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Case Report

A rare case of congenital aneurysm of the portal system at level of spleno-porto-mesenteric confluence complicated by massive thrombosis ☆,☆☆,★,★★,†

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ABSTRACT

Portal venous aneurysm is a rare and potential dangerous vascular pathology, which can result in thrombosis or rupture. It may be congenital or acquired. Acquired form can be related mainly to portal hypertension, chronic hepatic disease, and trauma. We present a peculiar case of a congenital aneurysm involving the hepatic portal system in nearly all its extra-hepatic components: the main portal trunk, the spleno-porto-mesenteric confluence and the distal segment of splenic, superior, and inferior mesenteric veins, in a 20-year-old male patient. The aneurysm was complicated by massive thrombosis in absence of further predisposing factors.

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Introduction

Aneurysms of the hepatic portal system constitute a rare clinical entity, with a case of splenic vein aneurysm being first described in the literature in 1953 by Lowenthal & Jacob [1]. No gender predilection and a mean age of 53 years at diagnosis have been reported [2].

Aneurysms of the portal system can be acquired or congenital. Acquired forms can have an acute onset as a result of abdominal trauma [3] or can be chronic for hepatic cirrhosis and portal hypertension [4]. Vessel wall weakening due to inflammatory conditions such as pancreatitis or local degenerative alterations have also been implicated in the genesis of the acquired form [5]. Previous surgery has been reported as a predisposing acquired factor, too [6]. Congenital aneurysms etiology still remains partly unknown. Congenital forms typically arise from aberrant development of the vitilline veins during the embryonic period or from an inherent weakness in the wall of the vessel. [5–7].

This pathologic entity may be asymptomatic or can lead to severe conditions such as colicky abdominal pain, jaundice due to compression “ab extrinseco” of adjacent biliary tract, and digestive bleeding secondary to portal hypertension [8–10]

The diagnosis is usually achieved by ultrasonography, computed tomography and/or magnetic resonance imaging [11]. In the asymptomatic cases the portal venous system aneurysms are discovered accidentally thanks to diagnostic imaging performed for other clinical reasons [4,12]. Particularly, the diagnosis of an aneurysm of the portal venous system is carried out with high accuracy with contrast enhanced CT (ce-CT) by finding a dilatation of more than 2 cm in diameter [13].

Most portal venous aneurysms remain stable on follow-up, however, when an increased diameter is observed or complications such as thrombosis and/or rupture occur, surgical treatment should be considered. The most common surgical treatments include splenectomy and aneurysmorrhaphy [14].

Herein, we present the case of a young male showing a huge congenital aneurysm of spleno-porto-mesenteric venous system complicated by massive thrombosis.

Case report

A 20-years-old male presented to the emergency room with persistent abdominal pain. On physical examination, the pain was localized to the upper quadrants, however the abdomen was treatable, and no other particular clinical signs were disclosed. The patients did not have prior clinical records. Laboratory tests showed: deficiency of coagulation factors VII (27, *normal range: 50%-150% of the activity in pooled normal plasma*), and X (42, *normal range: 50%-150% of the activity in pooled normal plasma*), increased bilirubin (total: 2.12 mg/dL, *normal range 0,0-1,2 mg/dL*; direct: 0.56 mg/dL, *normal range* 0,0-0,3 mg/dL; indirect 1,56 mg/dL, *normal range: 0,0-0,9 mg/dL*) and a mild reduction of cholinesterase (5194 IU/L, *normal range: 5300-12900 IU/L*); in addition, the INR appeared increased (1.57, *normal range: 0.80-1.20*), and the D-Dimer was frankly positive (2862 ng/mL, *normal range: <270 ng/mL*); the remaining main parameters were unremarkable.

The patient underwent ce-CT of the chest and abdomen, which disclosed a huge aneurysmatic dilatation involving the portal trunk (axial diameters 30 × 40 mm), the distal splenic vein (axial diameters 37 × 39 mm) and the distal tract of both superior and inferior mesenteric vein (respectively SMV and IMV), complicated by a massive thrombosis extending to the main intra-hepatic portal branches (Figs. 1A–C). Moreover the following findings were observed: enhancement of vasa vasorum of the portal walls, porto-systemic shunts characterized by vicariant activation of the capsular venous branches with an enlarged right hepatic vein (Fig. 2A), splenomegaly with longitudinal diameter of 15 cm (Fig. 1B), pancreatic tail structural changes characterized by poor glandular representation and large intra and extra-glandular fluid collections

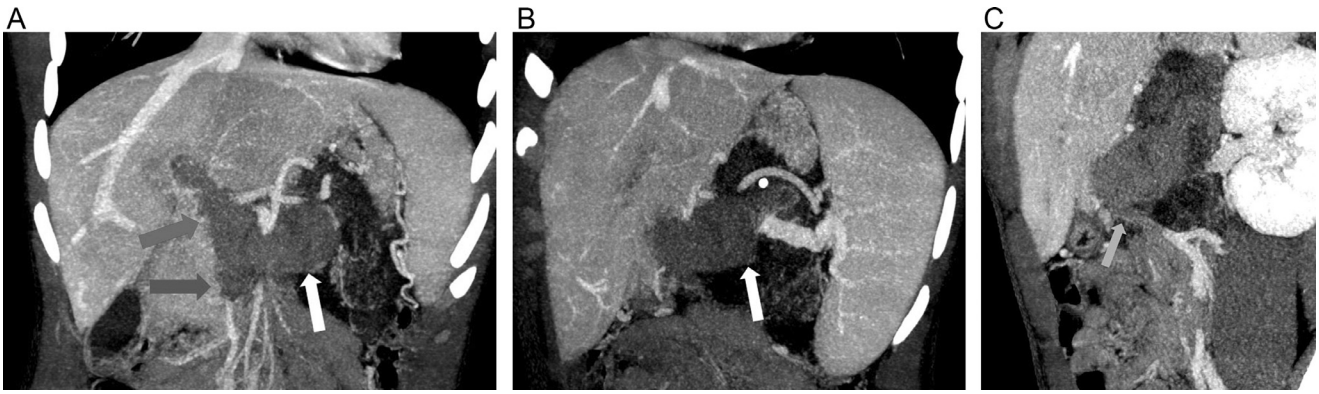

Fig. 1 – (A-C) Contrast-enhanced portal phase CT scan: The coronal MIP images depicts a large venous aneurysm with a superimposed massive thrombosis involving the portal vein (*red arrow*), the superior mesenteric vein (*blue arrow*), the splenic vein (*white arrow*), and the inferior mesenteric vein (*green arrow*). Enlarged spleen is also observed (Color version of the figure is available online.)

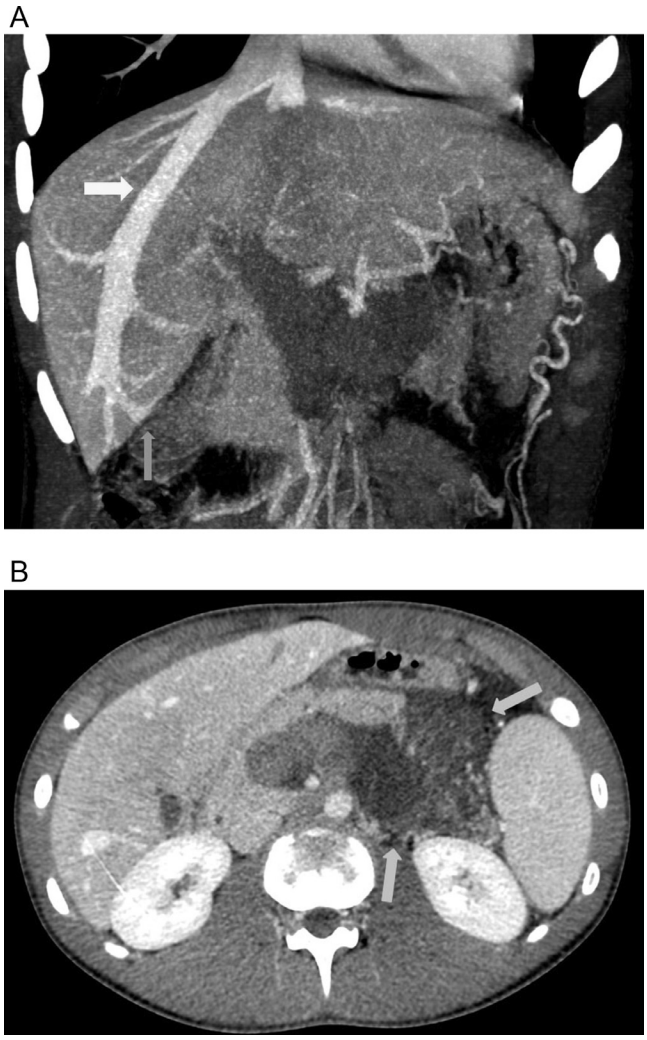

Fig. 2 – (A and B) Contrast-enhanced portal phase CT scan: The coronal oblique MIP (a) and the axial (b) images depicts: (A) vicariant activation of the capsular venous branches (*blue arrow*) with an enlarged right hepatic vein (*yellow arrow*); (B) large intra and extra-glandular fluid collections at pancreatic tail level (*orange arrows*) (Color version of the figure is available online.)

(Fig 2B). As a result, a therapy with low-molecular-weight heparin (LMWH) was adopted. Nine days later, the follow up ce-CT confirmed the previous findings except for partial peripheral re-habitation of spleno-portal thrombosis and the activation of peri-portal cavernomatosis.

During the hospitalization, the physicians excluded the presence of previous causes of acquired portal venous aneurysm including acute, and chronic pancreatitis on the base of the unremarkable clinical history, as well as, of thrombophilic diseases on the base of an unremarkable panel for genetic, immunologic, and serum coagulation disorders. As a consequence, the hypothesis of congenital aneurysm complicated by superimposed thrombosis was proposed; the pancreatic findings were interpreted as secondary to portal venous chronic pathology rather than the cause.

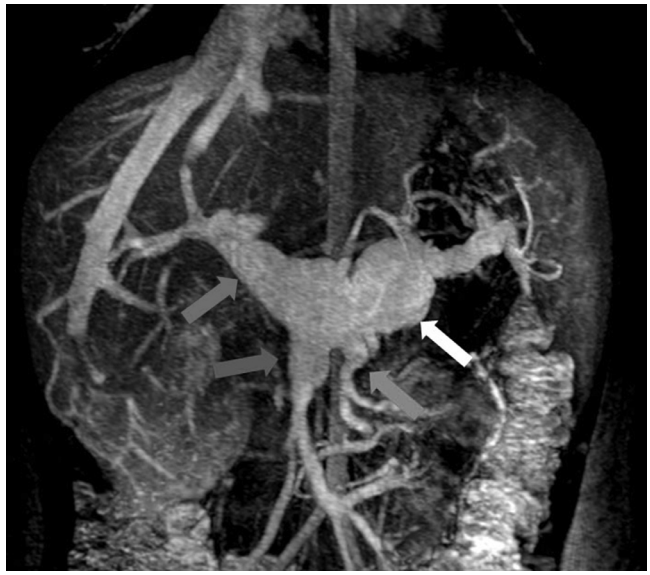

Fig. 3 – Contrast enhanced portal phase MRI scan. The coronal MIP image shows a complete recanalization of the portal venous system with persistent aneurysmal dilatation of portal vein (*red arrow*) and of the distal segment of superior mesenteric vein (*blue arrow*), inferior mesenteric vein (*green arrow*), and splenic vein (*white arrow*) (Color version of the figure is available online.)

Fifteen days later the patient was discharged recommending anticoagulant home therapy. After 3 months, the patient underwent a contrast enhanced MRI of the abdomen, which disclosed a complete recanalization of the porto-spleno-mesenteric aneurysm (Fig. 3). Subsequently, after 6 months, the anticoagulant therapy was suspended. Unfortunately, 25 days later, the patient returned to emergency room for the onset of intense abdominal pain. The ce-CT documented the extensive thrombotic recurrence of the porto-spleno-mesenteric aneurysm. The pancreatic findings were stable respect to the previous CT scans. Currently, the patient is asymptomatic under chronic therapy with new oral anticoagulants (NOACs), and undergoes periodic clinical follow-up.

Discussion

Venous aneurysms are vascular malformations rarely observed in the clinical practice, more frequently localized in the neck, and lower limbs [14]; among the visceral venous aneurysms, the portal venous system represents the most frequent affected site [12,13,15]. Portal vein aneurysm was first described by Barzilai and Kleckner in 1956 and since then less than 200 cases have been reported in English literature. [16]. *Machado* et al. reported that 76.9% of visceral venous aneurysms are located in the extrahepatic area [17]. Specifically, another study reported that the 57% of extrahepatic portal venous aneurysms involves the porto-spleno-mesenteric confluence, the main portal vein, and the portal bifurcation in descending order [18]. Furthermore, it seems to be a

relationship between the position and the size of these aneurysms, the largest being more frequently found outside the liver, as their growth is not hindered by the surrounding hepatic parenchyma [17].

Our case is peculiar because it is the first to report the involvement not only of the confluence but also of all the components of portal system. In our patient, the CT examination revealed the presence of a huge aneurysm at the spleno-porto-mesenteric confluence involving the portal trunk, the SMV, the IMV, and the splenic vein. Moreover a massive superimposed thrombosis of the aneurysm with complete obliteration of the lumen extending to the main intrahepatic portal branches was observed. This thrombotic condition was considered acute, because of the clinical setting, the absence of portal cavernomatosis, and the presence of "vasa vasorum" enhancement on the first CT examination [19,20]. On the other hand, the presence of porto-systemic shunts characterized by vicariant activation of the capsular venous branches with an enlarged right hepatic vein and the splenomegaly suggested a portal hypertension condition pre-existing to the thrombosis [21].

The lack in the clinical history of the main triggering causes of acquired aneurysm, ie liver cirrhosis, intrahepatic and post-hepatic portal hypertension non–cirrhotic causes, surgical interventions, abdominal trauma or abdominal inflammatory processes, as well as, the absence of a coagulopathy diseases, suggested the final diagnosis of congenital aneurysm of portal system complicated by thrombosis with associated pre-existing chronic condition of pre-hepatic portal hypertension. Moreover the stability of the venous aneurysm over time, as shown at the follow up imaging, supported successively the diagnosis. Furthermore, the pancreatic findings were considered secondary to previous misunderstood vascular insults related to the venous portal aneurysm [22–23]. This hypothesis was supported by the following considerations: 1) the segment of splenic vein adjacent to the pancreatic tail showed signs of hypertension but no aneurysmal dilatation; 2) the slowing of blood flow due to the increase in portal system luminal caliber has probably led to chronic unrecognized venous pancreatic ischemia, worsened by recurrent thrombo-embolic episodes [24]. However, the hypothesis of inflammatory portal wall involvement caused by recurrent mild idiopathic pancreatitis cannot be denied.

In conclusion, portal aneurysm is a rare condition that can lead to severe complication as thrombosis or rupture, therefore it's crucial for the radiologist to know its main imaging findings, and clinical features, to improve the patient's outcome. Although exceptional, the hypothesis of a congenital form should be assessed, both with a thorough review of the clinical history of the patient, and a complete screening panel for other possible causes for aneurysm or thrombosis.

Patient consent

Informed consent was obtained by the patient for publication of this case.

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