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A CASE OF THALASSEMIA DIAGNOSED WITH AUTOIMMUNE HEMOLYTIC ANEMIA

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A 37-year-old female patient with a diagnosis of thalassemia major was admitted to the emergency department with complaints of fatigue, nausea, vomiting, and abdominal pain. Laboratory tests revealed elevated liver enzymes and pancytopenia, prompting her hospitalization. It was noted that the patient had not received chelation therapy for the past three months and had a history of irregular use of chelating agents. Her laboratory values were as follows: WBC: 2,460/mm^3, Neutrophils: 700/mm^3, Hemoglobin: 5.2 g/dL, MCV: 62.8 fL, Platelets: 15,000/mm^3. Due to her symptomatic presentation, the patient received cross-matched erythrocyte and platelet suspensions for transfusion. CRP was 0.8 mg/dL; coagulation and renal function tests were within normal limits. The patient had indirect hyperbilirubinemia, LDH: 984 U/L, vitamin B12: 467 pg/mL, folate: 9.53 pg/mL, and ferritin: 956 ng/mL. Both direct and indirect Coombs tests were initially negative. Tests for hepatitis markers, EBV, TORCH, and HIV were also negative. Parvovirus evaluation could not be performed. Peripheral blood smear revealed schistocytes, fragmented erythrocytes, and target cells, thrombocytopenia but no atypical cells. The patient underwent abdominal ultrasonography, which showed hepatosplenomegaly, with the spleen measuring 19 cm. Chest X-ray revealed pleural effusion, and thoracic and abdominal CT scans were planned. Thoracic CT revealed mass-like lesions in the vertebral area with unclear distinction, areas of pneumonic consolidation, and pleural effusion. Intravenous cephalosporin therapy was initiated for presumed pneumonia. ANA and anti-dsDNA tests were sent and returned negative. A PET-CT scan was planned. As the patient's cytopenias persisted despite ongoing transfusion needs, a bone marrow biopsy was performed. Bone marrow aspiration revealed increased cellularity and erythropoiesis without any abnormal findings. PET-CT demonstrated vertebral involvement attributed to extramedullary hematopoiesis; no malignant uptake was detected. Methylprednisolone was initiated at 1 mg/kg. Although platelet levels increased, anemia persisted. Repeated Coombs tests later returned strongly positive (+3) for both direct and indirect Coombs. Direct Coombs was positive for both IgG and C3. The patient had an elevated LDH (1200 U/L) and decreased haptoglobin levels. Due to steroid-refractory autoimmune hemolytic anemia, Rituximab 375 mg/week was administered for four doses, and the steroid dosage was tapered off. After two months, lab results showed WBC: 5,150/mm^3, Hemoglobin: 9.2 g/dL, Platelets: 168,000/mm^3. With a now negative direct Coombs test and a post-transfusion ferritin level of 2,322 ng/mL, chelation therapy was reinitiated. The patient, diagnosed with infection-related autoimmune hemolytic anemia, continues to receive monthly transfusions of cross-matched erythrocyte suspensions, Türkiye. In this patient with thalassemia major who developed infection-associated autoimmune hemolytic anemia, rituximab was initiated due to steroid resistance and a favorable response was achieved. Conclusion: This patient, who developed infection-associated autoimmune hemolytic anemia and was reinitiated chelation therapy, continues to receive monthly transfusions of cross-matched erythrocyte suspensions

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COLD AGGLUTININ DISEASE ASSOCIATED WITH COVID-19 INFECTION IN A PEDIATRIC PATIENT: A RARE CASE PRESENTING WITH SEVERE HEMOLYTIC ANEMIA AND LOBAR PNEUMONIA

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Objective: Cold agglutinin disease (CAD) is a form of autoimmune hemolytic anemia caused by antibodies—typically immunoglobulin M (IgM), and less frequently IgA or IgG—that target antigens on the surface of erythrocytes. Although the etiology may involve infections or immunologic disorders, most cases are idiopathic. The clinical picture results from hemolysis triggered by antibodies that become active at cold temperatures, leading to degenerative changes in the erythrocyte membrane and autoagglutination. This causes a drop in erythrocyte count and hematocrit, while MCV, MCH, and MCHC values appear markedly elevated. Peripheral blood smears often reveal erythrocyte agglutination. Here in, we present a case of cold agglutinin disease secondary to COVID-19 infection. **Case Presentation:** A 14-year-old previously healthy girl was initially treated with amoxicillin-clavulanate for upper respiratory tract infection symptoms, including fever and cough. Her symptoms worsened, and she tested positive for COVID-19 at an outside hospital. She was diagnosed with lobar pneumonia, and significant anemia noted during follow-up prompted her referral to our institution, Türkiye. Upon admission to our pediatric intensive care unit, three consecutive hemogram samples were clotted and could not be analyzed. Venous blood gas revealed hemoglobin (Hb) of 4.2 g/dL. Biochemical analyses showed LDH: 724 U/L (range 110-295 U/L), total bilirubin: 1.85 mg/dL (range 0.3-1.2 mg/dL), direct bilirubin: 0.29 mg/dL (range 0-0.2 mg/dL), and haptoglobin: 0.38 g/L (range 0.35-2.5 g/L). Direct Coombs test was negative. Peripheral smear demonstrated erythrocyte agglutination clusters. Blood samples were delivered to the laboratory in warm water immediately after collection to prevent in vitro agglutination. Repeat tests showed Hb: 8.2 g/dL, MCV: 100 fL, and a markedly elevated MCHC of 683 g/dL. Quantitative cold agglutinin testing could not be performed due to technical limitations at our center. In addition to pneumonia treatment, the patient was started on methylprednisolone at 2 mg/kg/day for presumed cold agglutinin disease. She

was discharged on day 10 of treatment and her steroid therapy was tapered and discontinued by day 21. At follow-up on day 21, the patient's hemoglobin had increased to 13.9 g/dL, and no erythrocyte agglutination was observed on peripheral smear. **Conclusion:** This case highlights a rare pediatric presentation of cold agglutinin disease associated with COVID-19 infection, complicated by severe hemolysis and lobar pneumonia. Early recognition and a multidisciplinary approach including corticosteroids and supportive care played a critical role in the patient's favorable outcome.

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CASE REPORT: WIDESPREAD BONE INVOLVEMENT AFTER ALLOGENEIC TRANSPLANTATION IN A PATIENT WITH BIPHENOTYPIC ACUTE LEUKEMIA

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Objective: Biphenotypic acute leukemia (BAL) is a rare hematologic malignancy characterized by blasts expressing both myeloid and lymphoid markers, and is generally associated with poor prognosis. The advancement of cytochemical and immunophenotypic diagnostic techniques has improved recognition of such rare leukemias, which account for approximately 5% of adult acute leukemias. Despite recent developments, challenges remain in the diagnosis and treatment of BAL. The European Group for the Immunological Characterization of Leukemias (EGIL) and the World Health Organization (WHO) scoring systems, primarily based on flow cytometry, are widely used for diagnosis. Due to disease heterogeneity, there is no standardized chemotherapy for BAL; however, because of the high relapse risk, allo-HSCT is recommended as soon as remission is achieved. Following allo-HSCT, extramedullary relapse occurs in 3–12% of acute leukemia patients. In this study, we present a case of BAL with isolated widespread bone involvement occurring after allo-HSCT. **Case report:** A 33-year-old male patient was diagnosed with B/Myeloid biphenotypic acute leukemia in January 2024. Flow cytometric evaluation showed aberrant myeloid markers, while cytogenetic analysis did not reveal FLT3-ITD, t(8;21), t(9;22), or inv(16) mutations. He received induction therapy with 3+7 Idarubicin & Cytarabine, which failed to achieve remission. FLAG-Mito reinduction therapy was administered, but bone marrow evaluation still showed 8% blasts, and the patient was considered refractory. On March 24, 2024, he underwent allo-HSCT from an HLA-matched sibling donor after a myeloablative conditioning regimen with Fludarabine and Treosulfan. Post-transplant chimerism was 96%, and remission was achieved. In December 2024, the patient presented with left knee pain. Imaging revealed a bone lesion in the proximal left tibia, and biopsy confirmed BAL relapse. Bone marrow biopsy was normal. PET-CT revealed widespread skeletal involvement, including bilateral humeri, right clavicle, right scapula, sternum, L2 vertebra, left sixth rib, sacrum, pelvic bones, right femur, and proximal bilateral tibiae. Due to severe pain, palliative radiotherapy (2000 cGy to the left tibia and 800 cGy to the left sixth rib) was administered. As there was no bone marrow involvement, the patient was started on Decitabine (20 mg/m^2/day for 5 days) combined with Venetoclax (200 mg for 14 days per cycle, reduced due to concomitant posaconazole use). After four cycles, PET-CT demonstrated complete remission. Donor lymphocyte infusions (DLI) were administered in four doses (2.42×10^7/kg total). The patient remains in remission with mild chronic GVHD (grade 1–2). **Discussion:** Biphenotypic acute leukemia is a rare subtype of acute leukemia, most commonly presenting with a B/Myeloid phenotype. High-dose chemotherapy protocols derived from ALL or AML regimens are generally used, and allo-HSCT is recommended for patients achieving remission. Extramedullary relapse after allo-HSCT has been reported with variable incidence, most often accompanied by bone marrow relapse. Isolated extramedullary relapse without marrow involvement is rare. A European multicenter study reported isolated extramedullary relapse in 0.65% of cases after allo-HSCT, while another study of 287 patients identified such relapse in approximately 4%, most frequently in the CNS, skin, bone, pelvis, and breast. In our case, the patient relapsed nine months after allo-HSCT with widespread isolated bone involvement. Treatment with hypomethylating agent Decitabine combined with Venetoclax achieved remission, and subsequent DLI helped maintain disease control. There is limited literature regarding isolated bone relapse in BAL after allo-HSCT, highlighting the uniqueness of this case. **Conclusion:** Biphenotypic acute leukemia is a rare disease with poor prognosis and no standardized therapy. Treatment approaches usually involve high-dose chemotherapy regimens for ALL or AML followed by allo-HSCT. Although extramedullary relapse after allo-HSCT is known, isolated widespread bone involvement is extremely rare. Our case demonstrates successful treatment with Decitabine and Venetoclax, followed by donor lymphocyte infusions.

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"Kappa Light-Chain Multiple Myeloma Without Serum M-Spike: A Diagnostic and Therapeutic Challenge in an Elderly Patient"

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Introduction: Light-chain multiple myeloma (LCMM) accounts for a subset of myeloma cases characterized by the absence of an M-protein spike on serum protein electrophoresis. This diagnostic challenge often delays recognition and treatment. We present the case of a 75-year-old woman with kappa-dominant LCMM, where conventional marrow and serum