

Elderly female with Autoimmune hemolytic anemia

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ABSTRACT

Autoimmune hemolytic anemia (AIHA) is a rare disease with an estimated prevalence of around 17/100,000. It is often difficult to diagnose and treat AIHA, especially in elderly. A 60-year-old female was admitted with the complaints of low grade fever, on-off for 6 months, progressive fatigue and dyspnea on exertion. She was transfused with three units of blood within these 6 months. Examination revealed pallor, edema, hemic murmur, and palpable liver. Hb was 2.9 gm%, T Bil 5.2 mg/dl, ESR 160 mm, and reticulocyte count 44.05%. Direct Coombs test was positive, anti-nuclear antibody (ANA) and Anti ds DNA were positive. A diagnosis of systemic lupus erythematosus (SLE) with AIHA was considered and patient was transfused with two units of packed red cells and put on steroid (prednisolone) at 1 mg/kg body weight daily. After 3 weeks, her Hb had increased to 10.4 gm% with gross clinical improvement.

Key Words: Anemia, autoimmune, reticulocyte, steroid, warm antibody

INTRODUCTION

Autoimmune hemolytic anemia (AIHA) is characterised by the destruction of red blood cells (RBCs) in the presence of anti-RBC autoantibodies. It is a rare disease with an estimated prevalence of around 17/100,000.^[1] It is a relatively uncommon cause of anemia. It is often difficult to diagnose and treat AIHA. Correct diagnosis rests on a proper understanding of the pathophysiology and interpretation of blood tests. There is often a need to start therapy rapidly and also to transfuse blood which can be challenging. Anemia in elderly is either considered to be nutritional or indicative of internal malignancy, and therefore uncommon diagnoses are usually relegated for later thoughts. This report is about one such anemic patient who remained undiagnosed for several months in spite of multiple blood transfusions. This shows the low index of suspicion for AIHA in this population.

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CASE REPORT

A 60-year-old female was admitted with complaints of low grade fever, on-off for 6 months, progressive fatigability, and dyspnea on exertion. There was no history of orthopnea, paroxysmal nocturnal dyspnea (PND), chest pain, syncope, apparent bleeding, vomiting, leg swelling or joint pains. She was diagnosed as anemic and was transfused with 3 units of blood within these 6 months. She was non-hypertensive and non-diabetic. Examination revealed pallor, icterus [Figure 1], edema, normal neck veins, pulse rate 96/min, BP 110/60 mm Hg and weight 40 kgs. Chest was normal, cardiovascular system (CVS) showed hemic murmur and abdomen examination showed palpable liver. Central Nervous System (CNS) was within normal limits. Blood investigations revealed Hb 2.9 gm%, TLC 8100, platelets 1.37 lakhs, RBS 121 mg%, S. Creat 0.6 mg%, Bl. Urea 17 mg%, T Bil 5.2 mg/dl, DBil 0.6 mg/dl, IBil 4.6 mg/dl, AST 112 U, ALT 31 U, T Prot/Alb 7.1/3.1 mg/dl, ESR 160 mm, MCV 117, MCH 34.5, MCHC 29.3, LDH 452 U/L and reticulocyte count 44.05%. Peripheral smear showed anisocytosis, poikilocytosis, microcytic hypochromic RBC,

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Figure 1: Pallor and icterus visible in the patient (original Figure)

target cells, tear drop cells, microovalocytes and normal leucocytes. Serum Vitamin B12 level was 220 ng/L, S Folate level 10.3 µg/L, S. Iron 104 µg/dL, and S. Ferritin 740 ng/mL. Direct Coombs test was positive. Anti-nuclear antigen (ANA) and anti ds DNA were positive. Complement C4 was 26 CAE units. In view of fever, the antigen test for malaria was negative and widal showed insignificant titre. Stool for occult blood was negative. Urine test was normal. Ultrasonography (USG) whole abdomen showed hepatosplenomegaly and mild hydronephrosis both sides. Chest X-ray was normal. Hence a diagnosis of systemic lupus erythematosus (SLE) with AIHA was considered. Patient was transfused with two units of packed red cells. She was put on steroid (prednisolone) at 1 mg/kg body weight daily. Supportive therapy with omeprazole and paracetamol were also given. Patient showed marked improvement on steroids. The hemoglobin level increased and patient was better symptomatically. Patient was discharged on a tapering course of steroid. On OPD follow-up after three weeks, her Hb had increased to 10.4 gm% and Tbil/AST/ALT were 1.3 mg% /53 U/36 U while reticulocyte count was 12.09%.

DISCUSSION

AIHA can be classified on the basis of optimal temperature for autoantibody binding to RBC into: Warm antibody AIHA (WA-AIHA), cold antibody AIHA (CA-AIHA) or AIHA due to biphasic autoantibody (paroxysmal cold haemoglobinuria, PCH). About 10% of patients suffering from SLE develop an AIHA.^[2] WA-AIHA is a rare disease with an incidence of 1:100,000.^[3] It can be primary (idiopathic) or secondary to lymphoproliferative disease (lymphoma), autoimmune diseases (SLE) or acute leukaemia. Incidence of CA-AIHA is lower than WA-AIHA.^[3] Clinical features include pallor, fatigue, dyspnea, palpitations and jaundice. Hemoglobinuria

is rare. Proper history along with examination and laboratory investigations are essential for making the diagnosis. Laboratory features of hemolysis are indirect hyperbilirubinaemia, reticulocytosis, increased levels of lactate dehydrogenase (LDH) and decreased haptoglobin. Immunological tests which can detect the condition are indirect antiglobulin test (IAT) and the direct antiglobulin test (DAT). Therapy for WA-AIHA includes transfusion, drugs and splenectomy. Compatibility is a very important factor for blood transfusion. Steroids are often the mainstay of therapy in AIHA. Steroids decrease the production of auto-antibodies by B-cells.^[4] Prednisolone is started at 1 mg/kg/day and depending on the clinical response tapered slowly. In case of steroid failure or intolerable side effects, steroid sparing therapies like cyclophosphamide (100 mg/d) or azathioprine (100-150 mg/d) can be administered as monotherapy or in combination with steroids.^[5] Immunosuppressants such as cyclosporine or mycophenolate mofetil have also been tried and shown results.^[6] Splenectomy can help by preventing RBC destruction and the production of auto-antibodies. Other therapies like anti-C20 antibody Rituximab has been reported to achieve remission by decreasing autoantibody production by targeted destruction of B cells.^[7] Intravenous Immunoglobulin has been shown to provide temporary improvement in the disease.

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Conflicts of interest

There are no conflicts of interest.

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