ORIGINAL RESEARCH PAPER

Obstetrics & Gynaecology

A RARE CASE OF PRIMI GRAVIDA WITH HEREDITARY SPHAEROCYTOSIS

KEY WORDS:

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INTRODUCTION

Hereditary spherocytosis (HS) is a common inherited erythrocytic membranopathy , mostly presenting as hemolytic anemia. Here we are presenting acase of a PRIMIGRAVIDA woman, who was a diagnosed case of hereditary spherocytosis requiring multiple blood transfusions in the antenatal period.

Hereditary spherocytosis (HS) is a common type of hemolytic anemia. In an unselected population, the frequency is around 1 in 5000. The problem lies in the red cell membrane proteins; spectrin, ankyrin, band 3 protein and protein 4.] The abnormal red cell membrane proteins give rise to an unusual susceptibility to lysis. Peripheral smear shows spherocytes. Most individuals have mild or only moderate disease. Typical cases present with family history and in 75% of patients, inheritance is autosomal dominant. The disease can affect pregnancy in many ways.

Here we are presenting a case of primigravida 37.6weeks of gestation with PROM WITH hereditary spherocytosis.

OBJECTIVE- HOW TO DEAL WITH A CASE OF HEREDITARY SPHAEROCYTOSIS IN PREGNANCY .

ANTENATAL HISTORY

A 30 year old patient, PRIMIGRAVIDA,married for 3.5 years registered in antenatal outpatient department at 9 weeks of gestation with a hemoglobin of 8.5 gm %. She was diagnosed as a case of hereditary spherocytosis 8 years back. She was icteric. Her father was initially diagnosed with HSas he had landed in haemolytic crisis and splenectomy was done,post that she and her 3 sisters were diagnosed cases of HS and 1 of of them had undergone splenectomy also. Hematologists were consulted and they advised continuation of 1000 mcg of folic acid and regular monitoring of red blood cell indices. ** The current pregnancy was a spontaneous conception. At 11 weeks of gestation, she presented with complaints of slight breathlessness and fatigue. On examination, pallor was present, and pulse was around 100 bpm, with a respiratory rate of 26 cycles per minute. Abdominal examination showed mild spLENOMEGALY and a live fetus with no uterine activity. She was admitted for blood transfusion and monitoring. Hemoglobin was 7.2 gm % and platelet count was 100,000/ cc and serum bilirubin was SLIGHTLY elevated. She was stabilized with diuretics, and non-invasive oxygen support. Haematologist was consulted and The diagnosis of early hemolytic crisis was considered and she was transfused with 1 unit packed red cells over 2 days. Post transfusion hemoglobin was 8.6 gm %. She was then discharged. She followed up in the antenatal outpatient department AND was otherwise asymptomatic.

At 36 weeks of gestation, she came with pain in abdomen and leaking per vaginum. Abdominal examination showed live fetus with regular heart rate pattern and good uterine activity. Vaginal examination she was 1fl at internal os with a vertex presentation.cerviprime induction with trial of labour was given to the patient LDH 654 She was eventually taken up for C SECTION IN VIEW OF prolongeg pprom with failure of induction with poor bishops score . and delivered a female child of 3176 gm with APGAR score of 9/10. Neonate was shifted to NICU for weight gain and screening. Mother was given one unit packed red cells peripartum. Postpartum period was otherwise uneventful and she was discharged on day 7 of post op. Pt was advised tab flovite 5 mg bd to be continued And cap homocheck bd for 1 month

Discussion

HS IS AN AUTOSOMAL DOMINANT DISORDER .Pregnancy in HS is usually uneventful. However, some specific problems can arise. . In the case series by Pajor et al, they had noticed a 68% rate of term birth and 21% rate of spontaneous abortion. In our patient, there was one episode of early hemolytic crisis in the 19 week of gestation, necessitating blood transfusion. Pregnancy is a well known trigger for hemolytic crisis, as shown by HO-Yen et al.

In the case series by Pajor et al, hemolytic crises was seen in 30% of the cases. In our patient, apart from anemia and elevated liver indices , there were no major problems. In the case series by Maberry et al also, the main problem that was encountered was anemia. patients can developheart failures The causes of cardiac failure in spherocytosis can be severe anemia, and rarely due to cardiac hemosiderosis, as described by Fujino et al.[8] Our patient overcame the hemolytic crisis without requirement for splenectomy. However, two cases of hemolytic crisis necessitating splenectomy have also been described in literature.They have mentioned that pregnancies with HS, who have undergone splenectomy have lesser problems compared to those without. Association between hydrops fetalis and spherocytosis has also been described.[10] usually overall neonatal outcomes are good, as opined by Maberry et al.[6] The neonatal OUTCOME WAS GOOD AND no features of hydrops fetalis. Patient was to asked to follow up after week with cbc report