Porto-Sinusoidal Vascular Disease

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Introduction

Porto-sinusoidal vascular disease (PSVD) is defined as a vascular liver disease characterized by the absence of cirrhosis and the presence of specific histological features in the liver biopsy such as obliterative portal venopathy, nodular regenerative hyperplasia and incomplete septal fibrosis, with or without portal hypertension. PSVD is usually asymptomatic until complications of portal hypertension arise including portal vein thrombosis and variceal bleeding. The diagnosis of PSVD is based on liver biopsy. PSVD is largely under-recognized and underestimated due to lack of awareness among patients and physicians.

Case

A 41-year-old female with the history of chronic hepatitis B virus infection presents with intermittent right upper quadrant pain which happens several times daily and last for several minutes. Magnetic resonance imaging (MRI) shows multiple heterogeneous mass-like areas in the right lobe measuring up to 7.5 x 3.7 cm. The liver enzyme AST and ALT are slightly elevated. No evidence of cirrhosis or portal hypertension is identified. The liver biopsy shows architectural distortion with irregular distortion of portal tracts and central veins. There is marked dilation of centrilobular veins and patchy centrilobular sinusoidal dilation with focal absence of portal vein. The liver parenchyma has multiple subtle nodular areas highlighted by reticulin. There is minimally increased portal tract extracellular collagen deposits with no expansion in the periportal area and no significant subsinusoidal or pericentral fibrosis. The glutamine synthetase is positive in the centrilobular perivenular areas and does not show the characteristic map-like pattern, which is not suggestive of a focal nodular hyperplasia. The overall findings are favored the diagnosis of porto-sinusoidal vascular disease with nodular regenerative changes.

Results

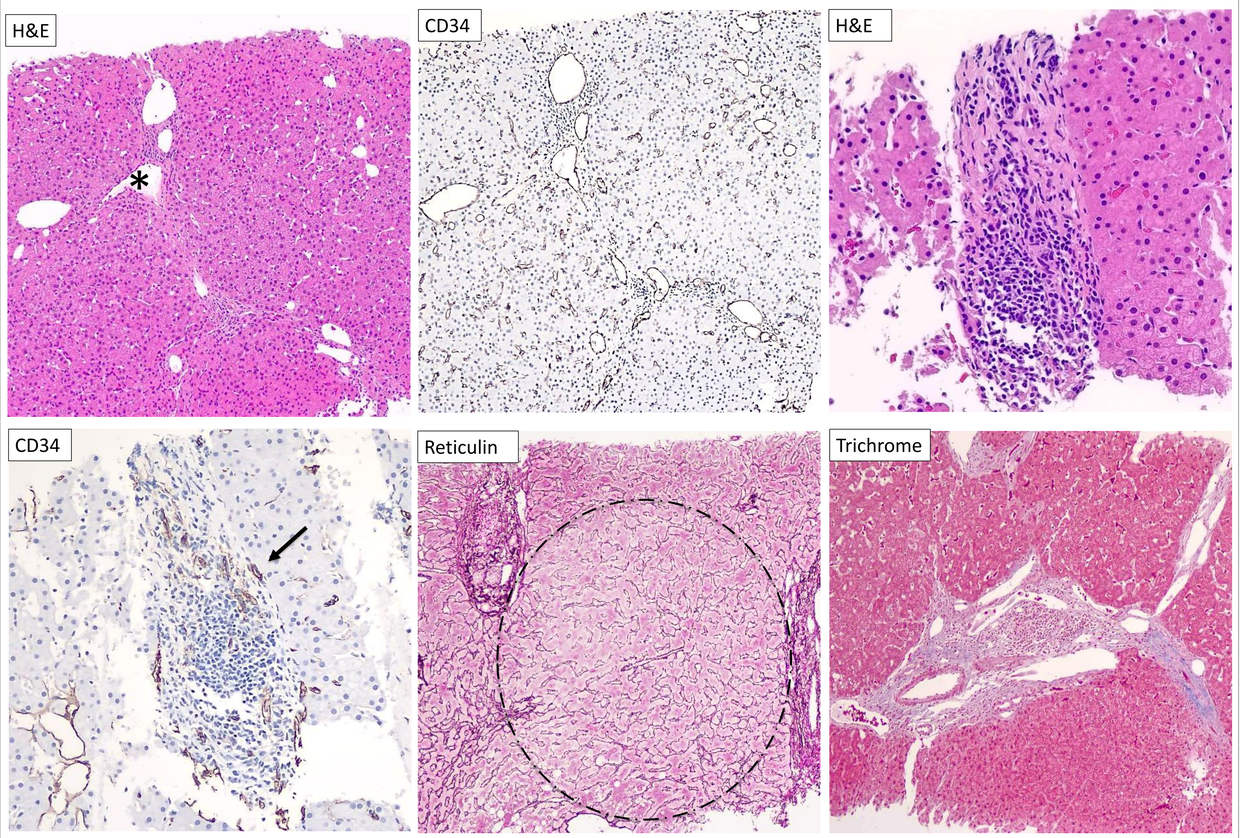

Figure 1. Porto-Sinusoidal Vascular Disease (PSVD). The biopsy cores of liver lesion show disrupted architecture with an irregular distribution of portal tracts, dilation of central and portal veins, sinusoidal dilation (CD34 immunostaining) and herniation of portal venules to the liver parenchyma (*). There is focal obliteration of the portal veins, replaced with several aberrant periportal venules with endothelial lining, as highlighted by CD34 stain (Black arrow). The hepatic parenchyma reveals several subtle nodular areas, better seen on reticulin staining (dotted circle) with no circumferential fibrosis. Trichrome stain highlights collagen deposition with no significant periportal expansion nor bridging fibrosis.

Discussion

The diagnosis of PSVD needs at least one of three specific histological features including obliterative portal venopathy, nodular regenerative hyperplasia and incomplete septal fibrosis or one specific sign for portal hypertension with absence of cirrhosis in more than 20 mm liver biopsy. In addition, the non-specific histological features including various portal tract abnormalities (such as multiplication, dilation of arteries, periportal vascular channels, and aberrant vessels), irregular distribution of the portal tracts and central veins, non-zonal sinusoidal dilation, and mild perisinusoidal fibrosis, are also critical for the differential diagnosis from other liver nodular hyperplasia entities such as focal nodular hyperplasia and nodular regenerative hyperplasia.

The treatment of PSVD focuses on managing portal hypertension-related complications and the long-term prognosis is better than cirrhosis, is associated with age, specific signs of portal hypertension, including ascites, and underlying conditions.

Due to lack of the awareness, it is critical to consider PSVD when there is portal hypertension with normal or slightly elevated liver stiffness and portal pressure values, and histological hallmarks.

References

1. De Gottardi A, Rautou P-E, Schouten J, Rubbia-Brandt L, Leebeek F, Trebicka J, et al. Porto-sinusoidal vascular disease: proposal and description of a novel entity. Lancet Gastroenterol Hepatol 2019;4(5).
2. Jin SJ, Choi WM. Porto-Sinusoidal Vascular Disease: A Concise Updated Summary of Epidemiology, Pathophysiology, Imaging, Clinical Features, and Treatments. Korean J Radiol. 2023 Jan;24(1):31-38. doi: 10.3348/kjr.2022.0668. PMID: 36606618; PMCID: PMC9830138.
3. Barisic-Jaman M, Milosevic M, Pastrovic F, Skrtic A, Grgurevic I. Porto-sinusoidal vascular disease: a new definition of an old clinical entity. Clin Exp Hepatol. 2023 Dec;9(4):297-306. doi: 10.5114/ceh.2023.133107. Epub 2023 Nov 24. PMID: 38774199; PMCID: PMC11103802.3. De Gottardi A, Sempoux C, Berzigotti A. Porto-sinusoidal vasculardisorder. J Hepatol. 2022;77(4):1124-1135.