accounting for 2-3% of all leukemias in the pediatric population under the age of 15. (1) It is defined by the presence of a translocation (9;22), a cytogenetic abnormality associated with the disease. We report one of these rare cases because of its unusual frequency. **Case Report:** Fourteen year male child came to the pediatric hematology policlinic complaints of abdominal distension, bone pain and weakness. Clinical examination revealed mucocutaneous pallor and hepatosplenomegaly. The complete blood count received on the day of admission showed hyperleukocytosis at 178000/μL, normocytic normochromic anemia at 10,8 g/dl and thrombocytosis at 281000/μL. When the blood smear was examined, it was seen that there were myelocytes, metamyelocytes and promyelocytes, neutrophils and 4% myeloid-appearing blasts. Subsequent bone marrow aspiration showed hyperplasia of the neutrophilic granulocytic lineage at all stages of maturation, with promyelocyte, hyper granular myelocyte, metamyelocyte. (Figure 1) Cytogenetic analysis of the bone marrow as part of the etiological work-up confirmed the presence of the Philadelphia chromosome. Molecular testing for the BCR-ABL1 fusion transcript by RT-PCR on EDTA whole blood detected 64% (IS). The patient was admitted to the pediatric hematology service and started on hydroxyurea treatment. After the genetic diagnosis was confirmed, he was treated with Imatinib, a first-generation tyrosine kinase inhibitor (TKI). In the molecular evaluation performed at the 3-month follow-up, BCR-ABL1 fusion transcript was detected as 5% (IS) by RT-PCR. **Discussion:** Chronic myeloid leukemia (CML) is a rare hematological malignancy in the pediatric population. For treatment, our patient benefited from specific Imatinib therapy. According to the literature, Imatinib is the first-line drug.

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Adult Hematology Abstract Categories

Coagulation Diseases
PP 14

PAGET SCHROETTER SYNDROME AND HOMOZYGOUS FACTOR V LEIDEN MUTATION: A CASE PRESENTATION

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Case Report: Thrombosis in the deep veins of the upper extremity accounts for only 5% of symptomatic cases but constitutes approximately 50% of hospital-acquired thromboses. The vast majority of upper extremity thromboses, result from the presence of permanent venous catheter. Unprovoked cases are often secondary to "effort" thrombosis. Here, we present a case of Paget-Schroetter syndrome combined with a homozygous mutation of factor V Leiden. A 19-year-old female patient presented with pain and swelling in her right arm. The report of the right arm venous Doppler ultrasound indicated the presence of thrombus within the lumen at the proximal and distal segments of the basilic vein at the fossa cubiti level. The patient was found to have a homozygous mutation of factor V Leiden, and it was learned that she had been undergoing intense training and was engaged in water polo for the last two months. She had no history of medication use or chronic illnesses, nor any previous history of thrombosis. The patient was started on low molecular weight heparin for three months. A control Doppler ultrasound showed that the existing thrombus had resolved. It was recommended that the patient continue on her current anticoagulation with a new generation oral anticoagulant for one year. During this period, the patient, who ceased sports activities, did not develop any new thrombosis. The combination of young age, intense physical activity, especially in sports that utilize the upper extremities, and risk factors such as the factor V Leiden mutation strengthens the diagnosis. In the pathophysiology of this syndrome, vascular microtravma and exercise, muscle hypertrophy and thrombophilias contribute to the condition. Low molecular weight heparin and new generation oral anticoagulants are effective in preventing thrombosis formation and in inhibiting the growth of existing thrombus. Thrombolytic therapy may be considered in cases of large thromboses or severe symptoms.

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PP 15

DESENSITIZATION TO RIVAROXABAN IN A PATIENT WHO EXPERIENCED ANAPHYLACTOID SHOCK AFTER ANTICOAGULANT USE: CASE REPORT

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Case Report: Over the last two decades, new anticoagulants have been developed to prevent and manage thromboembolic diseases, including direct-acting anticoagulants like rivaroxaban, which is used for venous thromboembolism prevention, stroke prevention in atrial fibrillation, and ischemic heart disease. Here, we present the experience of a case with a history of multiple thromboses and an anaphylactoid reaction to anticoagulants, who was able to continue prophylaxis without allergic reactions after rivaroxaban desensitization. A 42-year-old female patient visited the hematology outpatient clinic to obtain a prescription for a new anticoagulant due to a supply issue with her current medication, fondaparinux..

Her medical history included thrombosis in both upper and lower extremities ten years earlier, along with heterozygous mutations for factor V Leiden and MTHFR, necessitating lifelong anticoagulant therapy. She had previously experienced anaphylactic shock from enoxaparin, warfarin, tinzaparin, and rivaroxaban, which led her to use fondaparinux without issues. When faced with a supply problem prescribed apixaban, she suffered anaphylactic shock thirty minutes after administration, requiring epinephrine treatment. Following this, the allergy and immunology department recommended a desensitization protocol for rivaroxaban, crucial for her ongoing anticoagulation. After a one-day desensitization, she successfully continued treatment with 20 mg of rivaroxaban without any allergic reactions during follow-up visits. Desensitization is a technique that allows patients with drug hypersensitivity reactions to safely maintain drug therapy by creating temporary tolerance, especially for IgE-mediated reactions. It works by inhibiting mast cell activation and reducing the release of inflammatory mediators, often resulting in decreased skin sensitivity and potentially negative skin test results after the procedure. In this case, the patient had a grade 3 early-type drug allergy, and while literature on desensitization for new-generation oral anticoagulants is scarce, the successful desensitization to rivaroxaban suggests that it may be an effective option for similar patients in the future.

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PP 16

INTERVENTIONAL PROCEDURE IN HEMOPHILIA A PATIENT WITH EXTENDED HALF-LIFE FACTOR THERAPY- CIRCUMCISION- CASE REPORT

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Case Report: Hemophilia A is a hereditary bleeding disorder due to factor VIII deficiency. With the advances in the treatment of hemophilia in recent years, the average life expectancy of patients has reached the healthy population. Along with prolonged life, additional diseases and intervention requirements are developing in this patient group. Due to the developments, management of patients going under interventions are more clear and easier. In this case, a patient who underwent an intervention with extended half-life factor therapy was presented. Forty-three-year-old male patient with severe hemophilia A was evaluated on request for circumcision surgery while using prophylactically extended half-life factor therapy 2×1000 Units / week. Tranexamic acid was started one day preoperatively to the patient whose basal factor level was below 1% and whose inhibitory level was negative. Body weight of the patient was 63 kg. Extended half-life factor VIII preparation (efmorogtocog alfa) loading dose of 3000 units was administered befrore half an hour of the procedure. aPtt was detected for 30 seconds and factor VIII level was 55% 30 minutes after loading dose. The patient was given appropriate sedative treatment to prevent pre-perioperative erection. The operation was carried out without any problems. 1500 Units 12 hours after the loading dose, and 24 hours after this dose was performed. The patient was discharged without complications without bleeding. Factor therapy was continued with prophylaxis dosing. Tranexamic acid was continued for 7 days. No complications were observed. Interventional procedures of hemophilia patients can be performed without complications with a multidisciplinary approach under appropriate dose and scheme factor therapy. In the case, an interventinal procedure was made by giving an extended half-life factor to a severe hemophilia patient who could not have a circumcision operation for many years due to previous hesitations of both patient and surgeons. ,

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Adult Hematology Abstract Categories

Lymphoma
PP 17

RARE CASE! SECOND PRIMARY MALIGNANCY IN LANGERHANS CELL HISTIOCYTOSIS, A JAK2+ CASE

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Objective: Langerhans cell histiocytosis (LCH) is a rare inflammatory myeloid neoplasm characterised by the infiltration of CD1a+CD207+ myeloid dendritic cells and immune cells, thus described as an inflammatory myeloid neoplasm that clonally expands. LCH is a histiocytic neoplasm affecting both paediatric and adult populations, with an estimated incidence of 3 to 5 cases per million children and 1 to 2 cases per million adults. LCH can involve all organ systems, with symptoms ranging from single organ disease to multi-system disease. While it can appear in any organ system, LCH has a particular affinity for bones, skin, lungs, and the pituitary gland. In 2016, LCH was reclassified from a reactive disorder to an inflammatory myeloid neoplasm following the identification of the recurrent BRAF V600E mutation in half of the cases and the observation of clonality. Recently, additional BRAF mutations that activate the MAP kinase pathway have been demonstrated, shedding more light on the pathogenesis of LCH. Several studies have suggested a high prevalence of second primary malignancies, including haematological and solid organ neoplasms, in LCH patients. **Case Report:** A 58-year-old male patient, with a known history of hypertension and hypothyroidism, presented to a medical facility in Germany in 2011 with skin lesions on the chest and neck swelling. Following lymph node and skin punch biopsies from the sternum, the patient was diagnosed with LCH, with imaging