

Acquired FXIII deficiency has been described in association with malignancies or autoimmune disorders. We report two cases of acquired FXIII deficiency associated with hematologic malignancies. The first patient is a 60-year-old male with CMML who presented 4 weeks after confirming his diagnosis with non-traumatic anterior abdominal wall hematoma. Workup revealed FXIII deficiency. He was treated with FXIII replacement and other supportive measures. The hematoma resolved and patient was maintained on factor replacement. Unfortunately, his disease transformed to AML and he succumbed to death after starting AML therapy despite achieving complete remission. The second patient is a 24-year-old male patient post haploidentical transplant for intermediate risk AML. He developed hemorrhagic cystitis day 36 post-transplant and non-traumatic subdural hematoma on day 60 post-transplant. Workup revealed FXIII deficiency. He was treated with factor replacement and the subdural hematoma resolved with improvement of the hemorrhagic cystitis. Both patients had RUNX1 mutation which regulates expression of F13A1 in megakaryocyte this can decreased platelet expression of F13A1 in patient with RUNX1 haplo deficiency which lead to platelet dysfunction. FXIII deficiency should be considered for patient with unexplained bleeding with normal routine workup.

<https://doi.org/10.1016/j.htct.2021.10.1031>

## LYMPHOMA

### PP 22

#### A CASE OF MULTI REGIONAL PRIMARY MUSCLE LYMPHOMA

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**Objective:** Primary extranodal non-Hodgkin's lymphoma (eNHL) usually presents at an early stage, as an extranodal organ involvement along with draining lymph nodes only or the predominant site is extranodal. As an eNHL, primary skeletal muscle lymphoma is very rare. The usual clinical picture is local swelling and pain with or without systemic symptoms. MRI features are distinctive and FDG-PET/CT may help to evaluate the stage and monitor the response to the treatment. **Case report:** A 56-year-old male, presented with a one-month history of swelling and pain on his left ankle. There was no history of trauma or any physical strain. A mass lesion was palpated on the calcaneus bone. MRI showed diffuse muscle involvement. The clinical picture was not consistent with infection or hematoma. The blood cell count and biochemical investigations were within normal limits. Serology for hepatitis B, C and HIV were negative. Biopsy was decided. **Methodology:** Histological examination revealed CD19, CD20, bcl-2 and bcl-6 positive B-cell lymphoma with a Ki67 proliferation index of 95%. Myc, bcl-2, and bcl-6 gene rearrangements were not detected. Diffuse large B cell lymphoma was

diagnosed. FDG-PET/CT showed lesions in multiple regions only limited to skeletal muscles but no other organ involvement. He had no adverse risk factors but bulky lesion (11cm sized lesion). After 6 courses of R-CHOP protocol, he had complete anatomic and metabolic response. **Conclusion:** Healthy skeletal muscles do not have lymphatic system. Lymphomatous involvement of muscles occurs by 3 pathways as dissemination via the haematogenous or lymphatic pathway, extension from adjacent organs, such as the bones or lymph nodes, and de novo primary extranodal disease. Most of the histology primary skeletal lymphomas have the aggressive B-cell immunophenotype. In general, treatment is similar to nodal lymphomas. In conclusion, we aimed to contribute in experience with this rare eNHL type.

<https://doi.org/10.1016/j.htct.2021.10.1032>

### PP 23

#### A RARE CASE: POSTTRANSPLANT NK/T CELL LYMPHOMA

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**Case report:** We wanted to present our patient who was diagnosed with NK/T cell type PTLD after kidney transplantation, to contribute to the literature. Posttransplant lymphoma in NK cell phenotype (EBV unrelated) was detected in biopsy taken from the lesions that developed in mouth 11 years after kidney transplantation. It was detected as stage 1E with the examinations. As a result, early recognition of such rare cases and start treatment and reducing immunosuppressive agents are important

<https://doi.org/10.1016/j.htct.2021.10.1033>

### PP 24

#### A VERY RARE CAUSE OF DIARRHEA IN A CHEMOTHERAPY-INDUCED NEUTROPENIC PATIENT: PELLAGRA

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**Case report:** Pellagra is a systemic disease caused by a deficiency of vitamin B3. A 19-year-old male patient, who was diagnosed with Burkitt's lymphoma was admitted to the hematology clinic for the second cycle of R-CODOX-M chemotherapy treatment. The patient at risk of malnutrition developed dermatitis, diarrhea and demans during treatment. The

cause of diarrhea in the neutropenic patient is mostly in the form of infective diarrhea. Diarrhea due to vitamin deficiency should be kept in patients with malnutrition .

<https://doi.org/10.1016/j.htct.2021.10.1034>

PP 25

#### CAVITARY PRIMARY PULMONARY LYMPHOPLASMOCYTIC LYMPHOMA COMPLICATING HENOCHE–SCHÖNLEIN PURPURA

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**Introduction:** Non-Hodgkin lymphoma (NHL) may occur in the chest, often as secondary involvement but occasionally as primary disease. Low-grade pulmonary B-cell lymphoma is the most frequent form. The diagnosis based on histological examination of surgical samples. Henoch–Schönlein purpura (HSP) as a systemic vasculitis typically less commonly affects adults. Triggers including infections, medications and malignancy for HSP have been recognized. **Case report:** We report a patient presenting with HSP who had primary pulmonary lymphoplasmocytic lymphoma (PPLL) as an underlying malignancy. **Case:** 57-year-old male patient developed chest pain with a hemoglobin level 5.9g/dL. Symptoms resolved after erythrocyte transfusions. He has been diagnosed as having type 2 myocardial infarction. The detailed investigation contributed to warm autoimmune hemolytic anemia (AIHA) diagnosis. Steroid was started. He had high erythrocyte sedimentation rate. Further workup revealed bilateral multiple hilar lymphadenopathies and nodular cavitary pulmonary lesions on torax CT. The clinical picture and laboratory evaluation were not consistent with invasive fungal infection and tuberculosis. Purified protein derivative (PPD) skin test was negative. Bronchoalveolar lavage did not reveal any atypical cell and culture positivity. Thoracoscopic lymph node excision was performed. Histologic investigation showed plasma cells in the paracortical area with a slight increase in kappa to lambda ratio (3:1). A fine needle aspiration biopsy of lung tissue revealed lymphoplasmocytosis. PET-CT documented cavitary nodular lesions and hilar lymphadenomegalies but no other suspicious lesion. Biopsy sample from one lesion sized 18 × 12 mm with SUVmax 5 revealed plasma cell infiltration with an IgG kappa phenotype. PPLL was diagnosed. Meanwhile AIHA responded to steroid but recurred during dose tapering. PPLL treatment with bortezomib and rituximab based regimen was decided. AIHA went in remission but relapsed after one year with HSP associated clinical picture.

He had severe abdominal pain with intestinal wall thickness. Biopsy samples from kidney showed IgA vasculitis and from skin granular type of IgA and C3 deposition in the walls of small diameter vessels in the papillary dermis. Pulse steroid followed by cyclophosphamide controlled the clinical picture. **Conclusion:** We wished to highlight that in adults presenting with HSP may be a sign of underlying malignancy relapse.

<https://doi.org/10.1016/j.htct.2021.10.1035>

PP 26

#### ANTICARDIOLIPINIC ANTIBODIES IN NON-HODGKIN LYMPHOMA

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**Objective:** Identification of hemostasis changes in patients with non-Hodgkin’s lymphoma (NHL) and anticardiolipin antibodies (aCL). **Methodology:** The study included 83 patients (men-34, women-49) with a mean age of 63.2 years, with NHL, investigated complex, by research of lupus anticoagulant (LA) by Turbidimetry; antiβ2glycoprotein I IgG, IgM and aCL antibodies, by ELISA method. Hemostasis disorders were evaluated according to the type of NHL, stage, tumor size. **Results:** aCL were detected in 10 (12%) patients: 6 patients with aggressive type lymphoma and 4 patients with indolent type lymphoma, with advanced stage B cell NHL in 60%, mean age 52.8 years. LA was present in 80% of cases, unlike aCL IgG antibodies (10%) and antiβ2glycoprotein I IgG (10%). Hemostasis disorders were found in 6 (60%) patients: thrombotic events-at 4 (40%) patients with Mantle cell lymphoma (1 patient), Small lymphocytic lymphoma (1 patient), lymphoblastic lymphoma (2 patients). Local stage (I and II) of the lymphoma was in 75%, but with a large size of the tumor (> 11 cm), and hemorrhage at 2 (20%) patients with stage IV Small lymphocytic lymphoma, in which immune thrombocytopenia developed. **Conclusion:** The presence of antiphospholipid antibodies, in particular of lupus anticoagulant, advanced age, generalized stage, and large tumor size are risk factors for the development of hemostasis diseases in NHL patients, especially thrombosis.

<https://doi.org/10.1016/j.htct.2021.10.1036>

PP 27

#### A CASE OF STAGE 1E DIFFUSE LARGE B-CELL LYMPHOMA PRESENTED WITH KNEE INVOLVEMENT

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