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Case report

PREGNANCY OUTCOME IN POST SPLENECTOMY PATIENT WITH HEREDITARY SPHEROCYTOSIS – A CASE REPORT

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Abstract- Hereditary spherocytosis (HS) is a rare type of hemolytic anemia in southeast-Asian population whereas it is common among western countries like north Europe. Hereditary Spherocytosis (HS) is most common red cell membrane disorder with varying degrees of clinical severity, membrane protein defect and modes of inheritance. In majority of cases transmission is autosomal dominant .Severity of the disease is classified as mild, moderate and severe forms. 3-5% of the patients have severe disease and often complicated by cholelithiasis. Though severe form is an indication for splenectomy presently evidence favouring the conservative approach to splenectomy. There are only few case report series concerning the pregnancy complicated by HS. Though hemolytic crisis was managed conservatively in some cases, outcome of pregnancy was more favorable after splenectomy than before. We report a case of HS with post splenectomy complicated pregnancy with good maternal and fetal outcome.

Keywords- Hereditary spherocytosis, Splenectomy, Cholecystectomy.

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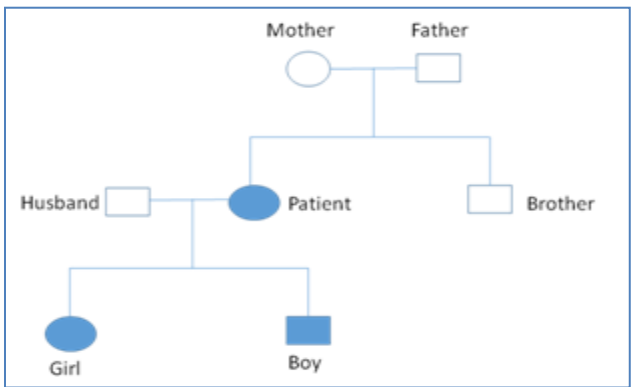
Introduction

Case report:

Mrs. M, 25 years G2P1L1 attended our antenatal clinic for prenatal care at 18 weeks of period of gestation. She was a known case of hereditary spherocytosis and had undergone splenectomy. In her past history she had recurrent episodes of jaundice, fever with abdominal pain from five to six years of age onwards for which she was evaluated and diagnosed to have hereditary spherocytosis at 10 years of age. Following this she underwent splenectomy and cholecystectomy for severe disease and cholelithiasis. Since then she was on penicillin prophylaxis. No similar complaints in family [1,2].

Pedigree chart



As per history and available data all her family members were evaluated and found to be unaffected with hereditary spherocytosis. Her first child was 4 and half years, female child. Her antenatal period was uneventful then. No history of blood transfusion. She had spontaneous onset of labor at term and it was a normal vaginal delivery. Baby developed anemia and jaundice at 1 ½ years of age and was diagnosed to have hereditary spherocytosis. Since then baby was on regular blood transfusions. In present pregnancy, she attended our OPD at 18 weeks and 2 days period of gestation. Since then she was been on regular antenatal checkups. All her investigations were all normal and she maintained her hemoglobin between 10-12 gm% [Fig-1]. Sonological study showed normal fetal growth. She had history of fever at 37 weeks and 2 days for which she got admitted and evaluated. Investigations were normal and she became a febrile in two days. Later, she had spontaneous onset of labor at 38 completed weeks. Her labor progressed well and delivered a live term male baby with weight of 2.8 kg. No postnatal complications. Baby developed icterus 31 hours after birth, which progressed and baby became anemic. On 8th postnatal day baby was transfused with packed cells and was tested positive for hereditary spherocytosis [Fig-2]. Later baby improved and was discharged in good condition [3-5].

Discussion

HS is a hemolytic membranopathy and in majority the transmission is autosomal dominant and one parent usually has the disease. However, in one fourth of the conditions, the cases occur sporadically and half of these are genuine recessive forms. Remaining cases are possibly due to spontaneous new mutation. In our patient, there were no hematological abnormalities in any of the family member. She developed the disease sporadically and is transmitting in an autosomal dominant fashion, which is evident by affecting both her children with moderate disease. There are very few case series and reports concerning pregnancy complicated with hereditary spherocytosis. Hemolytic crisis can be the first clinical sign of the disease and its association with pregnancy is well established. But the

information regarding the course of the disease during pregnancy is limited. These hemolyticcrises can be controlled by blood transfusion, treatment of the precipitating cause or by splenectomy.

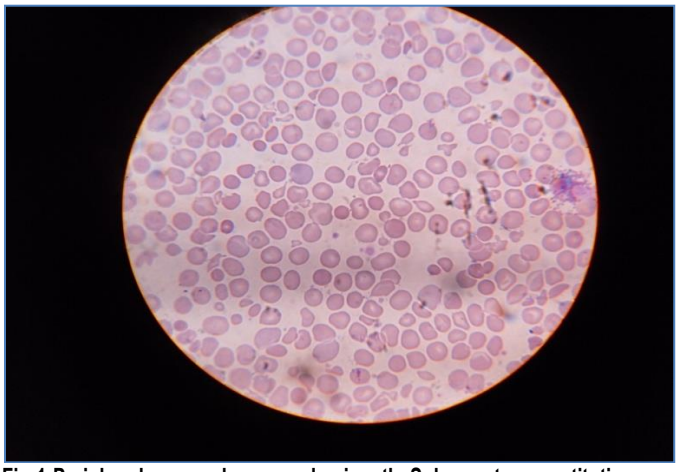


Fig-1 Peripheral smear shows predominantly Spherocytes constituting over 50% of the RBCs. Mild – 1-15/HPF, moderate 15-30/HPF, severe >30/HPF. Anisocytosis, schitiocytes and polychromatophils are seen.

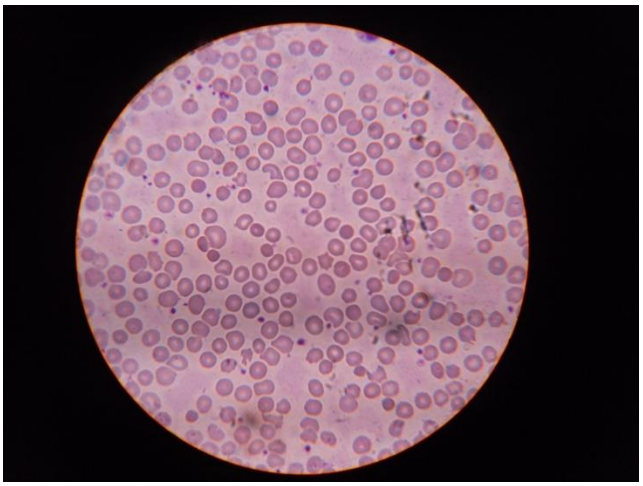


Fig-2 Peripheral smear shows predominantly microspherocytes (which are small RBCs without any central pallor). Spherocytes are uniform in size and density. RBCs show mild anisocytosis and polychromatophilia.

Moore [1] has reported two cases of splenectomy done during pregnancy and in his review 5 out of 23 had splenectomy for HS during pregnancy. In another review by Macberry none of the cases with hereditary spherocytosis required splenectomy during pregnancy [2]. Pajor [3] in his review of 8 patients has conservatively treated all patients with hematological support. Sometimes the clinical scenario can even warrant emergency splenectomy as reported by Allram [4].

In our case she has already undergone splenectomy during her childhood for severe disease and later was kept on antibiotic prophylaxis. Both her pregnancies were following splenectomy and were uneventful.

Gershovitz [5] analysed 150 women who had undergone splenectomy and reported splenectomy is significantly associated with pregnancy and labor complications, such as cesarean delivery, pneumonia during pregnancy, complications of anesthesia and sedation during labor. According to him, splenectomy is an independent risk factor for preterm delivery. But our patient never had any post splenectomy complications. So with splenectomy our patient had both subjective and objective benefits and had good pregnancy outcomes.

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