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Case Report

RECURRENT SEVERE ANEMIA IN A SETTING OF AUTO IMMUNE HEMOLYSIS

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ABSTRACT
Auto-immune haemolytic anaemia (AIHA) is an acquired type of haemolytic anaemia, caused by autoantibodies directed against the red cells. We hereby report a case of chronic severe anemia, which was caused by immune hemolysis. Treatment with prednisolone resulted in significant improvement of the haematological parameters. AIHA should be kept in mind while evaluating chronic anemia.

INTRODUCTION
Auto Immune Hemolytic anemia (AIHA) is the clinical condition caused by autoantibodies, which bind to the red cell surface resulting in extravascular hemolysis. RBC destruction is mediated via the complement system or the reticuloendothelial system. Macrophages recognise F_c receptor of Ig and/or C_{3b} and phagocytise a portion of the red cell membrane, each time it passes through the spleen. The recurrent loss of cell membrane results in shortening of the red cell life span and chronic anemia.
AIHA are classified into warm AIHA, cold AIHA, mixed type and drug induced type. Warm antibody type accounts for 70% of all AIHA. It is of Ig G type and usually does not fix the complement. AIHA can be a primary disorder (50%) or secondary to lymphoproliferative diseases, other systemic autoimmune diseases, viral infections, immune deficiency states, etc. Direct Coomb's test which detects Ig and/or Complement bound to the surface of the red cell is the diagnostic test for AIHA.

CASES
A 28 years old female patient presented with complaints of weakness, breathlessness on exertion and palpitations of 15 days duration. She was previously admitted elsewhere on several occasions (within past 3-4 months) with similar complaints, and had recieved multiple blood transfusions. As patient was from low socioeconomic status and peripheral smear had shown microcytic hypochromic picture, she was treated as iron deficiency anaemia (traditional approach). There was no history of blood loss/bleeding diathesis, no history of fever, joint pain or rash, and no history of recent drug intake.
On clinical examination, patient was moderately built and nourished, with severe pallor and mild icterus. There was post Polio residual paralysis of left lower limb. There was no clubbing, Lymphadenopathy or edema. Pulse was 110/min, regular. BP was 110/70 mm Hg. Cardiovascular and respiratory system examination revealed no significant abnormality. Per abdomen examination revealed mild Splenomegaly (2 cm below left costal margin) no Hepatomegaly.
Investigations revealed Hb% of 3.3 g/dl, Total corrected leukocyte count = 8200/ccm, Platelet count = 1.8 lakhs, ESR = 80 mm/ 1st hr, reticulocyte count was 39.5 % (fig 2), reticulocyte production index of 5.4 (RI above 3 is suggestive of haemolytic anaemia). Hematocrit -19.4%, Mean corpuscular volume- 127.6 fL (↑), Mean corpuscular haemoglobin = 36.2 pg, Mean corpuscular haemoglobin concentration- 28.4g/dL, Red cell distribution width (RDW-CV) was 26% (↑). Peripheral smear showed numerous spherocytes, nucleated RBCs (200 nRBC/100WBC) and polychromasia (fig.1) indicating a haemolytic process. Serum bilirubin= 2.5 (direct=0.6), CPK= 411 U/L, LactateDeHydrogenase (LDH) = 2775 U/L (NR up to 400 U/L). Serum Haptoglobin was below 6.63 (30-200 mg/dl). All these findings pointed towards haemolytic anaemia.
Hb Quantification using HPLC (high performance liquid chromatography) was normal. Serum G6PD activity was normal. Anti nuclear antibody was negative. Direct Coomb's test (using gel card) was

positive (4+) at both 37^0 C and at room temperature (fig. 3). Indirect Coomb's test was negative. Based on these findings, a diagnosis of warm antibody type immune haemolytic anaemia was made.

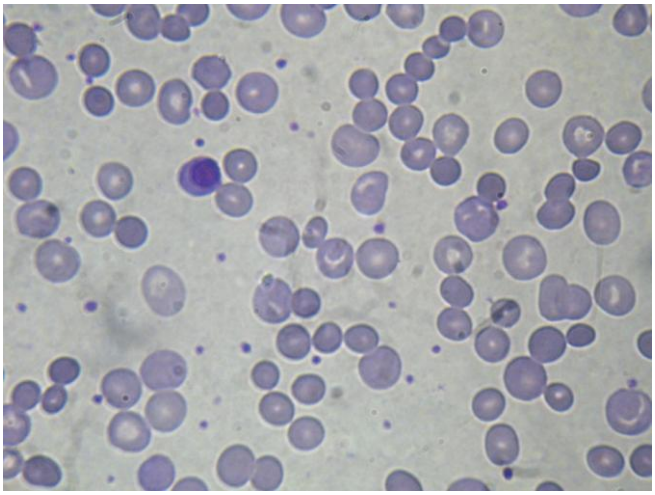

Figure 1: Peripheral smear showing spherocytes, one normoblast and polychromasia.

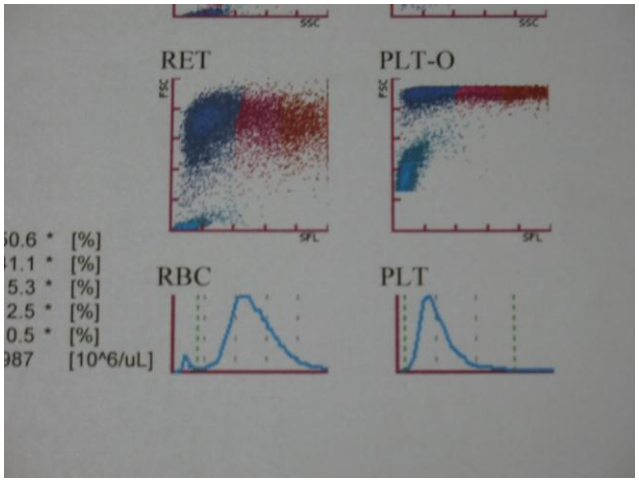

Figure 2: Hemogram showing marked reticulocytosis

Figure 3: Positive Direct Coomb's test

Blood urea was 23 mg/dl, Creatinine was 0.7 mg/dl, Ultrasonography of abdomen was normal, HIV and HBsAg were negative, and VDRL was non-reactive. IgG levels were 1540 mg/dl (NR 700-1600 mg/dl) IgM levels were 169 mg/dl (40-230 mg/dl). C_3 levels were 129 mg/dl (90-180 mg/dl). Ebstein Barr virus VCA (Viral Capsid Antigen) and EBNA (Nuclear antigen) was negative. TORCH screening (Toxoplasmosis, Rubella, Cytomegalovirus and Herpes simplex virus) was negative. A diagnosis of warm antibody type auto immune haemolytic anaemia (AIHA), probably of idiopathic type, was made and patient was started on steroid therapy. She was transfused with three units of blood.

After 1 week, repeat haemoglobin was 11 gm%. Peripheral smear showed reduction in the number of normoblasts (6-8 nucleated RBC /100 WBC).Repeat Reticulocyte count was 18%. Serum LDH level at the end of one week was 593 U/L reflecting decrease in haemolytic activity.

DISCUSSION

The course of warm antibody AIHA varies with age. In children, AIHA is usually a self limited disease, arising 1-3 weeks after a viral infection and disappearing within 1-3 months. In adults, the disease may have a variable manifestation. Severe AIHA can be a medical emergency. Red cell transfusion poses a special problem as the transfused cells are rapidly destroyed, but can be life saving and in the meantime steroids can exert their effect. This unique situation requires good liaison and understanding between the clinical unit and the serology lab. Thus it is very important to diagnose this condition and treat accordingly. Untreated, chronic auto immune hemolysis progresses to severe anaemia and associated complications. No association between past poliomyelitis and autoimmune haemolytic anaemia was found. Hence we report this case to emphasise the need to completely evaluate a patient, as it has several therapeutic implications.

REFERENCES

Bottiger LE and Werterholm B (No Year). Acquired haemolytic anaemia-Incidence and Etiology. *Acta MedIca Scandinavica* **193**(3) 223-236.

Cotran Ramzi S *et al.* (2005). Robbins and Cotran Pathologic basis of diseases. *St. Louis, Mo: Elsevier Saunders* 637.

Garratty G and Petz LD (2002). Approaches to selecting blood for transfusion to patients with auto immune haemolytic anaemia. *Transfusion* **42** 1390.

Gupta S, Szeiszen A, Nakhl F *et al.* (2011). Severe refractory auto immune haemolytic anaemia with both warm and cold auto antibodies. *Journal of Medical Case Reports* **5** 156.

Kumar P and Clark M (2005). Clinical Medicine (6^{th} Ed) Elsevier Saunders 437.

Nowak –Wegrzyn A, King KE, Shirey RS *et al.* (2001). Fatal warm auto immune haemolytic anaemia. *Journal of Pediatric Hematology Oncology* **23**(4) 250-252.

Morqensztern D, Kharfan-Dabaja MA *et al.* (2002). Warm antibody auto immune haemolytic anaemia developing after TTP. *Acta Haematologia* **108**(3) 154-156.