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Case Report

Successful delivery in a case of sickle cell disease with bilateral avascular necrosis: a case report

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ABSTRACT

Sickle cell disease is an inherited hemoglobinopathy. The patients with sickle cell disease had a small life years, before the advancement of medical science and development of newer drugs. These have improved the life expectancy among the sickle cell disease patients. Females are reaching reproductive age and are expectant of becoming mothers. But pregnancy in a sickle cell disease carries major fetal and maternal complications. Managing the pregnancy from conception to delivery is a difficult task and needs a team effort. Here we presented a case report of a female who got pregnant without any planning and was already on hydroxyurea. On top of that she was not willing to get her pregnancy terminated.

Keywords: Sickle cell disease, Bilateral avascular necrosis, Hydroxyurea, Pregnancy in sickle cell disease

INTRODUCTION

Sickle cell disease (SCD) is a common hemoglobinopathy that is inherited. The risk of complications with this disease is high, leading to early mortality. Earlier the life expectancy in these patients was low, but now with the advancement in medical science and development of newer drugs, the life expectancy has increased and some even reaching age more than 50 years.[1] With increasing life years, women are reaching their reproductive age and are wanting to conceive. Pregnancy in patients with SCD is not a smooth ride. It is associated with adverse maternal and fetal outcomes. High maternal and fetal morbidity and mortality are seen in SCD, such as, preterm delivery, intrauterine growth restriction, abortions, not appropriate for gestational age, neonatal death, etc. Above that if the patient is suffering from any other comorbid condition, it becomes much more difficult in managing such patients. Pregnancy in women with SCD is a challenge and requires a team effort of a gynaecologist, fetal medicine specialist and an hematologist. These women are followed from preconception to the delivery following strict guidelines for a successful delivery. The field of fetal medicine equipped with advanced prenatal diagnosis and preimplantation genetic diagnosis is proving a boon in delivering a healthy baby in patients with SCD.

Here we presented a case study of a pregnancy in a young female suffering from SCD having associated avascular necrosis of bilateral femoral head.

CASE REPORT

A 25-year-old woman presented to us with 6 months amenorrhea on 18 December 2020. She was a known case of SCD on hydroxyurea. She became pregnant without planning. Since last 5 years she was taking treatment for SCD from a hematologist and had suffered multiple complications due to microvascular thrombi. Hematologist advised that hydroxyurea should be stopped 3 months prior to planning for conception and since pregnancy had occurred without planning, the patient should undergo

medical termination of pregnancy. He also explained about the teratogenic effects of hydroxyurea and then asked the patient to seek consultation with a fetal medicine specialist.

Her obstetric ultrasound was done which showed single live intrauterine fetus of 21.4 weeks gestation, weighing around 409 gm, with no obvious fetal defects. Her cervical length was ≤25 mm with normal uterine artery Doppler. She was advised for medical termination of pregnancy, but the patient and the family were not willing for termination and wanted to continue with the pregnancy.

So, a team comprising of a gynaecologist (myself), a fetal medicine specialist and a hematologist (already taking take of SCD) decided that we will put our best to get the optimal fetal and maternal outcomes in this case.

It was in June 2016, that the patient complained of fatigue on exertion with periodic episodes of pain. She consulted a hematologist. Her hemoglobin was very low (Hb 7.9 gm%), raised TLC (36600 /cumm) and Rh (D) was positive. Her abdominal sonography revealed non-visualization of spleen due to infarct/atrophy secondary to SCD. She underwent MDCT of abdomen, which revealed small shrunken spleen (likely secondary to hemolytic disorder). Mild short segment dilatation of one of the ileal loops non-specific, serro-adhesive. A diagnosis of beta SCD was made, patient was given hepatitis B and pneumococcal vaccine and started on hydroxyurea 500 mg.

Then in October 2016, patient had complaint of difficulty in walking with severe pain in the hip region. MRI hip showed bilateral femoral head avascular necrosis (left side grade 3 and right side grade 1). She was put on medication. She was maintaining well and on proper medication schedule.

Then in December 2020, she presented with 6 months amenorrhea (unplanned pregnancy). Patient was admitted in hospital due to cervical incompetence. McDonald's stitch was given in view of short cervix (≤25 mm). She was in 22.1 weeks of gestation. Hb electrophoresis done was normal.

In January 2021, she contracted vaginitis, which was managed conservatively. Growth scan done showed normal fetal growth, amniotic fluid volume and fetal dopplers. Good fetal movement on the scan. 2D Echo was also normal.

In March 2021, investigations done showed raised ESR (52 mm at the end of first hour), raised serum ferritin (398 ng/ml), raised iron level (193 ug/dl) and rest of TIBC parameters were normal. Hb was low (9.0 gm%), raised TLC (26000 /cumm), high neutrophils (86%) and low lymphocytes (10%).

Her pregnancy progressed gradually and on 15 March 2021 high risk lower segment cesarean section was planned. Ultrasound obstetric done on 25 March 2021 showed single intrauterine gestation corresponding to a gestational age of 34 weeks 6 days with cephalic presentation and normal liquor. She was admitted on 26 March 2021. Her vitals were stable, fetal heart rate was present and uterus was relaxed. Cervical sutures were removed on 26 March 2021 and 1 unit of packed cell transfusion was given.

On 27 March 2021 she underwent lower segment cesarean section and delivered a healthy 2.5 kg female child. Both the mother and baby were safe and healthy.

No postoperative maternal and fetal complications were noted.

DISCUSSION

SCD is an inherited autosomal recessive disorder and affects the hemoglobin structure. The hemoglobin cell takes the shape of a sickle, thereby reducing the oxygen carrying capacity.[2,3] These cells are sticky and many cells stick together and blocks the small blood vessels, affecting the normal movement of the oxygen and its delivery. The blockage also causes pain. These sickled cells also damage the spleen, thereby increasing the risk of contracting infections.[4] This disease is very much prevalent in parts of India.

Pregnancy in a SCD carries high maternal and fetal complications. Higher perinatal complications are due to reduced placental circulatory flow.[5] Some studies have reported increase in maternal mortality, thromboembolic events, antepartum hemorrhage and necessitating delivery by cesarean section.[6,7]

The management of pregnant women having SCD requires more planning and a team effort from preconception to delivery. But in our case, the patient was already pregnant without planning and hence, it was more difficult to plan and make a guideline. Also, she was on hydroxyurea prior to her conception, which could have complicated the pregnancy further.

Our patient had received blood transfusion during the pregnancy without any transfusion related adverse events, but there is a controversy on whether blood transfusion be given prophylactically in these women with SCD.[8] With blood transfusion, there is reduction in circulating sickle cell hemoglobin and increase in normal hemoglobin in the blood stream.[9]

In our case, the patient experienced only few complications during the pregnancy period, while the study done by Elenga et al reported higher prevalence of obstetric complications in their SCD women.[9]

The limitation of the present case was that the patient had not contacted the hematologist prior to the planning for conception, or should have come within 1-2 months of pregnancy confirmation as would have given the team a better planning for the patient. Additionally, patient was suffering bilateral avascular necrosis of femoral head, this added to our burden of planning. Even though the odds were against us, the final outcome was good.

CONCLUSION

Pregnancy in a SCD patient is a roller coaster ride. The patient may need many hospitalizations and consultations till her delivery. In such case, a team effort of a gynaecologist, fetal medicine specialist and a hematologist is very essential for a good maternal and fetal outcome.

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