multicenter study. **Methodology:** Forty-five stage I–II FL patients received 8 cycles of Rituximab (375 mg/m^2) and IF irradiation (30/40 Gy). Progression-Free Survival (PFS) 1-year from treatment start is the primary endpoint. Secondary endpoints were complete response rates, toxicity, quality of life with protocol defined visits up to month 15. **Results:** For the primary endpoint, PFS at 1-year was 85% for the intention-to-treat set. Long-term data were captured in selected sites and evaluated as post hoc analysis in the Per Protocol (PP) set: PFS was 78% at 1-year with a median follow-up of 15 months, respectively. There were 17/45 recurrences in the PP set, of which 14 were outside the radiation volume only. There were 9 serious adverse events (3 related to the therapy) during the first 15 months. **Conclusion:** IF radiotherapy combined with Rituximab is well tolerated and highly efficient with low rates of recurrence in the first years in early-stage DLBCL. The efficacy is comparable with more aggressive therapy approaches without compromising the quality of life and maintains for an extended follow-up of more than 3 years.

https://doi.org/10.1016/j.htct.2025.103895

Adult Hematology Abstract Categories

T-Cell Lymphoma

PP 18_Case report

CASES OF PRIMARY CUTANEOUS ANAPLASTIC LARGE CELL LYMPHOMA TREATED WITH SYSTEMIC OR LOCAL THERAPY

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Introduction: Primary Cutaneous Anaplastic Large Cell Lymphoma (PC-ALCL) is a CD30+ peripheral T-cell lymphoproliferative disorder without systemic involvement. It accounts for approximately 8% of cutaneous lymphoma cases. Most patients with PC-ALCL present with slow-growing, solitary or grouped skin nodules, and in some cases, regional lymph node involvement is observed. **Case-1:** A 61-year-old male patient presented to our clinic with swelling and edema of the right lower lip, along with a 57 cm draining ulcerative skin lesion in the suprasternal region. The patient had previously received six cycles of treatment for T-cell lymphoma at an outside center. A PET-CT scan identified a soft tissue lesion in the skin/subcutaneous tissue at the level of the right thyroid lobe and isthmus, measuring 57*15*52 mm, with a maximum SUV of 34. Biopsies taken from the lower lip and suprasternal skin were reported as primary cutaneous anaplastic large cell lymphoma. CD30 expression was found to be 95%. The patient was started on brentuximab vedotin along with the GDP protocol. After two cycles of treatment, improvement in the skin lesions was observed. **Case-2:** A 61-year-old male patient presented to our clinic with a complaint of a lesion on the anterior surface of the right tibia, measuring approximately 10*10 cm. The biopsy taken from the lesion was reported as CD30+ primary cutaneous anaplastic large cell lymphoma. A PET-CT scan revealed moderately hypermetabolic lymph nodes with thick cortices in the right iliac and inguinal lymphatic chains. Radiotherapy was applied to the area of the primary lesion and the regional lymph nodes, and improvement in the skin lesions was observed with treatment. **Discussion:** We aimed to discuss the outcomes of applying localized radiotherapy or systemic treatment to two patients with PC-ALCL who presented to our clinic: one with a relapse and the other with a new diagnosis. Brentuximab vedotin is effective in this disease. Since disease control could not be fully achieved with localized radiotherapy alone, we believe that the combination of systemic therapy and radiotherapy may be an important treatment option for these patients. Further large-scale case series are needed to guide treatment.

https://doi.org/10.1016/j.htct.2025.103896

Adult Hematology Abstract Categories

Myelodysplastic Neoplasms

PP 19_Case report

CLINICAL PRESENTATION AND OUTCOMES OF PATIENTS WITH MYELODYSPLASTIC SYNDROME INTRODUCTION

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Objective: Myelodysplastic Syndrome (MDS) is a clonal hematopoietic disorder that is characterized by dysplasia along with anaemia, thrombocytopenia or neutropenia and a risk of progression to Acute Myeloid Leukaemia (AML). In the United States, the yearly incidence of MDS is approximately 4 per 100 000 people, notably higher among older population rising tenfold by the age of eighty (80) years. Prognostic systems, such as the revised International Prognostic Scoring System (IPSS-R), offer rationally accurate estimates of survival at the population level. The goals of treatment in individuals having lower-risk MDS includes improving quality of life and minimizing Red Blood Cells (RBCs) and platelet transfusions. Therapeutic goals in patients having Higher-Risk MDS (HR-MDS), include decreasing the risk of transformation to AML and increasing survival. Haematopoietic Cell Transplantation (HCT) has the potential to cure MDS, but less than 10% of affected people undergo this treatment. Improvements in the understanding of MDS has resulted in newer management strategies for these patients. As a result, the treatment landscape for MDS patients is changing. All these advancements are expected to improve the survival rate of patients suffering from MDS. There is limited data on presentation and outcomes of MDS

patients in Low Middle Income Countries (LMICs). The Aim of our study is to assess the clinical presentation and treatment outcomes in patients with MDS in a low middle income country. **Methodology:** This is a single-centre retrospective cohort study with analytical design, which was approved by the hospital ethics committee (IRB-017/AFBMTC/Approval/2022). The study was conducted at the Armed Forces Bone Marrow Transplant Centre, a tertiary care facility located in Rawalpindi, Pakistan and included all consecutive patients having age > 15-years diagnosed with MDS as per revised WHO 2016 criteria from January 2019 till December 2023. The data of 128 patients was collected, followed-up and analyzed for disease and survival outcomes. Patients lost to follow up in the first 12 weeks of diagnosis or with insufficient extractable data were excluded from the study. The initial demographic and clinical information collected included age, gender, clinical presentation and laboratory parameters. Bone Marrow (BM) morphology and cytogenetics were used to establish the diagnosis of MDS. Subclassification was done using revised World Health Organization (WHO) 2016 classification of hematopoietic and lymphoid tumors. Patients were risk stratified using International Prognostic Scoring System (IPSS) and Revised-IPSS (R-IPSS) scoring as per available data. Initial treatment was stratified depending upon the aim of treatment (palliative, definitive), supportive treatments given were blood products and antibiotics. For palliative intent treatments included growth factors, immunosuppressants, lenalidomide, low dose cytarabine. For the purpose of study, definitive treatment was Hypomethylating Agents (HMA), venetoclax, intermediate/high dose cytarabine and Stem Cell Transplantation (SCT). Response to treatment was documented as per recommendations of the International Working Group (IWG) 2018 for low risk and IWG 2023 for high risk MDS. For non-transplant patients OS was defined as duration from date of diagnosis till death/last follow up and DFS as duration after being transfusion independent or complete remission till any event(death/relapse) or last follow up, while for transplant patients OS was calculated as duration from date of transplant till death/last follow up and DFS as duration from date of transplant till death, relapse or last follow up percentage and frequency was calculated for categorical variables and mean ± standard deviation or median with Interquartile Range (IQR) for the continuous variables. Survival statistics were calculated using Kaplan Meier analysis. Univariate and multivariate regression was used to document significant factors affecting survival. **Results:** This study evaluated the clinical outcomes of 128 patients (mean age: 52.5 ± 17.01 years; range: 16–90) with a male-to-female ratio of 3:1. Treatment intent was categorized as palliative (81 patients, 63.3%) or definitive (47 patients, 36.7%). Patients receiving definitive treatment had a mean age of 45.85 ± 14.45 years, whereas those in the palliative group had a mean age of 56.60 ± 17.23 years ($p = 0.0001$). Chemotherapy regimens included Hypomethylating Agents (HMA) plus venetoclax (27.3%), Low-Dose Cytarabine (LDAC) (5.5%), LDAC plus venetoclax (1.6%), and other combinations. Among those receiving curative intent treatment, 14.8% received AZA+VEN, 10.9% received decitabine+VEN, and 5.5% underwent upfront transplant. Complete Response (CR) was achieved in 11% of definitively treated patients, while 22% showed no response. Palliative treatments included erythropoietin (30.5%), ciclosporin (7%), lenalidomide (3.9%), and supportive care (14.8%). Among palliative patients, 7% achieved hematologic improvement (NTD), while 36.7% showed no response. Seventeen patients (13.3%) underwent Hematopoietic Stem Cell Transplantation (HSCT), with 11 receiving cytoreductive therapy pre-transplant. The stem cell source was Bone Marrow Harvest (BMH) for 13 patients and BMH plus Peripheral Blood Stem Cells (PBSC) for four patients. Myeloablative Conditioning (MAC) was used in 12.5%, and Reduced-Intensity Conditioning (RIC) in 0.8%. The median CD34 dose was $2.76 \times 10\text{^}6$. Neutrophil engraftment occurred at a median of 13 days, while platelet engraftment averaged 21.4 ± 3.82 days. Post-transplant complications included febrile neutropenia (12.5%), mucositis (10.2%), and Graft-Versus-Host Disease (GVHD) (acute: 3.1%, chronic: 5.5%). The Overall Survival (OS) rate was 42.5% with a median survival of 440 days (95% CI 161.7–718.2). OS varied by risk group, with very low-risk patients achieving 75% OS and very high-risk patients 0% ($p = 0.01$). Patients receiving MAC conditioning had a 56% OS rate ($p = 0.01$). Disease-Free Survival (DFS) was 22.7%, with a mean DFS of 456 days. Patients achieving Non-Transfusion Dependency (NTD) had an 80% DFS, whereas those with complete response had 50% ($p < 0.001$). The DFS for MAC recipients was 50%, while RIC patients had 0% ($p = 0.01$). Chronic GVHD was associated with improved DFS (57%) ($p = 0.02$). **Conclusion:** This study examined the clinical presentation and outcomes of MDS patients, following them until June 30, 2024, to assess Overall Survival (OS), Disease-Free Survival (DFS), and transplant-related complications, including Graft-Versus-Host Disease (GVHD). The study population had a mean age of 55.5-years, with a male-to-female ratio of 3:1, consistent with prior studies. Anemia was the most common presenting symptom (85.9%), with infections (27.3%) and bleeding (20.3%) also observed. Comparisons with existing literature revealed similar trends in demographic distribution and symptomatology. Low-risk MDS treatment included erythropoietin, thrombopoietin receptor agonists, lenalidomide, and hypomethylating agents, while high-risk patients were candidates for allogeneic Hematopoietic Stem Cell Transplantation (HSCT). Palliative treatment was given to 51.6% of patients, with 7% achieving non-transfusion dependency. In contrast, 33.6% received curative therapy, achieving an 11% Complete Response (CR) rate, aligning with reported outcomes. HSCT was performed in 17 patients, with neutrophil and platelet engraftment occurring at medians of 13 and 21 days, respectively. Acute GVHD was noted in 3.1% of patients, lower than reported rates, while chronic GVHD was seen in 5.5% of cases. OS in the study cohort was 37.5%, with DFS at 21.1%. Literature comparisons demonstrated variable survival rates depending on risk stratification and treatment modality, with OS ranging from 12.1% in high-risk MDS to 75% in low-risk patients. The study's limitations included its geographically specific population encompassing all MDS subtypes and varied treatment approaches, with follow-up durations ranging from 6 to 54 months. Larger, more controlled studies are

recommended to improve result reliability and clinical applicability.

https://doi.org/10.1016/j.htct.2025.103897

PP 20_ Case report

A CASE OF FAMILIAL PORFIRIA

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Introduction: Porphyria is a group of metabolic diseases caused by hereditary defects in the enzymatic system of heme biosynthesis. We present three cases of familial porphyria in two sisters (50 and 45-years-old) and a brother (39-years-old). The clinical symptoms of these patients were analyzed, and it was found that all patients have the same symptoms of the disease since childhood: constantly dark urine, burn-like changes on the skin of the face and ears, long-lasting wounds, moderate splenomegaly, and deformity of the joints on the fingers. **Clinical case:** Here, we present the some clinical and laboratory data one of the three patients. This patient was initially diagnosed with scleroderma, but due to the splenomegaly, she was referred to a hematologist to clarify the diagnosis. It was found that since childhood she had black urine, poorly healing wounds, and burn-like changes on the face and on the hands, which most often appeared after exposure to the sun. A physical examination revealed splenomegaly (+2–3 cm). The urine was initially intensely yellow-colored – when exposed to bright sunlight, the color changed to a dark yellow color. Total porphyrins in daily urine were 1.93 mg/L (norm 0–0.15 mg/L), uroporphyrinogen in single probe urine is 13 mg/L (norm 0–2 mg/L), and δ-aminolevulinic acid is 16.7 mg/L (norm 0.1–4.5 mg/L). The patient's sister and brother also were examined for quantitative tests for porphyrins in the urine. Both had similar porphyrin samples in their urine: total porphyrins in single probe urine – 1.23–1.3 mg/L, uroporphyrinogen in single probe urine – 12–13 mg/L, δ-aminolevulinic acid – 17.1 mg/L. **Conclusions:** Increased excretion of porphyrin precursors is one of the permanent signs of the disease and is observed not only during acute manifestations of the disease but also during the period of remissions, among the people with the latent form of the disease. Analyzing the results of our discussions, we can draw the following conclusions. Porphyria is a rare disease with very variable clinical symptoms, which causes certain difficulties in its timely diagnosis. To verify the diagnosis, even in the presence of a characteristic clinical picture, it is necessary to study the excretory profile of porphyrin metabolism with quantification of porphyrin precursors and fractions, as well as a comprehensive genetic examination.

https://doi.org/10.1016/j.htct.2025.103898

Adult Hematology Abstract Categories

Myeloproliferative Neoplasms

PP 21_ Case report

CHIC2 DELETION-ASSOCIATED HYPEREOSINOPHILIA AND SUBSEQUENT JAK2 V617F POSITIVE THROMBOCYTOSIS

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Background and aim: Myeloid/lymphoid neoplasms with eosinophilia and tyrosine kinase gene fusions (MLN-TK) are myeloid or lymphoid neoplasms driven by rearrangements involving genes encoding specific tyrosine kinases. These BCR::ABL1-negative diseases have long been recognized for their sensitivity to tyrosine kinase inhibitors. Herein, we present an interesting case diagnosed as myeloid neoplasm with abnormality of PDGFRA. **Case presentation:** A 63-year-old female patient with known glaucoma, hypothyroidism, and vitiligo was admitted to our clinic 8-years ago with fatigue. The patient had splenomegaly and eosinophil-predominant (10×10^9/L) leukocytosis (70.8×10^9/L). Secondary causes of eosinophilia (rheumatologic, infectious and immunologic) were excluded. Although not directly attributed to the patient's hypereosinophilia, echocardiographic left ventricular contraction abnormality was observed. Bone marrow aspiration and biopsy were hypercellular and showed 35%–40% eosinophils. With a preliminary diagnosis of hypereosinophilic syndrome, the patient was treated with steroids and then with hydroxyurea. Karyotype analysis was normal and FISH for t(9;22) was negative. The FISH panel revealed a 76% CHIC2 (4q12) deletion, but was negative for abnormalities of PDGFRB, FGFR1, and FIP1L1::PDGFRA fusion. The patient was switched to imatinib treatment. While the patient was followed in hematological remission for a long time with imatinib, thrombocytosis (807×10^9/L) was detected in the patient six months ago. The patient had suppressed erythropoietin level (1.94 mu/mL) and JAK2 V617F mutation. Low-dose hydroxyurea was combined with imatinib. Hematological remission was regained. **Discussion:** The majority of MLN-TK cases associated with PDGFRA rearrangements have cytogenetically cryptic deletion of 4q12 resulting in FIP1L1::PDGFRA. Although FIP1L1::PDGFRA fusion could not be demonstrated in this patient, it was thought that the patient had a myeloid neoplasm with abnormality of PDGFRA class due to CHIC2 deletion and typical clinical findings. The fact that the patient responded to treatment for many years seems to be evidence of this. The detection of JAK2 mutation during follow-up raised the question of whether this clone was present in the patient from the beginning or was acquired later.

https://doi.org/10.1016/j.htct.2025.103899