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Blood Transfusion in Autoimmune Hemolytic Anemia— A Practical Problem

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Autoimmune hemolytic anemia (AIHA) is characterised by pallor, icterus, hepatosplenomegaly and a positive

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Received for publication: February 22, 1991;

Accepted: December 8, 1992

Coombs' test. The overall incidence of the condition is 1 : 80,000(1), but the incidence in children is much lower since the majority of patients reported were over 40 years of age(2). We present a case of AIHA and highlight the problems faced while transfusing blood in this patient.

Case Report

An 11-year-old girl was admitted with progressive pallor for 3 months and breathlessness for 15 days. There was no history of fever, anorexia, drug intake or injections, recent blood transfusion, joint pains or urinary complaints. Examination revealed pallor, icterus and signs of congestive heart failure with hepatomegaly of 5 cm and splenomegaly of 3 cm. There was no lymphadenopathy or petechial spots.

Investigations revealed a hemoglobin level of 3 g/dl, reticulocyte count of 45%, normal total and differential leucocyte and platelet counts. Peripheral smear was suggestive of severe hemolytic anemia, direct Coombs' test was positive, hemoglobin electrophoresis was normal and tests for G-6PD deficiency were negative. The serum bilirubin level was 3 mg/dl predominantly conjugated with normal levels of liver enzymes, immunoglobulins and complement. Antinuclear factor was negative and chest X-ray showed cardiomegaly. A diagnosis of idiopathic AIHA was made. The patient received three blood transfusions of 'O' Rh positive blood and prednisolone in the dosage of 2 mg/kg/day. Follow-up revealed rising hemoglobin levels and steroids were tapered to 5 mg/day when hemoglobin reached 10 g/dl.

Four years later, the child was again admitted with pallor and breathlessness. Examination showed pallor and features of congestive cardiac failure. She had lost her previous records and stopped steroids for

the past 6 months. Investigations revealed a hemoglobin of 5 g/dl, reticulocyte count 18% and positive direct Coombs' test. Peripheral smear showed evidence of hemolysis and antinuclear factor was negative. Packed cell transfusion after grouping and cross matching, was instituted with group 'AB' Rh positive blood, but resulted in mismatched transfusion with hemoglobinuria. The patient's previous admission records were traced and revealed that she was typed as group 'O' Rh positive previously. A packed cell transfusion with group 'O' Rh positive blood given slowly with strict monitoring was uneventful and subsequently two more low volume transfusions were given. She also received prednisolone 2 mg/kg/day and folic acid 5 mg/day. The child has been on regular follow-up for the past 1 year, with hemoglobin level maintained at 12 g/dl and reticulocyte count 1%, on alternate day dosage of 5 mg prednisolone.

Discussion

AIHA is rarely reported in children(2) and most are of the idiopathic variety(3). Abhyankar *et al.*(4) described 5 cases of AIHA in Indian children. None of these cases had any complications following blood transfusion.

Blood transfusion is required most frequently in the initial phase of therapy of AIHA. It should be reserved for cases in whom anemia is symptomatic(5). Unnecessary transfusion should be avoided since there is poor red cell survival, risk of severe transfusion reactions, and the possibility of sensitising the patient to red cell antigens present on donor erythrocytes.

Obtaining truly compatible blood for transfusion in AIHA may present major problems. There is risk of mistyping the patient's red cells because the bound IgG

may "block" specific typing sera or may lead to agglutination in high colloid media(6). In almost all cases of warm antibody hemolytic determinant that is basic to the Rh complex(7), or a complex that includes components not defined by specific Rh allo-antibodies used in red cell typing(8). The antibodies react against all red cells bearing the Rh antigen and is negative only to Rh null red cells. In most cases compatible blood cannot be obtained for transfusion and the "least incompatible" red cells are used for transfusion therapy, determined empirically by cross matching samples of blood from different donors against the patient's serum and selecting that which has the least *in vitro* activity. Recently, a mononuclear phagocytic assay has been described, that may be useful in distinguishing clinically significant red cell allo-antibodies(9). Rosenfield and Jagthambal(6), have noted that red cell survival in the sensitized individual is described by an exponential curve.

The rate of destruction is proportional to the total cells presented at any given time. The greater the volume of transfusate, the greater the absolute number of erythrocytes destroyed. Therefore, small periodic transfusions of 50 ml/m² of packed red cells per transfusion are recommended.

In the uncommon instances in which the patient has immune hemolysis mediated by cold agglutinin or biphasic antibody, a controlled temperature warmer for administering blood at 37°C must be used to prevent severe hemolytic reactions(10).

In conclusion, a diagnosis of AIHA should be considered in all patients with hemolytic anemia and transfusion therapy should be given carefully and only when indicated.

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NOTES AND NEWS

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 New Delhi 110 029.

or **Miss Madhumita Roy Choudhury,**
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