What the Eye Cannot See:

A Case of Persistent Altered Mental Status in a Patient with Cholestatic Liver Disease

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Background

- Altered mental status (AMS) is a common symptom in patients with liver disease, with a wide list of differential diagnoses.
- Knowledge of etiologies of AMS unique to patients with hepatic dysfunction is vital in order to help recognize, diagnose, and treat the underlying cause in a timely manner.

Case Presentation – Part 1

- A 46-year-old man who was recovering from a recent COVID infection was transferred to our hospital for further evaluation of acute liver injury and AMS. His past medical history was otherwise included diabetes and hypertension. He had no history of alcohol, tobacco, or illicit drug use.
- On arrival, his labs were notable for AST of 408 U/L, ALT of 620 U/L, ALP of 5942 U/L, TB of 11.0 mg/dL, and INR of 1.1.
- His work-up included:
  - An MRCP that showed segmental biliary ductal dilation with associated restricted diffusion and peribiliary enhancement concerning for sclerosing cholangitis.
  - An ERCP that revealed a 3cm biliary cast that was removed, with note of diffuse rarefaction of ducts throughout the entire biliary tree.
  - A liver biopsy that revealed centrizonal cholestasis with portal-based bile ductular reaction and mild bile duct injury.
- Despite adequate empiric treatment of suspected infection and hepatic encephalopathy, his AMS persisted.

Pseudo-normonatremia was suspected.

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Case Presentation – Part 2

- His BMP was notable for a Na of 143 mEq/L.
- A send-out lipid panel that was obtained to work-up his dyslipidemia revealed a total cholesterol of 1018 mg/dL, triglycerides of 420mg/dL, and the presence of lipoprotein X.
- A VBG was obtained showing a Na of 157 mEq/L and serum osmolality was 322 mmol/kg, confirming true hypernatremia.
- He was slowly treated with hypotonic solutions with significant improvement in his mentation.
- He was tried on ursodeoxycholic acid without improvement of his cholestasis.
- On follow-up one year later, he has persistent cholestasis and is currently being considered for liver transplant.

Discussion

- He was diagnosed with COVID-related ischemic/secondary sclerosing cholangitis and disappearing bile ducts with persistent cholangiopathy, presenting with severe cholestasis, accumulation of lipoprotein X, and pseudonormonatremia.
- When faced with severe cholestatic liver disease, clinicians should keep in mind the possibility of accumulation of lipoprotein X and its association with hyperviscosity and spurious electrolyte abnormalities.
- In such cholestatic patients, clinicians should rely on obtaining blood gas analyses for accurate electrolyte measurement as they utilize direct ion-sensitive electrodes (ISE) to measure electrolytes, whereas routine BMP's utilize indirect ISE that are liable to spurious results in the presence of hyperlipoproteinemia/lipoprotein X.