

# An Anomalous Cause of Portal Hypertension

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## ABSTRACT

Portal hypertension is a syndrome marked by an increase in the pressure of the portal vein. Portal hypertension can be diagnosed clinically or if the measurement of the hepatic venous pressure gradient is greater than 5 mm Hg. Cirrhosis is the most common etiology in Western countries, but there are other causes which lead to presinusoidal portal hypertension. We present a patient with a rare cause of portal hypertension.

## INTRODUCTION

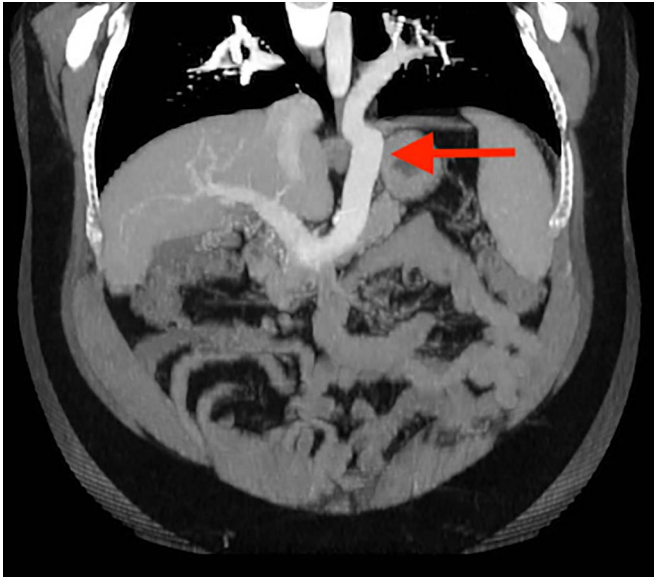
Portal hypertension is a syndrome in which there is a pathological increase of portal venous pressure. Clinically, it leads to complications such as variceal bleeding and ascites. Portal hypertension can be diagnosed clinically or, if there is any doubt, by measuring the hepatic venous pressure gradient. A gradient greater than 5 mm Hg is diagnostic. Cirrhosis is the most common etiology in Western countries, but there are other causes, including schistosomiasis and portal vein thrombosis, which lead to presinusoidal portal hypertension that are often forgotten. We present a patient with a rare cause of portal hypertension.

## CASE REPORT

A 39-year-old man with a medical history of obesity, hypercholesterolemia, hypertension, and no significant alcohol use was referred to hepatology for concern for nonalcoholic steatohepatitis in the setting of abnormal results in liver tests, splenomegaly, and thrombocytopenia. Five years before, he had been told that he had abnormal liver tests and abdominal ultrasound showing fatty liver. Triple-phase computed tomography (CT) the following year showed prominent undulation of the undersurface of the right hepatic lobe suggestive of focal fat and splenomegaly. He had not undergone noninvasive fibrosis testing. At his initial clinic visit, physical examination was unremarkable and without signs of chronic liver disease. Notable laboratory data included aspartate aminotransferase 48 U/L, alanine aminotransferase 98 U/L, alkaline phosphatase 176 U/L, international normalization ratio 1.0, and platelet count  $106 \times 10^9/L$ .

The original CT scan and an updated triple-phase CT were reviewed and showed venous communication between the left pulmonary veins and the portal vein's confluence suspicious for partial anomalous pulmonary venous return (PAPVR) (Figure 1). A pulmonary arteriogram showed evidence of pulmonary venous drainage to the portal system (Figure 2). Transjugular biopsy demonstrated a portosystemic gradient of 10 mm Hg and a wedged pressure of 22 mm Hg. Portal tracts had increased and irregularly distributed venules, fibrous expansion, increased chronic portal inflammation, and mild ductular reaction (Figure 3). The fibrosis extended into bridging fibrosis (Stage III). The lobules had sinusoidal and centrilobular vein dilatation.

Preoperative echocardiogram showed normal left ventricular size and function. The right ventricle (RV) appeared mildly dilated with normal systolic function. Cardiac catheterization pressures, in mm Hg, were significant for normal right atrial mean of 8, RV 56/11, pulmonary artery 49/23 (mean 32), and mean pulmonary capillary wedge pressure of 7. The systemic vascular resistance was 11 Wood



**Figure 1.** CT scan showing PAPVR to the central splenic vein near the junction with the superior mesenteric vein and portal vein. CT, computed tomography; PAPVR, partial anomalous pulmonary venous return.

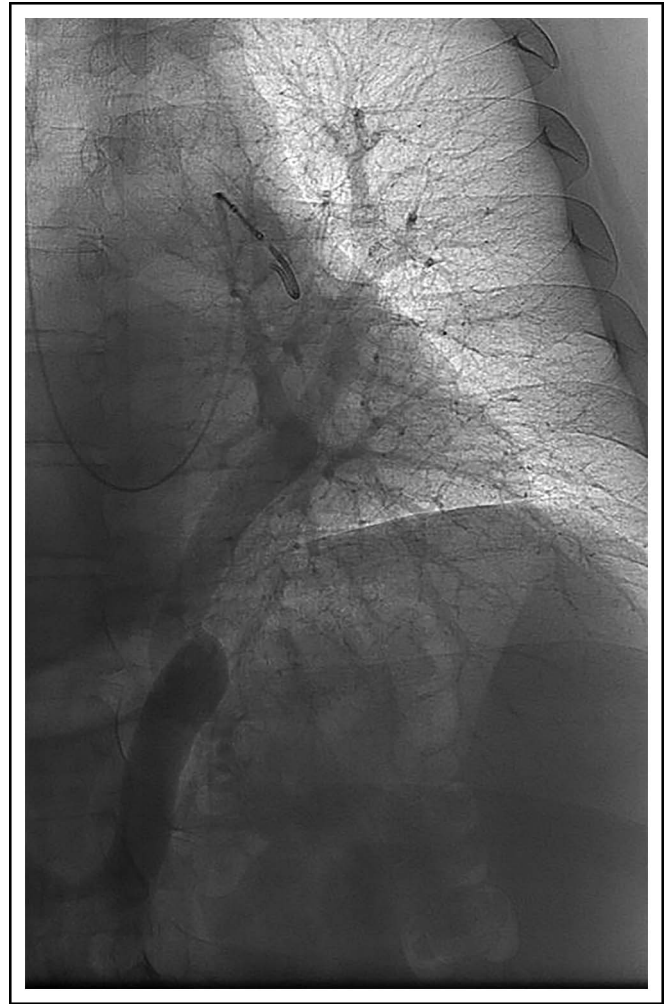
Units, and the peripheral vascular resistance was 3 Wood Units. This was consistent with mild pulmonary hypertension and slightly elevated pulmonary vascular resistance.

Based on these findings, it was felt that he had presinusoidal portal hypertension because of roughly half of the pulmonary blood flow returning to the portal circulation. He underwent surgical repair of PAPVR with ligation and division of the vertical vein and direct anastomosis of the vein to the left atrium. He continued to have mildly elevated results in liver tests and thrombocytopenia. Triple-phase CT a year after surgery showed mild improvement in moderate splenomegaly. Transjugular biopsy demonstrated improvement of the porto-systemic gradient (7 mm Hg) and wedged pressure (11 mm Hg). It demonstrated improved fibrosis, Stage Ia (peri-sinusoidal) down from Stage III. The portal venule abnormalities, portal chronic inflammation, portal ductular reaction, and sinusoidal and centrilobular vein dilatation resolved (Figure 3).

## DISCUSSION

We present a case of noncirrhotic portal hypertension from excess inflow into the portal vein from PAPVR of a left pulmonary vein draining into the portal vein rather than the left atrium. To our knowledge, this is the first case report of a PAPVR leading to portal hypertension. There has been at least 1 case of secondary cirrhosis and portal hypertension because of right-sided heart failure from PAPVR.<sup>1</sup>

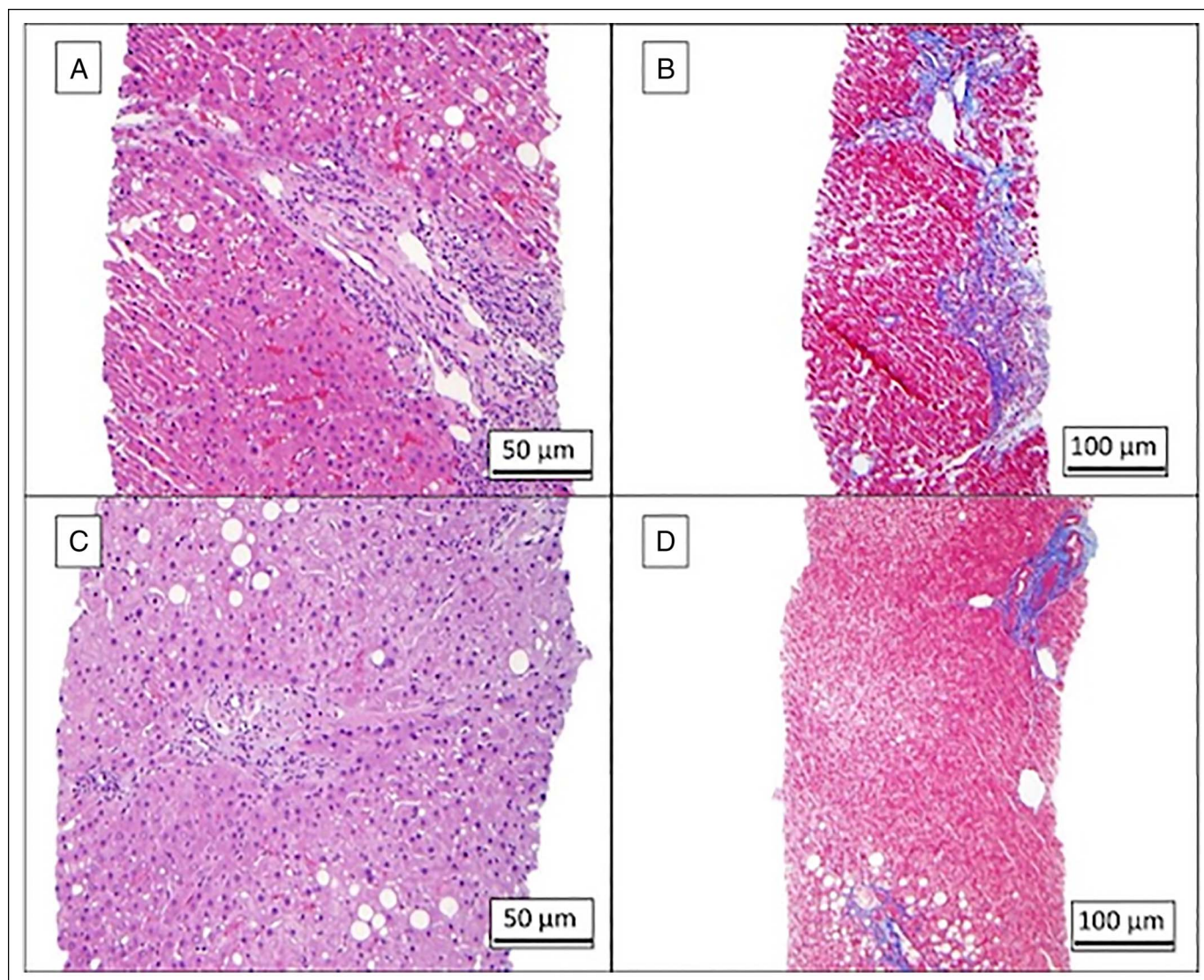
Anomalous pulmonary venous return can be total (involving all pulmonary veins) or partial. Total anomalous pulmonary venous connection is generally diagnosed during infancy, but PAPVR can go undiagnosed because it is often asymptomatic.<sup>2</sup> Incidence is



**Figure 2.** Pulmonary arteriogram demonstrating pulmonary venous drainage into the portal system.

0.1%–0.4%, but only about 20% require surgical correction when it causes significant right heart enlargement or pulmonary hypertension. Many cases involve the right pulmonary veins draining into the superior vena cava and can be found in association with an atrial septal defect. However, anomalous pulmonary veins can drain to the coronary sinus, azygous vein, and, rarely, below the diaphragm to the inferior vena cava or portal vein.

This particular constellation of portal tract changes has not been well-described in the setting of increased portal inflow and is instead usually seen with hepatic venous congestion.<sup>3</sup> The aberrant appearance and distribution of the venules are indicative of a vascular process, and the ductular reaction, chronic inflammation, and fibrosis suggest an acute-on-chronic injury which was reversed after PAPVR repair. Increased portal vein inflow is a rare cause of portal hypertension, but generally considered in vascular malformations such as hepatic arterioportal shunts.<sup>4</sup> This is the first case report of PAPVR leading to increased portal vein inflow and portal hypertension; it is important to keep on the differential when considering causes of increased portal vein inflow and noncirrhotic portal hypertension.



**Figure 3.** Liver biopsy images before surgery showing that (A) the portal tracts contain abnormal portal vasculature with signs of vascular remodeling, with mild chronic inflammation and ductular reaction (H & E stain) and (B) porto-portal bridging fibrosis is observed (trichrome stain). After surgical repair, images showing (C) that portal tracts are unremarkable (H & E stain) and (D) residual perisinusoidal fibrosis (trichrome stain). H & E, hematoxylin and eosin.

## DISCLOSURES

Author contributions: SA Brown, S. Healan, and D. Bichell wrote the article. VQ Trinh provided the pathology images. B. Frischhertz and A. Scanga revised the article for intellectual content. SA Brown is the guarantor of the article.

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Informed consent was obtained for this case report.

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