

Rare hepatocellular carcinoma presentation: Hepatoportal sclerosis

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Abstract

Hepatoportal sclerosis (HPS) is an idiopathic non-cirrhotic portal hypertension (INCPH) characterized by hypersplenism, portal hypertension, and splenomegaly. Hepatocellular carcinoma (HCC) is the most common form of liver cancer. Non-cirrhotic portal hypertension is an extremely rare cause of HCC. A 36-year-old woman was referred to our hospital with esophageal varices. All serologic tests for etiology were negative. Serum ceruloplasmin and serum Ig A-M-G were normal. In the follow-up, two liver lesions were identified on a triple-phase computer. The lesions had arterial enhancement but no washout in the venous phase. In the magnetic resonance imaging examination, differentiation in favor of HCC was considered at one of the lesions. Radiofrequency ablation therapy was first applied to a patient who had no signs of metastasis. Within 2 months, the patient underwent a living donor liver transplant. In explant pathology, well-differentiated HCC and HPS were considered the cause of non-cirrhotic portal hypertension. The patient has been followed without relapse for 3 years. The development of HCC in INCPH patients is still debatable. Despite the presence of liver cell atypia and pleomorphism in nodular regenerative hyperplasia liver specimens, a causal link between HCC and INCPH is yet to be established.

Keywords: Idiopathic non-cirrhotic portal hypertension; hepatocellular carcinoma; hepatoportal sclerosis.

Introduction

Idiopathic non-cirrhotic portal hypertension (INCPH) is a rare disease characterized by intrahepatic portal hypertension and splanchnic venous thrombosis in the absence of cirrhosis or other causes of liver disease. Symptoms of portal hypertension, such as gastroesophageal varices, variceal bleeding, and splenomegaly, are common in INCPH patients. In the presence of the precipitating factors, ascites and/or liver failure may develop.^[1]

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Liver cancer incidence is growing worldwide.^[2] Hepatocellular carcinoma (HCC) is the most common form of liver cancer. Most cases of HCC are associated with hepatitis B virus infection and nonalcoholic fatty liver disease. Other risk factors for developing HCC include hepatitis C, alcoholic liver disease, intake of aflatoxin-contaminated food, diabetes, and obesity.^[3] Non-cirrhotic portal hypertension is an infrequent cause of HCC. Here, we report a rare case of HCC based on hepatoportal sclerosis (HPS). On computed tomography (CT), evidence of significant portal hypertension was detected. There was no portal vein thrombosis.

Case Report

A 36-year-old woman with a 2-year history of liver cirrhosis presented with esophageal varices and was admitted to our hospital. Serum aminotransferase levels were 1.5–2 times higher than normal. All serologic tests for etiology were checked. The viral hepatitis panel and the autoimmune hepatitis panel were negative. Serum ceruloplasmin, serum alpha-1 antitrypsin, and serum IgA-M-G were normal. On abdominal ultrasound, splenomegaly (200 mm in diameter) was detected. A triple-phase CT scan demonstrated the increased caudate-to-right lobe ratio in the presence of preserved liver volumes with evidence of significant portal hypertension, including splenomegaly and varices. Her MELD-Na score was 7, and the Child-Pugh Score was A. There was no portal vein thrombosis. Non-cirrhotic portal hypertension was first considered in the patient.

In the 2-year follow-up, two liver lesions, 8 mm × 4.5 mm in segment IVA and 6 mm × 7 mm in segment VIII, were identified on triple-phase CT. The lesions had arterial enhancement but no washout in the venous or delayed phase. In the dynamic magnetic resonance imaging examination performed 2 years later, the increase in size in the 20-mm lesion seen in segment 8 includes a slightly hyperintense lesion in the T2 sequence and diffuse enhancement in the arterial phase in contrasted sequences. It was observed that the contrast enhancement continued moderately in the portal and venous phases. Differentiation in favor of HCC was considered. The alpha feta protein was 3 ng/mL. Radiofrequency ablation therapy was first applied to the patient who had no signs of metastasis, but multiple lesions were detected in the liver. Preparation for transplantation was started because LI-RADS 4 and 5 lesions were present.

After 2 months, the patient underwent a living donor liver transplant. In addition, well-differentiated HCC measuring 4 cm × 3 cm × 3 cm was seen, and also HPS was considered as the cause of non-cirrhotic portal hypertension in explant pathology (Fig. 1). Fibrous enlargement and short fibrous septa have been observed in some of the portal areas,

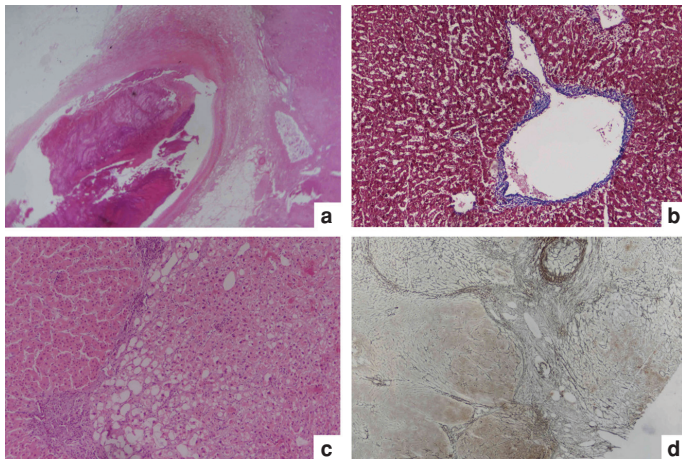


Figure 1. (a) Thrombus in the hepatic vein, (b) herniation to the parenchyma in the central vein (Masson's trichrome), (c) hepatocellular carcinoma, (d) loss of reticulin in hepatocellular carcinoma (reticulin stain).

which are also evident with Masson's trichrome in the surrounding liver tissue. In addition, enlargement of the central veins, pericentral fibrosis, mild thickening, and enlargement of the portal veins, as well as vascular proliferation, were observed. Non-cirrhotic portal hypertension was thought to be caused by HPS. The patient has been followed without relapse for 3 years.

Discussion

HCC has rarely caused by non-cirrhotic portal hypertension, particularly HPS. In contrast to cirrhosis, the risk of developing HCC is very low, and anecdotal reports of HCC in non-cirrhotic portal hypertension patients exist.^[4] The development of HCC in INCPH patients is still a topic of debate. Despite the presence of liver cell atypia and pleo-

morphism in nodular regenerative hyperplasia liver specimens, a causal link between HCC and INCPH is yet to be established.^[5]

The presence of NRH and HCC in the majority of patients could be attributed to other factors known to be linked to the development of NRH (e.g., portal vein thrombosis, chemotherapy, and radiotherapy), and the study failed to identify this disorder as the underlying cause.^[5]

Although the guidelines do not recommend routine HCC surveillance in INCPH, it should be kept in mind that HCC may develop in patients with NCPH.

Informed Consent: Written informed consent was obtained from the patient for the publication of the case report and the accompanying images.

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