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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

https://doi.org/10.1016/j.htct.2025.106171

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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