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Case Report

Pregnancy in sickle cell thalassemia: double trouble!

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ABSTRACT

This is a case report of 20-year-old multigravida (G2P1L0) belonging to tribal community presented at 30 weeks of gestation with severe anemia, fever, arthralgia and jaundice with multiple blood transfusions in the past and with previous pregnancy outcome being stillborn and was not evaluated for the same. A diagnosis of sickle cell β+Thalassemia (SCD crisis) was made and managed vigilantly by multidisciplinary approach and had full term vaginal delivery with good perinatal outcome and finally both mother and newborn were discharged in stable condition. This highlights the overall increase in maternal and fetal complications in pregnancy with sickle cell thalassemia. Thus health education, screening, early intervention with multidisciplinary approach and regular follow up prevents maternal morbidity and mortality.

Keywords: Sickle cell disease, Sickle cell crisis, Pregnancy, Multidisciplinary

INTRODUCTION

Sickle cell disease (SCD) is the most common autosomal hereditary disease worldwide and accounts for 14.5% of total newborns with SCD in India.[1] The term sickle cell disease (SCD) includes different genotypes, homozygous HbS sickle cell (SS) and double heterozygote sickle haemoglobin C (SC), sickle beta thalassemia (Sbβthal), sickle alpha thalassemia (Saαthal), sickle cell anaemia with high fetal Hb (SC+F). Affected people have a different change in each copy of Hb0gene – one that causes red blood cell to form 'Sickle' shape and second1that it is associated with reduced amount of normal Hb(β+ Thalassemia).[2] The presence of sickle shape red blood cell which often breakdown prematurely and reduction of mature RBCs leads to many signs and symptoms with variable severity. The physiological adaptation that occur in hematological, cardiovascular, pulmonary, renal and circulatory system during pregnancy can magnify in sickle cell disease and negatively influence pregnancy outcome with increased risk of hypertensive disorders (preeclampsia), venous thromboembolism, acute chest syndrome. Microvascular damage and decreased uteroplacental circulation leads to risk of spontaneous abortion and stillbirth and others like newborn abstinence syndrome and haemolytic disease of newborn.[3,4] Though pregnancy in sickle cell disease has greater fetal and maternal morbidity and mortality, it can be prevented by ensuring adequate preconceptional counselling, antenatal and postnatal care by multi-disciplinary approach.

CASE REPORT

A 20-year-old G2P1L0 in early third trimester (30+2 weeks) belonging to tribal community near Kodagu was referred from peripheral hospital in view of severe anaemia, fever and jaundice for further management.

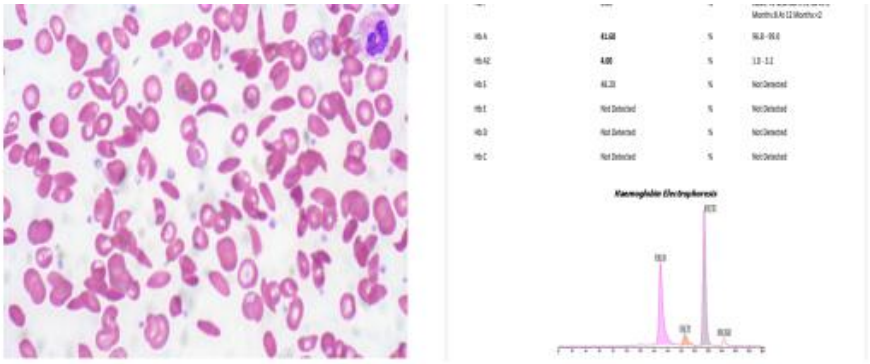
On admission, she complained of severe body ache, fever since 2 days and cough since 1 day. On examination she had grade 3 pallor, Icterus, bilateral lower limb edema present. Patient was tachypneic, febrile and tachycardia noted. Her investigations read: Hb-4.3g%, Platelets–1 lakh, WBC–23,370 cells/µl, LDH–1485, direct bilirubin-2.3, total bilirubin–3.2, peripheral smear- poikilocytosis, target cells, sickle cells, Howell-Jolly bodies, schistocyte

present. Hb electrophoresis showed HbF-6%, HbA1-40%, HbA2-4%, HbS- 60% indicating sickle cell β thalassemia.

Management

She was managed with multidisciplinary approach by obstetrician, haematologist and physician with oxygen supplementation, adequate hydration with IV fluids and started with prophylactic antibiotics and non-opioid analgesics. Patient received 3-pint packed RBC and Coomb's test (DAT) was done which read negative. She gave h/o repeated attacks of anaemia and jaundice in the past and history of blood transfusion which is suggestive of haemolytic anemia and was not evaluated for same and her previous pregnancy outcome was stillborn at 8 months.

Fever faded after 48 hr and improvement of symptoms noted after 4 days and recovery occurred within 10 days and was discharged and was followed up every week and monitored for any crisis and fetal growth restriction. At 36 weeks of GA, she presented with acute onset of chest pain and abdominal pain and was admitted in view of SCD crisis and managed symptomatically within a week patient spontaneously set into labour and delivered an alive male neonate of 2.4 kg birth weight with an APGAR – 1'-8/10, 5'-9/10 and intrapartum and postpartum was uneventful. Her lab value on PND1 showed Hb–6.5 g%, platelet–1.1 lakh, WBC–19,000 LDH – 984 and she was transfused with 1 pint PRBC and LMWH heparin was given and both mother and newborn were discharged in good condition on post-natal day 7.



A B

Figure 1: (A) Peripheral smear showing poikilocytes: sickel cells, target cells, Howell-Jolly bodies and schistocytes. (B) Hb electrophoresis: sickel cell βeta+ thalassemia.

DISCUSSION

Patients should be made aware of their disease by effective screening, reproductive health promotion and preconceptional counselling should be offered to all young adults with SCD as well as to women with SCD in the postpartum period.[5] Prevalence of sickle cell carriers varies in India from 1%–40% among various tribal group hence universal screening should be made available in tribal population free of cost.

The patients here are prone to various maternal complications like sickling crisis which includes both thrombotic and haemolytic type. The physiological changes of pregnancy which includes increased blood volume, increased metabolic demand and increased blood viscosity gets aggravated in SCD. The immune deficient status of pregnancy adds up to the existing burden resulting in more prone for infection acting like double edge sword.

Transfusion therapy is a major cause of improvement, however there is no agreement as to whether transfusion should be prophylactic or reserved for symptomatic cases.[6-8] To reduce haemolytic and thrombotic risk, the modern management is exchange transfusion to keep sickle cell percentage below 20% and haemoglobin level by 10 g/dl. Although our patient did not develop any atypical eythrocytic antibodies due to repeated blood transfusion which might have resulted in serious complications. Hence before blood products transfusion, careful cross matching to be done to minimise minor blood incompatability and alloimmunisation.[6] Multiple blood transfusions and ongoing haemolytic crisis resultsiin increase serum bilirubin levels as evident in this case.

Close monitoring of both the mother and the fetus is done to prevent repeated attacks of SCD crisis and for any fetal distress or fetal growth restriction.[9,10] A high index suspicion and good diagnostic acumen is necessary to obtain optimal result. Preterm labour is another complication but our patient had early term (37 weeks) labour.[11] The mean birth weight is usually lower in Sickle cell disease due to uteroplacental insufficiency, fetal growth reduction and preterm labour.[12] There is no standardized recommendation regarding treatment of painful crisis in pregnancy as most cases have to be individualized. Use of NSAIDs is generally avoided after 30 weeks due to increased risk of premature closure of ductus arteriosus but can be given in second trimester and postpartum.

Occurrence of SCD crisis during intra-partum period offers additional challenge to the care providers. The use of general anesthesia can result in significant increase in

sickling complication.[13] Therefore if caesarean delivery is indicated, regional anesthesia is preferred which was not same in our case as patient had spontaneous full term vaginal delivery. Post-partum period can trigger sickle cell Disease crisis and can have thrombotic episodes and pulmonary edema, hence post-natal LMWH prophylaxis was given.[11,14]

CONCLUSION

Overall pregnant women with SCD are more likely to experience antepartum, intrapartum and postpartum complications. Patient education, screening, early intervention and multidisciplinary approach, close follow up is important to minimise maternal morbidity and mortality. Therefore, we emphasize universal screening for in tribal community at free of cost due to high prevalence of hemoglobinopathies which helps in early diagnosis of high risk patients which can be managed vigilantly.

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