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.

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

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## 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 $2 \times 1000$ 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. ,

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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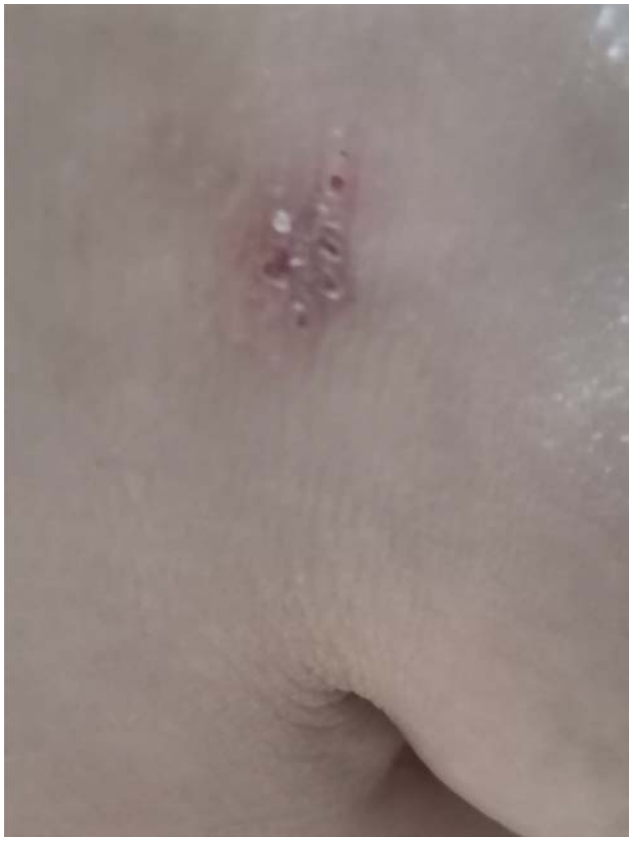
**Objective:** Langerhans cell histiocytosis (LCH) is a rare inflammatory myeloid neoplasm characterised by the infiltration of CD1a+CD207+ myeloid dendritic cells and immune cells, thus described as an inflammatory myeloid neoplasm that clonally expands. LCH is a histiocytic neoplasm affecting both 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

revealing involvement in the frontal bone of the skull, neck, spleen, axillary, liver, lungs, and skin. The patient was treated with steroids. In 2014, while on holiday in Istanbul, the patient was given 6 cycles of vinblastine in addition to steroids. Steroid treatment was completed over 5 years, followed by regular follow-up. In 2021, the patient presented to Ordu State Hospital with fatigue and skin rashes resembling LCH lesions. Investigations revealed thrombocytosis, erythrocytosis, and leukocytosis. Bone marrow biopsy was reported as normal, and a punch biopsy of the skin lesions showed no evidence supporting LCH. Cytogenetic tests, however, revealed a JAK2+ mutation, which had not been detected in previous tests. The patient was started on hydroxyurea, and imaging showed a 5 cm mass in the spleen, for which splenectomy was recommended, though the patient declined and sought further consultation. Our cytogenetic studies confirmed BCRABL polymerase chain reaction (PCR), PML/RARA, and AML/MDS panel negativity, with JAK2+ positivity. Erythropoietin levels were 6 mU/ml (normal range: 3.7-31.5), LDH was 218 u/l, sedimentation rate was 60 mm/hour, platelet count was 517,000/$\mu$l, and white blood cell (WBC) count was 13,000/$\mu$l. Physical examination revealed remnants of old skin lesions (Figure 1), and there were no palpable lymph nodes or masses. Imaging showed a significant mass in the spleen and involvement in the frontal bone, liver, lungs, stomach, and neck lymph nodes, similar to previous findings. During follow-up, the patient occasionally reported pain in both legs, and Doppler studies revealed widespread thrombosis, which the patient stated had been occurring for the past 1.5 years but was disregarded. Subcutaneous anticoagulants and anti-stasis treatment (Daflon 1000) were initiated, later transitioning to oral anticoagulants. Follow-up showed improvement in symptoms under hydroxyurea and anticoagulant therapy, but recurring thrombotic events were noted during subsequent check-ups while on oral anticoagulants. Figure 1: Skin findings and biopsy scar marks on the neck, sternum, and abdominal areas of the patient. **Conclusion:** Discussion Several case reports and smaller case series have observed that malignant diseases may occur before, concurrently with, or after LCH, with a frequency higher than by chance alone. Edelbroek, J. R., and colleagues linked the emergence of second malignancies in LCH to prior treatments with chemotherapeutic agents such as etoposide or vinblastine, with the second malignancies being identified as leukaemia and myelodysplastic syndrome (MDS). Another study by Goyal, Gaurav, and colleagues followed 1,392 LCH cases, showing that Hodgkin and non-Hodgkin lymphomas developed in children during follow-up, while adults developed MDS in early follow-up and had an increased risk of developing B-cell acute lymphoblastic leukaemia (B-ALL) after about five years. In children, the leading cause of death was infections, while in adults, it was second primary malignancies. In our literature review, we did not encounter any JAK2+ cases or studies following LCH, making the JAK2+ positivity observed in our LCH patient a potentially unique case. We did find that JAK2+ positivity has been observed in the follow-up of non-Langerhans cell histiocytosis. Given that LCH is rare and second primary malignancies are even more uncommon, identifying such cases remains challenging, and further clinical studies are clearly needed.

**Keywords:** Langerhans Cell Histiocytosis, JAK2+.



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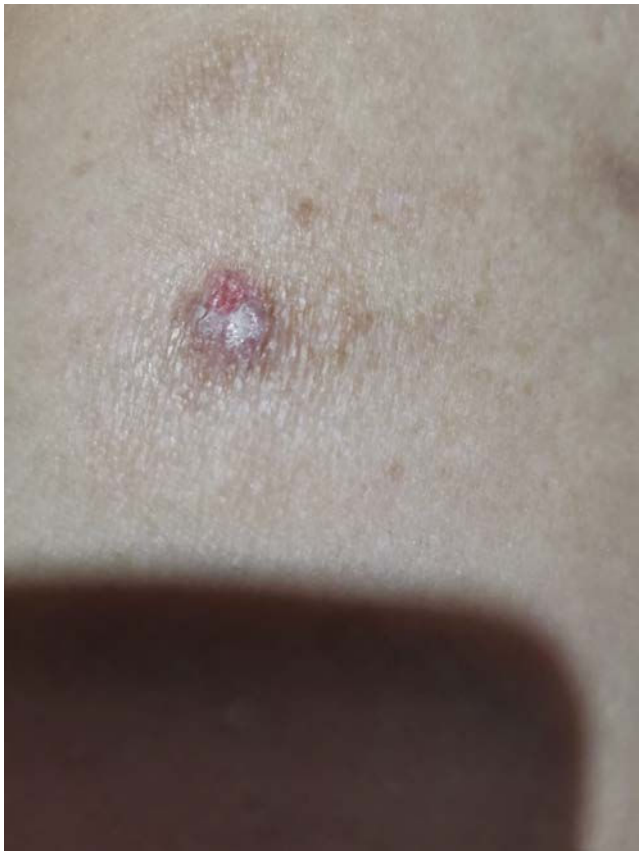

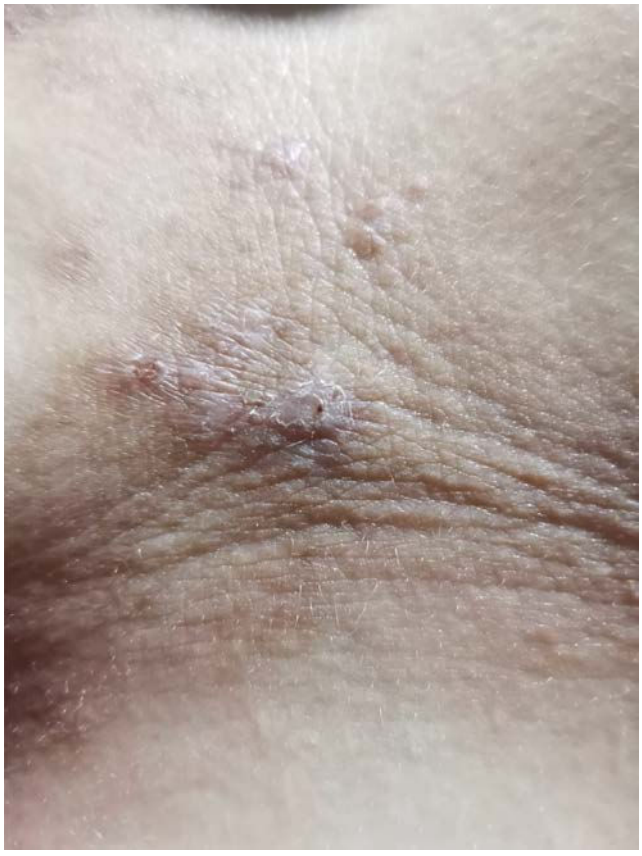

https://doi.org/10.1016/j.htct.2024.11.045

PP 18

## A CASE OF DIFFUSE LARGE B CELL LYMPHOMA PRESENTING AS OSTEOSARCOMA

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**Case Report:** Diffuse large B cell lymphoma (DLBCl) is the most common histologic subtype of non-Hodgkin lymphoma (NHl) accounting for approximately 25 percent of NHL cases. Additionally, Diffuse Large B Cell Lymphoma is the most common lymphoma. In the United States and England, the incidence of DLBCL is approximately 7 cases per 100,000 persons per year. In Europe as a whole, the incidence is approximately 4.92 cases per 100,000 persons per year. Like most other NHLs, there is a male predominance with approximately 55 percent of cases occurring in men. Incidence increases with age; the median age at presentation is 64 years for patients as a whole. IB, 45 years male patient. MRI scan taken in 2022 after a complaint of pain in right knee revealed a malignant tumoral lesion (osteosarcoma?) that caused intramuscular invasion in a segment of approximately 20 cm in the 1/2 distal femur and caused extensive cortical destruction in the distal. A biopsy was taken from the distal right femur. He was diagnosed with non-Hodgkin lymphoma and diffuse large B-cell lymphoma. Bcl-2, Bcl-6 and c-myc were found to be negative. After 4 cycles of R-CHOP protocol, PET-CT revealed minimal progression in the left clavicle and the IPI score was high. The patient's R-CHOP treatment was completed for 6 cycles with 2 cycles of intrathecal MTX. Afterwards, 2 cycles of maintenance rituximab were given. The patient, who subsequently went into remission, was followed up. This case shows us that NHL cases may present in a location such as primary bone tumor. The possibility of lymphoma should be considered in patients with atypical localization.

https://doi.org/10.1016/j.htct.2024.11.046

PP 19

## EFFICACY OF GLOFITAMAB IN PRIMARY REFRACTORY LYMPHOMA: A CASE REPORT

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**Objective:** Diffuse large B-cell lymphoma (DLBCL) constitutes 30% of non-Hodgkin lymphomas and is often curable with frontline chemoimmunotherapy. However, in some patients, remission cannot be achieved, and this situation necessitates the application of second, third or even fourth-line salvage therapies. The limited treatment options for relapsed or refractory (r/r) DLBCL underscore an unmet clinical need, which urges the development of new therapies for this patients. Glofitamab is a humanized IgG1 bispecific monoclonal antibody binds to CD20 on malignant B lymphocytes and to CD3 on cytotoxic T cells with promise for treating r/r DLBCL. Here we present a primary refractory DLBCL patient to whom we applied glofitamab treatment as the 5th line. **Case Report:** A 28-year-old male patient was diagnosed with stage IV germinal center DLBCL biopsy of sacral mass. The patient received dose-adjusted etoposide, prednisone, vincristine, cyclophosphamide, doxorubicin, and rituximab (EPOCH-R) as first-line treatment. However, progression was detected by 18F-Fluoro-2-deoxy-D-glucose (FDG) positron emission tomography (PET) with computed tomography (CT). Then, rituximab plus ifosfamide, carboplatin, etoposide (R-ICE), ifosfamide gemcitabine vinorelbine prednisolone (IGEV), salvage radiotherapy (RT), rituximab plus bendamustine (R-B) therapies were given, respectively. Since no response was obtained to all these treatments, glofitamab was started as the 5th line therapy. After the twelve cycles of glofitamab therapy, the patient achieved complete remission (CR). Four months post-treatment, he was still alive. **Discussion:** Glofitamab is approved as a third-line treatment for r/r DLBCL, inducing a CR in nearly 40% of patients in this situation. According to literature, CR can be maintained for years after completion of glofitamab treatment. Data from a follow-up in a cohort of patients who were treated with glofitamab showed a median duration of complete response of 34 months. Our case post-treatment fourth months was still alive. This case indicates that glofitamab is quite effective primary refractory DLBCL.

https://doi.org/10.1016/j.htct.2024.11.047