Case Report

Hereditary Spherocytosis with Pregnancy - A Case Report

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Introduction

Hereditary spherocytosis (HS) is a disorder of the red blood cell membrane protein that leads to sphere shaped red blood cells which are less resistant to stress and rupture easily leading to chronic hemolytic anemia. The incidence in northern Europe and America varies from 1:2000 to 1:50001. Indian incidence is not known. Both autosomal dominant and autosomal recessive modes of transmission have been identified. Diagnosis is made by anemia, increased reticulocyte count, MCHC, spherocytes in peripheral smear, elevated bilirubin and LDH levels, decreased MCV and negative ICT and DCT. There is increased osmotic and incubated fragility test. The confirmatory test for this disease is SDS-PAGE. The disease may be mild, moderate or severe. Mild disease needs no treatment. Moderate disease is treated with folic acid 5 mg/day, supportive treatment and regular annual check up. Severe disease warrants frequent hematological supervision and splenectomy. Information regarding the course of HS during

pregnancy is limited. We report a case of pregnancy with HS with massive splenomegaly detected during pregnancy. She had a successful laparoscopic splenectomy done during second trimester followed by successful pregnancy outcome.

Case Report

Mrs. R.J. 23-year-old female presented in the OPD in August 2004 complaining of generalized weakness, fatigue and sub fertility. She was married for four years, had normal menstrual cycles and one D & E done for blighted ovum four months back. There was a history of anemia for which she was not evaluated completely. On examination she had moderate pallor, no jaundice and no hepatosplenomegaly. The Hb was 8.6gm/dl, PCV-28.1%, TLC 6700/cmm, platelet count 89000/cmm, and serum iron studies and Hb electrophoresis were normal. Peripheral blood smear revealed features of megaloblastic anemia (Fig. 1) for which treatment was started. The patient reported back in Jan 2005 with 1 weeks of pregnancy. This time she was moderately pale, with no jaundice but had splenomegaly upto umbilicus. Her Hb, was 6.3gm/dl, PCV-22.5% MCV 107.1 fl, MCHC-28.0 gm/dl, reticulocyte count 15%, ICT & DCT were negative & Sr. LDH-779 IU. C3 and C4 level were normal. ANA was negative. Sr. bilirubin was 1.2mg% and osmotic fragility of RBCs was increased. A diagnosis of pregnancy with spherocytosis with massive splenomegaly and hemolytic crisis was made and a decision to have laparoscopic splenectomy was

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Table 1

Classification	Trait	Mild	Moderate	Severe
Hemoglobin (G/dl)	Normal	11-15	8-12	6-8
Reticulocyte Count (%)	Normal (<3)	3-6	>6	>10
Bilirubin (micromoles/l)	>17	17-34	>34	>51
Spectrin / erythrocyte (% of normal)	100	80-100	50-80	40-60
Splenectomy	Not required needed in childhood & adolescence	Usually not needed in childhood & adolesc.	Necessary in school age before puberty	Necessary delay until 6 years if possible

taken. She was transfused with four units of blood and vaccinated against pneumococcus, meningococcus and H. influenza. After two weeks of immunization laparoscopic splenectomy was done uneventfully at sixteen weeks of gestation. Retrieval of the spleen was done through left subcostal incision (weight of spleen 1.5 kg) with blood loss of about 300 ml. She was discharged on the fourth postoperative day. Penicillin-V 500 mg BD was given as postsplenectomy prophylaxis. After one week of surgery her Hb was 10.5 gm/dl, PCV 30% and platelet count was 420000/cmm. She developed PIH at 22 weeks of pregnancy and was started on tablet alpha methyl dopa 250 mg, thrice daily and was increased to 500mg thrice daily after 34 weeks

till delivery. From 34 weeks onwards she had significant proteinuria. So an elective LSCS was done on her at 36 weeks of pregnancy. She delivered a healthy male baby of 2.5kg. Her postnatal period was uneventful except that she had persistent hypertension which was controlled. She continued to have significant proteinuria even after the delivery for which she was under the care of nephrologist and eventually underwent renal biopsy in February 2006. Histopathology was suggestive of mesangio proliferative glomerulonephritis and she was put on steroids. Postpartum follow up was not available as the couple moved elsewhere .

Discussion

The methodology of diagnosis and clinical management of HS has changed over the past three decades. For many years making the diagnosis was usually followed almost automatically by splenectomy. There then followed a period when splenectomy fell from favor because of the risks of postsplenectomy sepsis. However presplenectomy immunization against *S. pneumoniae* (together with Hib and meningitis C vaccines if not already given as part of routine vaccination) together with prophylactic antibiotic therapy has reduced, although not totally eradicated, the risk. The disease may be mild, moderate or severe depending on various parameters which have been given in Table I. The indication for splenectomy depends on the severity of symptoms. Various authors have advised against splenectomy during pregnancy

Fig : Peripheral smear showing megaloblastic Anemia

because of the associated complications like serious sepsis, perioperative morbidity to the mother and the fetus and thrombocytosis and thrombosis, but none of the reported cases had such problems. Though splenectomy can be done at any time during pregnancy, second trimester is preferable. As such there is no consensus regarding the route but laparoscopic route should be preferred as there is less postoperative pain and ileus, less hospital stay and rapid return to normal activity. Immunization against pneumococcus, meningococcus and hemophilus influenza should be done at least two weeks prior to surgery. Antibiotic prophylaxis should be given for the first two years following splenectomy and life long in immunocompromised patients with phenoxymethyl penicillin-V 500mg twice daily or roxithromycin 150 mg daily (in case of penicillin allergy)¹

Moore² have reported two cases of pregnancy with HS who required splenectomy during pregnancy and reviewed 23 cases in literature out of which five had splenectomy during pregnancy. Maberry et al³ reported 23 cases with HS in 50 pregnancies but none of them required splenectomy during pregnancy. Pajor⁴ have reported eight patients with HS who were managed

conservatively with hematological support. Only once case of successful laparoscopic splenectomy in a morbidly obese pregnant woman with HS has been reported so far⁵.

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