Case Report

Pregnancy following splenectomy for spherocytosis

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Introduction

Although spherocytosis is an autosomal dominant trait in a fifth of the cases it appears to be due to new mutations.

Case report

A 27 years old primigravida was seen in out patient clinic when she was 16 weeks pregnant. She was a known case of spherocytosis. Because recurrent episodes of hemolytic anemia and jaundice she had multiple blood transfusions and underwent splenectomy at the age of 10 years. Since then she was put on tablet folic acid 5 mg daily and was given 1.2 mega units of injection benzathine penicillin intramuscularly once in 3 weeks for 5 years.

Following splenectomy she suffered from repeated worm infestations and had nearly 24 attacks of malaria over a period of 10 years. During the above period her hemoglobin ranged between 10 and 11 g/dL and the peripheral smear showed mild eosinophilia, moderate hypochromia, platelet clumps and microspherocytosis. None of her family members suffered from similar problems.

On examination she was not anemic and not jaundiced. Her pregnancy was corresponding to 16 weeks of gestation. In view of her previous medical history, besides the usual investigations she was periodically investigated for anemia, malaria and worm infestation. Throughout the antenatal period her hemoglobin ranged between 10.5 and 11 g/dL, PCV was 39% and leukocyte count 9,900/mm$^3$, with differential count P 65 L 29 E 6. Her peripheral smear showed microcytic hypochromic anemia with few spherocytes. She was given adequate folate supplementation and her pregnancy progressed well. As infection prevention is vital in cases that had splenectomy, she was given anthelmintics at 20 weeks, 28 weeks and 34 weeks and antimalarial prophylaxis with tablet chloroquine 300 mg once weekly from 20 weeks until 38 weeks of gestation.

She went into spontaneous labor at 39 weeks of gestation and delivered a healthy male baby weighing 2.7 kg. The baby was not icteric at birth, and had no evidence of splenomegaly. Cord blood showed hemoglobin of 26 g/dL and leukocytes 24,000/mm$^3$ with 83% polymorphs and 17% lymphocytes. Peripheral smear did not show spherocytosis. The postpartum period was uneventful and she was advised to continue with folic acid supplements. The baby was reviewed at
one year of age and was found to be healthy while hematological investigations did not show evidence of spherocytosis.

**Discussion**

Though in majority of cases, spherocytosis is inherited as an autosomal dominant trait, it has been shown that in about 20% of patients both parents are normal and it has been assumed that these cases are due to new mutations. Probably our patient also developed this condition due to new mutation as her parents, offspring and other family members were found to be normal.

In most of the cases spherocytosis is diagnosed during childhood and splenectomy is carried out. In these cases there are usually no significant maternal effects during pregnancy. As our patient had undergone splenectomy during childhood she did not develop significant antepartum or intrapartum complications. In those cases without splenectomy, life threatening aplastic and hemolytic crisis can occur during pregnancy because of accelerated red cell turn over. Megaloblastic anemia due to folic acid deficiency and hemolytic jaundice may present for the first time in pregnancy. These patients may require multiple transfusions and double the normal dose of folate supplementation.

Following splenectomy there is prompt cessation of hemolysis with return of hemoglobin to normal and disappearance of jaundice. Since our patient had splenectomy in childhood her hemoglobin remained between 10.5 and 11 g/dL throughout pregnancy. Agarwal et al reported a case where inspite of having had splenectomy previously, a pregnant mother presented with severe anemia and succumbed in the postpartum period.

In postsplenectomy patients, the major dangers are plasmodium falciparum malaria and other infections. Our patient was given injection penicillin for 5 years as prophylaxis against bacterial infection. However she developed recurrent episodes of worm infestation and malaria during her adolescence. In view of this she was given anthelmintics and antimalarial prophylaxis during pregnancy. It is recommended that in malaria endemic areas rigorous antimalarial prophylaxis must be given for life. Pneumococcal vaccination is also mandatory. Though the baby was reported normal at one year, her future offspring should be followed up to identify autosomal mode of inheritance so that genetic counseling can be offered.

**References**