

Thalassemia Major with Central Placenta Previa

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Introduction

Thalassemia major also called as Cooley's anemia was first described by Cooley and Lee in the year 1925. It has a co-dominant inheritance and its pathology lies in the decreased synthesis of beta globin chains resulting in an increased production of alpha chains, which subsequently leads to red cell destruction, ineffective erythropoiesis and anemia.

Statistics reveal that 250 million or 45% of the world's population are affected by thalassemia; out of these, 3.5% of the population in India are carriers of thalassemia. Over 1,00,000 people with thalassemia major are born annually worldwide of whom 8,000-10,000 are Indians. It is a common disorder with a widespread geographical distribution. It is a cause of concern for obstetricians as it complicates motherhood.

Since repeated blood transfusions are required in thalassemia, there is iron overload which in turn causes deposition of iron in the hypothalamus and the pituitary causing reproductive axis failure leading to delayed puberty, delayed sexual development and infertility. A known case of beta thalassemia major carrying a pregnancy to near term with a successful outcome is rare.

Case Report

Mrs. M, a 20 year old primigravida with 8 ½ months amenorrhea, a known case of beta thalassemia major on regular blood transfusions right from the age of one year, first came for the antenatal registration on the 18th of August 2000. Her menstrual history suggested that her last menstrual period was on 20th March, 2000 and her EDD was 27th of December, 2000. Her subsequent visits were quite irregular and she came on the 20th of November 2000 complaining of vaginal spotting for four days. She did not complain of pain in the abdomen and increase or decrease in fetal movement. Both her parents suffered from thalassemia minor and she had lost one sibling in childhood. She had received about 500 blood transfusions

and was irregular with iron chelation therapy. She had undergone splenectomy in childhood.

General examination revealed that vital parameters were within normal limits but for her pallor. Systemic examination did not reveal any abnormal findings. On abdominal examination, the uterus corresponded to 30 weeks gestation, with breech presentation. Fetal heart sounds were heard well and were regular. On speculum examination there was no evidence of bleeding. She was clinically diagnosed as a case of thalassemia major with 32 weeks pregnancy, IUGR, breech presentation and placenta previa. Ultrasonography showed 30 weeks pregnancy with central placenta previa type IV.

She was admitted and being a high-risk case, was closely monitored with regular non-stress tests. Tocolytics and antibiotics were started. After an uneventful period of a few days with minimal bleeding, she bled heavily on 28th of November 2000 warranting LSCS at 1.40 PM on the same day. A female baby was delivered without congenital anomalies weighing 1.56 kgs and having 15 gm of hemoglobin. Two units of blood were given intraoperatively to account for the operative blood loss and the underlying chronic anemia. The postoperative period was uneventful though during her hospital stay she received five units of blood. The baby was admitted in NICU in view of prematurity and low birth weight. Mother and baby were discharged in good condition on 27th December, 2002 in good condition. The baby weighed 2 kgs at the time of discharge. She had Thalassemia minor. Both baby and mother were fine when last seen on 17th September, 2003.

Discussion

Due to reduced fertility and short life expectancy, pregnancy is rare among homozygous β -thalassemia patients. During the last three decades, because of prolonged life expectancy and improvement in the quality of life by extensive blood transfusions and treatment with chelating agents a few pregnancies have been reported.

Since the first report in 1969 by Walker and Whelton¹, others have described their experience with homozygous β -thalassemia and pregnancy²⁻⁵. Our patient was totally dependent upon blood transfusion throughout her life. She had undergone splenectomy in childhood.

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Both the thalassemic pregnant mother and her fetus may face deleterious complications which should be anticipated, prevented and promptly treated. In thalassemic patients, the combination of severe anemia, chronic hypoxia and iron overload causing hemosiderosis eventually leads to cardiac arrhythmia and congestive heart failure. Cardiovascular changes related to pregnancy (anemia, increase in plasma volume and cardiac output) may aggravate cardiac failure in these patients. It is therefore essential in pregnant thalassemic patients to maintain a reasonably high mean hemoglobin concentration and prevent cardiac compromise. These goals are achieved with a strict transfusion regimen maintaining the hemoglobin above 10 gm/dl and thorough cardiovascular surveillance.

Severe anemia and hypoxia may also affect placental gas exchange thus altering intrauterine fetal growth and development. The relatively high rate of pregnancy loss and prematurity as well as the intrauterine growth retardation may result from these alteration. In spite of all these complications successful pregnancy outcome is possible.

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