blood smear performed in our clinic demonstrated approximately 7–8% schistocytes, polychromatic erythrocytes, and thrombocytopenia, consistent primarily with thrombotic microangiopathy Review of the patient's previous laboratory records revealed episodes of anemia and thrombocytopenia, accompanied by elevated LDH, indirect bilirubin, and reticulocyte counts during these periods. ADAMTS-13 antigen, activity, and inhibitor levels were subsequently evaluated. The patient received red blood cell transfusions and 3 units of fresh frozen plasma. Laboratory results showed ADAMTS-13 activity of 23.61% (reference range: 40–130%), ADAMTS-13 antigen 0.06 IU/mL (reference range: 0.19–0.81), and ADAMTS-13 inhibitor 3.36 U/mL, with the inhibitor interpreted as negative (<12 U/mL), borderline (12–15 U/mL), or positive (>15 U/mL). Given the severely reduced ADAMTS-13 antigen and negative inhibitor, the patient was diagnosed with congenital TTP and initiated on biweekly therapeutic plasma infusion (10 mL/kg). Two weeks later, follow-up blood tests showed a platelet count of 219×10^9/L, and peripheral smear findings had completely normalized (Figure 3). At the subsequent follow-up, ADAMTS-13 tests were repeated, revealing activity <0.20%, antigen <0.01 IU/mL, and inhibitor 2.96 U/mL, consistent once again with congenital TTP. The patient continues to be followed and managed in our clinic. **DISCUSSION:** The aim of this case report is to raise awareness among clinicians about this rare syndrome, which, if accurately diagnosed, can be effectively treated, whereas misdiagnosis may lead to fatal outcomes. In neonates and children, clinicians may suspect congenital TTP in the presence of jaundice, hemolytic anemia, and thrombocytopenia. This rare syndrome was first described in 1960 by Schulman in an eight-year-old girl who experienced recurrent thrombocytopenia episodes responsive to plasma infusions [7]. In 1978, Upshaw reported a similar case in a 29-year-old patient with recurrent thrombocytopenia attacks associated with microangiopathic hemolytic anemia (MAHA), also responsive to plasma infusions, documenting multiple MAHA episodes often triggered by acute infections or stressors such as pregnancy or surgery [8].Classically, TTP is characterized by a pentad of fever, microangiopathic hemolytic anemia, thrombocytopenia, and variable renal and neurological dysfunction (observed in 20–30% of patients).However, this full presentation is often absent inmost patients [9,10].The current standard treatment for congenital TTP involves prophylactic or on-demand infusions of fresh frozen plasma (FFP) or plasma-derived factor VIII–vWF concentrates containing ADAMTS-13 for replacement therapy [11,12]. Until recently, no drug had been specifically approved for routine prophylaxis in patients with congenital TTP. Recombinant ADAMTS-13 received FDA approval in November 2023 for prophylactic or on-demand ADAMTS-13 replacement therapy in both adults and children with congenital TTP [13].Scully et al. [14] recently reported a phase 3 study comparing recombinant ADAMTS-13 with standard therapy for prophylaxis in congenital TTP patients. The study demonstrated that recombinant ADAMTS-13 is an effective prophylactic treatment approach for these patients. No safety concerns were reported, and no neutralizing antibodies against ADAMTS-13 were detected. Our patient had previously been diagnosed with MDS by apediatric hematology clinic and monitored with intermittent blood transfusions.MDS in children (≤18 years) is exceedingly rare, with an incidence of 1–4 cases per million [15].Therefore, we approached the initial diagnosis cautiously. During an MAHA episode, detailed investigations led to the diagnosis of congenital TTP. The patient responded well to fresh frozen plasma therapy. Currently, she continues biweekly FFP infusions at 10 mL/kg in our center, has not experienced further microangiopathic episodes, and remains under close follow-up.

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PP 14

A CASE OF X-LINKED THROMBOCYTOPENIA CONFUSED WITH IMMUNE THROMBOCYTOPENIA

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Wiskott-Aldrich Syndrome (WAS) is an X-linked disorder characterized by severe thrombocytopenia, eczema, humoral and cellular immunodeficiency, and an increased susceptibility to lymphoid malignancies. The milder form is X-linked thrombocytopenia (XLT), characterized by persistent thrombocytopenia with minimal or no signs of eczema or immunodeficiency. An eight-year-old male patient was admitted to the hospital at six months of age with complaints of coughing and wheezing. Bone marrow aspiration was performed after thrombocytopenia was detected in a complete blood count, and the bone marrow was interpreted as technically hypocellular. Immun thrombocytopenia was suspected, and follow-up was recommended. Since he had no bleeding, he did not return to our clinic. When the patient was scheduled circumcision at the age of seven, his platelet count was 30,000/mm3, so he was referred to our clinic from anesthesia. In his medical history, he had severe eczema as a baby, which later improved, and he has no history of bleeding. In his family history, his uncles also had low platelet counts, and three of his uncles underwent splenectomy for this reason, after which their platelet counts returned to normal. On physical examination, the patient had no signs of dermatitis, petechiae, purpura, or ecchymosis. The liver and spleen were palpable at 2 cm. Laboratory tests: Hgb: 12.9 g/dl, Htc: 36.7%, white blood cells: 12,360/mm3, platelets: 30,000/mm3, MPV: 9.1 fl (normal), and platelet size appeared normal in the peripheral smear. Sedimentation was normal, and immunoglobulin values were normal for age. Based on these findings, a preliminary diagnosis of XLT was considered, and WAS genetics were sent. In the WAS gene, a c.223G>A (p.V75M) hemizygous mutation was detected. The patient was diagnosed with XLT based on clinical findings and this mutation in the WAS gene. Eltrombopag treatment was initiated but was ineffective, so a splenectomy was performed.Subsequently, the platelet count reached 323,000/mm3. No decrease was observed during follow-up. This situation can cause confusion with immune thrombocytopenia (ITP) and delay the diagnosis of XLT. Since XLT patients present with a milder clinical picture than

classic WAS, treatment selection should be based on the patient's clinical presentation. Studies have shown that IVIG or antibiotic prophylaxis has no effect on the frequency or severity of infections in these patients. Since our patient did not have a history of frequent or severe infections, we did not initiate these treatments. Studies have shown that the incidence of bleeding decreases significantly after splenectomy, but the incidence of infection increases. Our patient's platelet counts returned to normal after splenectomy. To reduce the frequency of infections, we administered encapsulated bacterial vaccines and started penicillin prophylaxis for our patient. Due to the permanent morbidity in XLT, hematopoietic stem cell transplantation (HSCT) is the definitive treatment method. However, considering the side effects of HSCT, this decision should be made according to the patient's clinical condition.

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Adult Hematology Abstract Categories

Stem Cell Transplantation

PP 15

COMPARISON OF FEBRILE NEUTROPENIA IN PATIENTS UNDERGOING AUTOLOGOUS STEM CELL TRANSPLANTATION WITH BEAM AND HIGH-DOSE MELPHALAN CONDITIONING REGIMENS

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Objective: Objective: Conditioning regimens used before autolog stem cell transplantation (ASCT) have a direct impact on post-transplant complications and infectious morbidity. The BEAM regimen (carmustine, etoposide, cytarabine, and melphalan) is frequently preferred for patients with Hodgkin and non-Hodgkin lymphomas, while high-dose melphalan is commonly used in multiple myeloma. This study aims to compare the incidence of febrile neutropenia (FN) in patients undergoing ASCT with either the BEAM or high-dose melphalan conditioning regimen. **Methods:** In this study, febrile neutropenic patients who underwent autologous stem cell transplantation between 2010 and 2023 at the Hematology Department of Bursa Uludağ University Faculty of Medicine were analyzed. We evaluated the patients' demographic and clinicopathological data, duration of FN episodes, depth of neutropenia, and length of hospital stay. Additionally, the causative pathogens of FN and FN-related mortality were also analyzed, Türkiye. **Result:** A total of 164 patients were included in this study. Seventy-three of the patients were female and 91 were male. There were 131 multiple myeloma, 23 Non-Hodgkin lymphoma, and 10 Hodgkin lymphoma. One-hundred thirty one (%80) received high-dose melphalan, 33 (%20) received BEAM. The median dose of CD34+ cells was similar in both groups (p=0,938). The duration of FN episode and length of hospital stay were significantly longer in the BEAM arm (p=0,001 and p=0,001). Invasive pulmonary aspergillosis was significantly more common in the BEAM arm (p=0,013). Of the bacteria isolated in culture, 29% (n=48) were gram-positive and 9% (n=14) were gram-negative. The most frequently isolated gram-positive bacteria were Staphylococcus epidermidis (n=29) and Staphylococcus aureus (n=7), while gram-negative bacteria were Klebsiella pneumoniae (n=5) and Pseudomonas aeruginosa (n=4). CRP and Pitt score were similar in bot groups (p=0,152 vs p=0,247). No significant difference in FN-related mortality was seen between the two arms (p=0,802), Türkiye. **Conclusion:** Conclusion: The BEAM regimen significantly increased the risk of invasive pulmonary aspergillosis, length of hospital stay, and duration of febrile neutropenia. These results suggest that, particularly in lymphoma patients, the risks of FN should be taken into account when selecting the BEAM regimen, Türkiye.

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PP 16

HEMATOLOGICAL APPROACHES IN AUTOIMMUNE ENCEPHALITIS: OFATUMUMAB EXPERIENCE

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Objective: Autoimmune encephalitis (AE) is a group of encephalitides caused by immune-mediated inflammatory disorders of the brain. While B cell–mediated autoimmunity is observed in many patients, some subtypes also involve T cell–mediated mechanisms. AE-related antibodies are classified into three groups: paraneoplastic antibodies, synaptic antibodies, and antibodies of uncertain significance. Paraneoplastic antibodies are frequently associated with systemic tumors and show poor responsiveness to immunotherapy. Synaptic antibodies, on the other hand, display variable associations with systemic tumors but are generally more responsive to immunotherapy. The diagnosis of AE is based on clinical features, radiological findings (such as abnormalities on T2 and FLAIR brain MRI), slow-wave activity in the temporal lobe, cerebrospinal fluid (CSF) pleocytosis, and the exclusion of alternative causes. Although antibody detection remains one of the best diagnostic tools, many cases may still be seronegative. Common paraneoplastic antibodies include anti-Hu, anti-Yo, anti-CV2, anti-Ma2, anti-Ri, anti-amphiphysin, ZIC4, and GAD65. Major synaptic autoantibodies include anti-NMDA, anti-AMPA, anti-GABA-B receptor, anti-CASPR,