

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 life-long 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.

<https://doi.org/10.1016/j.htct.2024.11.043>

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 before 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-operative 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 interventional 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. ,

<https://doi.org/10.1016/j.htct.2024.11.044>

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