Acute Sickle Intrahepatic Cholestasis from Sickle Cell SC Disease Causing Acute on Chronic Liver Failure

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INTRODUCTION

- **Acute on Chronic Liver Failure (ACLF)** is a disorder that results in grave systemic consequences.
- The dysregulated inflammatory response caused by an acute liver injury often results in multi-organ dysfunction.
- While alcoholic/viral hepatitis or infections are common causes of ACLF, hematologic disorders are unusual.
- We present a case of ACLF due to **sickle cell intrahepatic cholestasis** from **SC disease**.

CASE DESCRIPTION

- A 61-year-old man with history of HTN, sickle-cell trait, alcoholic cirrhosis presented for altered mental status.
- Initial labs showed **TBili 30.7, DBili >20.0**, **AST/ALT 308/123**, **ALP 532** and INR 1.6. Phosphatidylethanol level negative.
- **No** new history of medication/herbal use, new sexual contacts, or drug use.
- Workup **negative** for hepatitis A/B/C/E. ANA negative, ASMA 26.7, IgG 2742.
- MRI Abdomen showed no liver mass with **patent** hepatic vasculature/biliary tree.
- **LDH 531**, Haptoglobin <10, **reticulocyte count 11.2**, and **Coombs test was positive**.
- **Hb electrophoresis** showed **44.4% Hb C and 49.5% Hb S**, consistent with **SC disease**.
- Hb was 8.5 (baseline), with **no known crises** since youth.
- Liver biopsy findings were consistent with **sickle cell hepatopathy** (**sinusoidal dilation and congestion of sickled cells**, ballooning degeneration of hepatocytes, and **marked intracanalicular cholestasis**).

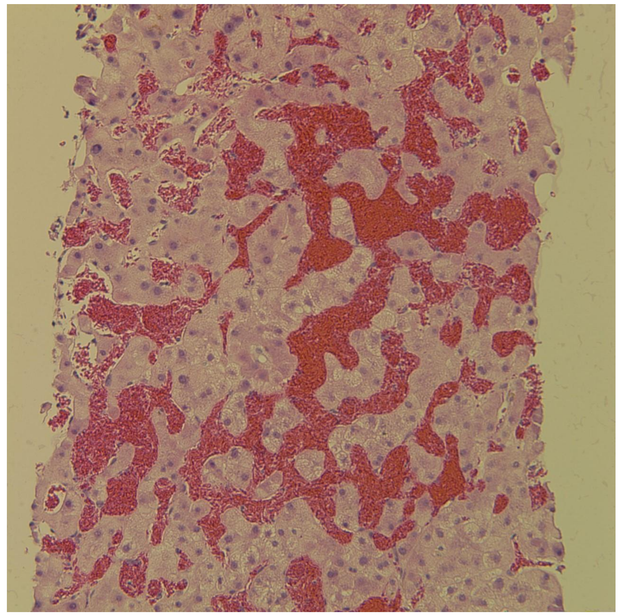

***Figure 1*: Sinusoidal dilatation of liver**

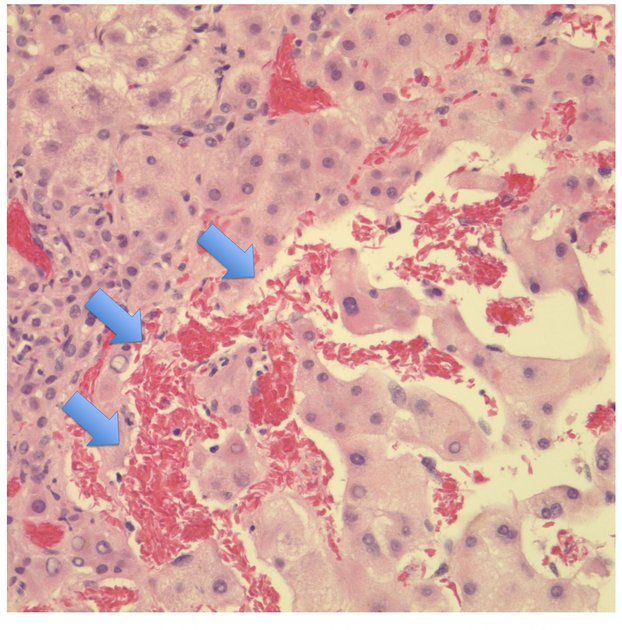

***Figure 2*: Sickled RBCs within sinusoids of liver**

CASE DESCRIPTION CONT.

- Patient had **exchange blood transfusion (EBT)** with improvement in Hb to 11.8 and decrease in retic count, LDH.
- Patient however continued to deteriorate, with **increasing aminotransferases into thousands**, worsening mental status, AKI, and coagulopathy.
- Patient made hospice care and eventually expired.

DISCUSSION

- **Hemoglobin SC (HbSC)** disease is a **less common form** of sickle cell disease (SCD).
- Patients with HbSC disease have milder symptoms than SCD but more severe than sickle cell trait.
- Acute sickle hepatic crisis occurs in **10%** of patients with SCD.
- **Acute sickle intrahepatic cholestasis** is the **most severe form of sickle hepatopathy**, associated with increased mortality.
- It is usually seen in SCD and **rarely in HbSC**.
- Clinical presentation is **more severe**, with significant hyperbilirubinemia **>15 mg/dL** and aminotransferase elevations **>1000 IU/L**, reflecting ischemic injury.
- **ALF/ACLF can evolve rapidly** with multi-organ failure.
- **EBT is considered** in these situations.
- Non-responders have **poor prognosis**.
- Clinicians should be aware of the liver complications that can arise in patients with sickle cell-related disorders, including HbSC, particularly in patients with chronic liver disease.

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