**Abstract**

Hereditary spherocytosis (HS) is a common inherited extravascular haemolytic disorder. It usually presents with anaemia, intermittent jaundice, abdominal pain, splenomegaly and sometimes features of cholelithiasis. Increased fragility of Red blood cells (RBC) due to defect in RBC membrane proteins leads to haemolytic anaemia, splenomegaly and hyperbilirubinemia. Here we report a case of 19-year-old male who presented with fever, right hypochondriac pain, jaundice and exertional tiredness. He was diagnosed as hereditary spherocytosis with acute cholecystitis due to cholelithiasis based on spherocytosis on blood picture, increased reticulocyte counts and direct and indirect bilirubin, positive osmotic fragility test and splenomegaly and gall stones on ultrasound scan of abdomen. He was managed with intravenous antibiotics and supportive measures for acute cholecystitis. Elective concomitant splenectomy and cholecystectomy was performed after immunization for capsulated organisms. He was commenced on lifelong oral penicillin prophylaxis post splenectomy.