

# Endoscopic Retrograde Cholangiopancreatography Management of Choledocholithiasis, Acute Cholangitis and Acute biliary Pancreatitis in a Patient with Sickle Cell Disease: A Case Report

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## Introduction

Choledocholithiasis can be either primary or secondary. Primary refers to the stones which originate in the bile duct and the secondary are due to the stones ejected from the gallbladder. Approximately 95% of choledocholithiasis is due to the migration of gallstones. Therefore, in the presence of gallbladder, or only recently removed, the origin of the stone in the common duct is impossible to prove. Primary choledocholithiasis is very uncommon in sickle cell disease (SCD), even though sickle cell disease patients are at a higher risk to develop pigmented stones due to hemolysis.<sup>1</sup> Acute cholangitis is a life-threatening condition, delay in diagnosis and treatment can lead to septic shock.<sup>2</sup> Obstruction of the pancreatic outlet due to the presence of stones in the bile duct leads to development of acute biliary pancreatitis.<sup>1</sup>

Five to 20% of general patients with cholelithiasis also have choledocholithiasis.<sup>3</sup> While SCD the choledocholithiasis ranges from 18% to 30%.<sup>4</sup> The frequency of cholelithiasis in patients with SCD is variable, ranging from 4% to 55%, and this increases with age.<sup>5</sup> Gall stones have been found to be prevalent in the

## Abstract

Acute cholangitis is a life-threatening condition caused by an ascending bacterial infection of biliary tree. Choledocholithiasis is the most common cause of ascending cholangitis. It can be caused by either primary or secondary bile duct stones.

Obstruction of the pancreatic outlet due to migration of bile duct stones trigger the pathophysiological mechanisms of acute biliary pancreatitis, such as intracellular enzyme activation, self-digestion, and activation of the inflammatory pathway. Sickle cell disease patients, due to hemolysis, are at high risk of development of pigmented gallstones. Development of primary choledocholithiasis in Sickle cell disease is very uncommon. Cholangitis if not timely managed can be fatal. Here, we report on a case of Sickle cell disease patients presenting with acute cholangitis and acute biliary pancreatitis which was successfully managed by Endoscopic Retrograde Cholangiopancreatography (ERCP).

**Keywords:** Sickle cell disease, Choledocholithiasis, ERCP

age group 11- to 29-years.<sup>6</sup> We report a case of acute cholangitis due to choledocholithiasis in a sickle cell disease patient who was successfully managed with ERCP.

## Case Presentation

We present a case report of 17-year male, Mr. S Gala originally from Assam, India belonging to tribal race is currently living with his father (Mr. P Gala), mother (Mrs. S Giri) and three siblings in Eastern Nepal. He is homozygous for SCD; however, sickle cell status of others is unknown. He was admitted at this center with complaints of pain in his upper abdomen, fever, increasing jaundice, nausea and vomiting for one week duration. However, he gave history of pain in the abdomen and fever on and off for last one month. For the same complaint there were multiple hospital visits. However, due to financial constraints he was not evaluated and taken back against medical advice on previous occasions.

During the last presentation (15th May 2025) the patient had high fever documented 105.1 °F, his blood pressure was 80/50 mmHg and oxygen saturation was 88% in room air. His hemoglobin was 11.1 g/dL, platelet 355×10<sup>3</sup> /mm<sup>3</sup>,

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total count 28,210 cells /mm<sup>3</sup> , total serum bilirubin 39.41 mg/dL, conjugated serum bilirubin 17.52 mg/dL, serum alanine aminotransferase level 221.5 U/L, serum aspartate aminotransferase 242.2 U/L, Alkaline phosphatase was 416.8 U/L , Lactate Dehydrogenase was 824.04 U/L, amylase 1399.07 U/L, Lipase 2312.50 U/L , urea 29.7 mg/dl, creatinine 0.56mg/dl, Na 127.75 mmol/L, K 3.81 mmol/L. Urine culture and sensitivity was done Acetobacter species was isolated. It was sensitive to gentamicin and nitrofurantoin only. Abdominal ultrasound showed hepatomegaly, choledocholithiasis with mild bilobar intrahepatic biliary radicals dilated (IHBRD). Other investigations human immunodeficiency virus, hepatitis B surface antibody, hepatitis C virus antibody, hepatitis A virus antibody, hepatitis E virus antibody, Malaria Parasite were negative. The C-reactive protein was normal (<6mg/L).

His Hb Electrophoresis report: Hb F 27.50%, Hb Adult 1.80%, Hb A2 2.10%, HB sickle 69.20%, Others 0.10%. All these features supported acute cholangitis, sepsis and acute biliary pancreatitis. The patient was initially resuscitated by iv fluids, Ringer’s Lactate (RL) bolus of 20 mL/kg in first 2 hours and followed by RL of 1.5 mL/kg/h over 24 hours to prevent hypovolemia, dehydration and acute kidney injury. The patient was given higher antibiotics (meropenem and metronidazole) for sepsis and acute cholangitis. The oxygen was supplemented via nasal prongs to maintain saturation. After initial resuscitation and patient stabilization, the next step for management was ERCP. It was planned for the next day of admission which was within 24 hours. Selective biliary cannulation was done by standard guide wire technique. Cholangiogram revealed filling defect in mid common bile duct (CBD). Sphincterotomy was done and followed by multiple balloon sweeps. A calculus along with sludges were expelled. Occlusion cholangiogram revealed CBD clearance. A straight plastic biliary stent of 8.5fr 8cm was deployed. The patient was kept in ward; antibiotics and iv fluids were continued. Next morning Total count 18,530 cells/mm<sup>3</sup>, total serum bilirubin 16.08 mg/dL, conjugated serum bilirubin 6.90 mg/dL, serum alanine aminotransferase level 64.76 U/L, serum aspartate aminotransferase 116.32 U/L, Alkaline phosphatase was 176.44 U/L. On the sixth day of admission the patient was discharged on oral antibiotics and was asked to follow up. The stent was removed after one month.



Fig 1: Cholangiogram showing filling defect (arrow)

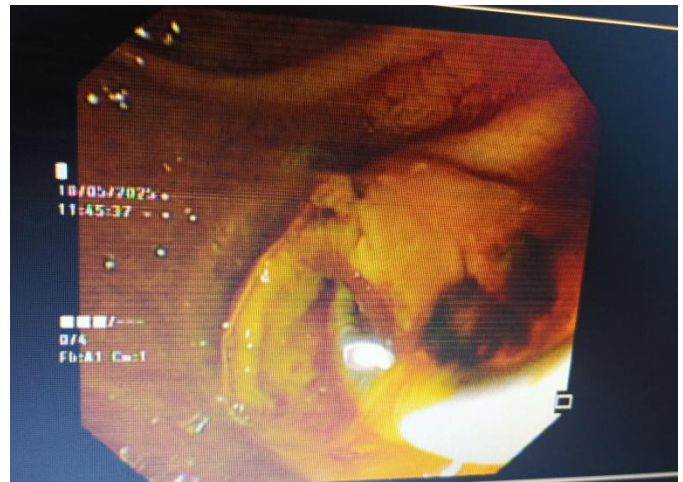


Fig 2: ERCP image showing extracted stone and sludges



Fig 3: ERCP with Plastic biliary stent

Discussion

Sickle cell disease is one of the most common inherited hemoglobinopathies worldwide which has huge social impact and has been recognized as a global public health problem by the World Health Organization. Mutation in chromosome 11 causes various diseases, including SCD with homozygous sickle hemoglobin (HbSS), HbSC, and HbS beta thalassemia. HbS polymerizes on exposure of deoxygenation, it is responsible for Vaso occlusive manifestations. The rigid polymers of HbS disrupt the RBC membrane, shortening the lifespan of RBCs and inducing hemolysis.<sup>7</sup>

SCD is more prevalent in the Western, Mid-Western or Far-Western region of Nepal. Around 97.7% of the patients and carriers of sickle cell disease belonged to the Tharu ethnic group and rest were non-Tharu- Yogi, B C, Raidas etc.<sup>8</sup> The patient’s parents had tribal roots from Assam, India and was brought up in Eastern Nepal. The tribals working in Chai Bagan (tea gardens) of North-East India (Assam) show HbS as the predominant haemoglobinopathy. In upper Assam, the HbS gene frequency was 5.11%.<sup>9</sup> The mean age of patients and carriers was 24.5 ± 12 yrs. Maximum number of patients 381 (30.5%) were in the age group 21-30 years. Only 156 (12.5%) patients and carriers were under the age of 11 years.<sup>8</sup> Gallstones are a frequent complication in SCD. The development of pigment gallstones

due to chronic hemolysis is age dependent: 15% under 10 years, 22% between 10 and 14 years and 36% between 15 and 18 years and reported prevalence of 50% by the age of 22 years.<sup>1</sup>

The sickle cell disease patient can present with “sickle cell crisis” which is the severe acute condition of Vaso-occlusive crisis (acute painful crisis), aplastic crisis, splenic sequestration crisis, hyper hemolytic crisis, hepatic crisis, dactylitis, and acute chest syndrome. Other acute complications include pneumonia, meningitis, sepsis and osteomyelitis, stroke, avascular necrosis, priapism, and venous thromboembolism.<sup>10</sup> Symptomatic biliary tract disease is difficult to diagnosis

in patients with SCD, who frequently can have acute abdominal pain, fever and jaundice, as symptoms of Vaso occlusive crisis.<sup>1</sup> SCD patients are prone to develop biliary sludge and bile duct stones.<sup>11</sup> Pigmented stones develop due to hemolysis in SCD. Bile stasis and infections predispose to stone formation.<sup>1</sup> ERCP is beneficial in preventing future development of bile duct stones in cases of dilated bile ducts.<sup>11</sup>

Acute biliary pancreatitis is managed early conservatively by appropriate intravenous fluid administration which minimizes the risk of hypovolemia and prevents dehydration and prevents organ failure. After initial resuscitation and stabilization of patient, the ERCP in acute biliary pancreatitis depends upon the clinical situation. When the patient has cholangitis, the evidence strongly supports performing ERCP as soon as possible and within 24 hours.

Cochrane systematic review and meta-analysis Pointed that the role of ERCP in acute biliary pancreatitis without cholangitis was a matter of debate.<sup>12</sup>

## Conclusion

SCD itself is a rare inherited hemoglobinopathy. Gallstones and bile duct stones can form due to hemolysis, infection and biliary stasis. Timely recognition and appropriate management of acute biliary cholangitis are critical for optimal outcomes. ERCP is the minimally invasive modality for management of choledocholithiasis and cholangitis.

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