



Sickle cell disease in pregnancy

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Abstract

Sickle cell disease (SCD) is a group of inherited single gene autosomal recessive disorder caused by single gene, which affects haemoglobin structure. SCD has its origin in sub-Saharan Africa and Middle East, hence it is most common in people of African descent, as well as in the Caribbean, Middle East, parts of India, South and Central America. Sickle cell anemia in pregnancy need to be addressed and has to be managed by both obstetric team and haematologist in co-ordination. Preconceptional counseling plays a key role in decreasing maternal and fetal complications in sickle cell anemia in pregnancy.

Keywords: sickle cell disease; pregnancy; preconceptional counseling; haemoglobinopathy

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Introduction

Sickle cell anaemia, or SS haemoglobinopathy, is a result of sickle cell gene homozygosis. Sickle cell disease also includes haemoglobin (Hb) S interactions with haemoglobin variants other than normal HbA. The most common Hb variants are C (SC haemoglobinopathy), D-Punjab, and the co-inheritance of beta thalassaemia trait (HbS/ β -thalassaemia) [1-3].

SCD in pregnancy is associated with both fetal and maternal complications and is associated with increased incidence of perinatal mortality [4-9], premature labour [4-10], fetal growth restriction [4-11], and acute pain crisis in pregnancy [4-7, 12, 13]. Studies also described an increase in spontaneous miscarriage [10], antenatal hospitalisation [11], maternal mortality [14], delivery by C-sections [11, 14], infection, thromboembolic events [15], antepartum haemorrhage [14].

Case presentation

A 22-year-old primigravida with 28 weeks of gestational age, known case of sickle cell anaemia

(homozygous SS) was referred from peripheral hospital in view of per vaginal bleedings since 12 hours (partially soaked 1 pad) associated with cramping lower abdominal pain, intermittent in nature and obstetric scan was suggestive of single live fetus in cephalic presentation of 28 week gestational age with placental infarcts.

Patient was a booked case in a peripheral hospital, Adilabad. She was diagnosed with sickle cell anemia at the age of 11 years and was not on any treatment for the same. She was started on hydroxyurea by the doctor after confirmation of pregnancy. She was taking iron, folic acid and calcium supplements throughout her antenatal period. Tetanus prophylaxis was done for the patient in peripheral hospital.

On the morning of 20th March 2016, she presented with, bleeding per vagina and abdominal pain, to the same hospital. Ultrasonogram (USG) was done, which showed single live fetus with composite gestational age corresponding to 28 weeks and estimated fetal weight, 984 grams. Fundal placenta had features suggestive of infarcts and fetal heart rate was 98/min. She was referred to KIMS hospital, Secunderabad for further management. At the time of presentation she was perceiving fetal movements well. There was a history of easy fatigability and generalised weakness.

There was no complaint of per vaginal bleeding during the pregnancy, earlier to this episode. There was no history of trauma or fever during pregnancy. There was no past history of acute chest pain or abdominal pain.

Her last normal menstrual period was on 9th September 2015 which corresponded to 26 weeks of gestational age but as per early scans she was 28 weeks, so it was considered as wrong LMP. She spontaneously conceived after one year of active married life, and it was a non-consanguineous marriage. Her previous menstrual cycles were regular with normal flow with no dysmenorrhea. There was no surgical history. There was no history of sickle cell anemia or other co-morbidities in the family.

On examination, patient was conscious and coherent. General condition was fair but clinically pallor was present. Pulse rate was 96/min and

oxygen saturation was 98% at room air, other vital parameters were normal. Systemic examination was unremarkable. Abdomen was globular; on palpation it was found to be tense, with uterus of 28 week size, with cephalic presentation. On auscultation fetal heart sound was present on left ischio-umbilical line. Emergency obstetric scan was done which showed single live fetus in cephalic presentation of 28 week gestational age, placenta was seen in fundal posterior position. Multiple densities with altered echotexture of varied sizes were noted in the placenta, most likely to be placental haemorrhage/ infarcts. She was shifted to medical intensive care unit (MICU) for close observation. Corticosteroid prophylaxis was given to the patient and was started on higher antibiotics. Patient was given one blood transfusion and one unit of fresh frozen plasma in view of severe anemia with haemoglobin 7.5 gm%. She was started on oxygen support. Second hourly fetal heart monitoring was done. Next day morning fetal heart sounds could not be detected, and an obstetric scan was done which showed intrauterine death (IUD). Patient was shifted to ward for further management. Patient was managed further in coordination with haematologist. Adequate hydration was maintained, oxygen support was given, hydroxy urea 500 mg was continued with folic acid supplementation, and extremes of temperature were avoided for the patient. Two more units of blood transfusion were given subsequently.

She was induced with prostaglandin. During induction patient developed transient tachycardia with hypotension and chest discomfort, in view of which she was shifted back to MICU. Bedside 2D ECHO showed mitral regurgitation, mildly dilated left atrium, normal sized chambers, good left ventricular and right ventricular function. In MICU close monitoring was done for the patient.

In MICU, she delivered a dead female fetus of 900 gms with one liter of retro-placental clots (Figure 1) at 2:50 am on 23rd March 2016. Post expulsion, patient was observed in MICU for 6 hours, after stabilization she was shifted back to ward. Patient was observed in ward for 4 days and was discharged on 5th day in stable condition with haemoglobin of 11.1 gm%. She was put on antibiotics with an advice to continue hydroxyurea 500 mg twice a day along with folic acid. She was counselled to avoid pregnancy for the next 6 months and was advised copper-T insertion after 6 weeks post-partum. She was given prenatal counselling.



Figure 1: Shows placenta with retroplacental clots.

Patient was reviewed in outpatient department (OPD) after one week and patient's condition was fair. Haematologist advised her to continue hydroxyurea 500mg twice a day, folic acid supplementation and review after a month.

Discussion

Pregnancy exacerbates the preexisting pathophysiological characteristics of sickle cell disease (SCD): anemia, increased risk of infection, vaso-occlusion and pro-coagulant profile [16]. Interestingly the plasma levels of the placental growth factor, which rise throughout the pregnancy, are already elevated at baseline in SCD and have been associated with the frequency of acute pain episodes [17]. Therefore pregnancy is associated with increased incidence of painful crisis, infections, pulmonary complications, thromboembolic events, and antepartum bleeding, even in women who previously had few symptoms [18].

Awareness of these risks make it mandatory to manage these patients in centres where excellence in obstetrics, sickle cell disease, intensive care and transfusion management are available. A preliminary step must be pre-conceptual counseling, testing the partner, and clearly explaining the risks of pregnancy. A complete work up of clinical condition of the mother is mandatory, including heart, lung, renal and ophthalmology examination. Pregnant women need to be followed by both obstetric team that has knowledge in the treatment of women with SCD and also by sickle cell team.

More research is needed, with good quality, population based observational studies and prospective designs, that are large enough to have sufficient power to assess rare outcomes, to ensure quality of case ascertainment and quality of information collected. A better understanding of the reasons for the death of mothers and babies will allow relevant strategies for interventional studies [19].

Conclusion

Sickle cell anemia in pregnancy needs to be addressed and has to be managed by both obstetric team and haematologist in co-ordination. Pre-conceptional counseling plays a key role in decreasing maternal and fetal complications in sickle cell anemia in pregnancy.

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Conflict of interest

There are no conflicts of interest.

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