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Case Report

Manual RBC exchange transfusion in a patient of sickle-beta thalassemia syndrome presenting in crisis: a case report

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ABSTRACT

Sickle cell disease and beta thalassemia are caused by abnormal haemoglobin (Hb) derived from mutation of the HBB gene encoding beta-globin. Compound heterozygous status for both mutations results in HbS/beta thalassemia (Sickle- beta thalassemia). Vaso-occlusive phenomena and haemolysis are the clinical hallmarks and major causes of mortality. Here we report a case of successful reduction of HbS level by manual RBC exchange transfusion. Capillary zone electrophoresis showed the case to be Sickle-beta thalassemia. A total of 3 units of 450ml whole blood units were used for manual exchange transfusion done in 2 sittings on consecutive days. Preexchange HbS level was 80.9% of total Hb. HbS level after 24 hours of the second procedure was 44%. In the absence of facility to conduct automated RBC exchange by a cell separator, to reduce HbS in patients presenting with acute complications of SCD and in patients with Vaso-occlusive Crisis, previous stroke, manual RBC exchange can provide a better relief.

Keywords: Exchange transfusion, Manual red cell exchange, Sickle cell anaemia

INTRODUCTION

Disturbances in haemoglobin synthesis are some of the most common human hereditary disorder. There is an increased prevalence among the African and Asian populations and more recently, immigration has led to an increase in the incidence of this disorder in Europe as well.[1]

SCD is a haemoglobinopathy characterised by an abnormal haemoglobin variant termed haemoglobin S. HbS causes irreversible filamentous precipitation which causes red blood cells to change shape causing circulation problems. Clinical symptoms include relapsing ischemic episodes, chronic haemolysis and a specific type of anaemia termed sickle cell anaemia.[2]

Homozygous Patients are in danger of increased perioperative mortality and have a reduction in the life expectancy. Hypothermia, hypoxia, acidosis and dehydration in the perioperative period can cause an acute exacerbation of the disease.[2]

Transfusion may be aimed at either to correct an aggravation of anaemia or to provide non-deformable red cells to deliver oxygen to ischemic tissues in cases of vaso-occlusion. In the latter setting, an attention must be paid not to raise haematocrit above 35%, in order to prevent hyper viscosity. A way to decrease HbS level without increasing haematocrit is to conduct phlebotomy with transfusion, which defines RBC exchange transfusion. It may be performed manually, exchanging patient's whole blood with packed red blood cells, or using a cell separator, with reinfusion of patient's plasma. Exchange transfusion may be advised in emergency, after an acute vaso-occlusive event (stroke, acute chest syndrome, acute organ failure). The most frequent indication of chronic transfusion program in children is the prevention of cerebro-vascular event, either of recurrence of stroke or of a first stroke in children with

elevated cerebral blood flow as measured by transcranial Doppler (TCD) screening test. In these conditions, hydroxyrea has also been administered.[3]

CASE REPORT

Our case was a 15 years old Indian male child who was first diagnosed at the age of one and half years as a homozygous form of sickle cell disease presenting with fever for few days and Hb level of 7gm/dl. At the age of 5 yrs pain appeared at left hypochondrium, persisted for few days and then subsided automatically, USG showed no abnormalities. At the age of 12 to 13 years, frequency of pain gradually increased. Initially episodic pain was experienced in 7 to 8 months interval, later decreased to 3 to 4 months interval. He was taking hydroxyurea from 5 years of age.

Patient presented to Haematology Department of our institute with complaint of severe pain in left thigh for 2 to 3 days, fever for 1 day, and 1 to 2 bouts of voluminous vomiting. He was given symptomatic treatment for pain, fever, vomiting and was planned for exchange transfusion. He was first hydrated with 500ml NS followed by phlebotomy with 450ml of bloodletting using 18" (gauge) needle through antecubital vein. This was followed by 500ml of NS infusion, and another 450ml of whole bloodletting. Then 2 units of ABO specific PRBC were transfused. Next day, maintaining hydration with 500ml NS, 450ml whole blood was let and 1 unit of ABO specific PRBC transfused. Patient's blood sample sent for Hb electrophoresis prior to phlebotomy on 18th September 2015 and 24 hours after the completion of second procedure on 19th September 2015. Manual RBC Exchange in both sittings was uneventful.

Laboratory results

18th September 2015 (before the 1st procedure)

CBC

Hb-8.4 gm%, Hct-25.1, RBC-3.93x106/µl, MCHC-33.5 gm/dl, MCV-67.3 fl, MCH-22.5 pg, Retic-3%., WBC-11.9x103/µl, N-81.3x103/µl, L-13.4x103/µl, E-3.7x103/µl, B-0.3x103/µl, PLT-285x103/µl,

Haemoglobin electrophoresis (Figure 1)- HbA-3.1%, HbF-10.4%, HbS-80.9%, HbA2-5.6%

20th September 2015 (24 hours after the 2nd procedure)

CBC

Hb-9.9 gm/dl, Hct-30.3%, RBC-4.09x106/µl, MCV-70.5 fl, MCHC-32.7 gm/dl, MCH-23.0 pg, WBC-11.71x103/µl, N-77%, L-12%, Mylocytes+ Metamylocytes = 5%, E-5%, B-01%, PLT-241X103/µl

Haemoglobin electrophoresis (Figure 2)- HbA-46.7%, HbF-5.2%, HbS-44%, HbA2-4.1%.

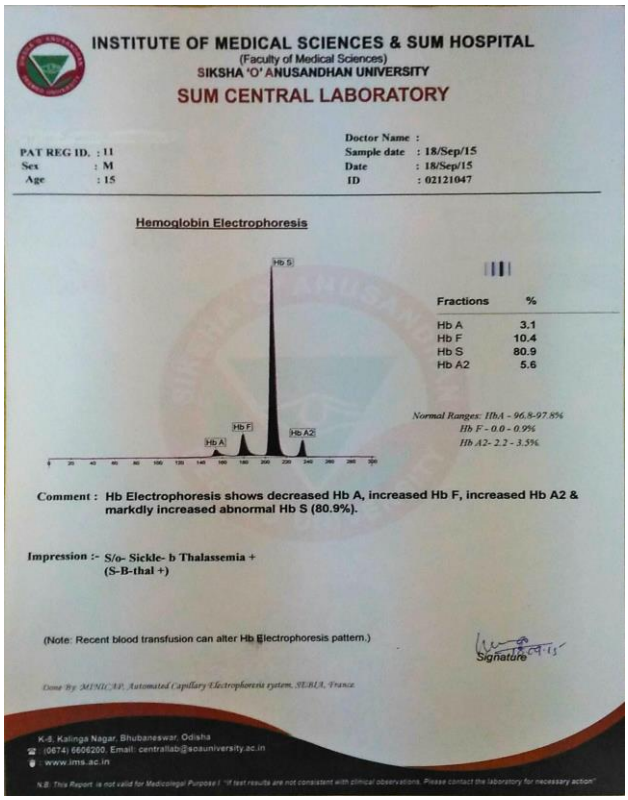

Figure 1: Hb Electrophoresis on 18th September 2015.

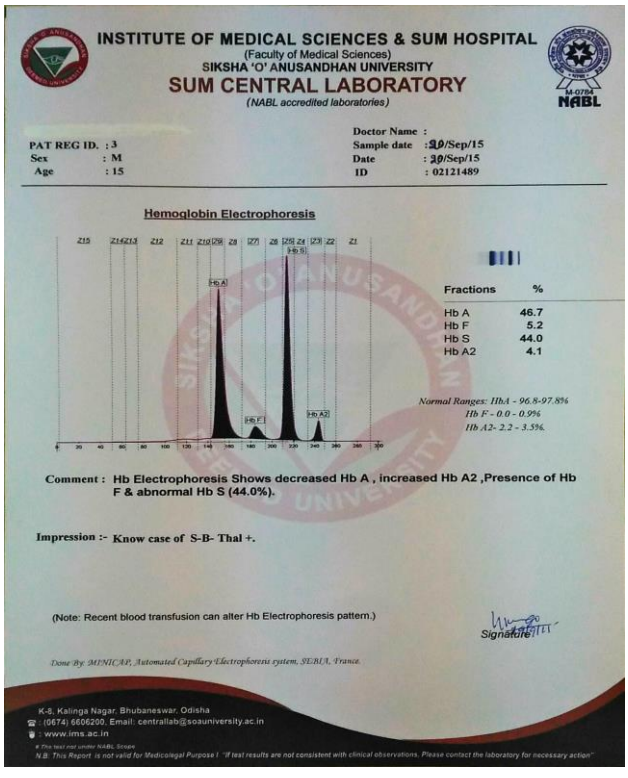

Figure 2: Hb Electrophoresis on 20th September 2015.

DISCUSSION

Simple transfusions are widely used in management of sickle cell disease complications. However, it can increase Hct, blood viscosity, and vaso-occlusion. Red cell partial exchange transfusion may provide the same effect on oxygen carrying capacity without increasing Hb level and viscosity of blood.[4]

Hb level of more than 10 gm/dl (haematocrit more than 30%) in the presence of substantial amounts of HbS more than 30% is associated with hyper viscosity. Some data indicate that phlebotomy to reduce haematocrit and blood viscosity (which may also address iron overload) can decrease the frequency of crisis in HbSc or HbS-B+ disease. Phlebotomy alone has also been used in sickle cell disease maintaining a base line haemoglobin level of within 9.5 gm/dl.[5] One approach of phlebotomy is to remove approximately 10 ml/kg body wt.

A comparative study of manual and automated red blood cell exchange transfusion in sickle cell disease patients showed that automated RBC exchange utilized more PRBCS in terms of volumes and units but requires half the time and performed less frequently. Treatment failure, defined as a SCD related complications that the red cell exchange was intended to prevent, did not occur in any case within defined period with exception of pain. No significant difference in adverse events was observed. The majority of adverse effects reported in the manual RBCX were termination of RBCX procedure due to poor IV access, blocked line or port, while majority of adverse events in the automated RBCX were attributed to dizziness or hypotension.[6]

CONCLUSION

SCD is a common hereditary affliction that is more widespread among people in Sub-Saharan Africa. Increased population migration has increased the prevalence of this disease in other countries as well. SCD causes progressive damage to multiple organs.[2] Manual RBC exchange is a simple and safe procedure that can be performed in the setting of acute complications of SCD and in patients with vaso occlusive crisis, previous stroke, to reduce HbS. It can be an additional therapy for refractory VOC and for respiratory improvement of acute thoracic syndrome, besides simple transfusion.[4] In this case we performed a partial RBC exchange transfusion in a child in sickle cell crisis. Now the patient is stable and relieved of painful crisis. In the absence of facility to conduct automated RBC exchange by a cell separator such patients can be provided better relief by manual RBC exchange.

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