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Case Report

Acute acalculous cholecystitis with portal cavernoma: A case report with literature review ☆,☆☆

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ABSTRACT

Portal cavernoma cholangiopathy (PCC) refers to morphological changes in the intrahepatic, extrahepatic biliary system, along with the gallbladder (GB), induced by portal cavernoma (PC). Acute acalculous cholecystitis (AAC) represents an infrequent clinical manifestation of PCC. Given the inadequacy of documentation within medical literature, AAC may go undiagnosed among patients with PC presenting symptoms of right upper quadrant pain. The current study aims to report a case of acute acalculous cholecystitis secondary to portal cavernoma, focusing on radiological findings, with a brief review of literature.

Introduction

Portal cavernoma is characterized by a network of tortuous and dilated collateral channels in cases of portal vein obstruction. These bridging vessels encircle bile ducts and project towards the lumen, inducing morphological changes in the intrahepatic, extrahepatic biliary system, as well as gallbladder. This condition is termed portal cavernoma cholangiopathy [1,2]. Studies have shown that biliary structural alterations are identified in approximately 80%-100% of patients with PC. Nevertheless, only a smaller percentage, ranging from 5% to 38% of individuals, exhibit symptoms [3,4]. Typically, clinical manifestations of PCC include cholestasis, cholangitis, and the development of gallstones [5]. Acute acalculous cholecystitis (AAC) constitutes an infrequent presentation of PCC, which remains insufficiently documented in existing literature. AAC merits recognition as a potential etiology of right upper quadrant pain in patients with PC. Conversely, AAC denotes a critical clinical entity usually intertwined with concomitant comorbidities, emphasizing the imperative acknowledgment of PCC as an uncommon underlying condition in cases of AAC.

Case report

A 70-year-old male presented to the emergency room with worsening right upper quadrant pain of 3-day duration. He

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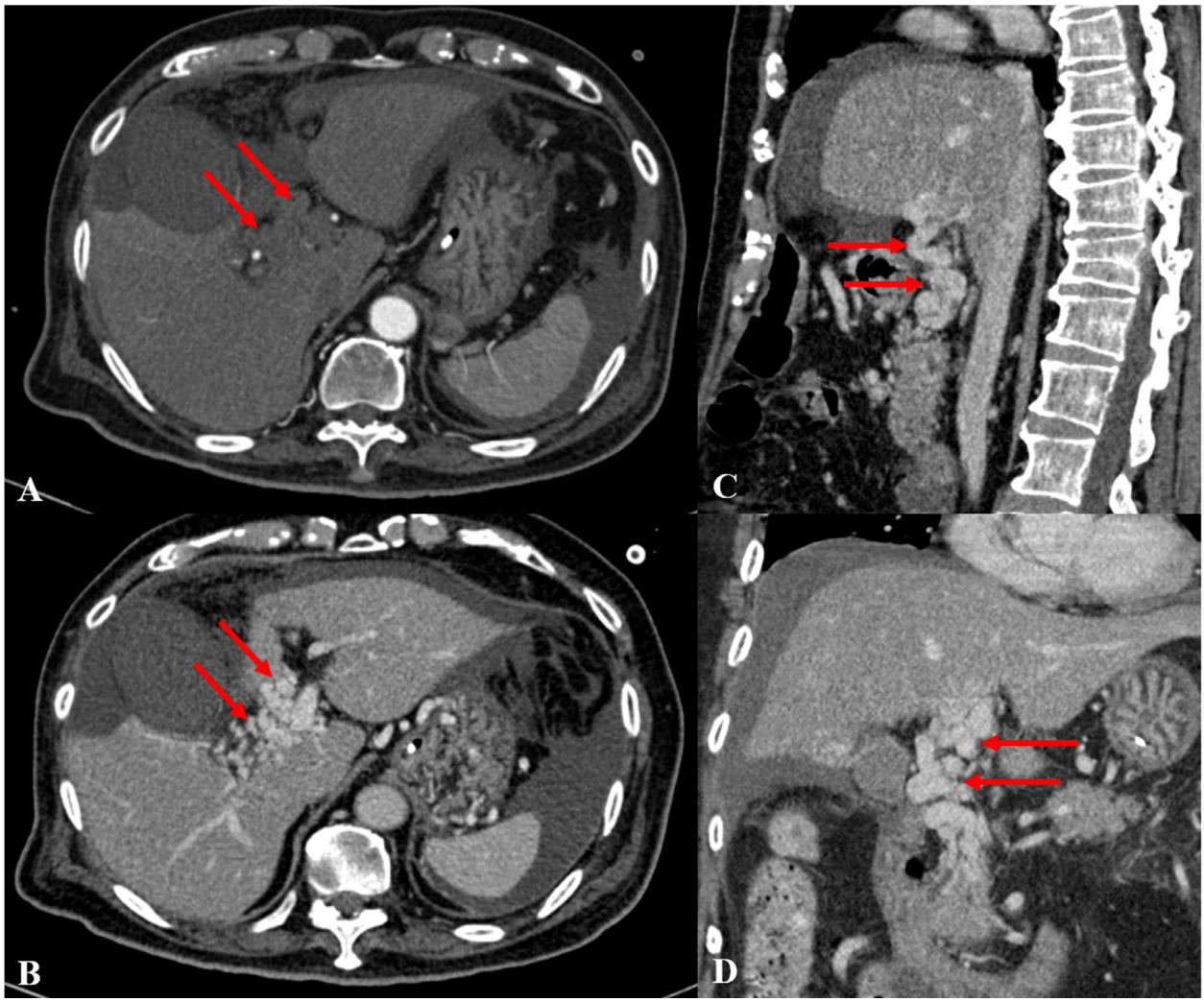

Fig. 1 – Axial CT scan through the hepatic hilum in the arterial phase (A) and portal venous phase (B). Sagittal (C) and coronal (D) reconstruction CT scan through the hepatic hilum in the portal venous phase. Numerous tortuous channels enhancing during portal venous phase and not during arterial phase (→), consistent with a diagnosis of portal cavernoma.

Table 1 – Laboratory tests.

Study	Result	Normal values
WBC	12.63 G/L	4-10 G/L
Total bilirubin	1.44 mg/dL	1.02 mg/dL
GGT	114 U/L	<40 U/L
CRP	22.8 mg/L	<5 mg/L

CRP, C-reactive protein; GGT, gamma-glutamyl transferase; WBC, white blood count.

described the pain as sharp, constant, worse with movement and alleviated by remaining still. He also had nausea, vomiting, and fever. On admission, he had the following initial vital signs: blood pressure 140/100 mm Hg, pulse rate 121 beats/min, respiratory rate 26 breaths/min, body temperature 38°C. Physical examination showed right upper quadrant tenderness. Laboratory tests performed on admission revealed elevated CRP level, leukocytosis, as well as high GGT, hyperbilirubinemia (Table 1).

The patient underwent abdominal contrast-enhanced computerized tomography (CT) examination. Numerous tortuous channels enhancing during portal venous phase and not during arterial phase were found in the hepatic hilum, consistent with a diagnosis of portal cavernoma (Fig. 1), whereas, no varicose vein was found in the gallbladder wall (Fig. 2). The extrahepatic biliary tree was directly compressed by PC (Fig. 3). The results also demonstrated gallbladder distension (with a transverse diameter of 5 cm), diffuse GB wall thickening (with a thickness of 5 mm) and absence of wall enhancement without the presence of GB calculus (Fig. 2). In addition, there were pericholecystic fat stranding and increased enhancement of the hepatic parenchyma of the gallbladder fossa resulting from reactive hyperemia (Fig. 4). Radiological features of portal hypertension such as ascites, portosystemic shunt, were noted.

Due to the presence of multiple dilated collateral veins in the hepatocystic triangle, PC was considered a relative contraindication for laparoscopic cholecystectomy. Gallbladder drainage via a percutaneous cholecystostomy tube was per-

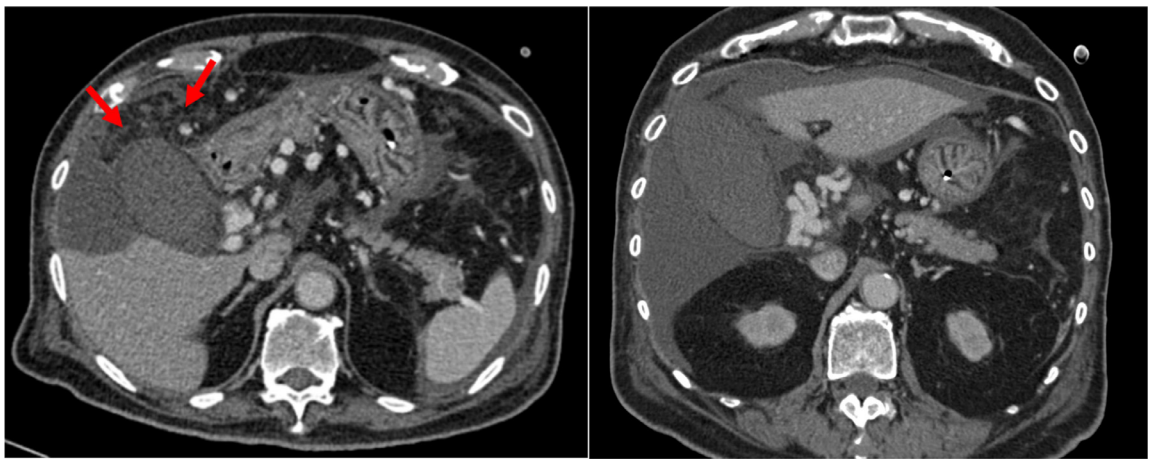

Fig. 2 – Axial CT scan through the gallbladder in the portal venous phase (A). Oblique axial reconstruction CT scan along the longitudinal axis of the gallbladder in the portal venous phase (B). Gallbladder distension (with a transverse diameter of 5 cm), diffuse gallbladder wall thickening (with a thickness of 5 mm), absence of wall enhancement, pericholecystic fat stranding (→) without the presence of gallbladder calculus. No varicose vein in the gallbladder wall.

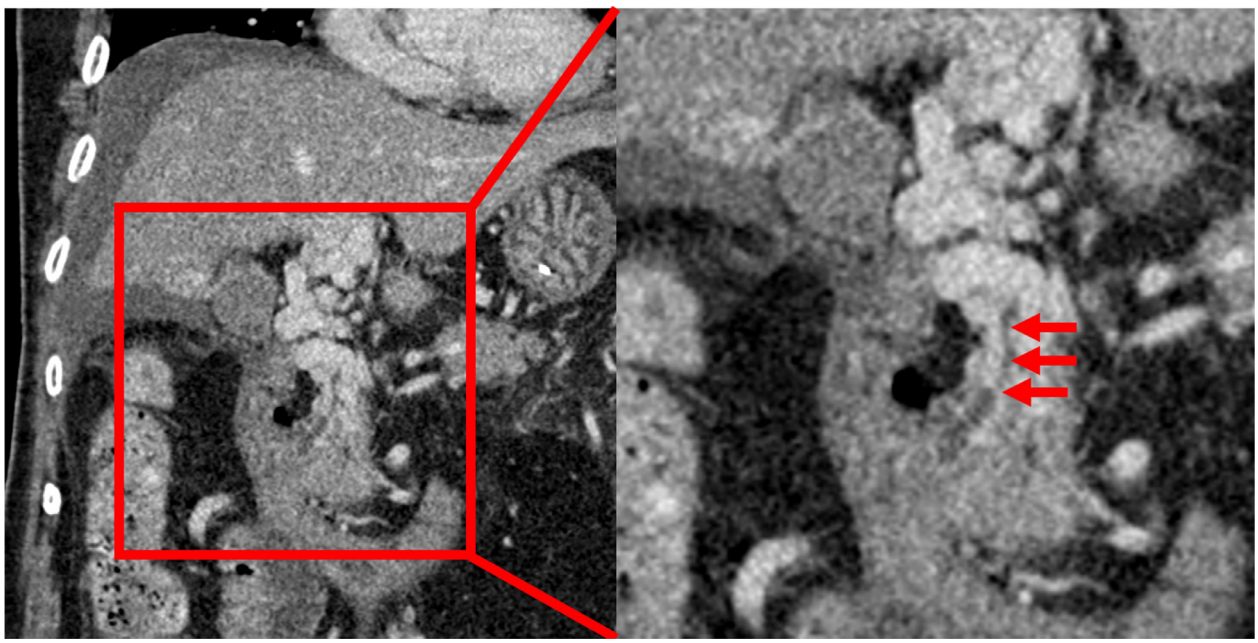

Fig. 3 – Coronal reconstruction CT scan through the common bile duct in the portal venous phase. The common bile duct (→) was directly compressed by the portal cavernoma.

formed in an interventional radiology suite under local anesthesia. The procedure was uneventful. Subsequently, the patient was transferred to the intensive care unit where his health status was closely monitored. Ultrasound examination also indicated no gallstones (not shown).

Discussion

Portal cavernoma is formed by a bunch of serpiginous and distended hepatopetal collaterals in response to portal vein obstruction. The veins that drain the biliary tract are arranged in the form of 2 plexuses. The paracholedochal venous plexus of Petren lies outside and parallel to the bile ducts whereas the epicholedochal venous plexus of Saint is a fine reticular plexus found on the bile duct wall. The cholecystic vein drains into the paracholedochal venous plexus. In patients with portal vein obstruction, the cholecystic vein, paracholedochal and epicholedochal venous plexuses dilate to provide an alternate route for blood flow around the obstructed segment of the portal vein [1,2,6]. The appearance and location of varices depends on the extent and location of obstruction in the portal system [7].

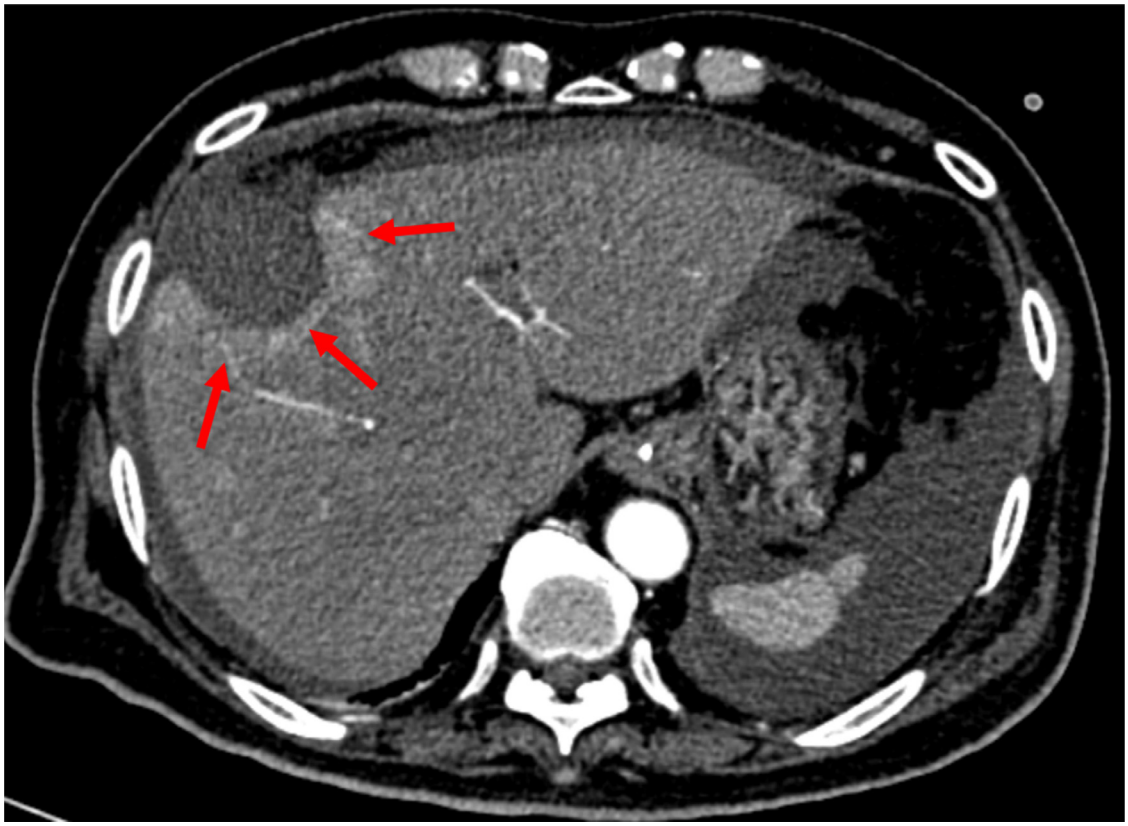

Fig. 4 – Axial CT scan through the gallbladder fossa in the arterial phase. Increased enhancement of the hepatic parenchyma of the gallbladder fossa (→) resulting from reactive hyperemia.

In this report, we describe a case of a 70-year-old male with findings of cholestasis related to portal cavernoma, referred to as portal cavernoma cholangiopathy. PCC includes morphological alterations in the intrahepatic, extrahepatic biliary tree, as well as gallbladder, induced by PC. There are 2 main mechanisms regarding the development of PCC: compression and ischemia. Collateral vessels in PC produce extrinsic pressure over the thin and flexible bile duct wall, which results in areas of indentation, irregular ductal contour, narrowing, duct angulation in association with upstream dilatation. Reduced contractile function of the GB due to the presence of varicose veins in the gallbladder wall also contribute to the development of bile stasis. Long-term local wall compression and portal hypertension may provoke ischemia of the bile duct [1,2,8]. Biliary structural changes are detected in approximately 80%-100% of patients with PC, although most of them are asymptomatic [3,4]. The symptoms and clinical presentations in connection with PCC include jaundice, cholangitis, cholelithiasis, cholecystitis… [5].

In our case, the CT results also meet the diagnostic criteria for acute cholecystitis: large perpendicular diameters of GB, increased GB wall thickness, pericholecystic fat infiltration and hyperenhancement of gallbladder fossa. The absence of gallstones was verified through both CT and ultrasound imaging. After contrast injection, there is no enhancement and no varicose vein in the GB wall, suggesting GB wall ischemia and thrombosed varix. In this context, the pathogenesis of acute acalculous cholecystitis appears to involve multiple factors. The initial factor is the obstruction of biliary outflow by PC as mentioned above, leading to biliary stasis [1,2,5,8]. GB stasis leads to the accumulation of bile salts with a build-up of intraluminal pressure, subsequently rendering the GB mucosa susceptible to ischemic injury. Thrombosed varix further advances the progression of GB epithelial ischemia. Besides, stagnation of bile may result in bacterial colonization, which in turn contributes to the inflammatory response [2,9,10].

Laparoscopic cholecystectomy is an effective treatment for AAC. However, this patient with multiple enlarged collateral veins in the hepatic hilum is considered a poor surgical candidate because of the high risk of bleeding; hence, a percutaneous cholecystostomy tube was placed to secure gallbladder drainage [9–12].

Conclusion

Portal cavernoma cholangiopathy includes morphological changes in the intrahepatic, extrahepatic biliary system, as well as gallbladder, induced by portal cavernoma. Acute acalculous cholecystitis is an infrequent presentation of portal cavernoma cholangiopathy and should be acknowledged as a potential cause of right upper quadrant pain in patients with portal cavernoma.

Patient consent

Written informed consent for the publication of this case report was obtained from the patient.

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