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# Treatment approach to a patient with Autoimmune Hemolytic Anemia (AIHA) in a teritiary care hospital - A Case Report

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## ABSTRACT

Autoimmune hemolytic anemia is a rare autoimmune disease in which autoantibodies are formed inside the body which target red blood cells leading to marked decrease in their life span. The auto-antibodies formed will bind to the circulating erythrocytes, leading to hemolysis and decreased survival of red blood cells. Since autoimmune diseases are rare and life threatening, the understanding about the disease and its management have an immense role for the public and health care system. Here we report a 45 year old female who admitted with the complaints of fever and headache in a private hospital. Her lab findings showed reduced levels of hemoglobin (severe anemia) and tested positive for both Direct Coomb's test and Indirect Coomb's test and was diagnosed as Autoimmune hemolytic anemia. She was managed with repeated blood transfusions, corticosteroids, antibiotics and antihistamines. She was discharged in a stable condition.

## INTRODUCTION

Autoimmune hemolytic anemia is a rare group of disorder that can occur when your immune system mistakes red blood cells as unwanted substances. These disorders affect women more often than men. Mostly the cause of autoimmune hemolytic anemia cannot be determined (idiopathic autoimmune hemolytic anemia). AIHA can also occur with another disorder, such as systemic lupus erythematosus or a lymphoma or it can be as a result of use of certain drugs like penicillin.[1,2]

As per the available case reports, the prevalence of AIHAs is estimated as 1 to 3 per 100000. It is usually reported in middle age adult patients. It is rare, affecting approximately 1 to 2 out of every 100000 people each year and only a few cases of AIHAs have been reported. The sudden onset, seriousness and unpredictable nature of the disease play a major role in its management.[3] More recently, AIHA has been identified as a greatly heterogeneous disease, due to several immunological mechanisms involved beyond antibodies, complement and antibody-dependent cell mediated cytotoxicity.[4, 5] Previously, steroids, immuno suppressant, and splenectomy were the treatment approaches of AIHA.

More recently, several new targeted therapies are increasingly used in the clinical practice. In most of the cases, the first line treatment for AIHA is corticosteroid therapy which is effective in 70-85% of patients that should be tapered over a time period of 6-12 months along with antibiotics, and blood transfusions.[5, 6] Here in this case also the patient had received standard care with corticosteroids, antibiotics and also repeated blood transfusions.

## CASE REPORT

A 45 year old female patient was admitted with complaints of fever on and off for 7 days with headache and nausea and also had past medical history of Type 2 Diabetes mellitus for last one year (on Inj HUMAN ACTRAPID-20Units) and Hypothyroidism for 22 years (on THYROXINE 125mcg). Upon general examination, the patient was sick, febrile (100° F) along with pallor. Her laboratory investigations showed a declined level of hemoglobin (4.5 g/dL), sodium (130mmol/L) and elevated levels of ESR (145mm/hr), CRP (8.2mg/L), total bilirubin (4.64 mg/dL), indirect bilirubin (4.49 mg/dL), SGOT (55 U/L) and iron (235 ug/dL).

Since the hemoglobin levels were not normalizing, there was a consultation by the physician and hence advised to carry out the Coomb's test. The test was performed and she was tested positive for both Direct Coombs Test (DCT) and Indirect Coombs test (ICT). Also there was a declined level of complement C3 (79.0mg/dL), increased level of LDH (1210U/L) and transferrin saturation (81%) and a borderline Anti-mitochondrial M2 antibody (AMA-M2). The patient was also tested positive for Dengue IgG (68.75), but Dengue IgM, Rapid malarial test and Widal test showed negative results.

Upon Peripheral smear test, it was found that the RBC showed agglutination and were predominantly normocytic normochromic cells. Many polychromatophilic cells, few spherocytes and nucleated RBCs were seen. Also platelet count was normal and was seen scattered singly and in clumps. Thus the test revealed a suggestive of Hemolytic anemia, possibly immune hemolytic anemia. USG Abdomen and pelvis showed enlarged spleen of about 13cms (Hepatosplenomegaly). Based on these subjective and objective data, the condition was diagnosed as Hemolytic Anemia (Autoimmune).

The patient was treated with the corticosteroid; Inj METHYLPREDNISOLONE in 100ml Normal Saline for 3 days and antibiotics; Inj AZITHROMYCIN 1g OD and Inj DOXYCYCLINE 100mg BD. Since the patient couldn't tolerate Azithromycin, it was changed to Inj PIPERACILLIN TAZOBACTAM 4.5g Q6H. The patient had the complaints of vomiting which were treated with Inj ONDANSETRON 4mg BD. Elevated body temperature was managed by using tablet PARACETAMOL 650mg thrice daily. She was also given protein supplements since she was too weak. Besides these medications, the physician advised to give washed RBCs since the cross matching sample was incompatible and thus packed Red Blood Cells (PRBC) were transfused for seven days.

Upon the last day in hospital, the patient's hemoglobin levels reached up to 9.2 g/dL from 4.5 g/dL. The patient was clinically stable. She was discharged with GLICLAZIDE, combination of DAPAGLIFLOZIN and METFORMIN, THYROXINE, FERROUS ASCORBATE and FOLIC ACID TABLETS with the recommendation of review after 10 days with LFT and Hb reports.

## DISCUSSION

Autoimmune hemolytic anemia (AIHA) is mostly caused by the defects in red blood cells associated with the defects in membrane, red cell enzyme deficiencies or any abnormalities in hemoglobin. These are due to the production of autoantibodies which acts upon the surface of the red blood cells.[9,10]

AIHA is diagnosed based on the occurrence of hemolytic anemia, jaundice, reticulocytosis, increased serum bilirubin levels, splenomegaly and also based on a positive result of Direct antiglobulin test[11]. As per the standard guidelines, Corticosteroids are the first line treatment. Corticosteroids, usually prednisone is given until hemoglobin levels reach 10g/dL in which the response can be seen during the second week of treatment[7, 8]. After the hemoglobin levels are stabilized, prednisone should be slowly and gradually tapered. Other options in the management of AIHA include Rituximab which has shown effectiveness in both idiopathic and secondary AIHA.[12] Immunosuppressive agents can also be considered in these cases. Those who are not responding to the corticosteroid therapy can be treated by performing splenectomy which has short time efficacy and a good initial response rate.

**Table 1 :** Units of PRBCs transfused

| DAYS | UNITS OF WASHED PRBCs TRANSFUSED |
|---|---|
| Day 1 | 1U PRBC transfused |
| Day 2 | 1U PRBC transfused |
| Day 3 | 2U PRBC transfused |
| Day 4 | 2U PRBC transfused |
| Day 5 | 2U PRBC transfused |
| Day 6 | 2U PRBC transfused |
| Day 7 | 2U PRBC transfused |

A case report by Fransisca Kristina Elisabet[1] presents a case of AIHA where a 50 year old woman came to hospital with the main complaint of weakness for 3 days. She has also the complaints of frequent join pain, headaches, lost appetite and abdominal pain. The patient was treated with blood transfusions. Her LDH profile was standard, but the ANA profile test was positive. She had been given ringer lactate 2000cc per 24 hours, methylprednisolone, ranitidine and folic acid tablets. After seven days of being hospitalized, she was discharged because of getting better and the levels of hemoglobin had reached 12.5 g/dL. Another report entitled Immune Hemolytic Anemia: A Report of two cases by Paramjit Kaur et.al[2] discusses two cases- the first case was a 20 year old female referred to hospital with complaints of icterus and breathlessness. She had mild hepatosplenomegaly. There was mild leukocytosis and film showed auto agglutination with the presence of nucleated red cells. She was treated with steroid therapy, antibiotics and diuretics. The second case was a 57 year old male presented with chest pain and breathlessness. Peripheral blood smear showed dimorphic blood picture with moderate anisocytosis and poikilocytosis with mild hypochromia, microcytes and Direct Antiglobulin Test was negative. Blood transfusions were done and also were treated with hematinics, diuretics.

In these cases presented above, the patients are managed with corticosteroids, antibiotics along with blood transfusions. Our patient presented with complaints of fever for 7 days on and off and further investigations showed declined hemoglobin, positive DCT and ICT test, low complement C3, and hepatosplenomegaly. The findings which favor the diagnosis of AIHA are low hemoglobin levels, reduced complement C3, positive DCT and ICT tests, USG Abdomen showing hepatosplenomegaly and hematology with a suggestive of hemolytic anemia (immune). The patient was treated with steroids and also done blood transfusions with washed RBCs and became stable.

In our case, the patient was effectively treated according to the standard treatment guidelines with corticosteroids, antibiotics, antihistamines along with repeated transfusion of packed red blood cells (PRBCs) which helped the patient to stabilize hemoglobin levels and became clinically stable. Since autoimmune diseases are rare and life threatening, it is important to take special attention and care by the health care providers for treating the patients and also it is very important to take an appropriate critical approach in evaluating risks and harms.

## CONCLUSION

Autoimmune hemolytic anemia (AIHA) is an acquired immune disorder characterized by production of antibodies that bind to surface of circulating erythrocytes leading to hemolysis and shortened red blood cells life span with removal of reticuloendothelial system. Normally AIHA is diagnosed through direct and indirect coombs test, low complement C3 and low hemoglobin levels. Corticosteroids, blood transfusions can be done to prevent the worsening of the condition.

## CONFLICT OF INTEREST

The author declares no conflict of interest.

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