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Case Report

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 cholelithiasis, cholecholelithiasis, skin ulceration etc. If any patient presentes with cholelithiasis to surgeon in younger age group evaluation for hereditary hemolytic anaemia should be required. Otherwise stone may develop in other sites in billiary tree which may be a serious complication.

Case report

A 30 year old gentleman hailing from serpur, Bogra was admitted in Delta Medical College Hospital in October, 2009 with the features of frontal bossing, 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 cholelithiasis.

At presentation lab investigations revealed haemoglobin 7.5g/dl, WBC $8x10^9$/L, Platelets $200x10^9$/L, ESR 30mm in 1st hour, reticulocytes count 6%, PT 14.2s, total bilirubin 3.90 mg/dl, SGPT 27 U/L alkaline phosphatase 110 U/L. Serum Ferritin level was 261.33µgm/L, HbsAg and Anti HCV were negative. Peripheral blood film revealed fair number of spherocytosis. Direct Coomb's test was negative. Haemoglobin electrophoresis was normal. Osmotic fragility test was increased. Abdominal ultrasonogram showed mild hepatomegaly and massive splenomegaly and no abnormal dilatation of intra or extra hepatic billiary tree. The diagnosis of hereditary spherocytosis was made and the patient underwent splenectomy after vaccination against pneumococci and meningococci. The spleen was removed measuring 30x20x15cm and biopsy showed thickened capsule with widening of splenic cords which were congested and with erythrophagocytosis. The sinuses were empty and increase in connective tissue fibres were seen focally.

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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.[1] The prevalence of this disease in Bangladesh is not known but in Northern Europe and North America, with a reported incidence of 1/ 5,000 births.[2,3] Most cases are inherited in an autosomal dominant fashion and approximately 25% of these

Fig 1: Previous cholecystectomy scar

Fig 2: Abdominal cavity showing spleen

Fig 3: Spleen after removal

cases may represent spontaneous mutations or recessive forms of the disease.[4,5] Disruptions of ankyrin, 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 biconcave to sphere and loses the ability to circulate freely through narrow capillaries in the body, so 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 infant in whom jaundice presents in the first 24 hours after birth or in whom jaundice persists beyond the first postnatal week. In the first few postnatal months, anaemia can develop in children who do not mount an adequate reticulocytes response. In later childhood, HS can present with anaemia, jaundice, and splenomegaly.[9] Older individuals develop bilirubin stones and may present with cholecystitis.[1] 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 sepsis.[2,8,10] Splenectomy dramatically improves red cell survival.[11] But there are controversy regarding absolute indications for splenectomy.[12] Cholecystectomy is recommended for cholelithiasis, and during cholecystectomy, splenectomy is also indicated otherwise stone may develop at other sites of biliary tree.[6,8]

Before splenectomy, it is essential to vaccinate the patient against different infectious diseases, with life-long oral penicillin V. Splenectomised patients may admit with septicaemia should be resuscitated and given intravenous antibiotics to cover pneumococcus, haemophilus and meningococcus.[1,10,11,12]

Conclusion

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

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