ORIGINAL RESEARCH PAPER

General Medicine

AN INTERESTING CASE OF LOBAR PNEUMONIA

KEY WORDS: Acute chest syndrome, Red cell exchange, Sickle cell anemia.

DR. Kalidindi Lakshmi priya* Postgraduate, Dept. of General Medicine Saveetha Medical college and Hospital, Chennai *Corresponding Author

DR. Mahendra Kumar Kalappan Professor, Dept. of General Medicine Saveetha Medical college and Hospital, Chennai

DR. Arun Kumar Bathena Assistant Professor, Dept. of General Medicine Saveetha Medical college and Hospital, Chennai

DR. Prasanna Karthik.S Professor, Dept. of General Medicine Saveetha Medical college and Hospital, Chennai

ABSTRACT

Sickle cell disease (SCD) is an inherited haemoglobinopathy that may present acutely as stroke, vaso-occlusive crisis or acute chest syndrome. Acute conditions require prompt diagnosis and intensive management to prevent morbidity and mortality. Here, we present a case of sickle cell anemia treated with therapeutic red cell exchange that manifested as acute chest syndrome. A 26 year old male who had history of high-grade intermittent fever and cough with expectoration was diagnosed as pneumonia and put on antibiotics. Peripheral smear showed sickle cells and HPLC showed HbS of 67%. we made a diagnosis of sickle cell anemia with acute chest syndrome. We treated the patient with therapeutic red cell exchange following which HbS reduced to 24% from the initial 67%. We observed significant clinical improvement suggesting red cell exchange as a useful therapeutic option for acute sickle cell anaemia.

INTRODUCTION

Sickle cell disease is an inherited haemoglobinopathy that may present acutely as stroke, vaso-occlusive crisis or acute chest syndrome. Acute conditions require prompt diagnosis and intensive management to prevent morbidity and mortality. SCD is a disease that most commonly affects people of the African descent with over 1 in 500 African Americans affected by it. The mutation involved is in the beta globin chain in the chromosome 11. Inheritance of two abnormal copies of the gene result in the most severe form of the disease. However, people with only one abnormal copy are said to have the sickle cell trait and are most often asymptomatic.

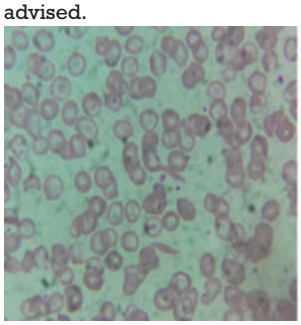
Since SCD is associated with painful crises, the main goal of therapy would be to reduce the occurrence of such episodes. Currently available treatment modalities are aimed at pain reduction and supportive care. Drugs like hydroxyurea are underutilized despite being efficacious.

One therapeutic modality that is also underutilized is red cell exchange transfusion, wherein the patient's red cells are replaced by healthy donor cells. This therapy effectively removes cells vulnerable to sickling, thereby preventing the occurrence of vaso-occlusive events. In addition, it also provides improved oxygen carrying capacity and a decrease in blood viscosity. Red cell exchange is of particular use in acute situations and has been shown to produce significant clinical improvement in many cases Here, we present a case of sickle cell anaemia that manifested as acute chest syndrome and was successfully managed with red cell exchange treatment. (1)

CASE REPORT

A 26 - year old male hailing from Orissa without any comorbidities came with history of high grade intermittent fever with chills and rigor for 4 days. He also had cough with expectoration for 2 days. He had a history of recurrent episodes of severe joint pains from childhood.

On examination, he was found to be febrile, pale and hypoxic requiring 4 litres of oxygen. Provisional diagnosis of pneumonia was considered and started him on empirical antibiotics along with symptomatic management. Laboratory investigations revealed anemia (Hb – 7.4gm/dl) and thrombocytopenia (Plt count – 12,000 cells/cu.mm). Peripheral smear showed normocytic normochromic RBCs with few target cells and sickle cells. We did an HRCT thorax which showed right lower lobe pneumonia. Infective workup including sputum analysis was done and bacteriological work-up were negative. HPLC showed HbS of 67% , HbF - 19.1%, HbA2 – 3.7% and USG abdomen showed contracted spleen. He was diagnosed with sickle cell anemia presenting with acute chest syndrome. Therapeutic Red cell exchange was done with three packed red blood cells. Post exchange transfusion, HbS was 31.8%. following this, two packed red blood cell transfusion were done on subsequent day and repeat HPLC showed HbS of 24%. His Hemoglobin-10gm/dl and platelets improved to 1.28 lakhs/cu.mm^3 after therapeutic red cell exchange. His hypoxia improved and he was symptomatically better. He was discharged on oral hydroxyurea tablet and genetic screening of the family was advised.



Peripheral smear showing sickle cells and nucleated RBCs

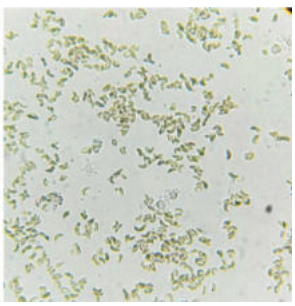


Image demonstrating positive sickling test

DISCUSSION

Sickle cell disease is an inherited disorder of the gene encoding the beta subunit of hemoglobin. The umbrella term of SCD includes other entities like sickle cell anemia, hemoglobin SC disease and hemoglobin sickle-beta-thalassemia. This said mutation occurs when the negatively charged glutamine in the sixth position of the beta globin chain is replaced by a neutral valine. The most severe forms of the disease have a homozygous mutation or the HbSS disease.

On the other hand, HbAS patients who are heterozygous are most commonly asymptomatic and diagnosis is often incidental.

This defect in the beta globin gene causes the hemoglobin to become a rigid, elongated polymer at times of stress, initially transiently but at one point, irreversibly. This irreversible 'sickling' results in hemolysis and vaso-occlusive crisis.(2) Stressors that could increase the risk of polymerization are hypoxia, oxidative stress, infection and dehydration. Characteristic features of SCA on peripheral smear area helpful in clinching a diagnosis. These include the presence of drepanocytes, Howell-Jolly bodies, target cells and nucleated RBCs.

In our patient, peripheral smear showed sickle cells and some target cells with normocytic, normochromic RBCs. His ultrasound showing a contracted spleen and HPLC positivity for HbS further helped confirming his SCA diagnosis. However, it is interesting to note that our patient presented to us with Acute Chest Syndrome (ACS). ACS is the most common cause of death in SCA patients and is characterized by cough and shortness of breath. The presence of fever, cough with expectoration in our case along with a HRCT diagnosis of lower lobar pneumonia pointed towards an infection. We opted for a red cell exchange transfusion for this patient. (3)

Red cell exchange transfusion is a therapeutic modality that provides the necessary oxygen carrying capacity and reduces the overall viscosity of blood. In SCD patients with a history of or risk for an acute stroke, RBC exchange transfusions are the conventional treatment, and they are also a clinical option for other complications. The rare indications for RBC exchange are severe erythrocytosis or hereditary hemochromatosis, intraerythrocytic pathogen infections such malaria or babesiosis, and severe erythrocytosis. This process becomes important in acute situations where it is necessary to decrease immediate complications. Situations like ACS, as in our case, require rapid improvement in oxygenation, which can be provided by a red cell exchange. (4–6)

Following therapeutic red cell exchange, we noted significant improvement in hypoxia and symptoms. The HbS had reduced from the initial 67% to 24%, in accordance with the reduction of hypoxia and symptoms.

Red cell exchange may be one of two types: manual and automated. Manual method requires little training and can achieve intermediate control of HbS. But this is limited by its time-consuming nature. Automated methods overcome this limitation, but require special equipment and extensive staff training. No matter the type, therapeutic red cell exchange is a relatively safe procedure. Side effects that may rarely occur include fatigue, nausea, dizziness, chills and tingling. Allergic reactions and blood-borne infections are extremely rare side-effects

Several case studies about red cell exchange for SCD have been published. Tsitsikas et al. have described a case of fat embolism in sickle cell disease treated with red cell exchange transfusion. Their patient had modest clinical improvement but a complete regain in consciousness after three cycles of the therapy. Similarly, Kassem et al., have successfully treated sickle cell associated acute liver failure with exchange transfusion. A case of sickle cell associated priapism in a 19 year old male was successfully managed by Ebraheem et al., with this therapy. They have reported a decrease in HbS from 45.8% to 11.7%, which is in accordance with our case. (7–10)

CONCLUSION

We have treated a patient of SCA presented initially as acute chest syndrome with therapeutic red cell exchange, and have observed significant clinical improvement. Red cell exchange, though a very effective therapy, still remains underutilized probably due to reduced awareness and resources. However, it is inarguable that this is a effective treatment for sickle cell patients presenting with crisis situations, and can be lifesaving.

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