Erythrocytapheresis in a Patient of Sickle Cell Disease Presenting in Crisis

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Received: March 17, 2020; Published: June 29, 2020

Abstract

Background: As per American Society of Apheresis ASFA guidelines, the indication for Erythrocytapheresis for sickle cell patient is category III for preoperative management and vasocclusive crisis [1]. Erythrocytapheresis with the help of automated cell separators have the benefit of reducing the Sickle hemoglobin (HbS) levels to < 30% with reduced risk of increasing blood viscosity and single procedure achieving target hemoglobin levels.

Case Details: A 17-year old lady who was a known case of sickle cell disease, presented to our institution with avascular necrosis of right femoral head. Due to the pain and limitations of her day to day activities she was planned for total hip arthroplasty. This patient underwent a successful Erythrocytapheresis using automated cell separator and was operated.

Conclusion: In a sickle cell patient planned for surgical intervention the procedure of Erythrocytapheresis can help in decreasing the chances of the patient developing any vaso-occlusive crisis during surgery and in the immediate post-operative period. This technique can help in improving the chances of a successful surgical procedure for such patients.

Keywords: Erythrocytapheresis; Vasoocclusive Crisis; Target Hematocrit

Introduction

Sickle cell disease is an inherited hemoglobinopathy with mutation of the beta globin gene. This mutation leads to formation of abnormal hemoglobin in red cells which tend to produce sickle shaped red cells under conditions of low oxygen tension in the blood. This morphological change in the red cells leads to alteration in their structure and function. It can lead to vaso-occlusive crises. SCD is associated with hemolysis with vasoocclusive crisis and reperfusion injury. Presently, SCD patients are transfused with packed red cells to correct anemia.

Erythrocytapheresis in sickle cell patients is an automated red cell exchange (RBC Exchange) procedure for removal of sickled red cells and replacing them with compatible sickle free packed red cells of donor. The donor RBCs selected for transfusion are ABO and Rh compatible, < 7 days old, HBS negative and preferably extended phenotype matched to prevent alloimmunization [2]. The sickle cells of patient are removed when the blood passes through the automated cell separator and the plasma is returned to patient. After this, HBS negative packed normal red cells are transfused. Thus, the patient has a higher proportion of normal non-sickled cells and a lower proportion of sickle cells. Automated red cell exchange procedures reduce the HBS levels to < 30 Thus theoretically, decrease the probability of the patient developing a vaso-occlusive crisis and as viscosity decreases and the chances of clinical deterioration also decrease. The major advantage of erythrocytapheresis is the rapid reduction of HbS erythrocytes without increasing the patient’s hematocrit or causing

Citation: Babita Raghuwanshi. “Erythrocytapheresis in a Patient of Sickle Cell Disease Presenting in Crisis”. EC Anaesthesia 6.7 (2020): 09-11.
fluid overload along with a reduced risk of iron accumulation. As the viscosity decreases, blood flow improves and vasocclusive crisis is reversed rapidly.

**Case Report**

A 17-year girl who was a previously known case of sickle cell disease presented to the Orthopedics department of our institute with gradually progressing right sided hip pain for the past 3 years. As she was an already diagnosed case of sickle cell disease, she was on regular treatment with hydroxyurea. At presentation, she had severe hip pain which had progressively increased to the present level over a three-month period. She was unable to stand or walk unsupported on the right side; her hip movements were grossly restricted, and her activities of daily living were significantly impaired. The radiographs showed multiple patchy areas of geographically altered marrow signal in the femoral head. These images were consistent with a diagnosis of avascular necrosis of the femoral head (AVN). HbS (Sickle hemoglobin) levels were 64% by HPLC. In view of her complaints a surgical intervention the form of total hip arthroplasty was planned. Assistance from the department of transfusion medicine was sought for optimizing the patient for this proposed surgery. She had never received any blood transfusion in the past. The patient was planned for preoperative erythrocytapheresis. The patient’s coagulation profile was normal. The height, weight and hematocrit of patient were recorded for RBC exchange calculation. Nadler's formula was used for calculating the blood volume. As per this formula the blood volume is calculated as $(0.3561 \times H^3) + (0.03308 \times W) + 0.1833$ [3]. The height $H$ of patient was as 161 cms and weight $W$ was 47 kg. The calculated total blood volume was 3223.

Pretransfusion testing workup was done. The patient’s blood group was B negative and extended Rh Phenotype was c e positive/E C negative. (E Tulip Lot No 1129031, e Lot no 1149071, C Lot no 1119011 and c Lot no 1139021). Direct Coomb's test, auto-control and allo-antibody workup was negative. The patient's hemoglobin concentration was 10.2 gm/dl with a hematocrit of 30%. Smear showed normocyctic normochromic red cells and target cells. HPLC showed SCD with a sickle cell percentage of 64%. The RBC exchange procedure was started with a pre-procedure HbS level of 64% to achieve a target HBS of 30%. Replacement volume required was calculated as 1188 ml.

FRC was calculated using the following formula: desired percentage of cells at conclusion of procedure/actual percentage of cells now $\frac{30}{64}$. Fraction of cells remaining (FCR) was calculated as 47%.

Five units of compatible B negative blood units with extended Rh matching < 7 days old, HbS negative and AHG Cross match compatible were selected for RBC exchange transfusion. The volume of RBC required as replacement volume for transfusion was calculated to be as 1188 ml and accordingly 5 units of compatible PRBC with an average volume of 240 ml/unit were selected. Complete blood counts of all the five donor B negative units was done and mean hematocrit was calculated to be 61. Anticoagulant (AC) to whole blood (WB) ratio was set to at 1:13 value. Temperature, pulse, blood pressure, Oxygen saturation was monitored before starting procedure and at every 15 minutes during procedure. Calcium gluconate infusion @ rate of 1 ml/min. The procedure was uneventful. The post procedure HBS level came down to 20.70%. Platelet count were reduced during RBC exchange. The pre and post procedure laboratory parameters have been presented in table 1.

<table>
<thead>
<tr>
<th></th>
<th>Pre procedure</th>
<th>Post Procedure</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hemoglobin</td>
<td>10.0</td>
<td>11.0</td>
</tr>
<tr>
<td>Hematocrit</td>
<td>30.0</td>
<td>32.3</td>
</tr>
<tr>
<td>RBC Count</td>
<td>3.26 X 10^3</td>
<td>3.49 X 10^3</td>
</tr>
<tr>
<td>WBC count</td>
<td>5400</td>
<td>7000</td>
</tr>
<tr>
<td>Platelet count</td>
<td>156 X 10^3</td>
<td>126 X 10^3</td>
</tr>
<tr>
<td>HPLC HbS</td>
<td>64%</td>
<td>20.70%</td>
</tr>
<tr>
<td>HBF</td>
<td>30.60%</td>
<td>11.20%</td>
</tr>
<tr>
<td>HBA</td>
<td>2.40%</td>
<td>59.20%</td>
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<tr>
<td>HBA$_2$</td>
<td>2.10%</td>
<td>2.80%</td>
</tr>
</tbody>
</table>

*Table 1:* Table showing the laboratory parameters of the patient before and after erythrocytapheresis.
Total hip replacement for the patient was performed successfully 5 days after erythrocytapheresis under general anesthesia with due precautions to maintain patient temperature, oxygen saturation and fluid balance.

Discussion

Surgical intervention and general anesthesia can cause hypotension, local hypoxia, acidosis and inflammation leading to sickling of PRBC and aggravation of vaso-occlusive crisis in a patient with SCD. Erythrocytapheresis is a simple day-care procedure for RBC exchange in these patients. It has emerged as a preferred treatment for patients with acute cell crisis and is an ASFA Grade 1 indication. However, Erythrocytapheresis for sickle cell patient is category III for preoperative management and vaso-occlusive crisis [1]. It is an effective method to reduce the burden of HbS in patients without altering the hematocrit of patient significantly as the PRBCs transfused during procedure adjusts the depleted RBC volume. Platelet reduction has been seen in other reports also [4]. The procedure is safe and allows emergency intervention to reduce viscosity in sickle cell crisis. In SCD transfusion of HbS negative blood can be performed to reduce vaso-occlusive crisis and increase oxygen carrying capacity of blood. These are done either as simple transfusion, manual exchange transfusion or automated RBC exchange transfusion.

The simple transfusions and manual exchange transfusions do not require any special training and equipment but are associated with risk of hyper-viscosity. The Automated RBC exchange requires special training and equipment but it has least risk of all the other procedures for producing hyper-viscosity with best control of HbS levels [5].

The transfusion therapy in SCD can be for reducing HbS burden and prevent sickle cell crisis by improving capillary perfusion. The reduction in sickle cell reduces incidence of vaso-occlusive crisis, improves oxygen carrying capacity as new red cells are transfused and hemodynamic stability and blood volume balance is maintained, and risk associated with circulatory volume alteration is reduced. Erythrocytapheresis requires trained apheresis personnel with insertion of central venous 3-way cannula for inflow and return lines and calcium infusion.

Conclusion

The RBC Exchange using continuous flow automated cell separators is especially helpful in young patients as hemodynamic stability is satisfactorily maintained in them. The RBC exchange of ABO compatible and Rh Phenotype matched blood facilitates depletion of HbS which reduces the probability of intra and post-operative complications and is therefore an effective treatment option.

Bibliography