and abdomen, debulking of recurrent done. Results: Conventional radiotherapy to the whole pelvis by External beam was started using the linear accelerating machine, with a total radiotherapy dose of 45 grays (Gy) in 25 fractions for five weeks. No local recurrences, Nor lymph node, or systemic metastasis in serial CT scans of chest /abdomen /pelvis and MRI pelvis since January 2022 up to now. Conclusion: Local radiotherapy could be considered as adjuvant therapy in recurrent GCTS due to the high recurrence rate, especially post-incomplete surgical excision.

#### https://doi.org/10.1016/j.htct.2023.09.072

#### PP 23

## A CASE OF DAPSONE-INDUCED HEMOLYTIC ANEMIA RELATED TO G6PD ENZYME DEFICIENCY

Ali Dogan<sup>1</sup>, Omer Ekıncı<sup>2</sup>,

Narin Yıldırım Dogan<sup>3</sup>, Sinan Demırcıoglu<sup>4</sup>, Cengiz Demır<sup>5</sup>, Cihan Ural<sup>1</sup>, Ramazan Esen<sup>1</sup>

<sup>1</sup> Van Yüzüncü Yıl University, Department of Hematology, Van

- <sup>2</sup> Medicana International Istanbul Hospital, Istanbul
- <sup>3</sup> Van Training and Research Hospital, Van
- <sup>4</sup> Necmettin Erbakan University, Department of Hematology, Konya
- <sup>5</sup> Gazi Yaşargil Training and Research Hospital, Diyarbakir

**Objective:** Hemolytic anemia defines a group of anemias occurring due to the shortening of normal red blood cell (RBC)

lifespan due to factors extrinsic to RBCs or structural changes in RBCs. As a result of the increase in RBC hemolysis, anemia and associated clinical symptoms become manifest. Hemolytic anemias can be categorized under two broad titles: hereditary and acquired. Here, we present a case diagnosed with pemphigus vulgaris who was determined to have Glucose-6-phosphate dehydrogenase (G6PD) deficiency based on the tests performed subsequent to hemolytic anemia that occurred during dapsone therapy. Case report: 66 year-old female patient presented to the dermatology polyclinic with raised erythema and bullous lesions in a butterfly distribution on the face involving the eyelids. The patient was diagnosed with pemphigus vulgaris based on punch biopsy and, as treatment, was started on  $2 \times 50$  mg dapsone (PO),  $1 \times 16$  mg methylprednisolone (PO) and corticosteroid pomades. Blood parameters at diagnosis were as follows: leukocyte,  $8.1 \times 10^9$ /L (4.4-11); hemoglobin (Hgb), 12.3 gr/dl (12-16); thrombocyte,  $270 \times 10^{9}$ /L (142-424); MCV, 86 fl (80-100); LDH, 210 U/L (135-214); ALT, 22 U/L (0-33); AST, 16 U/L (0-32); direct bilirubin, 0.5 mg/dl (0-0.3); indirect bilirubin, 0.8 mg/dl (0.1-0.9); creatinine, 0.59 mg/dl (0.5-0.9); folate, 10 ng/ml (5.4-24); vitamin B<sub>12</sub>, 310 ng/ml (210-910). The patient presented to the dermatology polyclinic 6 days after the onset of treatment due to fatigue, pallor, icterus of the sclerae. The patient was referred to the hematology polyclinic based on the following test results: Hgb, 3.8 gr/dl; leukocyte,  $11 \times 10^{9}$ /L; thrombocyte,  $222 \times 10^{9}$ /L; MCV, 108 fl; creatinine, 0.8 gr/dl; LDH, 810 U/L; indirect bilirubin, 6.4 mg/dl; direct bilirubin, 0.8 mg/dl. The patient's history and anamnesis did not include a similar condition that followed medication use or an operation. On physical examination; sclerae were icteric, skin was pale, and there was no organomegaly or peripheral lymphadenopathy. In addition, urine was dark in color. On peripheral blood smear; macrocytosis, anisocytosis-poikilocytosis, polychromasia and Heinz bodies were observed. Corrected reticulocyte was determined as 5.2% (0.5-2%); ANA, anti-dsDNA, direct Coombs (IgG) and indirect Coombs' tests were negative. The haptoglobulin level was determined as 8 mg/dl (30-200) and was below the reference range. As the present hemolytic anemia picture was reasoned to be associated with dapsone, the medication was stopped and 16 mg methylprednisolone was started. No pathological findings were determined on abdominal ultrasonography and lung radiography. Based on the perception that anemia was associated with dapsone, G6PD enzyme levels were examined. The patients' G6PD level was found as 3.52 IU/gHb (7.48-10.20 IU/gHb), and was below the reference. During follow-up, fatigue, subicterus and pallor improved. Hgb levels increased, LDH and indirect bilirubin levels showed a gradual decrease. Blood parameters after 10 days were as follows: Hgb 11,8, gr/dl; leukocyte,  $7.6 \times 10^{9}$ /L; thrombocyte,  $234 \times 10^9$ /L; MCV, 98 fl; creatinine, 0,6 gr/dl; LDH, 260 U/ L; direct bilirubin, 0.42 mg/dl; indirect bilirubin, 0.44 mg/dl. Conclusion: Dapsone is used widely in the treatment of various disorders, most notably, dermatological disorders. In G6PD deficiency, using dapsone is risky and is associated with a high probability of hemolytic anemia occurrence. In this case presentation, we aimed to stress that hemolytic anemia encountered in a patient on dapsone would be linked to G6PD enzyme deficiency.

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Pediatric Hematology Abstract Categories

General Hemostasis / Thrombosis / Vascular Biology PP 24

## UNRAVELING BLOOD DONOR DEFERRAL TRENDS: A REAL-WORLD SINGLE-CENTER STUDY

İbrahim Halil Açar<sup>1</sup>, Şule Menziletoğu Yıldız<sup>2</sup>, Birol Güvenç<sup>3</sup>

<sup>1</sup> Department of Hematology, Osmaniye State Hospital, Osmaniye, Turkey

<sup>2</sup> Blood Bank, Faculty of Medicine, Balcali Hospital,

Cukurova University, Adana, Turkey

<sup>3</sup> Department of Hematology, Cukurova University, Adana, Turkey

Backround: Enhancing blood safety and donor eligibility are vital in blood banking. We analyze our blood center's approach and Turkey's general strategy in this domain, focusing on identifying and mitigating the reasons for donor deferral. Materials and Method: We retrospectively evaluated data from 169,410 donors visiting Çukurova University Medical Faculty Blood Center from 2015 to 2021, including demographic, clinical, and laboratory information. We also compared this data with historical records from 2009 and 2011 obtained from Turkish conference papers. Results and Conclusions: Our analysis covered donors aged 18-65 years (mean 38 years) consisting of 91.1% males and 8.9% females. Blood type distribution was A Rh(+) 36.7%, O Rh(+) 29.5%, B Rh(+) 14.8%, and AB Rh(+) 7.6%. Only 3.6% of donors volunteered, while the rest had different donation reasons. A 72.3% successful donation rate was observed, but there was a 27.7% deferral rate, surpassing 2011's 25.3% and 2009's 18.2%. Deferrals were mostly due to anemia, recent medication use, elevated blood pressure, and vaccination history. Donor deferral aims to safeguard both donors and recipients against potential risks, underlining the importance of continual evaluation and management strategies to minimize deferral rates.

Key worlds: Blood Donor Deferral Blood Banking in Turkey Donor Rejection Causes Blood Donation Rates

## https://doi.org/10.1016/j.htct.2023.09.074

## PP 25

## EVALUATION OF THROMBOSIS RISK FACTORS AND PROGNOSIS IN CHILDHOOD THROMBOSIS

Mehmet Fatih Alpkiray<sup>1</sup>, Aysegul Unuvar<sup>2</sup>

<sup>1</sup> Istanbul University, Istanbul Faculty of Medicine, Department of Pediatrics, Division of Pediatric Hematology and Oncology, Istanbul, Türkiye <sup>2</sup> Istanbul University, Istanbul Faculty of Medicine, Department of Pediatrics, Istanbul, Türkiye

Objective: The aim of our study is to determine demographic data in patients with thrombosis in childhood to determine hereditary and/or acquired risk factors that cause thrombosis, to diagnose and treat thrombosis, to detect the complications related to thrombosis or treatment, to examine mortality and morbidity after thrombosis, and to evaluate the final status of the patients. Methodology: 160 cases diagnosed with thrombosis between the ages of 1 month and 18 years, who were followed up by the Pediatric Hematology and Oncology outpatient clinic of IstanbulSchool of Medicine, between 01-JAN-2012 and 01-JAN-2022 were analyzed, retrospectively. While obtaining the medical data of the patients, patient files and hospital information management systems were used. The obtained data were analyzed with IBM SPSS V23 computer program and p<0.05 was considered statistically significant. Results: Cerebral thrombosis was present in 33% of the cases, thrombosis in the lower extremity in 30.6% and upper extremity in 25.6%. At least one acquired or hereditary thrombosis risk factor was detected in 96.9% of the patients. Acquired risk factors were found in 81.2% of the patients, hereditary risk factors in 60.6% and both acquired and hereditary risk factors in 45% of the patients. Twenty (12.5%) patients were followed up without anticoagulant treatment.66.2% of the patients received prophylaxis Conclusion: In our study; the incidence of childhood thrombosis, acquired and inherited risk factors, treatment and complications of thrombosis were found to be compatible with the studies conducted in our country and in the world. Based on the frequency of inherited and acquired risk factors in every child with thrombosis, it is thought that these risk factors cannot be ignored. Conducting studies in a larger population, including the healthy control group, will contribute to the literature.

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#### Pediatric Hematology Abstract Categories

Red Blood Cell Disorders PP 26

## SLEEP QUALITY IN PATIENTS WITH B-THALASSAEMIA MAJOR

Ali Özdemir<sup>1</sup>, Funda Erkasar<sup>2</sup>, Şefika Toga<sup>3</sup>

 <sup>1</sup> Mersin City Training and Research Hospital, Pediatric Pulmonology Section
<sup>2</sup> Mersin City Training and Research Hospital, Pediatric Hematology Section
<sup>3</sup> Mersin City Training and Research Hospital, Department of Pediatrics

**Objective:** INTRODUCTION AND PURPOSE:  $\beta$ -thalassaemia major ( $\beta$ -TM) is characterized by chronic anemia due to a genetic deficiency in hemoglobin production. The clinical findings of the disease include hepatosplenomegaly, enlargement and thinning of the bones with flattening of the nasal

root, protrusion of the forehead and other facial bones resulting abnormal facial appearance. In this study, we aimed to examine sleep apnea and abnormal sleep quality in patients with  $\beta$ -TM that might occur as a result of structural facial defect. Methodology: METHODS AND MATERIALS: Two separate sleep-related questionnaires, pediatric sleep (PSQ) and pediatric sleep habits (PSHQ), were used to patients with  $\beta$ -TM who were followed in the pediatric hematology section of our hospital. Same questionnaires were applied to children in pediatric outpatient clinic who had no history of any chronic illness as a control group. The families included to the study were asked to fill questionnaires under the supervision of a clinical nurse. Results: FINDINGS: A total of 50 children with  $\beta$ -TM and 47 children as a control group were included in the study. No significant difference was found among the characteristics (age, gender, family education level) of both groups. Additionally, there was also no statistical difference between the total sleep duration of patients with  $\beta$ -TM and the control group. Similarly, no statistical difference was observed among the groups in the pediatric sleep apnea questionnaire. However, there were statistically significant higher scores in patients with  $\beta$ -TM compared to control group in the pediatric sleep habits questionnaire. In addition, the findings in the habit questionnaire scores did not change when the groups were compared by segregated age (i.e. 3-10 years old and 10-17 years old). Conclusion: DISCUSSION: The current study concluded that sleep apnea risk was not increased in patients with  $\beta$ -TM, but sleep quality was poor. No definite information exists about the cause of sleeprelated disorders in patients with  $\beta$ -TM. Probably, the atypical facial structure resulting from nasopharyngeal extramedullary increased hematopoietic activity predisposes to sleeprelated problems in patients with  $\beta$ -TM. It was also shown that the uvula-glossopharyngeal dimension was shorter in patients with thalassemia than in patients with no thalassemia. There is limited information in the literature with regard to sleep-related problems in children with  $\beta$ -TM. In a study consisted 120 patients with severe  $\beta$ -TM, the prevalence of obstructive sleep apnea was reported 8.3% and habitual snoring was 15.8%. Furthermore, an increase in periodic limb movement during sleep secondary to sleep fragmentation disorder had also reported in the same study.

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#### Pediatric Hematology Abstract Categories

Leukemia PP 27

## IS THERE AN ASSOCIATION BETWEEN PULMONARY EMBOLISM AND THE USE OF PEG-ASPARAGINASE IN CHILDREN WITH LEUKEMIA?

Emine Yılmaz Orulluoğlu<sup>1</sup>, Zühre Kaya<sup>1</sup>, Merve Yazol<sup>2</sup>, Büşra Topuz Turkcan<sup>1</sup>, Serap Kirkiz Kayalı<sup>1</sup>, Ülker Koçak<sup>1,3</sup>

<sup>1</sup> Gazi University Faculty of Medicine

 <sup>2</sup> Gazi University Faculty of Medicine, Department of Pediatric Hematology
<sup>3</sup> Gazi University Faculty of Medicine, Department

of Radiology

We present two leukemic children who developed pulmonary thromboembolism (PTE) after using PEG-asparaginase.The first child, an eight-year-old boy, was diagnosed with T-acute lymphoblastic leukemia (ALL). The second child, a 6-year-old boy, was diagnosed with B-ALL.They developed PTE following induction phases of BFM protocol's.They were given PEGasparaginase at a dose of 2500IU/m<sup>2</sup>. Heparin was successfully used in both cases.Physician may consider prophylactic anticoagulants during induction.

#### https://doi.org/10.1016/j.htct.2023.09.077

## PP 28

A PEDIATRIC CHRONIC EOSINOPHILIC LEUKEMIA CASE SUCCESFULLY TREATED WITH STEM CELL TRANSPLANTATION AFTER TRANSFORMATION TO ACUTE LYMPHOBLASTIC LEUKEMIA

Hasan Fatih Cakmaklı<sup>1</sup>, Hatice Erkol Tuncer<sup>1</sup>, Esra Pekpak Sahınoglu<sup>2</sup>, Elif Unal Ince<sup>1</sup>, Talia Ilen<sup>1</sup>, Mehmet Ertem<sup>1</sup>

<sup>1</sup> Ankara University Faculty of Medicine Department of Pediatric Hematology <sup>2</sup> Gaziantep University Faculty of Medicine Department of Pediatric Hematology and Oncology

Chronic eosinophilic leukemia (CEL) is an extremely severe and rare disease in childhood with a very poor prognosis, frequently transforms to acute leukemia in a few years, and once transformed median survival time is only 2 months. Here we present a 9-year-old boy with CEL, transformed to acute lymphoblastic leukemia 17 months after diagnosis and successfully treated with chemotherapy and unrelated stem cell transplantation, he is still in remission after 7 years without any chronic morbidities.

#### https://doi.org/10.1016/j.htct.2023.09.078

## PP 29

A COMPARATIVE STUDY OF CONVENTIONAL BLOOD CULTURE METHOD VS SEPSIS QPCR MX-30 <sup>®</sup> PANEL IN PATIENTS WITH PEDIATRIC LEUKEMIA

F.Burçin Kurtipek<sup>1</sup>, Ayca Koca Yozgat<sup>1</sup>, Zeliha Güzelküçük<sup>1</sup>, Bedia Dinç<sup>1</sup>, Dilek Gürlek Gökçebay<sup>1</sup>, Namık Yaşar Özbek<sup>1</sup>, Neşe Yaralı<sup>2</sup>

<sup>1</sup> Sağlık Bilimleri Üniversitesi, Ankara Bilkent Şehir Hastanesi Çocuk Hematoloji ve Onkoloji Kliniği <sup>2</sup> Yıldırım Beyazıt Üniversitesi, Ankara Bilkent Şehir Hastanesi Çocuk Hematoloji ve Onkoloji Kliniği

Objective: Acute leukemia is the most common pediatric hematological malignancy. Blood stream infections (BSI) are severe complications in these patients during chemotherapy. In patients with leukemia, early detection of the infectious agent and rapid initiation of appropriate treatment increase the success of treatment and reduce the death rate. In this study, we aimed to compare the causative microorganism and detection time with classical blood culture and sepsis qPCR MX-30 panel Methodology: Patients aged <18 years, diagnosed with acute leukemia from March-July 2023 were enrolled. Clinical presentations, demographic features, and microbiological findings were retrospectively reviewed. Blood culture and sepsis PCR panel were taken simultaneously from the first day of febrile neutropenia or fever persisted. Results: In total, 327 samples of 48 patients evaluated. No causative agent was detected in both blood culture and sepsis PCR panel in 262 (%80.2) samples. Although blood culture was negative in 19 (%5.8) samples, the sepsis PCR panel identified some microorganisms. Culture positivity was detected in 29 (%8.8) samples, while the sepsis PCR panel results were negative. Simultaneous identification was detected in 17 (%5.2) samples. Conclusion: In our study, we found sepsis panel sensitivity as 90% and positive predictive value as 93%. Although conventional blood culture is a more accessible, inexpensive and reliable method for detecting the causative agent in leukemia patients, it will be useful due to early results with the sepsis qPCR MX-30 panel.

#### https://doi.org/10.1016/j.htct.2023.09.079

**Pediatric Hematology Abstract Categories** 

Hemoglobinopathies (Sickle Cell Disease, Thalassemia etc...) PP 30

## EVALUATION OF GLUCOSE-6-PHOSPHATE DEHYDROGENASE DEFICIENCY IN PATIENTS WITH SICKLE CELL ANEMIA

Şule Çalışkan Kamış <sup>1</sup>, Defne Ay Tuncel <sup>1</sup>, Begül Yağcı-Küpeli <sup>1</sup>

<sup>1</sup> Adana City Training and Research Hospital

**Objective:** The aim of this study was to evaluate patients with a diagnosis of Sickle Cell Anemia (SCA) for Glucose-6-Phosphate Dehydrogenase (G6PD) enzyme deficiency. **Methodology:** In our study, patients diagnosed with SCA who presented to the Pediatric Hematology and Oncology Clinic at the Adana Faculty of Medicine, Health Sciences University, Adana City Training and Research Hospital, between August 1, 2022, and August 1, 2023, were evaluated. G6PD enzyme data from routine tests performed for the patients were recorded from the patient files or the hospital system. **Results:** A total of 23 patients diagnosed with Sickle Cell Anemia (SCA) were included in the study. 65.2% (n=15) of the patients were female, and 34.8% (n=8) were male. The ages of the cases ranged from 4 to 30 years, with a median age of 12. Among the cases, 20 were within the age range of 0-18 years (87%), while 3 cases (13%) were over 18 years old. The median G6PD value was found to be 26.28 U/g Hb (2.22-36.98). G6PD deficiency was detected in 2 patients (8.7%), while it was not detected in 21 patient **Conclusion:** Screening for G6PD deficiency is necessary in patients with Sickle Cell Anemia (SCA) to prevent deterioration of their condition during treatment. The co-inheritance of both diseases can worsen hemolysis in SCA patients. Therefore, caution should be exercised in drug selection for SCA patients with G6PD enzyme deficiency.

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Pediatric Hematology Abstract Categories

Stem Cell Transplantation PP 31

## VIRAL INFECTIONS IN PEDIATRIC HEAMTOPOIETIC STEM CELL TRANSPLANT PATIENTS

Irem Bozkurt<sup>1</sup>, Ikbal Ok Bozkaya<sup>1</sup>, Ozlem Arman Bılır<sup>1</sup>, Mehtap Kanbur<sup>1</sup>, Namık Yasar Ozbek<sup>1</sup>

#### <sup>1</sup> Ankara Bilkent City Hospital

Objective: The aim of this study is to determine the frequency and causative virus of viral infections seen after hematopoietic stem cell transplantation (HSCT) in pedaitric patients, the effect of the immunosuppresive agents and antiviral prophylaxis to viral infections, to evaluate the efficacy of antiviral treatment used for viral infections, the impact of viral infections on mortality after HSCT. Methodology: 295 pediatric HSCT patients between April 2010-August 2022 from a Children's Stem Cell Transplantation Unit were included. Patients' demographic info, HSCTrelated data, GVHD prophylaxis regime, antiviral prophylaxis after HSCT, the time span of prophylaxes applied, 27 different viral infections diagnosed from serum, stool and nasopharyngeal swab samples after HSCT, their frequencies and their timespans, patients' mortalities were documented from patients' files. Results: 68% of 295 patients were documented with a viral infection, most common isolates are CMV 26%, EBV 11%, ADV 9%, COVID-19 9%, BKV 7%, VZV 6%. Mortality rates are CMV 27%, EBV 38%, ADV 47%. Virus detection after HSCT is 1,10 months for CMV, 2,33 for EBV, 1,16 for ADV, 11 for VZV, 1 for BKV. The most common co-infections documented are CMV/EBV. For CMV treatment 69% valgancyclovir, 54% gancyclovir, 7% foscarnet is used. 53% of VZV infections were seen after acyclovir prophylaxis is stopped. Conclusion: HSCT is a curative treatment for a variety of hematological diseases, immune deficiencies, solid organ tumors, some genetic and metabolic disorders. With preparations before HSCT and the GVHD prophylaxis after HSCT, patients become immunosuppressive and susceptible to opportunistic viral infections. Viral infections have an impact on mortality, and it is beneficial to know the

common viral agents, when they are detected, viruses that are frequently detected together, and their treatment responses.

#### https://doi.org/10.1016/j.htct.2023.09.081

#### Pediatric Hematology Abstract Categories

Quality improvement / Patient safety PP 32

## EVALUATION OF MENSTRUATION RELATED QUALITY OF LIFE IN ADOLESCENTS WITH ABNORMAL UTERINE BLEEDING

Mine Dedeoğlu<sup>1</sup>, Neşe Yaralı<sup>1</sup>, Alkım Akman<sup>2</sup>, Demet Taş<sup>1</sup>

 <sup>1</sup> Ankara Yıldırım Beyazıt University Medicine Faculty Bilkent City Hospital
<sup>2</sup> Ankara Bilkent City Hospital

Objective: Abnormal uterine bleeding (AUB) is a common menstrual problem in adolescent girls. Every adolescent with AUB should also be evaluated for bleeding disorders. This study evaluated adolescent girls with AUB, with and without bleeding disorders, as well as their coping skills and menstruation specific quality of life compared to their peers. Methodology: The research was conducted in Ankara Bilkent City Hospital, Department of Pediatric Hematology and Adolescent Health as a prospective cross sectional study. The aim of this study was to determine coping skills and menstruationrelated quality of life of adolescent girls with AUB according to Pediatric Bleeding Questionnaire Scoring and Menstrual Assessment Chart. 167 patients with AUB and 165 control group, were included in our study. Each patient was evaluated by the hematology department in terms of bleeding disorder. The participants completed the Adolescent Coping Scale (CEIBO), the Children's Quality of Life Scale (PedsQL) and a scale developed by the researchers to determine the directly menstruation related quality of life (MRQL). Results: Bleeding disorder was found in 10.1% of adolescents diagnosed with AUB. When the CIBS sub-dimensions were compared between the patient and control groups, no significant difference was found between them (p=0,056). In adolescents with AUK; total quality of life score, and quality of life score related to school and physical health functionality were found to be statistically significantly lower than the adolescents in the control group (p=0,004; p=0,007). When the adolescents with AUK were compared with the adolescents in the control group, there was no significant difference between the social functionality and emotional functionality quality of life subdimensions (p=0,116; 0,063). Menstruation related quality of life was found to be significantly lower in adolescents with AUB (p<0,001). The quality of life of adolescents with severe AUB was found to be lower than those with moderate and mild AUB (p=0,026) .When the total PedsQL scores were compared between the patient, control, the patient group's score was significantly lower than the control group (p=0,012). However, there was no significant difference between the patients

with and without bleeding disorders in terms of quality of life and other scales. (p>0,05) Menstruation related quality of life was found to be significantly lower in adolescents with AUB than in those with bleeding disorders and the control group. (p<0,001). **Conclusion:** Although the coping skills of adolescents with AUB are similar to their peers, their quality of life is significantly impaired due to heavy menstrual bleeding. In addition to the treatment for the anemia, it is important to reduce their bleeding for their comfort in their school and social life. Also MRQL, which has been specially developed for this research, can be used for screening purposes due to its short and consistent results in primary health centers, pediatric clinics and hematology clinics.

## https://doi.org/10.1016/j.htct.2023.09.082

#### Pediatric Oncology Abstract Categories

Neuroblastoma PP 33

## NEUROBLASTOMA AND ASSOCIATED DISORDERS, A SINGLE CENTER EXPERIENCE

Arzu Yazal Erdem<sup>1</sup>, Selma Çakmakcı<sup>1</sup>, Seda Şahin<sup>1</sup>, Derya Özyörük<sup>1</sup>, Neriman Sarı<sup>1</sup>, Suna Emir<sup>2</sup>, İnci Ergürhan İlhan<sup>1</sup>

<sup>1</sup> Ankara Bilkent City Hospital <sup>2</sup> Atılım Üniversitesi Tıp Fakültesi

Objective: The genetic factors involved in development of neuroblastoma are not yet well understood. The most common somatic genomic alterations in neuroblastomas are recurrent chromosomal copy number alterations. In addition a number of genes with germline mutations commonpolymorphisms have been identified that raise the risk of developing neuroblastoma, it is unclear what role they play. With this aim, we investigated the syndromes, diseases and abnormalities accompanying our neuroblastoma patients. Case report Methodology: The files of patients with neuroblastoma in Ankara Dışkapı Children's Hospital, Ankara Oncology Hospital, and Ankara City Hospital between 1993 and 2023 were retrospectively analyzed. Data collected from the files included the age, sex, pathological findings, physical examination findings, imaging findings and follow-up time. Results: The files of 194 patients diagnosed with neuroblastoma were retrospectively evaluated, and distinct abnormalities and syndromes were noted in 11 patients (0.56%). The patient characteristics were presented in the Table1. Heterochromia have been known in association with NB. Neuroblastomas are rare per se in the setting of NF1 (0.2% of all NBs) and even if compared to the overall frequency of malignancies in NF1 (i.e., 14.7%). Paraneoplastic syndromes including opsoclonusmyoclonus-ataxia syndro Conclusion: Here we report on a new patient with Kabuki syndrome and a germline variant in KMT2D who developed a neuroblastoma. Including our patient literature review identifed 19 patients with Kabuki syndrome and a malignancy. Although we found no strong arguments pointing towards KS as a tumor predisposition

syndrome, based on the small numbers any relation cannot be fully excluded. As the genetics of neuroblastoma become understood in syndromic patients, steps towards intervention may be successful.

Patient no	Age at diagnosis/ gender	Syndrome/ disease	Histology	Follow-up time (year)
1	8y,F	MMR+NF type 1	GNB	3
2	1,5y, M	Heterochromia	NB	13
3	2,5y, F	Heterochromia	NB	13
4	2у, М	Hypotonic infant	GNB	6
5	12y, F	Hereditary sferocytosis	GN	12
6	1y, M	Vertebral fusion anom- alies, syndactily	NB	3,5
7	9y, F	Congenital C3 deficiency	GNB	3
8	1.5y, F	Congenital adrenal hyperplasia	GNB	2,5
9	7y, F	Kabuki syndrome	GNB	0,25
10	2y, F	OMAS	GNB	5
11	1y, M	OMAS	GNB	12

Abbreviations: GN: ganglioneuroma GNB: ganglioneuroblastoma NB: neuroblastoma NF: neurofibromatosis MMR: mental motor retardation OMAS: opsoclonus myoclonus ataxia syndrome

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#### Pediatric Oncology Abstract Categories

Rare Tumours and Histiocytosis PP 34

## TWO RARE CASES OF SUBGLOTTIC HEMANGIOMA TREATED WITH PROPRANOLOL

Melda Berber Hamamcı<sup>1</sup>, Şule Yeşil<sup>1</sup>, Firdevs Aydın<sup>1</sup>, Gülcan Erbaş<sup>2</sup>, Deniz Tuğcu<sup>2</sup>, Şifa Şahin<sup>2</sup>, Zuhal Bayramoğlu<sup>3</sup>, Yasin Ateş<sup>2</sup>, Serap Karaman<sup>2</sup>, Hikmet Gülşah Yıldız<sup>2</sup>, Hakan Kocaman<sup>4</sup>, Elif Dede<sup>5</sup>, Ayper Somer<sup>5</sup>, Ayşegül Ünüvar<sup>2</sup>, Zeynep Karakaş<sup>2</sup>

<sup>1</sup> Ankara Etlık Cıty Hospital

- <sup>2</sup> Istanbul Faculty Of Medicine, Pediatric Oncology
- And Hematology Department
- <sup>3</sup> Istanbul Faculty Of Medicine Radiology
- Department
- <sup>4</sup> Istanbul Faculty Of Medicine Pediatric Surgery Department

<sup>5</sup> Istanbul Faculty Of Medicine, Child Infection Department

**Case report:** The 22-month-old male and 15-day-old female patients presented with persistent stridor since birth. Tracheoscopy of the first patient revealed a 90% obstructing hemangioma in the subglottic area, while the second patient's CT scan showed a hemangioma at the subglottic level. Both patients were initiated on propranolol therapy. These cases highlight the significance of subglottic hemangioma as a treatable cause of stridor in infants and emphasize the importance of propranolol treatment.





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#### PP 35

## A RARE INTERSECTION: COEXISTENCE OF BREAST CANCER AND SICKLE CELL DISEASE IN A 40- YEAR-OLD FEMALE - A CASE REPORT

Birol Güvenç<sup>1</sup>, İdil Yürekli<sup>2</sup>, Berksoy Şahin<sup>3</sup>

 <sup>1</sup> Department of Hematology, Cukurova University, Adana, Turkey
<sup>2</sup> Department of Anatomy, Faculty of Medicine,

Cukurova University, Adana, Turkey

<sup>3</sup> Department of Medical Oncology, Cukurova

University, Adana, Turkey

Background: Breast cancer, a prevalent malignancy in women, and sickle cell disease (SCD), a genetic disorder affecting red blood cells, are both well-understood individually. However, their coexistence is rare and presents unique challenges in diagnosis, treatment, and management. The complex interplay between these two conditions necessitates a tailored approach to care. The report focuses on a case of coexistence of breast cancer and sickle cell disease in a 40 - year-old female. Case Presentation: A 40-year-old female patient, diagnosed with SCD and managed with 20 mg/kg hydroxyurea, experiencing 1-2 mild painful crises annually and requiring 1-2 units of transfusion yearly, presented with swelling in the right breast in October 2022. Initial MRI revealed widespread edematous changes in the right breast parenchyma and multiple lymph nodes in the right axilla. Follow-up ultrasound in December 2022 detected an ill-defined hypoechoic area in the right breast and lymphadenopathies. A tru-cut biopsy confirmed invasive ductal carcinoma. PET scan showed no metastatic focus, but cranial imaging revealed an aneurysmatic dilation in the left ICA cavernous segment. The patient's biopsy material was re-examined, showing 90% positive estrogen receptor, 60% positive progesterone receptor, Cerb2:1 positive, E-cadherin positive, and a Ki-67 proliferation index of 10%. The patient underwent neoadjuvant chemotherapy followed by modified radical mastectomy surgery, and adjuvant RT was planned with radiation oncology. Comments: The coexistence of breast cancer and SCD in this case underscores the importance of an integrated approach to diagnosis and treatment. The rarity of this coexistence in the literature highlights the need for further research to understand the specific interactions between these diseases. The case also emphasizes the necessity of collaboration between oncology, hematology, and other specialties to develop effective therapeutic strategies tailored to the unique needs of patients affected by both conditions.

Keywords: Breast Cancer Sickle Cell Disease Sicle Cell Anemia Invasive Ductal Carcinoma Lymphadenopathies,

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Pediatric Oncology Abstract Categories

Survivorship and Late side effects PP 36

## SECONDARY BRAIN TUMORS IN THE SURVIVORS OF CHILDHOOD LEUKEMIA

Nida Erbaş <sup>1</sup>, Mehmet Kantar <sup>2</sup>, Eda Ataseven <sup>2</sup>, Serra Kamer <sup>3</sup>, Cenk Eraslan <sup>4</sup>, Yeşim Ertan <sup>5</sup>

<sup>1</sup> Ege University School of Medicine Department of Pediatrics

<sup>2</sup> Ege University School of Medicine Department of Pediatrics Division of Pediatric Hematology-Oncology

<sup>3</sup> Ege University School of Medicine Department of Radiation Oncology

<sup>4</sup> Ege University School of Medicine Department of Radiodiagnostics

<sup>5</sup> Ege University School of Medicine Department of Pathology

Long term survivors of leukemia increasingly experience late effects many years after treatment. Secondary malignant neoplasms (SMNs) after ALL treatment are AML, myelodysplastic syndrome, lymphomas, CNS tumors, carcinomas and sarcomas. In the literature, CNS tumors, either meningioma or non-meningioma tumors constitute 21.5% of the SMNs in a large pediatric leukemia series. The latent period is median 15 years for meningioma and 8 years for other CNS tumors. Here in, we report three leukemia survivors of whom two developed meningiomas and one glioblastoma multiforme in the long-term period. A 3-year-old girl with T-cell ALL was treated by ALL BFM-95 protocol between 2017-2019. She also received 12 Gy of prophylactic cranial irradiation before maintenance treatment. In April 2019, at the age of 22, she developed headache, vomiting and blurred vision. CT and MRI scans revealed an extraaxial mass in the right frontal region which was compressing lateral ventricle. She, then, underwent a total excision of the tumor. The pathology was atypical meningioma (grade II). No further therapy was given. A 3year-old with T-cell ALL was given ALL IC-BFM 2002 protocol between 2010-2012. He also received 12 Gy of prophylactic cranial irradiation before maintenance treatment. The patient remained disease-free until June 2017 when he presented with generalized tonic-clonic seizures. His MRI scan showed an intraaxial lesion in the right frontal region. He underwent a biopsy that revealed an anaplastic astrocytoma. He was started cranial irradiation and temozolomide treatment. In the follow-up, the tumor progressed and the patient deceased. A 3-year-old girl with AML-M2 was treated by AML-BFM-98 protocol between 2005-2007. Before maintenance treatment she was given prophylactic cranial irradiation as 18 Gy. In 2020, she developed headache and somnolance at the age of 19. She, therefore, underwent a cranial MRI scanning that demonstrated a frontal mass. She was operated and the mass was removed totally. The pathology was grade I meningioma. She was given no further treatment. The incidence of secondary brain tumors in ALL is higher than that in AML. The exact causative mechanism is uncertain, however irradiation itself or genetic predisposition may be responsible for the pathogenesis of these type of tumors. In our two meningioma cases, there was no clinical signs of neurofibromatosis as an underlying genetic predisposition to secondary cancer. Histopathologically, gliomas are more common tumors than meningiomas in ALL survivors. More cases of high-grade gliomas were reported than low-grade gliomas in this population. WHO grade-I meningiomas are also frequent subtypes in ALL survivors.. In a large series of AML-BFM-87 and AML-BFM-93 treatment protocols, the authors reported only one case without histology detail. The cases presented here have highlighted the importance of long-term follow-up of leukemia survivors in terms of development of secondary cranial neoplasms.

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