Journal of
Medical Pharmaceutical
And Allied Sciences

www.jmpas.com
ISSN NO. 2320 - 7418

CASE REPORT

A RARE CASE OF THALASSEMIA INTERMEDIA WITH MULTIPLE ALLOANTIBODIES

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Keywords
Alloimmunization, phenotyping, adsorption, elution

Received
06 December 2016
Reviewed
08 December 2016
Accepted
14 December 2016

ABSTRACT

Presence of anti red cell antibodies remains a major problem in Thalassemia patients. Beta-Thalassemia major patients do commonly suffer from alloimmunization which is rarely seen in Thalassemia intermedia patients. Association of multiple antibodies and antibody against high frequency blood group antigen further complicates the transfusion therapy. Advance immunohematological tests like adsorption, elution, and phenotyping are necessary along with antibody typing and identification. The present case is a 24 year male Thalassemia intermedia patient, whose blood sample when cross matched found incompatible with more than 100 units of PRBC. Various tests like DCT, ICT, 3 cell panel, 4 cell panel, 10, 11, 16 cell panels, adsorption, elution, minor phenotyping were done to resolve the case. We found multiple alloantibodies with no autoantibody. The present case emphasises on importance of minor phenotyping before first transfusion in all multitransfused patients, and importance of immunohematological tests in resolving blood incompatibility.

INTRODUCTION

Thalassemia is a congenital genetic disorder. It is an autosomal recessive disorder which affect more than 4,00,000 newborns per year worldwide.1 It has high frequency extending from the Mediterranean basin through the Middle East (Iran), India and Southeast Asia.2 In India, the carrier rate of Beta Thalassemia varies from 3-17%.3 This disease is treated lifelong with red blood cell (RBC) transfusion4 and iron chelation therapy. Development of alloimmunisation to minor blood group antigens causes difficulties in treatment. Prevalence of alloimmunisation in Thalassemia patients has been reported to be 5 - 30% in the world, which is mostly contributed by the alloimmunization to minor blood group antigens.5 Among Asians, the incidence of red cell alloimmunization, is 22%.5 Development of multiple alloantibodies further complicates the transfusion therapy. Present case is a case of Thalassemia intermedia requiring blood infrequently whose transfusion therapy became complicated due to presence of multiple alloantibodies including antibody against high frequency blood group antigen

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CASE REPORT

A 24 year male, known case of beta Thalassemia intermedia presented to The Department of Transfusion Medicine SCB medical college Cuttack for compatible blood for transfusion. Current haemoglobin of the patient was 6.1 gm/dl. He was a diagnosed case of Thalassemia intermedia. Blood transfusion requirement were less frequent, first transfusion taken when he was 8 years old. Initial requirement were once a year, further increase to one unit transfusion every 6 to 8 months. In last 16 years he had around 25 blood transfusions. Most of the transfusions were whole blood, non leucoreduced.

In last 6 months his blood requirement increased and he required around four blood transfusions. There was ongoing haemolysis; haemoglobin was not increasing with blood transfusion. He had jaundice; total bilirubin was raised to 5.6 mg/dl.

Blood sample was received for grouping and cross matching. Cell grouping showed no agglutination with anti sera A & B. There was 4+ reaction with anti D. Serum grouping showed agglutination with A, B, O, cells. Blood group determined to be ‘O’ positive. With anti H there was 4+ agglutination. This ruled out Bombay blood group. Crossmatching performed with many

units of ‘O’ positive blood but all showed incompatible at saline phase (Immediate spin method), 370temperature and at AHG Phase. Further immunohematological work up was done with patient blood. Direct Antiglobulin Test (DAT) showed negative, autocontrol negative and Indirect Antiglobulin Test (IAT) showed positive, 3+ reaction.

Anti body screening was performed with 3 cell panel, Orthodiagnostic column agglutination test showed pan positive. With 11 cell panel also pan positivity noted.

Blood sample was sent to reference lab. The sample was tested with IMMUCOR’s 4 cell screen panel, 10 cell panel and 16 cell panel. Phenotyping was done for patient’s cell. We found that the patient had R2R2 (DccEE), Kell negative, ‘M’ negative, ‘S’ negative ‘N’ positive, ‘s’ positive, ‘Jka negative, Jkb positive, fya positive, fyb negative.

Antibody identification was done and found out to be antibody reactive at wide thermal amplitude (37 degree C to 22 degree Celsius) with underlying alloantibody. Adsorption / elution showed underlying alloantibody “anti - e” detected by PEG adsorption. ‘M’ anti body also detected in the patient plasma. This ‘M’ antibody was clinically significant as it was detected at 370. Final impression was made that, the patient had alloimmunisation to multiple antibody.

Patient was advised for “R2R2 Blood (e-antigen negative i.e. DccEE), M negative AHG cross matched pack cell under very close observation. Since R2R2 blood group is rare phenotype it was very difficult to get such donor. We screened seven hundred units of to get a ‘O’ positive R2R2 blood unit.

M-antigen negative, R2R2 PRBC of same blood group was chosen for the patient and was transfused. Transfusion was uneventful, and patient improved subsequently.

DISCUSSION:

Prevention and treatment support of Thalassemia patient with alloantibody remains a challenge always. Alloimmunization may develop due to the RBC antigenic difference between the blood donor and the recipient, and immune status of recipient. There can be a variable degree of disparity amongst the donors and the recipient as far as minor blood group antigens are concerned, which are not tested for routine transfusions. Therefore alloimmunization can take place during the transfusion therapy. Due to above risk of alloimmunization, patients receiving

multiple transfusions should be typed for clinically important blood group antigens including ABO, Rh, Kell, Kidd and Duffy systems, etc.6 Percentage of alloimmunization in thalassemia patient was reported from Taiwan (37%), 4 Arab (30%).7

Situations become more difficult when multiple alloantibodies are associated. Such cases are suspected when a compatible match is not found during crossmatching. These are ideal cases when advance immunohematological test are required like adsorption, elution along with antibody screening and identification. Phenotyping also became essential in most cases. Regular transfusion of extended red cell phenotype matched blood is a very effective regimen for such patients.8

The overall frequency of 'e' antigen is higher than 'E' in all population 9, 10. In English population the frequency of e is 98% and E is 29%.11 Overally 'e' antigen is considered as high frequency antigen. R2R2 (DccEE) Blood group incidence is very low in all populations. The R2R2 phenotypes of Indian donors as per Makroo R.N. et al and Garg et al was found to be 0.8.,12,13 when an antibody developed against these antigen it is very difficult to get a matched blood. Maintenance of rare blood group registry will help to overcome these difficulties.

Antigen negative blood units must be transfused whenever any clinical significant antibody is present in the patient. Here we detected two clinical significant antibodies ie Anti 'e' and Anti 'M'. Rh extended phenotyping helps to resolve these cases but not a replacement for antigen negative blood.

Considering cost effectiveness, Minor blood group phenotyping is to be done once a life time. It helps always in resolving alloimmunised multi transfused patients. There by it increase transfusion interval, decreases transfusion burden thus saves money, resource and manpower.

CONCLUSION

Alloimmunisation is not an uncommon problem faced in Thalassemia patient and become complicated when associated with multiple alloantibody and antibody against high frequency blood group antigen. Any grouping discrepancy or incompatible cross matching report in these patients should be taken seriously. Blood sample in all these case should be subjected to advance immunohematological tests like phenotyping, elution, and adsorption for alloantibody specificity. Alloimmunization

preventive measures like AHG crossmatch blood, leucoreduction, pack cell RBC should be a routine protocol. Compulsory implementation of these protocols will decrease alloimmunization in thalassemics and improve their life longevity.

DECLARATION

- Informed consent was obtained from the patient in the study.
- All the author declares that they have no conflict of interest.
- No fund received for the research work undertaken in the above study.

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