

## Pregnancy with Sickle Cell Anemia and Eclampsia

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**Abstract:** Pregnancy in women with sickle cell disease is a high-risk condition which is associated with increased maternal and fetal morbidity and mortality. We present you a case of sickle cell anemia in pregnancy complicated by eclampsia. Sickle cell anemia in pregnancy is not a common entity encountered. In India, according to hospital-based study, average frequency of sickle cell anemia in pregnancy is around 5%. Based on survey in Gujrat in various ethnic groups, prevalence of sickle cell anemia was 1.2% and sickle cell trait was 15.6%<sup>[1]</sup>. Specific management program and protocols are essential for better outcome of pregnancy in women with sickle cell disease.

**Key Words:** Sickle cell anemia, Pre-eclampsia, Eclampsia, Pregnancy

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### I. Introduction

Sickle cell disease is most commonly inherited single gene autosomal recessive hemoglobinopathy with structural abnormality affecting the Beta chain of Hb A and is known to give rise to life threatening complications. About 300,000 children with sickle cell disease are born every year.<sup>[2,3]</sup> Sickle cell disease is common in Africa, Middle east countries, parts of India, Mediterranean countries, Caribbean islands and also in South and Central America. Sickle cell disease in pregnancy is commonly encountered these days because more women are expressing their desire to reproduce.<sup>[4]</sup> This condition has adverse effects on pregnancy such as miscarriage,<sup>[5]</sup> prematurity, fetal loss, Intra-uterine growth restriction, increased incident of pre-eclampsia, recurrent infection, sickle cell crisis and post-partum hemorrhage therefore needing antenatal hospitalization.<sup>[6]</sup> There is increased incidence of pre-eclampsia in patients of sickle cell anemia. However, the cause is still not clear but at present researchers believe that clotting factors and inflammation may have a role to play. We describe a case of sickle cell anemia complicated by eclampsia during pregnancy. Pre-eclampsia complicated by generalized tonic clonic convulsions is referred as eclampsia. Sickle cell anemia often gives rise to painful vaso-occlusive crisis due to vascular occlusion by capillary thrombosis leading to infarction. Organs commonly affected are kidney (renal medulla), hepatosplenomegaly, lung (infarction), heart (failure), neurologic complications (seizures, stroke) with superadded infections. One such case is presented here.

### II. Case Report

An 18-year-old woman presented to MGM Hospital, Navi Mumbai, as a Primigravida with 35 weeks gestation with complaints of generalized weakness. On physical examination, patient looked pale, her pulse rate was 100 beats per minute, blood pressure was 110/70 mmHg. Systemic examination was found to be normal. Patient was advised admission and all related investigations were sent. Strict antenatal monitoring was done to ensure fetal well-being.

Laboratory values were as follows:

#### COMPLETE BLOOD COUNT:

Haemoglobin 7.8gm%

Total leucocyte counts 6300/mm<sup>3</sup>

Platelet 1.96/mm<sup>3</sup>

#### LIVER FUNCTION TEST:RENAL FUNCTION TESTS:

Sr. Bilirubin (Total) 0.28mg/Dl

Sr. Bilirubin (Direct) 0.14 mg/dL

SGOT 43.5 IU/L

SGPT 16.5 IU/L

Sr. Urea 14.3 mg/dL

Sr. Creatinine 0.7 mg/dL

Sr. Uric acid 3.8 mg/dL

Alkaline Phosphatase 166.7 IU/L

Serum Ferritin 40.5 ng/ml (Normal range is 11-300 ng/ml), Serum Iron 54 microgram/dL (Normal range is 60-180 microgram/dl), HPLC of Hemoglobin suggestive of sickle cell anemia and peripheral smear showing microcytic hypochromic anemia with sickle cells. Coagulation profile and ECG were normal. High Performance Liquid Chromatography of spouse was within normal limits.

The patient was transfused with one packed cell with strict monitoring of pulse, blood pressure and fetal heart sound throughout the transfusion. Obstetric ultrasound showed a single live intrauterine pregnancy of 32 weeks with breech presentation with early changes of utero-placental insufficiency. Patient has 3 episodes of fever 24 hours after blood transfusion with complaints of minimal pain in abdomen. Prophylactic antibiotic in form of Inj. Ceftriaxone 1gram twice a day with adequate antipyretics and analgesics were started. Laboratory investigations tested negative for dengue, malaria and typhoid. Blood and urine culture sensitivity tested negative for microbes. However, with due course of treatment, fever subsided within 48 hours.

During the hospital stay, there was a rise in blood pressure up to 160/100mmHg for which T. Labetolol 100mg twice a day was started. Repeat obstetric ultrasound was done after a week showed a single live intrauterine pregnancy of 33 weeks 3 days with breech presentation with growth lag of 3 weeks with symmetrical intra-uterine growth restriction. Two doses of betamethasone given to patient and elective lower segment caesarean section was planned at 37 weeks of gestation.

Patient was maintaining blood pressure of 130/90 mmHg on Tab. Labetolol 100mg twice daily but later had an episode of convulsion. On examination, her pulse rate was 106/minute, blood pressure was 150/90mmHg after convulsion. In view of convulsions and high blood pressure, diagnosis of Eclampsia was made, and the patient was placed on MgSo4 as per 'Pritchard regimen'. She was given 4g of magnesium sulphate as a 20% solution intravenous, followed by 10 g of 50% magnesium sulphate solution, injected deeply intramuscular in upper outer quadrant of each buttock. Decision was made to take her for emergency LSCS under spinal anesthesia. She was kept warm with adequate hydration during intra-operative period. Patient tolerated the procedure well. However, she had another episode of convulsion in post-operative room and was then shifted to ICU.

Inj. Levetiracetam was started and MRI Brain was done after neurology opinion. MRI Brain was suggestive of acute non-hemorrhagic infarct in right high frontal lobe associated with posterior reversible encephalopathy syndrome. Tab. Aspirin 75mg was started and to be continued up to 6 weeks postpartum. Laboratory investigation showed a fall in hemoglobin from 8.7gm% to 6.4gm% for which two packed cells was transfused.

Within 48 hours of ICU stay, patient's condition improved after joint management and was shifted to ward maintaining blood pressure of 120/80mmHg without anti-hypertensives hence, labetalol was tapered off. Patient was discharged on post-operative day 10.

### **III. Discussion**

Patient was admitted in view of breathlessness which was suspected to be secondary to anemia. As sickle cell disease is associated with maternal complication such acute chest syndrome, stroke (both infarctive and haemorrhagic), acute anemia leading to fetal complications, patient was given adequate oxygen supply and blood transfusion to avoid hypoxemia. Iron supplementation is advised only if there is laboratory evidence of iron deficiency anemia. The pathophysiology is probably a consequence of polymerization of abnormal hemoglobin in hypoxic situations, leading to formation of fragile and rigid sickle cells. These cells undergo easy breakdown leading to hemolytic anemia and vaso-occlusive crisis.

This patient also had an additional complaint of recurrent burning micturition and bacteria present in urine routine and microscopy on admission. These patients are generally hypo-splenic, therefore, immune-compromised and are at risk of infection. Patient was started on course of nitrofurantoin for 5 days. UK guidance is that daily penicillin prophylaxis is given to patients with sickle cell disease with guidelines for all hypo-splenic patients.<sup>[7,8]</sup>

Mild pain was managed with adequate rest, oral fluids and paracetamol or weak opioids. An adequate hydration of 60ml/kg was advised in 24 hours intravenously along with plenty of hydration. During the lower segment caesarean section, adequate warm fluids were given to the patient intravenously to avoid crisis. Women with sickle cell disease should avoid precipitating factors like heavy exercise, exposure to extremes of temperature and dehydration.

Low dose aspirin 75mg once daily is generally started from 12 weeks of gestation to reduce the risk of pre-eclampsia. As this patient was not registered with any medical center, early detection of pre-eclampsia and intrauterine growth restriction could not be done in earlier trimester.

Patient had episodes of convulsion precipitated by thrombus formation during ante-natal period at 37 weeks when elective lower segment caesarean section was planned and also post-partum. Generally, low-molecular weight heparin is advised during ante-natal hospital admission to reduce risk of thrombosis and in

cases of acute painful crisis. Anticonvulsant drugs and anti-platelet agents were started for this patient. Low-molecular weight heparin and aspirin was advised for a period of 6 weeks following caesarean section.

Post-partum investigation showed falling trend of hemoglobin for which two packed cells was transfused to the patient to avoid further deterioration. "Top-up" transfusion is recommended in acute anemia. A hemoglobin level of 6gm% or a fall of about 2gm% from baseline indicates towards need of transfusion. Exchange transfusion is indicated for acute stroke.<sup>[9]</sup>

This patient who was a case of sickle cell anemia with eclampsia along with infarct which was probably precipitated because of thrombosis. However, strict vigilance and timely management helped patient to recover.

#### **IV. Conclusion**

Hospitals should have a clear protocol for management of pregnancy with sickle cell disease. A multi-disciplinary team should be appointed comprising of obstetricians along with hematologist. However, recent advanced techniques and achievement in fields of prenatal and preimplantation genetic diagnosis, proper antenatal care with strict vigilance has led to good maternal and fetal outcomes in cases of sickle cell anemia. Pre-eclampsia and eclampsia are associated with maternal complications and protocols for early detection and management should be instituted from basic health care level for good maternal and perinatal outcome.

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