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Case Report

Paediatrics Section

Hepatopathy with Common Hepatic Duct Stenosis Diagnosed with Endoscopic Retrograde Cholangiopancreatography in a Sickle Cell Anaemic Female Child: A Case Report

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ABSTRACT

Hepatic complications are common in sickle cell disease, particularly among patients with the Sickle Cell Anemia (SS genotype). Liver injuries may result from viral hepatitis, iron overload due to transfusions, or vascular issues caused by sickled red blood cells. Clinical manifestations include sequestration crises, ischaemic hepatocyte injury, pigment gallstones, and biliary obstruction. In rare cases, these complications can progress to coagulopathy, jaundice, encephalopathy, or acute liver failure requiring transplantation. Acute sickle cell intra or extrahepatic cholestasis, though rare, is often fatal and presents with severe jaundice, Right Upper Quadrant (RUQ) pain, elevated liver enzymes, and coagulopathy. In the present case report, an 11-year-old girl with known sickle cell anemia (SS genotype) presented with yellowish discoloration extending to her palms and soles, abdominal pain, and palpable liver and spleen, accompanied by hepatic insult indicated by increasing levels of liver enzymes and bilirubin. She was diagnosed with Common Hepatic Duct (CHD) stricture using advanced radiological intervention, specifically Magnetic Resonance Cholangiopancreatography (MRCP). This condition was managed through Endoscopic Retrograde Cholangiopancreatography (ERCP). Following this procedure, she was discharged with a diagnosis of acute sickle cell hepatopathy, exhibiting a spectrum of hepatobiliary involvement. After approximately 30 days, a decreasing trend in her bilirubin levels was noted. Patients with sickle cell disease are more susceptible to hepatic complications, which can lead to significant morbidity and even mortality. Clinical manifestations can range from asymptomatic conditions to acute hepatic crises and chronic liver disease. Advanced investigations can be helpful for early intervention, which, in turn, can lead to better disease management and prevention of adverse outcomes.

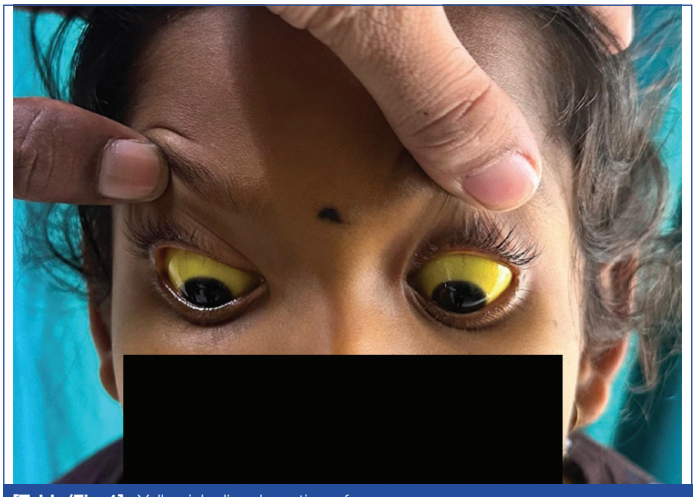
Keywords: Abdominal pain, Acute presentation, Biliary tree obstruction, Jaundice, Magnetic resonance cholangiopancreatography, Sickle cell crisis

CASE REPORT

An 11-year-old girl known to have sickle cell anemia visited the Outpatient Department with chief complaints of fever, vomiting, abdominal pain, and yellowish discoloration of her body and eyes for the past seven days [Table/Fig-1]. As recounted by her mother, she had been well seven days prior but then developed moderate-grade fever that was intermittent, without diurnal variations. The fever was alleviated with medication and was associated with vomiting and abdominal pain. She experienced 3-4 episodes of non-bilious vomiting daily, which contained food particles but was not blood-tinged or foul-smelling. The vomiting was associated with intermittent, colicky abdominal pain primarily in the RUQ.

She also exhibited increasing yellowish discoloration of her eyes and body, beginning with her eyes and spreading to her trunk and limbs. She was born to parents who are second-degree cousins, as the first child in their family. She was delivered in a government facility via normal vaginal delivery at a weight of approximately 2.90 kg, at term gestation, and cried immediately after birth without requiring Neonatal Intensive Care Unit (NICU) admission. Her mother attended all antenatal check-ups and reported no significant antenatal history. There was a family history of normal haemoglobin gene (A) and one sickle cell gene (S) {AS genotype (recessive)} in both the mother and father. The girl was diagnosed with sickle cell anemia (SS genotype) at two years of age through haemoglobin electrophoresis.

On examination, the patient exhibited icterus, hepatomegaly, and splenomegaly, accompanied by RUQ pain. She was unable to eat,



[Table/Fig-1]: Yellowish discolouration of eyes.

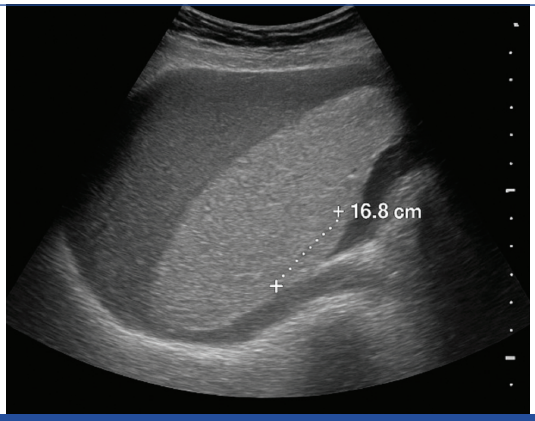
experienced weakness, and had multiple episodes of vomiting over the past four days.

During the general examination, she presented with pallor and icterus, and yellowish discoloration extending to her palms and soles. The abdominal examination revealed a soft abdomen, with a palpable liver 3-4 centimeters below the costal margin and a spleen 2 centimeters below the costal margin, along with diffuse abdominal pain.

Laboratory investigations revealed a haemoglobin of 7.8 grams per decilitre (g/dL), platelet count- 4.73 per microliter, Haematocrit

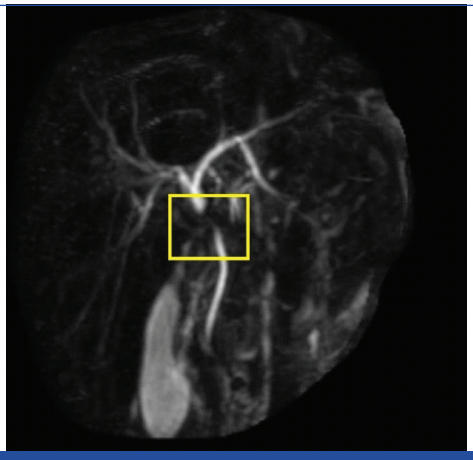
(HCT)- 23.1%, total White Blood Cells (WBC)-14000 cells per microliter, liver function test Alanine Aminotransferase (ALT)-4900 units per litre (U/L), Aspartate Aminotransferase (AST)- 13000 units per litre (U/L), Total bilirubin- 19.6 mg/dL, conjugated-16.6 mg/dL, unconjugated 3.0 mg/dL, International Normalized Ratio (INR)-1.5, and a positive Hepatitis A reactivity.

An abdominal ultrasound suggested mild hepatosplenomegaly, with the liver measuring 16.8 cm in craniocaudal dimension [Table/ Fig-2]. Provisional diagnosis of acute hepatopathy in sickle cell disease was made and treatment was initiated for the same. N acetyl cysteine drip (100 mg/kg/d Intravenous (IV) infusion up to 72 hours), Inj. cefotaxime (150 mg/kg/day, 8 hourly × 7 days), Inj. Amikacin (15 mg/kg/day × 7 days), Syp. Lactulose (10 mL/day bis in die {BD (twice daily)} × 15 days), Vitamin A (200,000 International Unit (IU)/dose Once Daily (OD) Per Os { PO (by mouth)} × 2 days), Vitamin D (60,000 IU/week × 4 weeks), Vitamin E (400 IU × 15 days), Vitamin K (5 mg, 24 hourly × 3 days) were initiated. The patient was monitored for signs of altered sensorium. After 10 days, liver profiles were repeated, showing a decreasing trend in ALT (43 to 23 mg/dL) and AST (147 to 130 mg/dL). However, total bilirubin levels decreased from 52.3 mg/dL to 27.1 mg/dL and then increased to 34.4 mg/dL, with conjugated bilirubin remaining elevated (33.4 to 32.0 mg/dL). Given the clinical presentations and laboratory features-RUQ pain, hepatomegaly, jaundice, and rising conjugated bilirubin-suspicions arose for a blockage in the hepatobiliary tree or an icteric phase of hepatitis infection. Continued monitoring over 5-7 days showed no decrease in serum bilirubin.



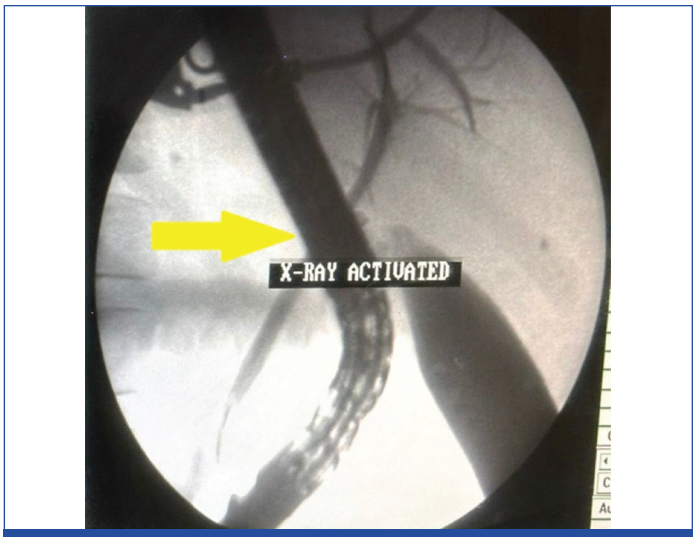
[Table/Fig-2]: Ultrasound image of the (abdomen-pelvis), showing mild hepatosplenomegaly with liver size marked in cranial-caudal dimension.

Differential diagnoses considered included hepatic sequestration, acute hepatic crisis, and cholelithiasis. However, without clinical supporting evidence, a blockage in the hepatobiliary tree was suspected. The patient was advised to undergo MRCP, which indicated liver enlargement and demonstrated short-segment abrupt narrowing of the intrahepatic segment of the CHD, approximately 4.4 mm in length, with noted ductal irregularities [Table/Fig-3].



[Table/Fig-3]: MRCP showing the focal area of stricturous narrowing involving the CHD and subtle ductal irregularities.

This led to a final diagnosis of hepatopathy with CHD stenosis. A gastroenterology consult was made, and the patient was scheduled for an ERCP. The ERCP was successfully performed under total intravenous anesthesia due to obstructive jaundice and stenosis of the CHD [Table/Fig-4].



[Table/Fig-4]: ERCP, endoscope probe near CHD.

During the procedure, conducted with an Olympus endoscope, the oesophagus and stomach appeared normal, and a small downward-facing ampulla was noted in the duodenum. Selective Common Bile Duct (CBD) cannulation was achieved, and bile was aspirated and sent for culture. The bile was greenish, and the cholangiogram revealed a suspicious CHD stricture near the biliary confluence. Endoscopic papillotomy was completed with a 10 Frenchx10 cm stent placement, allowing for free flow of bile.

The child was monitored with liver function tests. Over the next two months, total bilirubin levels decreased: from 49.9 mg/dL to 46.0 mg/dL for conjugated bilirubin, and from 3.9 mg/dL to 2.1 mg/dL, eventually reaching 0.9 mg/dL. The patient was discharged after 29 days with noted improvements, including resolution of icterus, reduced hepatomegaly, no more vomiting, and resumed normal food intake. On follow-up after three months, the serum bilirubin levels were within normal limits and the stent was removed. Tab folic acid (5 mg/OD) and tab hydroxyurea (250 mg/OD) were advised to be continued until the next follow-up.

On follow-up after three months, serum bilirubin levels were within normal limits, and the stent was removed. The patient was advised to continue folic acid (5 mg once daily) and hydroxyurea (250 mg once daily) until the next follow-up.

DISCUSSION

Sickle cell disease is a haemoglobinopathy characterised by a mutation in the sixth position of the beta-globin gene, where glutamic acid is replaced by valine. This change leads to the formation of abnormal haemoglobin, which decreases solubility and results in sickle-shaped erythrocytes [1]. Sickle cell anemia, particularly in the HbSS (Haemoglobin and 2 beta globin subunits of sickle cell) homozygous condition, can lead to a range of hepatic and hepatobiliary pathologies. These pathologies arise from various insults to the hepatobiliary system, primarily due to the sickling of red blood cells within hepatic ducts, biliary systems, or the liver's vasculature [2]. Sickle cell anaemia may result in multiple hepatobiliary complications like gallstone formation due to hyperbilirubinemia, which may get complicated by common bile duct obstruction and cholangitis [3]. It may present as a clinical spectrum of asymptomatic patients to an acute crisis with acute liver failure to chronic liver disease. In a similar case, a known patient with sickle cell anemia (SS pattern) presented with acute RUQ pain, yellowish discoloration of the body, generalised weakness, and hepatosplenomegaly.

Clinical presentations	Scenario	Management
Asymptomatic, RUQ pain, Hepatomegaly, Jaundice, Elevated bilirubin levels, Cholangitis	Biliary obstruction/ Gallstones	Cholecystectomy for symptomatic bile duct stones ERCP for hepatobiliary clearance
Vaso occlusive crisis, Right upper quatrant pain, Elevated bilirubin and liver enzymes (ALT >300)	Acute sickle hepatic crisis	Supportive ERCP (If stenosis or blockage in the hepatobiliary system- consider exchange blood transfusion)
Vaso occlusive crisis, RUQ pain, Leukocytosis, Fever, Striking jaundice, Very high bilirubin (ALT in 1000s), Coagulopathy, Renal failure	Sickle cell intrahepatic cholestasis	Full supportive management, exchange blood transfusion
Enlarging liver, RUQ pain, Anaemia, Reticulocytosis	Hepatic sequestration	Supportive transfusion

[Table/Fig-5]: Management recommendations in acute sickle hepatopathy.
ALT: Alanine aminotransferase; RUQ: Right upper quadrant [12].

The incidence of sickle cell hepatopathy is variable. Liver function may be abnormal in sickle cell patients without indicating intrinsic liver damage; the reported prevalence of hepatopathy is around 10% [4]. More severe hepatopathy is often noted in patients with homozygous genetics (SS) [2]. The most severe type of sickle hepatopathy is acute intrahepatic cholestasis, which is uncommon. Its pathophysiology may involve blockage of sinusoids from sickle red blood cells, leading to obstruction in the hepatobiliary system. This obstruction can result in ischemia or hepatocyte injury [3-5]. The patients might present with RUQ pain, fever, marked jaundice, deranged coagulation profile, renal failure, elevation of liver enzymes, and full supportive management, also consider exchange transfusion and further investigation is required to rule out an obstruction in the biliary system by MRCP and should be managed accordingly. Pancreatitis, cholangitis, acute chest syndrome, pneumonia, and painful crisis are reported as common complications of ERCP for the management of choledocholithiasis in sickle cell patients [6].

Laboratory investigations typically indicate hyperbilirubinemia and elevated liver enzymes, suggesting hepatocellular damage. Sickle cell disease can impact multiple organ systems, and gastrointestinal involvement often manifests through clinical presentations of vascular infarcts resulting from vascular occlusions [3,5,7,8]. Hyperbilirubinemia in sickle cell patients might complicate due to obstruction of the CHD, along with severe hepatic issues like acute liver crises [5]. Sickle cell hepatopathy, resulting from biliary obstruction and hyperbilirubinemia, can be diagnosed through radiological interventions such as MRCP, followed by treatment with ERCP [9]. In cases of sickle cell disease, liver function tests may be mildly elevated, in contrast to sickle cell hepatopathy, where clinical presentations can vary widely and pose diagnostic challenges [9-11]. Management recommendations for acute sickle hepatopathy have been outlined in a referenced [Table/Fig-5] [12]. Timely diagnosis of hepatic complications in pediatric patients with sickle cell anemia is crucial to prevent severe complications and mortality. In this case, a complication in the sickle cell disease patient (SS) type was noted, which resulted in acute hepatic insult with biliary tree pathology resulting in acute hepatopathy, which was further resolved with the help of radiological support, magnetic radio-pancreatography and interventional endoscopic cholangiopancreatography.

CONCLUSION(S)

Patients with sickle cell anemia, especially those with the SS pattern, are particularly vulnerable to a range of hepatobiliary problems stemming from both sickling and non-sickling complications. This spectrum of disorders presents overlapping symptoms and can be classified as sickle cell hepatopathies. Management should be tailored according to the dominant clinical presentation, utilising thorough assessment of the patient's clinical data, laboratory results, and radiological findings.

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