

## Acute Idiopathic Cold Antibody Autoimmune Hemolytic Anemia (AIHA) ---An Unusual Medical Emergency.

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Sir,

A 65 year old male presented to us with history of 2 episodes of hematemesis in 24 hours. There was history of intermittent abdominal pain after meals. Initial hemoglobin was 5 mg%. Upper GI endoscopy revealed bleeding gastric ulcer with organized clot. Conservative management was done with intravenous proton pump inhibitors. Physical examination revealed marked pallor. There was no lymphadenopathy, splenomegaly, edema, rash, petechiae or bruises. Other system examination was normal. The patient was transfusion of 2 PRBC. First transfusion was uneventful. Crossmatching for the second transfusion showed agglutination of red blood cells. Icterus developed in next 6 hours. Subsequent investigations revealed haemoglobin (Hb)—3 g/dl. TLC -5400/mm<sup>3</sup> with normal differential counts, with evidence of microcytosis (MCV—66 fl) and increase in MCH (20.9 pg) and MCHC (22.7 g/dl) also. Coagulation profile was normal. Agglutination of red blood cells was noted on peripheral smear examination with separation on warming the slides. Smear showed anisopikilocytosis with predominant microcytes, hypochromia and nucleated red blood cells. Malarial parasite was absent. Reticulocyte count

was 10%. Direct anti globulin test (DAT) was strongly positive. Urine for haemoglobin was positive. Total bilirubin was 6.2 mg%, Direct 1.2 mg%, indirect 4.0 mg%. Serum LDH 967 IU/L (normal value range is 105 - 333 IU/L). Liver function tests showed a total serum bilirubin of 5.5 mg/dl with an indirect fraction of 4.2 mg/dl and normal enzyme levels. Urine analysis was normal. A diagnosis of AIHA cold antibody type with intravascular hemolysis was made.

For severe anemia transfusion with packed red cells was again planned. Group specific packed red cells which were 'least incompatible' was transfused. Auto agglutination of red blood cells at room temperature with reversal on warming the slide with a strongly positive DAT with evidence of hemolysis confirmed the diagnosis as AIHA due to cold auto antibodies. Serum glucose levels and thyroid function tests were normal. Bone marrow aspiration showed reactive erythroid hyperplasia and there were no abnormal cells. Serology for Human immunodeficiency virus, Hepatitis A, B, C viruses and Mycoplasma pneumonia were negative. Rheumatoid factors, anti nuclear antibody and lupus cells were negative.

## DISCUSSION

Autoimmune hemolytic anemia (AIHA) occurs when a patient produces pathologic antibodies that attach to and lead to the destruction (hemolysis) of their red blood cells (RBCs) with consequent anaemia. AIHA is the most common form of acquired hemolytic anemia. Autoimmune haemolytic anemia (AIHA) is a group of acquired haemolytic anemias resulting from development of auto antibodies directed against antigens on the surface of patient's own red blood cells [1]. AIHA are of 2 types based on temperatures at which auto antibodies react with red blood cells. Warm antibody AIHA occurs predominantly in children. They belong to IgG class, react at temperatures  $\geq 37^{\circ}\text{C}$ , do not require complement for activity and do not produce agglutination in vitro. In cold antibody AIHA antibodies are of IgM class which react at temperatures  $< 37^{\circ}\text{C}$ , require complement for activity and produce spontaneous agglutination of red blood cells in vitro [2]. In most cases, CAHA is a primary disorder that typically becomes apparent at 50 to 60 years of age.

The onset of AIHA is very often abrupt and can be dramatic like in our case. The hemoglobin level can drop dramatically. The massive red cell removal will produce jaundice. Intravascular hemolysis further manifests with increased free hemoglobinemia, increased hemoglobinuria, hemosiderinuria. Typically, intravascular hemolysis is rapid and aggressive.

Severe acute AIHA can be a medical emergency. The immediate treatment includes transfusion of red cells. But if the antibody involved is unspecific, all the blood units cross-matched will usually become incompatible. In these cases it is rationale to transfuse incompatible blood, the explanation for this ironic treatment is that the transfused red cells will be destroyed no less but no more than the patient's own red cells, and the patient remains alive. [3,4]

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