Sickle Cell Hepatopathy In A Patient with Markedly Elevated Bilirubin

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Abstract

- The vaso-occlusive events that characterize a sickle cell pain crisis are known to affect multiple organ systems.
- Sickling in the hepatic sinusoids leads to ischemia, cholestasis, and sequestration, which collectively contribute to acute liver dysfunction often referred to as acute sickle cell hepatic crises (ASCHC), a form of sickle cell hepatopathy.
- As one of the rarer complications of sickle cell disease (SCD), ASCHC may often go under-recognized.

History & Physical

- 20-year-old male with a history of SCD presented for evaluation of intermittent epigastric and right upper quadrant abdominal pain.
- His pain was of abrupt onset and associated with yellowing of the eyes and skin.
- Vital signs unremarkable on presentation without fever.
- Exam notable for epigastric and right upper quadrant tenderness without rebound or guarding. Noted to have overt scleral icterus and jaundice.

Initial Workup

- Given concern for painful jaundice presenting workup was broad and included liver profile.
- As highlighted by Table 1, the patient was noted to have markedly elevated direct bilirubin of 34.6 in setting of elevated alkaline phosphatase and elevated transaminases.
- Hemoglobin was stable at 9 (baseline), however, LDH elevation and increased reticulocyte count were concerning for active hemolysis.
- Right upper quadrant ultrasound showed evidence of increased hepatic echogenicity concerning for hepatitis; mild dilation of common bile duct (CBD) was seen.

Liver Biopsy

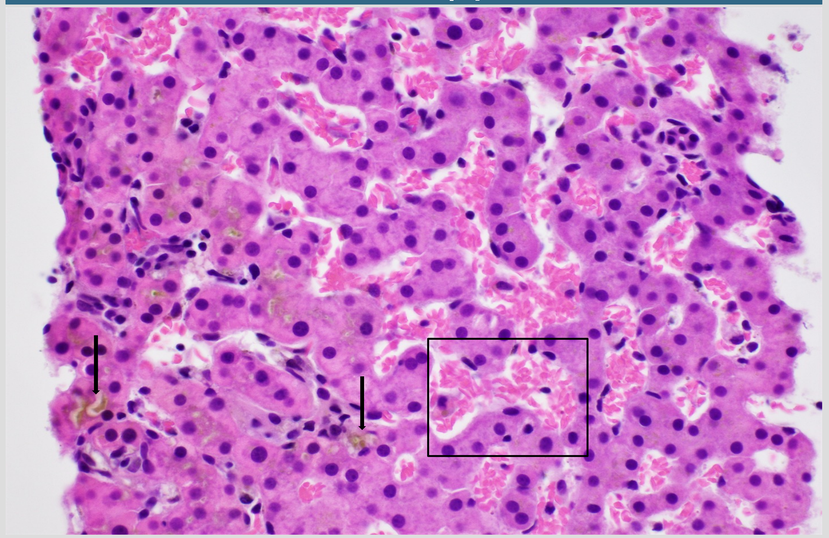

Figure 1. **Liver biopsy, H&E 200x magnification**: Liver parenchyma with dilated sinusoids, readily visualized sickled red blood cells (box), and canalicular cholestasis (arrows).

Liver Profile	Day of Presentation	Day 2 of Admission	Day 3 of Admission
Alkaline Phosphatase	211 U/L	247 U/L	219 U/L
Total Bilirubin	35.8 mg/dL	31.6 mg/dL	**12.9 mg/dL**
Direct Bilirubin	34.6 mg/dL	28.8 mg/dL	**7.2 mg/dL**
ALT	186 U/L	200 U/L	161 U/L
AST	126 U/L	146 U/L	83 U/L

Table 1. Liver profile trend during admission. Patient with improvement of elevation in transaminases and direct bilirubin with symptomatic treatment only.

Hospital Course

- Day 1:
  - Follow up MRCP ordered given ongoing concern for obstruction. MRCP with evidence of mild intrahepatic and extrahepatic dilation and mild CBD dilation.
  - Patient continued to be symptomatic. Pain treated with IV pain medication.
  - Workup up for viral and autoimmune hepatitis found to be normal. No evidence of Wilson's disease or hemochromatosis.
- Day 2:
  - Patient with complete resolution of abdominal pain with pain medication alone.
  - Despite improvement direct bilirubin continues to be significantly elevated (Table 1).
  - Given non-revealing workup decision made to pursue liver biopsy.
- Day 3:
  - Liver biopsy completed. Pathology consistent with acute sickle cell hepatic crisis (Figure 1).
  - Liver profile with significant improvement (Table 1).
  - Patient continues to be asymptomatic and is thus discharged.

Discussion

- Acute sickle cell hepatic crisis (ASCHC), a subset of sickle cell hepatopathy (SCH), is often difficult to diagnose.
- Guidelines are lacking with regards to the specific treatment for ASCHC.
- ASCHC has the potential to be concurrent with a more severe form of the disease known as sickle cell intrahepatic cholestasis (SCIC), which if untreated can be fatal.
- Both ASCHC and SCIC can present with jaundice, as in our patient, but only SCIC progresses to organ failure.
- As more cases are described, the optimal treatment modalities for these rarer complications of sickle cell disease will only increase.

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