Case Report

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Manual red cell exchange transfusion to avert sickle cell related complications

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Abstract:

Sickle cell-beta thalassemia is a double heterozygous state. Red cell exchange (RCE) transfusion reduces the concentration of sickle cells without increasing the hematocrit or whole-blood viscosity. It can be performed manually or by erythrocytapheresis. RCE transfusion is an effective tool for both acute and chronic complications of sickle cell disease. In patients unaffording erythrocytapheresis, even manual RCE can give favorable results. A 37-year-old male, a known case of sickle cell-beta^{+} thalassemia ($\beta^{s}\beta$+), presented with avascular necrosis of right femur and humeral head. He was posted for the right hip arthroplasty and shoulder hemiarthroplasty. Successful manual RCE transfusions were done. The hemoglobin S levels decreased postmanual RCE procedures, and the patient was operated successfully.

Keywords:

Complications, erythrocytapheresis, manual, red cell exchange, sickle cell disease, thalassemia

Introduction

Sickle cell disease (SCD) and thalassemia are autosomal-recessive genetic-inherited disorders. Sickle cell-beta thalassemia can be of two types: sickle cell-β^{o} thalassemia (no production of hemoglobin [Hb]-A chains) or sickle cell-β^{+} thalassemia (some amount of HbA chains are produced).[1]

The clinical features of these patients result from chronic variable intravascular hemolysis, microvascular ischemia, and organ damage.[2] Here, a case of alloimmunized sickle cell-β^{+} thalassemia who presented with vaso-occlusive crises underwent manual red cell exchange (RCE) transfusion, is reported.

Case Report

A 37-year-old male, a resident of Nanded region of Maharashtra, issue of a nonconsanguineous marriage, admitted in May 2015 with chief complaints of right shoulder pain and restricted range of movement for the past 1 year along with difficulty in walking for the past 4 years.

He was a known case of sickle cell-β^{+} thalassemia, had received 10–20 blood transfusions intermittently. History revealed that he suffered from an episode of avascular necrosis (AVN) of the right hip joint in the year 2011, when total hip replacement was attempted, but due to on table complications, the procedure had to be abandoned halfway, resulting in pseudoarthrosis.

Family workup of the patient revealed that his father and mother had sickle cell and beta thalassemia trait, respectively. The patient was vitally stable, with mild pallor on examination. Peripheral smear showed normocytic normochromic with occasional target cells, sickling test was positive, and high-performance liquid chromatography confirmed the presence of sickle cell-β^{+} thalassemia with HbS level of 72.3%.

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Final diagnosis made was AVN of the right shoulder with pseudoarthrosis of the right hip. The treatment planned was right shoulder hemiarthroplasty along with a corrective surgery for the pseudoarthrosis of hip joint. The patient was advised to undergo RCE transfusion to decrease his HbS levels for which he was referred to the Department of Immunohematology and Blood Transfusion.

Pretransfusion workup of the patient revealed his blood group to be AB Rh (D) positive with the presence of anti-c alloantibody (IgG). Phenotype of the patient was found to be R_1R_1 (DCe/DCe) (Dia-med Bio-rad, Switzerland). Direct antiglobulin test, autocontrol was negative. Due to patient built-up, vascular access, monetary, and infrastructural constraints, manual RCE was planned. The amount of blood to be exchanged was calculated by the formula as follows:[3]

$$\text{PRBC volume} = \frac{1.5 \times \text{Patient's Red Cell Volume}}{\text{Average volume of donor units}}$$

According to the calculations, three units of whole blood were to be venesected and replaced by saline and three units of packed red blood cells (PRBCs). Two PRBCs were required to be reserved for transfusion intraoperatively. Hence, total five units of PRBCs were required for the patient. The frequency of c antigen is around 80% and hence percent compatible is 20%, according to formula as follows:[4]

$$\text{No. of units to be tested to find compatible unit} = \frac{\text{No. of units required}}{\text{Percent compatible}} = \frac{5}{0.20} = 25$$

Successfully 5 compatible units were found after cross-matching with 30 units of PRBCs.

Manual RCE transfusion was performed over a period of 2 days. On day 1, the first unit of blood was venesected and replaced by 500 ml normal saline over 15–30 min, and then the second unit of blood was venesected and replaced by 1 PRBC. On day 2, the third unit of blood was venesected and replaced by two PRBCs. The characteristics of PRBCs used were compatible, sickle– negative, PRBCs <7 days old. The temperature, pulse, blood pressure, and oxygen saturation were monitored prior/postvenesection as well as before, 15 min, and 1 h after starting blood transfusions.

All transfusions were uneventful. Post-RCE HbS level decreased to 43.8%, which even though not ideal, was accepted due to time and procedural constraints. The patient was operated immediately on the 3rd day post-RCE. Surgery was performed under general anesthesia. Special precautions were taken during the surgery to maintain the patient's temperature, oxygen saturation, and fluid balance. One unit of PRBC was transfused intraoperatively and one unit postoperatively. The surgery was successful and the patient is doing well.

Discussion

Blood transfusions in SCD can be simple or exchange transfusions. RCE procedure has seen an increased popularity in these last 1-2 decades,[5] since it allows for an effective treatment for both acute sickling crisis unresponsive to conventional therapies and a prophylactic treatment for high-risk patients. With RCE, hematocrit and HbS can be adjusted rapidly and simultaneously, allowing for intervention in an emergency and eliminating the risks associated with alterations in viscosity and patient's blood volume.[6]

For acute complications of SCD, the goal of transfusion therapy is to reduce the posttransfusion HbS level to <30%; for chronic complications, the goal is to maintain the pretransfusion HbS level at <30%–50% while maintaining the Hb level at ~10 g/dL. Rapid lowering of HbS levels can only be achieved by acute RCE.[7] Exchange can either be performed manually or by automated cell separators. Erythrocytapheresis can be performed using different cell separators, operating either with continuous or discontinuous flow.[6] The former is preferable when low-weight patients (namely pediatric patients) are treated, since these devices allow for a lower extracorporeal blood volume throughout the procedure. With the ready availability of automated cell separators and ease in technical performance, erythrocytapheresis is being used increasingly to treat acute and chronic complications of RBC disorders, particularly in patients with SCD. As opposed to automated RCE, manual RCE is labor intensive, prolonged, and perhaps less safe and efficient than erythrocytapheresis, but can be beneficial for cases with monetary, infrastructural, or venous access constraints.[8]

The two primary goals of transfusion are to correct the low oxygen-carrying capacity caused by severe anemia and to improve microvascular perfusion by decreasing the proportion of sickle red cells in the circulation.[8] RCE is recognized as the most rapid method for lowering HbS levels, offering potential advantages over simple transfusions, but its role in the treatment of SCD, except in severe crises, is still controversial and it is not widely adopted. Advantages of RCE are that the exchange prevents the removed sickle cells from participating in new vaso-occlusive events, reduces hemolytic complications, provides with added oxygen-carrying capacity, reduces iron accumulation, better control of blood volume and viscosity.[9]

In SCD, both transfusion methods (simple transfusion and erythrocytapheresis) offer similar benefits in maintaining target HbS levels for long-term transfusion therapy. Although simple transfusion is available worldwide and is simple to perform, erythrocytapheresis is not universally available, requires experienced personnel to perform, and may require a central venous catheter/port. The distinctive benefits of chronic erythrocytapheresis are prevention of iron overload. In addition, erythrocytapheresis may avoid the risk of circulatory volume alterations and hemodynamic distress and, thus, is a safer procedure than other methods of transfusion. However, to date, the advantages and efficacy of RCE have not been substantially documented through clinical trials, especially compared with simple transfusion or manual versus automated RCE.[7] All SCD patients undergoing major surgery are prepared in advance with transfusion to correct their anemia to a Hb of approximately 10 g/dl and HbS percentage to approximately 30%.[2,3,9]

Conclusion

Despite all the constraints in this patient, provision of compatible c-negative PRBC units (for RCE and surgery) and manual RCE could facilitate hemiarthroplasty surgery safely. Manual RCE could be considered as an effective alternative to erythrocytapheresis in unaffording patients.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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