Fulminant liver failure due to Budd-Chiari syndrome triggered by strenuous exercise

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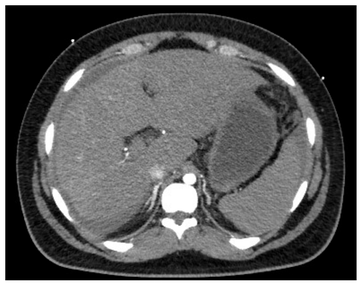
Introduction

This case illustrates the complexity of diagnosis of Budd-Chiari syndrome, especially with an atypical presentation.

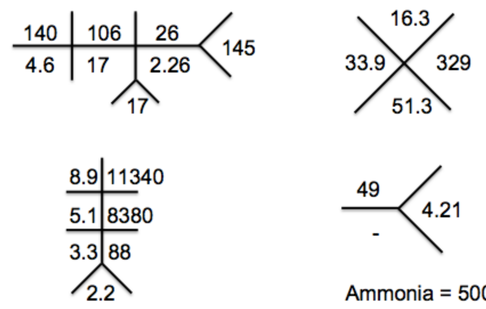
Case Description:

- A 27 year old man presents to OSH with abdominal pain, nausea, vomiting, diarrhea & confusion 24 hours after an all day police academy fitness test
- PMHx/PSHx: None
- Medications/Allergies: None
- SocHx: No EtOH, tobacco or illicits
- Physical Exam: Notable for hypothermia, tachycardia and hypoxemia requiring 2L nasal cannula
- Initial Labs: notable for creatinine 1.7, lactate 11.8, total bilirubin 1, AST 116, and ALT 95. Normal PT/INR
- Initial CT A/P concerning for possible SMV thrombus and patient was transferred to our facility

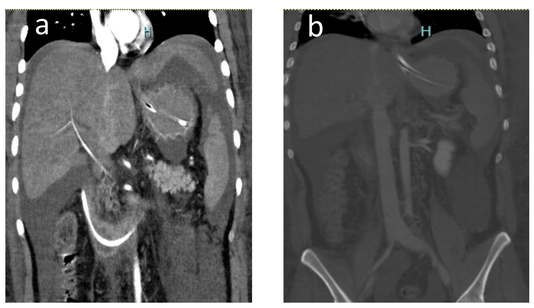


CT A/P with contrast done on hospital transfer negative for SMV/PV thrombus. Showing diffuse fatty infiltration of the liver

- Patient underwent exploratory laparatomy following transfer with no evidence of bowel ischemia and normal appearing liver
- Laboratory tests 48 hours following transfer:



- Transjugular liver biopsy demonstrated perivenular coagulative necrosis involving 80% of the hepatic parenchyma, and a hepatic venogram noted thrombosis of all hepatic veins



Representative coronal sections from CT Triple phase showing patent hepatic arterial anatomy (a) and interval thrombosis of the main portal vein extending into the L portal vein, along with diminutive hepatic veins (b)

- OLT was performed on hospital day six. During the surgery the patient was confirmed to have extensive thrombus involving the portal system as well as the superior mesenteric vein, and thrombus in the hepatic veins.
- Testing for hypercoagulable state was positive for JAK2 V617F mutation, and he was begun on treatment with hydroxyurea.

Discussion:

- The development of Budd-Chiari syndrome is likely due to his hypercoagulable state combined with dehydration from strenuous exertion.
- Lack of hepatomegaly on imaging had led to uncertainty regarding the diagnosis of Budd-Chiari, but the occlusion of the portal vein resulted in decreased blood flow to the liver and prevented the development of hepatomegaly.
- His initial development of lactic acidosis & leukocytosis is felt to be the result of marked hepatic necrosis from the Budd-Chiari syndrome but had led to confusion over the possibility of septic shock or bowel ischemia.