INTRODUCTION

To date thirty-three different blood group systems (including the ABO and Rh systems) have so far been recognized by the International Society of Blood Transfusion (ISBT)4. The commonest is ABO followed by the Rhesus (D) system. Rare blood groups are also present in a very small proportion of population and so are occasionally encountered in clinical practice2. To name a few of these rare groups are MNS, P, Lutheran, Kell, Lewis, Duffy, Kidd, Diego, Colton, Cromer, Dombroch and Gerbich3.

We report a case of a patient whose scheduled open heart surgery had to be deferred because of cross match failure. All the available techniques in a modern well equipped blood bank failed to detect the culprit antigen responsible for it. The specific antigen was subsequently diagnosed from a European institute. The patient was operated at a later date and is now leading a healthy life.

CASE REPORT

A 15 year old female patient was booked for open heart surgery for closure of Atrial Septal Defect at Armed Forces Institute of Cardiology-National Institute of Heart Diseases, Rawalpindi (AFIC-NIHD). Patient’s haemoglobin was 9 grams/dl and four units of O Rh positive packed red cells were requested for surgery. Blood bank staff reported that the donor red cells were incompatible by standard test tube cross match (Saline phase, LISS phase and Coomb’s phase). Further work up with direct anti-globulin test (DAT) was negative, which was done by using poly specific anti-globulin (AntiIgG and C3d) along with negative auto control. Antibody screening procedure with commercially available 11 cell panel (BIO-RAD Diamond GmbH) could not identify the antibody responsible for incompatible cross match. The surgery was deferred.

The first issue before the surgical and haematology team was the decision of sending the patient’s blood sample for detection of rare blood group antigens to a European centre of excellence. Finally the blood sample was sent to the National Heart, Lung and Blood Institute (NHLBI), England. After a month, NHLBI informed that the patient had rare adult antigen on RBC’s and I alloantibody in its serum at 4-C. This antibody reacts with all (or nearly all) adult red cells and is called cold agglutinating auto antibody (CAA). NHLBI also offered to ship the desired cross match blood units. The cost of one unit was approximately PKR 100,000 and the patient needed 3 units. The cumulative cost was too much to bear by the ill affording parents.

Another problem hovering over the treating team was the fact that during open heart surgery, patients are cooled down to 32°C for cardiac protection. This patient had a high chance that during cooling, her own blood might agglutinate resulting into a fatal outcome.

After intense deliberation with anaesthesiologists and perfusionists, a plan for “not cooling” during the cardio pulmonary bypass was made and rather other local methods of cardio-protection like local cold saline bathing of heart were agreed upon. Meanwhile, haematology team came up with the novel idea of collecting the parents’ blood sample and cross matching it with their daughter’s blood. Luckily, her father’s blood did. Father was put on erythropoietin for stimulating erythropoiesis and he was asked...
to donate 1 unit of blood per week for 3 weeks. At the end of the third week, we had successfully collected 3 units of blood with no ill effect on father. Surgery was planned for the next day. The patient was “not cooled” during cardio pulmonary bypass and the team was extra cautious in achieving haemostasis. So no blood transfusion was required during or after surgery.

The patient was discharged and went home on 5th post-operative day of a successful surgery. Her follow up was unremarkable and now she is living a normal and a healthy life. This was a unique case, the first case of its kind in Pakistani medical literature in which a normothermic cardiac surgery was done at AFIC/NIHD Rawalpindi to avoid CAA reaction.

**DISCUSSION**

Cold agglutinating autoantibodies (CAA) are usually of IgM-class. They may be acute or chronic, benign or pathogenic. Specificity of CAA is most often for the li antigen system. Expression of the I and i antigens changes with age. Newborns express i antigen on their red blood cells, while children and adults primarily express I antigen4. Low titers (≤256) of CAA are common in the general population and have not been implicated in disease. However, higher titers (≥512) of CAA are more likely to cause haemolysis.

For detection of CAA, modified blood testing should be performed4. Blood samples having suspicion of CAA should be collected and maintained at 37°C until the plasma and RBCs are separated. Cold agglutinins react with human red cells but they differ in the strength of the reaction with different cells. Most cold agglutinins react more strongly with red cells of adults donors as compared to those of newborns or infants. These antibodies are also called anti-I and the antigen is known as the I-antigen5,6.

**CONCLUSION**

This case taught us the value of brainstorming in order to find solutions of medical problems. Asking experts when in doubt opened up hitherto untapped therapeutic and diagnostic vistas for us.

**CONFLICT OF INTEREST**

The authors of this study reported no conflict of interest.

**REFERENCES**