Case Report

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Transfusion support in a severe autoimmune hemolytic anemia patient associated with systemic lupus erythematosus and antiphospholipid syndrome

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Abstract:

A positive direct antiglobulin test is a criterion for the diagnosis of systemic lupus erythematosus (SLE). In general, severe hemolysis is absent in SLE. Sometimes, these patients may show hemolysis when presenting with antiphospholipid syndrome (APS). It is essential to exclude an underlying alloantibody along with autoantibody. We had reported a case of a 24-year-old female SLE along with an APS patient requiring transfusion support with underlying allo-anti-S antibody. We provided two units of S antigen-negative best-matched units to the patient who tolerated it well and showed improvement.

Keywords:

Alloantibody, antibody screening, best-matched blood, blood grouping discrepancy

Introduction

Antiphospholipid syndrome (APS) is an autoimmune condition characterized by thrombosis and pregnancy loss[1] and associated with systemic lupus erythematosus (SLE) in about half the cases.[2] Autoantibody generally generated against self-red blood cell (RBC) antigens. In contrast, alloantibody formed due to exposure to foreign RBC antigens. Direct antiglobulin test (DAT) is generally positive without hemolysis in SLE and also attached with APS.[3,4]

Autoantibody reacts with most of the donor RBCs, making it almost impossible to find out compatible units. In that case, first, we have to rule out alloantibody and second find out best-matched blood units for transfusion. In general, blood requirement is rare in SLE patients. However, there may meet a blood requirement if hemoglobin (Hb) value less due to severe hemolysis making oxygen craving to patients. Here, we presented a case report of a 24-year-old female, a known SLE patient, and APS, who required transfusion due to severe hemolysis. The case report was written after obtaining appropriate consent from the patient and her guardian.

Case Report

A 24-year-old female came to the emergency with a history of fever in the last 3–4 days, nausea, and generalized weakness. She was diagnosed with autoimmune hemolytic anemia (AIHA) 1 year back and had treatment with prednisolone 30 mg. Her vitals were recorded as temperature – 104.6°F, blood pressure – 100/60 mmHg, and pulse – 146 bpm at the time of admission.

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The patient had allegedly had 3–4 transfusions over few months and several vague miscarriage histories a year back. Furthermore, the patient had been taking prednisolone irregularly and had stopped it completely 1 month ago. The patient's initial investigations came as Hb 2.9 g/dl, total leukocyte count 11490/cumm, total platelet count 1.62 lakh/cumm, serum creatinine 1.59 mg/dl, serum urea 56 mg/dl, and lactate dehydrogenase 1124 IU/L. She was found to be positive for antinuclear antibody and antiphospholipid antibodies.

The patient was admitted to the isolation ward, started on Inj Methyl prednisolone 1 g iv, and blood requisition was sent to the department of transfusion medicine and blood center for emergency release of two units of packed RBC (PRBC). Blood grouping and Rh typing were initially done with solid-phase red cell adherence assay (Neo Blood Bank Analyzer, Immucor Inc., Georgia, US). There was a discrepancy in the blood grouping, and hence, it was repeated by the gel card method (Column Agglutination Technology, Tulip Diagnostics Ltd., Goa, India). The blood group was still not confirmed due to the interference of cold autoantibody, as shown in Figure 1. Using the prewarming technique and warm saline washing of patient red cells, the blood group was confirmed as O Rh D positive using conventional test tube technique (CTT). Crossmatching was done with gel card and showed incompatibility. Polyspecific DAT showed 4 + agglutination reactions, and antibody screening with red reagent cells shows pan-positivity with different reaction strengths in gel card [Figure 2]. Both monospecific IgG and C3d were performed by gel card (ID "Coombs Anti-IgG, C3d" Direct and Indirect Antiglobulin Testing, DiaMed, Cressier, Switzerland), and were 4 + reactions. Antibody identification was conducted with 11 cell panels (ID-DiaCell Panel, DiaMed, Cressier, Switzerland) which showed 2 + reactions in panels 1, 2, 4, 6, and 11, while the rest of the panels showed a 4 + reaction [Figure 3]. At this point, we suspected alloantibody might be present due to different strengths of reaction in both screening and identification panels. Hence, allogeneic adsorption was done with R1R1, R2R2, and rr PRBCs by the CTT method. After complete adsorption, antibody screening and identification were performed. We found anti-S alloantibody, and two units of S antigen-negative "O" Rh (D)-positive best-matched PRBCs were transfused. Intravenous methylprednisolone was also given to the patient. The patient was responded well to the transfusion, and posttransfusion Hb came up to 7 g/dL.

Discussion

A DAT-positive reaction points to the presence of IgG and/or complement attached to the red cells. It can be positive in several cases, but in our case, it was mainly due to the presence of an autoimmune disease (SLE). It has also been seen to show a cross-reaction of antiphospholipid antibodies with phospholipid epitopes from the red cell membrane, which gives a positive DAT reaction in patients with APS.[5]

Hemolysis of red cells occurs when immunoglobulin and/or complement are attached to the RBCs.[6] In

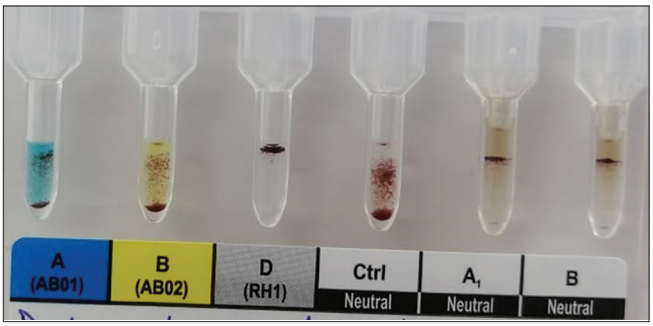

Figure 1: Blood grouping and Rh D typing showing miss match

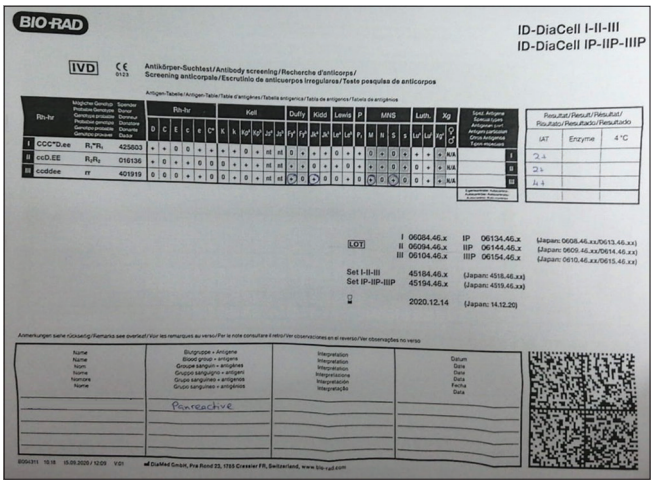

Figure 2: Antibody screening with reagent red cell shows pan positive with different strengths of reaction

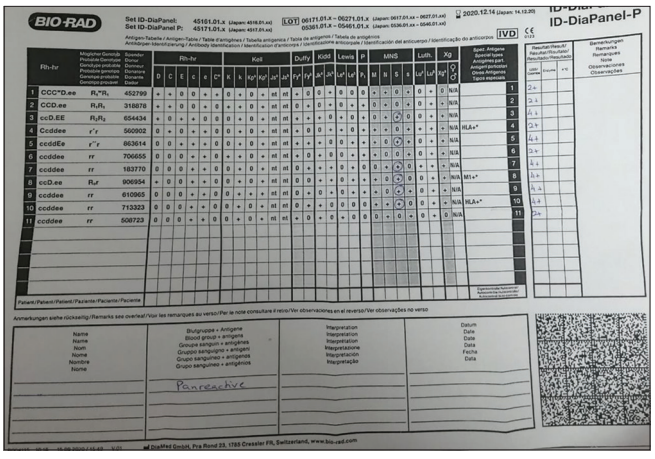

Figure 3: Antibody identification result

our case, the hemolysis of the red cells was due to the presence of both IgG and complement.

Blood grouping includes both forward and reverse grouping methods as each method serves as a check on each other. Blood group discrepancy occurs when both methods do not match with each other. On occasions, the reason for this discrepancy in blood grouping and Rh typing may be due to autoantibodies present in the blood. This may lead to mismatch during crossmatching, which may lead to the transfusion of incorrect ABO blood group. This may result in transfusion reactions in the patient. Hence, finding the correct blood group is of utmost importance.

In our case, we used the prewarming technique,[7] which helped us resolve the blood grouping discrepancy, and the blood group was found to be "O" Rh (D) positive.

In AIHA cases, the immune system is abnormal, resulting in autoantibodies directed against the patient's red cells, which leads to anemia. This warrants the help of transfusion to correct the anemia. Due to frequent transfusions, the patient develops alloantibodies which further complicate subsequent transfusion therapy. Of the two, the autoantibodies interfere with pretransfusion testing and, in cases of recent transfusion (i.e., within the last 3 months), mask the alloantibodies present in the blood. Hence, to identify the alloantibodies present, we had to do differential adsorption.[8,9] We adsorbed the patient's autoantibodies and proceeded to test for the presence of any alloantibodies present in the blood.

The alloantibody we identified was anti-S after testing the adsorbed serum with screening and identification panels. In other cases similar to ours, there are reports of alloantibody anti-S along with autoantibody in SLE.[10,11] It is challenging to find match blood due to interference of autoantibodies. In general, the best-matched blood is transfused to the patient with autoantibody after excluding the alloantibody.

In our case, we transfused two units of S antigen-negative best-matched "O" Rh (D)-positive PRBC to the patient. She responded well to the transfusion and showed no adverse reaction.

Conclusion

Clinicians are generally reluctant to give a best-matched blood transfusion to patients with autoantibody-induced hemolysis of red cells. Transfusion medicine specialists should help the clinician understand that this is the best treatment plan for the patient and safe blood transfusion. In our case, we communicated with the clinician efficiently, and two units of S antigen-negative "O" Rh (D)-positive best-matched PRBCs were transfused to the patient.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient had given her consent for her images and other clinical information to be reported in the journal. The patient understands that her name and initials will not be published and due efforts will be made to conceal her identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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