The Moment HVPG Changed our Mind: Role of Liver Hemodynamics in Vascular Liver Disease

O Momento em que o Gradiente de Pressão Venosa Hepática Muda a Nossa Mente: O Papel da Hemodinâmica Hepática na Doença Vascular do Fígado

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Abstract:

Portosinusoidal vascular liver disorder (PSVD) is an uncommon cause of clinically significant portal hypertension that can be misdiagnosed as cirrhosis, leading to delayed management. We report a case in which low liver stiffness, portal hypertension manifestations, and hemodynamic assessment reoriented the diagnosis.

A 61-year-old man previously diagnosed with MetALD and prior oxaliplatin-based chemotherapy presented in 2017 with variceal upper gastrointestinal bleeding. Although imaging suggested cirrhosis with portal vein thrombosis, liver stiffness was low (LSM 7.7 kPa) and HVPG was 9 mmHg. Inconsistencies in the diagnostic work up led us to perform a liver biopsy, which showed portal abnormalities with focal sinusoidal dilatation and no fibrosis.

Even when other potential causes of liver disease are present, in the absence of a liver biopsy, the underlying aetiology cannot be assumed to be the driver of portal hypertension; in this setting, liver elastography together with invasive hepatic hemodynamic measurements can help reorient the diagnosis toward PSVD and guide management.

Keywords: Elasticity Imaging Techniques; Hypertension, Portal; Liver Diseases; Vascular Diseases.

Resumo:

O distúrbio vascular portos-sinusoidal hepático (DVPS) é uma causa incomum de hipertensão portal clinicamente significativa, podendo ser erroneamente diagnosticado como cirrose e conduzir a atraso na abordagem terapêutica. Reporta-se um caso em que o baixo valor de elastografia hepática, as manifestações de hipertensão portal e a avaliação hemodinâmica permitiram reorientar o diagnóstico. Homem de 61 anos, previamente diagnosticado com MetALD e com história de quimioterapia baseada em oxaliplatina, apresentou em 2017 hemorragia digestiva alta por rotura de varizes. Embora a imagiologia sugerisse cirrose com trombose da veia porta, a rigidez hepática era baixa (7,7 kPa) e o gradiente de pressão venosa hepática, de 9 mmHg. As inconsistências na investigação motivaram biópsia hepática, que revelou alterações portais com dilatação sinusoidal focal e ausência de fibrose. Mesmo na presença de outras causas potenciais de doença hepática, a etiologia subjacente não deve ser presumida como determinante da hipertensão portal sem confirmação histológica; neste contexto, a elastografia e a avaliação hemodinâmica invasiva são fundamentais para reorientar o diagnóstico para DVPS e orientar o tratamento.

Palavras-chave: Doenças do Fígado; Doenças Vasculares; Elastografia; Hipertensão Portal.

Learning Points

1. Portal hypertension does not necessarily indicate cirrhosis. Porto-sinusoidal vascular disorder (PSVD) should be considered in patients with discordant clinical and non-invasive findings.
2. Low liver stiffness despite portal hypertension is a diagnostic red flag. This finding should prompt evaluation for non-cirrhotic vascular liver diseases.
3. Hepatic hemodynamic assessment can reorient the diagnosis. HVPG measurement may help distinguish PSVD from cirrhosis.
4. Liver biopsy remains essential in uncertain cases. Histology can confirm PSVD and exclude cirrhosis.

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Introduction

Portosinusoidal vascular liver disorder (PSVD) is a rare entity characterized by abnormal vascular architecture in the absence of cirrhosis that is often associated with portal hypertension.[1,2] This entity is most frequently associated with immunological disorders, haematological disorders, and certain medications, among others, but it can remain without any identifiable cause in up to 31.7% of cases.[2] However, as clinical manifestations include portal hypertension complications, it might be mistaken for cirrhosis. We present a case illustrating how PSVD can be suspected in this setting, supported by elastography and hepatic venous pressure gradient (HVPG) measurements.

Case Report

We present the clinical case of a 61-year-old man who presented at our tertiary referral hospital for upper gastrointestinal bleeding in 2017. His past medical history included arterial hypertension, type 2 diabetes mellitus, and colorectal cancer (right hemicolectomy for pT4N0M0 colon cancer in 2009, followed by a three-month course of adjuvant chemotherapy with XELOX (capecitabine and oxaliplatin); surgical removal of pT1/T2N0M0 rectal cancer in 2013) with negative oncologic follow-up. He also had metabolic dysfunction and alcohol-related liver disease (MetALD), with consumption of 4 alcohol units/day and no previous evidence of advanced disease. His chronic medication included metformin 1000 mg daily and enalapril 20 mg daily.

Upon presentation with hematemesis, he underwent urgent esophagogastroduodenoscopy (EGD), with evidence of three large oesophageal varices with active bleeding, and severe portal hypertensive gastropathy (PHG); endoscopic band ligation (EBL) with five bands was performed, achieving good endoscopic control. Furthermore, complex conservative treatment of portal hypertension-related bleeding, including antibiotics and terlipressin, was initiated. During work-up evaluation, blood exams were performed, and the results are reported in Table 1. Etiological work-up for liver disease excluded viral, autoimmune, and cholestatic liver disease, as well as iron or copper accumulation, alpha-1-antitrypsin deficiency, thyroid disorders, and celiac disease.

Moreover, imaging with ultrasound (US) and computed tomography (CT) scan showed signs of advanced liver disease with irregular liver surface and portal hypertension with splenomegaly, porto-systemic collateral circulation, and partial thrombosis of the main portal vein (approximately 50% of the circumference), extending into the right portal branch and into the superior mesenteric vein. The splenic vein was patent, and no ascites was present. The initial diagnostic approach was oriented toward cirrhosis attributed to MetALD, with Child–Pugh class B (8 points) and active bleeding meeting high-risk criteria; therefore, he was evaluated for pre-emptive TIPS. However, during the prior work-up, vibration-controlled transient elastography (VCTE) performed with FibroScan® (Echosens, Paris, France) showed a low liver stiffness measurement (LSM) of 7.7 kPa (interquartile range [IQR], 1.1 kPa); and HVPG was 9 mmHg with the presence of venovenous collaterals, which may lead to underestimation of HVPG measurements (Fig. 1).

Given the discordance between portal hypertension-related complications (variceal bleeding) and low LSM and

Table 1: Blood exams at presentation.

Leucocytes (x 10^9/L)	3.47	AST (IU/L)	31
RBC (x 10^{12}/L)	2.25	ALT(IU/L)	15
Haemoglobin (g/L)	70	GGT (IU/L)	33
MCV (fL)	95	ALP (IU/L)	58
Platelets (x 10^9/L)	82	Total bilirubin (mg/dL)	3
PT (%)	78	Direct bilirubin (mg/dL)	0.6
PT (sec)	12.8	Albumin (g/L)	29
INR	1.14	Creatinine (mg/dL)	0.96
aPTT (sec)	24.9	Sodium (mmol/L)	139
Fibrinogen (g/L)	1.8	Potassium (mmol/L)	4.3

RBC – red blood cells; MCV – mean corpuscular volume; PT – prothrombin time; INR – international normalized ratio; aPTT – activated partial thromboplastin time; AST – asparagine aminotransferase; ALT – alanine aminotransferase; GGT – gamma glutamyl transferase; ALP – alkaline phosphatase.

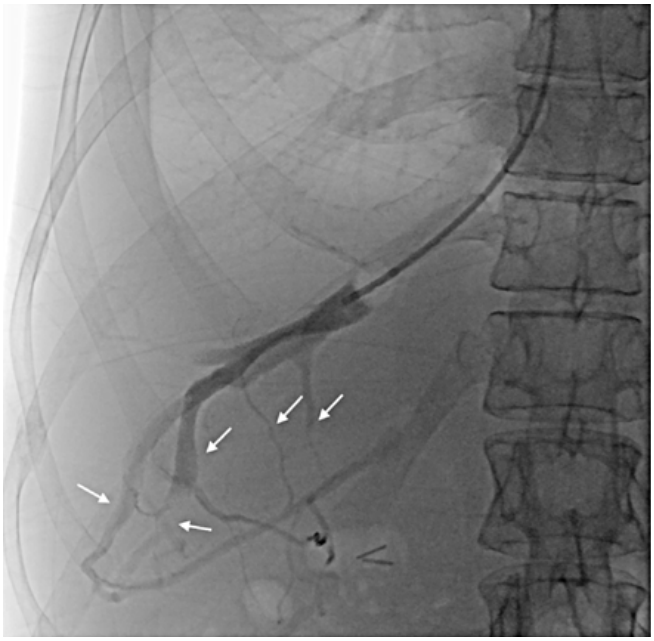

Figure 1: A wedged fluoroscopic venography of the right hepatic vein reveals five large veno-venous communications.

HVPG values, PSVD was suspected. TIPS was not performed because the high-risk criteria for considering pre-emptive TIPS do not apply to vascular liver disease, and haemostasis was achieved at the initial EGD. A transjugular liver biopsy was performed at the time of HVPG measurement, showing the absence of portal veins in six portal spaces and abnormal portal vein distribution in the other four, with focal sinusoidal dilatation. Mild macrovesicular steatosis was present, and no fibrosis was identified. These findings supported the exclusion of cirrhosis and the diagnosis of PSVD. A thrombophilia work-up was then performed, which did not reveal an underlying prothrombotic disorder.

Overall, the case illustrated a portal hypertension syndrome mimicking cirrhosis from a radiological standpoint, which was reoriented by low elastography, HVPG not compatible with CSPH despite variceal bleeding, and characteristic histology consistent with PSVD in the context of prior oxaliplatin exposure.

The patient was discharged with anticoagulation (low molecular weight heparin) secondary prophylaxis with beta-blockers and further endoscopic band ligation. However, one month later, he was readmitted due to recurrent variceal haemorrhage, which could not be controlled with band ligation and required placement of an oesophageal stent as bridge therapy. In consideration of the recurrent episode of variceal bleeding and the failure to achieve endoscopic control, a transjugular intrahepatic portosystemic shunt (TIPS) was required. During the procedure, hemodynamic assessment at portal puncture showed severe portal hypertension with a portal pressure gradient (PPG) of 20 mmHg (portal pressure 32 mmHg, inferior vena cava pressure 12 mmHg). A PTFE-covered stent (10 mm × 7 cm + 2 cm) was placed and dilated to 8 mm, achieving a descent of PPG to 6 mmHg (portal pressure 21 mmHg, inferior vena cava pressure 15 mmHg). The 24-hour catheterization without sedation confirmed sustained PPG reduction to 7 mmHg (Fig. 2). Ultrasound and catheterization follow up at 1 month after TIPS placement demonstrated a patent TIPS, with a portal pressure gradient of 7.5 mmHg. Liver stiffness measurements also remained low throughout follow up (LSM 6–8.1 kPa). The patient remained compensated and free of portal hypertension–related complications up to the present day.

Discussion

In this clinical scenario, the patient exhibited signs that are generally consistent with the natural history of liver cirrhosis in the context of previously diagnosed MetALD. Firstly, there was a persistent alteration in liver biochemical parameters, and secondly, imaging revealed a nodular liver surface on ultrasound and computed tomography. In addition, imaging findings demonstrated features of portal hypertension, including the presence of perigastric and periesophageal collateral circulation as well as splenomegaly. The patient subsequently experienced two episodes of portal hypertension-related bleeding, which ultimately led to the creation of a transjugular intrahepatic portosystemic shunt. As the patient had MetALD, it could have allowed for a plausible etiological classification of the chronic liver disease.

However, several red flag features were present that prompted a more in-depth diagnostic work-up for PSVD. Firstly, the patient had a history of colorectal carcinoma, specifically treated with adjuvant XELOX chemotherapy (capecitabine and oxaliplatin), a regimen known to be associated with vascular liver injury.[3] Secondly, there was an unexpectedly low liver stiffness measurement (7.7 kPa) and a hepatic venous pressure gradient of 9 mmHg, despite clear radiological and endoscopic signs of portal hypertension.[3,4] Altogether, these findings led appropriately to a liver biopsy. Histopathological examination revealed the exclusion of cirrhosis and was compatible with obliterative venopathy.

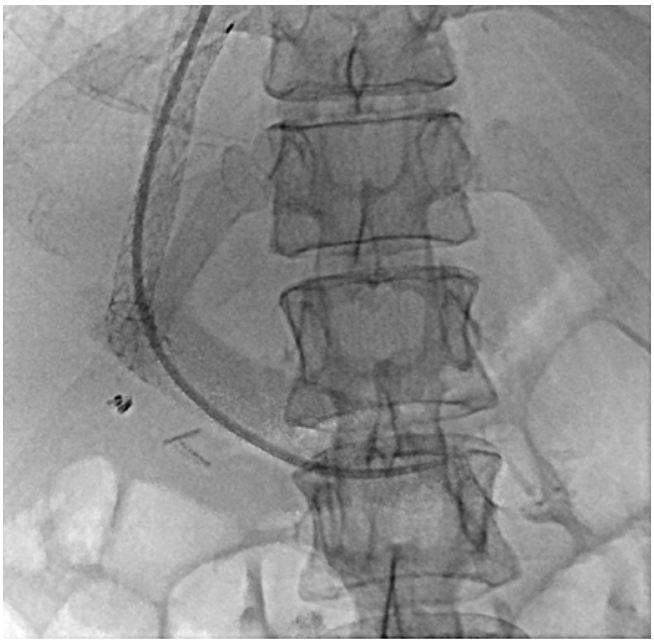

Figure 2: A fluoroscopic venography demonstrating patency of portosystemic intrahepatic shunt.

In this clinical case, the patient had a liver biopsy that excluded cirrhosis, and met the VALDIG diagnostic criteria for PSVD with specific signs of portal hypertension with variceal bleeding, collateral circulation, as well as specific histopathology signs with obliterative venopathy.[1,5]

In conclusion, this case underscores that even in patients with well-established risk factors for chronic liver disease, portal hypertension complications are not synonymous with cirrhosis in the absence of a liver biopsy. A complete evaluation using non-invasive liver stiffness measurement (LSM) and invasive hepatic venous pressure gradient (HVPG) assessment is critical to accurately orient the diagnosis. ■

Contributorship Statement

XMC, AB, JG, VHG – Design of the case, data collection, analysis and interpretation of the data, and drafting of the manuscript.
All authors approved the final version to be published.

Declaração de Contribuição

XMC, AB, JG, VHG – Conceção do caso, recolha de dados, análise e interpretação dos dados, e redação do manuscrito.
Todos os autores aprovaram a versão final a ser publicada.

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