treatment will be continued until ADAMTS13 activity level of ≥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 ≥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

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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 decreasedfrombaselineateachstudyvisitintheepoetinalfagroupandonlyatthe12thmonth visitinthedarbepoetinalfagroup.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

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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≥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

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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–life-threatening 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 × 14mm 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

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Kikuchi Fujimoto Disease (KFD) is known as NecrotizingHistiocytic Lymphadenitis. It is a self-limiting clinicalsituation 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 thepresence 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 13-year-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 andsimultaneous 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

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