

A Case of idiopathic noncirrhotic portal hypertension

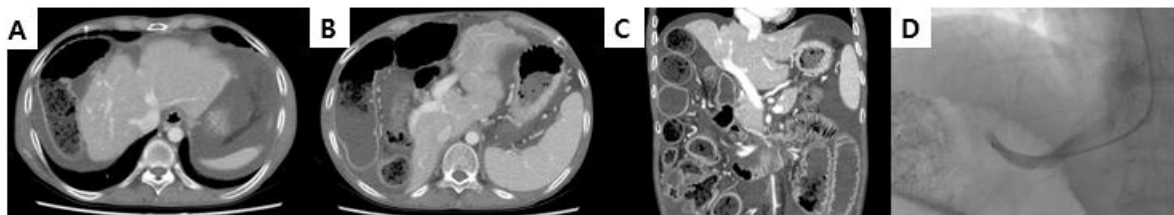
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Background: Portal hypertension develops when there is resistance to portal blood flow. Liver cirrhosis is the most common cause of portal hypertension, but portal hypertension can also occur in the absence of liver cirrhosis (noncirrhotic portal hypertension). Herein, we report a case of idiopathic noncirrhotic portal hypertension (INCPH).

Case: A 38-year-old male was referred for further evaluation of abdominal pain, distension and diarrhea. There was no history of alcohol abuse and chronic viral hepatitis B or C. The patient had been treated at outside hospital under the diagnosis of crohn's disease with infliximab every 2 months since 2016. He had a blood pressure of 118/65mmHg, a pulse of 59 beat per minute, a temperature of 36.5°C, and a respiratory rate of 18 breaths per minute. Laboratory test showed low platelet count ($63 \times 10^3/\mu\text{L}$), slightly decreased albumin (3.3 g/dL), and prolongation of PT INR (1.46). Small bowel endoscopy showed mucosal edema, diffuse distension and angiodysplasia. Abdominal CT demonstrated right hepatic lobe atrophy, splenomegaly and esophageal varix, but patent portal and hepatic vein. Hepatic vein wedge pressure was 8 mmHg. Liver biopsy showed distorted portal vasculature and occasionally invisible portal vein without cirrhotic change (periportal and perisinusoidal fibrosis). The patient is currently treated with diuretics and beta blocker for symptom control.

Discussion: INCPH is characterized by the presence of portal hypertension in the absence of cirrhosis on liver histology, the exclusion of other obvious causes of portal hypertension. One possible proposed event in the development of INCPH is a portal venopathy, caused by thrombosis or obliteration due to hypercoagulopathy, endothelial injury, or autoimmune injury. The management of INCPH is focused on the prevention and management of complications of portal hypertension, such as varix bleeding, ascites, and portal hypertensive gastropathy. INCPH should be a differential diagnosis in patient presenting with symptoms and signs of portal hypertension.



- A) CT imaging of Hepatic vein area
- B) CT imaging at portal bifurcation area
- C) Coronal view
- D) Transjugular liver biopsy