

Therapeutic Red Cell Exchange Transfusion as an Adjuvant Therapy for Management of Sickle Cell Crisis in Adults

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Abstract

Sickle cell crisis is an acute clinical condition, caused due to capillary occlusion by the deformed red blood cells, leading to vaso-occlusive status. Vaso-occlusion is an emergency condition requiring intensive care admission and carries a high mortality. Sickle cell crisis is usually managed with hydration, analgesics, and supportive care. Therapeutic red cell exchange transfusion is advised as an adjuvant, for the management of sickle cell crisis, and it is mainly practiced in the pediatric population. We report an adult case of sickle cell crisis managed with therapeutic red cell exchange transfusion in Intensive Care Unit and successful outcome in patient management.

Keywords: Intensive Care Unit, red cell exchange transfusion, sickle cell crisis

INTRODUCTION

Sickle cell disease (SCD) is caused by a mutation in the β -globin gene that changes the sixth amino acid from glutamic acid to valine. Sick cell hemoglobin (HbS) polymerizes in deoxygenated condition to form a gelatinous network of fibrous polymers that stiffen the red blood cell (RBC) membrane, which lose the pliability needed to traverse small capillaries. These rigid, adherent cells clog small capillaries and venules leading to episodes of microvascular vaso-occlusion and premature RBC destruction. This vaso-occlusive component usually dominates the clinical course and manifestations as episodes of ischemic pain (i.e., painful crises) and ischemic malfunction or frank infarction in the spleen, central nervous system, bones, liver, kidneys, and lungs.^[1] Provocative factors include infection, fever, excessive exercise, anxiety, abrupt changes in temperature, hypoxia, or hypertonic dyes.

CASE REPORT

A 26-year-old female was referred from an outside hospital to our hospital in view of persistent chest pain and abdominal pain for 3 days. The patient was a known for SCD, not on regular medication, last Hydroxyurea was taken a year ago, and last blood transfusion was also done a few years back. The patient was referred in view of hypotension and worsening clinical condition. On examination in the emergency department, she

was conscious, afebrile, tachycardic, and hypotensive. Fluid resuscitation was started, appropriate cultures were sent, and broad-spectrum antibiotics started. On arrival to Medical Intensive Care Unit (ICU), she was hemodynamically unstable; bilateral crepitations were present on auscultation. She was started on vasopressors in view of persistent hypotension and connected to noninvasive ventilation (NIV) for respiratory support. Arterial blood gas analysis showed severe metabolic acidosis and laboratory parameters showed grossly deranged renal function test and liver function test (LFT), a likely diagnosis of sickle cell crisis was made.

The patient had persistent anuria and severe metabolic acidosis; hence, the patient was planned for sustained low-efficiency dialysis (SLED). Peripheral smear showed sickle cells, spherocytes, and plenty of nucleated RBCs, which was suggestive of sickle cell crisis and the sickling test was also positive [Figures 1 and 2]. Packed RBC (PRBC) transfusion was done to maintain Hb level >10 g%, and Hb electrophoresis was also sent. Liver viral markers, dengue,

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Figure 1: Peripheral smear demonstrating sickle cells, spherocytes and plenty of nucleated red blood cells

malaria, leptospira, and scrub typhus tests were negative. The patient had persistent severe chest pain suggestive of acute chest syndrome in spite of PRBC transfusion; hence, she was planned for red cell exchange transfusion during SLED.

During the exchange transfusion, whole blood was removed, which was replaced by PRBC and fresh frozen plasma (FFP) in equal proportion, at the same time of SLED. The patient tolerated the exchange transfusion without any complications and showed good clinical improvement after exchange transfusion. Two more cycles of exchange transfusion were done along with SLED and patient clinical condition further improved. The patient was off vasopressors, off NIV, chest pain had reduced, and LFT and LDH levels started improving. The patient gradually started producing urine and was shifted with to the ward in stable condition with dialysis catheter *in situ*. In the ward, her renal function improved further and she was discharged home with oral medications.

DISCUSSION

RBC transfusion is recommended as the first line of management, for SCD to keep the HbS levels below 30%.^[2,3] The Hb electrophoresis report of the patient showed HbS of 49.7%. We initially managed the patient with simple RBC transfusion, but the patient clinical condition did not show any improvement; hence, we considered for RBC exchange transfusion. The American Society for Apheresis (ASFA) recommend RBC exchange transfusion for SCD with acute chest syndrome or acute stroke as grade 1C recommendation.^[4] ASFA recommends acute SCD with multiorgan failure, splenic/hepatic sequestration, and intrahepatic cholestasis as grade 2C recommendation.^[4] Our patient had acute chest syndrome which is a grade 1C recommendation; we proceeded for exchange transfusion as an adjuvant therapy along the ongoing supportive therapy. Two methods of exchange transfusion are available: manual



Figure 2: Image showing positive sickling test

exchange transfusion (MET) and automated exchange transfusion (AET), At our ICU, we performed MET as AET was not available, MET consists of removal of whole blood of the patient, by either phlebotomy or other vascular access followed by replacement of normal adult (HbA) PRBC. Since the patient was undergoing SLED, we performed the exchange transfusion on the dialysis circuit. We removed 15 mL/kg of patient blood through the dialysis circuit and replaced the same volume of PRBC and FFP in equal proportion through a central line. The guidelines recommend that one red cell volume exchange transfusion can replace 10%–15% of the patient's original RBCs.^[5] The patient showed clinical improvement, and we performed a total of 3 cycles of exchange transfusion on each day.

CONCLUSION

Sickle cell crisis is an emergency condition requiring aggressive management in ICU. Our case emphasizes the role of RBC exchange transfusion in the management of the sickle crisis, in an adult patient with acute chest syndrome and multiorgan organ failure.

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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