A Case Report of a Post Cholecystectomy Patient with Anaemia, Jaundice and Splenomegaly

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Abstract

Splenomegaly with anaemia and mild jaundice are not an uncommon cases for haematologists but occasionally they may present with some other problems like cholesterolosis, choleodochochiasia, skin ulceration etc. If any patient presents with

Case report

A 30 year old gentleman hailing from Surup, Sagra was admitted in Delta Medical College Hospital in February, 2017 with the history of frontal headache, pallor, jaundice and recurrent pain in left upper abdomen and mildly enlarged liver and hugely enlarged spleen for the last 4 years. He denied any positive family history of same disease. He received 12 unit of red cell transfusion and had a history of cholecystectomy 10 years back due to cholecystitis.

At presentation lab investigations revealed haemoglobin 7.6g/dl, WBC 6x10^9/L, Platelets 200x10^9/L, BSR 50mm in 1st hour, reticulocytes count 3%, PT 14.3s, INR 3.90 mg/dl, SCFT 27 U/L alkaline phosphatase 110 U/L. Serum Parath level was 261.24µg/L, LmbsAg and Anti HCV were negative. Peripheral blood film revealed high number of spherocytes. Direct Coomb’s test was negative. Haemoglobin electrophoresis was normal. Genetic fragility test was increased. Abdominal ultrasound showed mild hepatomegaly and massive splenomegaly and no abnormal dilatation of intra or extra hepatic biliary tree. The diagnosis of hereditary spherocytosis was made and the patient underwent splenectomy after vaccination against pneumococcal and meningococcal. The spleen was measured measuring 30x20x15cm and biopsy showed thinned capsule with widening of splenic  uncle which was unexpected and with erythropoietic. The sinuses were empty and increase in connective tissue fibres were seen focally.

Discussion

Hereditary spherocytosis(HS) is one of the most common red cell membrane disorders of RBC membrane leading to chronic anaemia and a common cause of haemolysis and haemolytic anaemia, clinical features range from asymptomatic to fulminant haemolytic anaemia. The condition was first described in 1871. The prevalence of the disease in Bangladesh is not known but in Northern Europe and North America, with a reported incidence of 1/ 5,000 births. Most cases are inherited in an autosomal dominant fashion and approximately 25% of these

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Fig 3: Spleen after removal

cases may represent spontaneous mutations or recessive forms of the disease. Dysmorphology of erythrocyte, band 3, or the other structural proteins lead to common secondary defects in spectrin assembly, resulting in an unstable red cell membrane. 6, 7 In HS, RBC becomes less stable, and forms the ability to circulate freely through narrow capillaries in the body, as they trapped in the spleen and engulfed by macrophages causes haemolysis and ultimately develop gallstones. 8 In HS, clinical manifestations may vary. HS can present soon after birth. It should be suspected in infants who present with jaundice and anemia, as well as in adults with recurrent infections. In later childhood, HS can present with anemia, jaundice, and splenomegaly. 9. Older individuals develop hepatomegaly and may present with cholelithiasis. 10 Splenectomy is the definitive treatment of this disease, which should be performed after the age of 5 years to avoid increased risk of post-splenectomy septicaemia. 11 After splenectomy, patients may develop with anemia, neutropenia, and thrombocytopenia. 13, 14

Splenectomy is recommended for cholelithiasis, and during cholelithiasis, splenectomy is also indicated. The condition may develop at other sites of splenic tissue. 15

Before splenectomy, it is essential to screen the patient against different infectious diseases, with life-long vaccine. Vaccination against hepatitis B, pneumococcus, meningococcal meningitis, and tuberculosis is important. 13, 14

Conclusion
Cholelithiasis is a common problem in surgical practice but is very rare. If any patient present with this problem in early childhood evaluation before cholelithiasis is required because patients with hereditary spherocytosis are more prone to develop anaesthetic hazards and perioperative infection. This patient also requires splenectomy in the same sitting otherwise stones may form in any part of the biliary tree afterward.

References