Budd-Chiari Syndrome Leading to Cirrhosis in a Young Woman

Hailey Olds
Eric Scher

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Budd-Chiari Syndrome Leading to Cirrhosis in a Young Woman

Hailey Olds BS, Dr. Eric Scher MD

Henry Ford Health System
Medicine, Detroit, Michigan

Abstract

Introduction: Advanced liver disease is rather unusual in young adults because cirrhosis typically develops gradually over years of persistent liver injury. Cirrhosis is commonly caused by alcohol abuse, viral hepatitis, or nonalcoholic fatty liver disease. Other potential etiologies include autoimmune, hepatitis, veno-occlusive, and genetic disorders such as Wilson disease or hemochromatosis. This case report suggests that veno-occlusive conditions such as Budd-Chiari syndrome should be considered in young adults presenting with cirrhosis, especially those with hypercoagulable risk factors and lack of other contributors such as alcohol use and viral hepatitis. Cirrhosis is rare in this age group, but hypercoagulable states may manifest in this manner.

Case: A young woman with a past medical history of hepatitis steatosis and polycystic ovarian syndrome controlled with oral contraceptive pills was transferred from an outside hospital for escalation of care for ascites and diffuse hepatic hematopoiesis of unknown etiology. The patient initially presented with right upper quadrant pain, nausea, and vomiting. This was attributed to cholecystitis, and a laparoscopic cholecystectomy was performed. During the procedure, the liver appeared cirrhotic and ascitic fluid was present. A CT scan showed diffuse hepatic hematopoiesis with mesenteric lymphadenopathy. After extensive testing, no clear etiology for her advanced liver disease was discovered. During admission to our hospital, she was found to have Budd-Chiari syndrome and bilateral lower lobe pulmonary embolism. Her oral contraceptives were discontinued, anticoagulation was started, and a direct intrahepatic portocaval shunt procedure was performed to relieve portal hypertension.

Discussion and Conclusion: In young patients with rapidly progressing liver disease, Budd-Chiari syndrome is a rare but important diagnosis to consider, especially in those with hypercoagulable risk factors. This patient was likely in a hypercoagulable state secondary to her oral contraceptive use. Presentation can range from asymptomatic (15-20%) to acute fulminant liver failure (5%); course may be acute, subacute, or chronic. When a patient presents with acute liver failure, Budd-Chiari syndrome should be suspected in patients with hepatomegaly, right upper quadrant pain, and ascites. Usual diagnosis with imaging (ultrasound, CT scan, or MRI) is treatment: address the underlying cause, start anticoagulation, and relieve portal hypertension if present. Patients should be monitored for disease progression outpatient treatment.

Case Presentation

- Young female in her early 30s with PCOS well controlled with OCPs and hepatitis steatosis presents to OSH with RUQ pain, nausea, and vomiting.
- OSH Course:
  - Ultrasound of the RUQ showed a thickened gallbladder wall and decreased gallbladder ejection fraction.
  - Attributed presentation to acute cholecystitis, scheduled cholecystectomy.
  - During the procedure, the liver appeared cirrhotic with ascitic fluid was present.
  - Drained 2 L of ascitic fluid and biopsied the liver.
  - Continued drainage from port sites post-operatively.
  - Developed chyle enteric pain and tachycardia (HR 144 beats per minute).
  - Workup for cirrhosis showed no clear cause.
  - Two more paracenteses performed, draining 4 and 3.5 L, respectively.
  - Transferred to Henry Ford Hospital (HFH) for escalation of care.
- FHF Course:
  - On presentation, patient had re-accumulation of fluid in the abdomen and tachycardia was noted (130-140 beats per minute).
  - Various laboratory testing and imaging was performed (see Table 2).
  - Diagnosed with Budd-Chiari Syndrome, started on anticoagulation with heparin while inpatient and taken off OCPs.
  - Interventional Radiology performed a direct infraportal portoocaval shunt (DIPS) procedure to relieve portal hypertension.
  - Discharged on apixaban for long term anticoagulation, plan to follow up with hepatology outpatient.

PMHx: PCOS, mild asymptomatic hepatic steatosis seen on imaging a few years prior, GERD

FMHx: mother had a saddle pulmonary embolism while on oral contraceptive pills after an ankle fracture

SHx: drinks 1–2 glasses of wine per week. Does not smoke or use illicit drugs. No recent travel

Table 1. Patient history.

<table>
<thead>
<tr>
<th>Laboratory Test</th>
<th>Result</th>
</tr>
</thead>
<tbody>
<tr>
<td>Electrolytes</td>
<td>Hyponatremia (129 mg/dL), mild hypokalemia (3.4 mg/dL), hypocalcemia (7.8 mg/dL), hypomagnesemia (1.2 mg/dL), normal BUN, Cr, phosphorus</td>
</tr>
<tr>
<td>Complete Blood Count</td>
<td>Elevated WBCs (11,600/L), microcytic anemia (Hb 9.5 g/dL, MCV 62.6 μm³), normal platelets (265,000/L)</td>
</tr>
<tr>
<td>Liver Panel</td>
<td>Normal ALT and AST (9 U/L and 19 U/L), low albumin (3 g/dL), normal total bilirubin (1.1 mg/dL), normal alkaline phosphatase (123 U/L)</td>
</tr>
<tr>
<td>Coagulation Studies</td>
<td>Mildly elevated INR (1.52), normal PTT (35 sec)</td>
</tr>
<tr>
<td>Autoimmune Hepatitis Panel</td>
<td>Negative for LKM Ab, mitochondrial M2 Ab, and smooth muscle Ab</td>
</tr>
<tr>
<td>Viral Hepatitis Panel</td>
<td>Negative for Hepatitis A, Hepatitis B, Hepatitis C, EBV, CMV</td>
</tr>
<tr>
<td>Liver biopsy</td>
<td>Venous outflow obstruction leading to chronic congestive hepatopathy</td>
</tr>
<tr>
<td>Acute Fluid Studies</td>
<td>Clear yellow fluid with serum-to-ascitic fluid albumin gradient &gt; 1.1 g/dL suggesting the presence of portal hypertension, no organisms seen</td>
</tr>
<tr>
<td>EKG</td>
<td>Sinus tachycardia, normal rhythm</td>
</tr>
<tr>
<td>Pregnancy Test</td>
<td>Negative</td>
</tr>
</tbody>
</table>

Table 2. Patient laboratory values while at HFH.

Imaging

CT abdomen with liver protocol:
- Thrombosed right, middle, and left hepatic veins with hepatic congestion and ascites.
- Bilateral lower lobe pulmonary embolisms.

Figure 1. CT of the liver in Budd-Chiari syndrome. Arrows show thrombi in the hepatic veins. Liver is notably congested as a result.

Differential Diagnosis:
- Budd-Chiari Syndrome is the most likely diagnosis based on the liver biopsy, presence of portal hypertension, and CT findings.
- Although the patient had a history of hepatitis steatosis, it is unlikely that this would progress to severe diffuse hepatic cellular disease so rapidly.
- Alcohol abuse and viral hepatitis are other major causes of liver disease in the United States, but the patient had no history of excessive alcohol consumption and viral serologies were negative.
- Other possible causes of cirrhosis that are less likely in this case: autoimmune hepatitis, Wilson’s disease, hemochromatosis.

Budd-Chiari Syndrome:
- Relatively uncommon, but when diagnosed, the patient is usually hypercoagulable.
- In primary Budd-Chiari syndrome, a thrombus forms in the hepatic veins, preventing blood from exiting the liver. Treatment involves addressing the underlying cause, starting anticoagulation therapy, and relieving portal hypertension if present.

References

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