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Case Report

Radiological evaluation of extrahepatic and intrahepatic portal vein aneurysms: A report of two cases ☆

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ABSTRACT

Portal vein aneurysm (PVA) is a rare vascular entity. Here, we describe cases of 2 separate patients who presented with congenital and acquired causes of PVA respectively. The first patient presented with vague abdominal pain and was incidentally diagnosed with PVA, whereas the cause in the second patient was iatrogenic. With a limited number of cases published to date, there is little data on the natural history of the disease. Herein, we will discuss the radiological findings aiding us in reaching our diagnosis and also the probable mimickers of the disease, with a brief overview of its possible causes, complications, and the currently available management options.

Introduction

PVA refers to the aneurysmal or variceal dilatation of the portal vein [1]. Less than 200 cases have been reported so far worldwide [2]. However, the frequent use of radiological investigations is probably the cause behind increasing case reports in recent years. PVAs can be entirely asymptomatic and only discovered incidentally, or can be symptomatic due to their complications or mass effect over the neighboring viscera. They can have both congenital and acquired causes and can present at different locations within the portal venous system. Herein, with the help of 2 cases we will discuss the radiological findings, causes, possible complications and brief overview about the treatment of this entity.

Case 1

A 60-year-old female patient presented with vague epigastric pain and dyspepsia for a week. She had no history of any liver disease. Her abdomen was soft and nontender. No signs of

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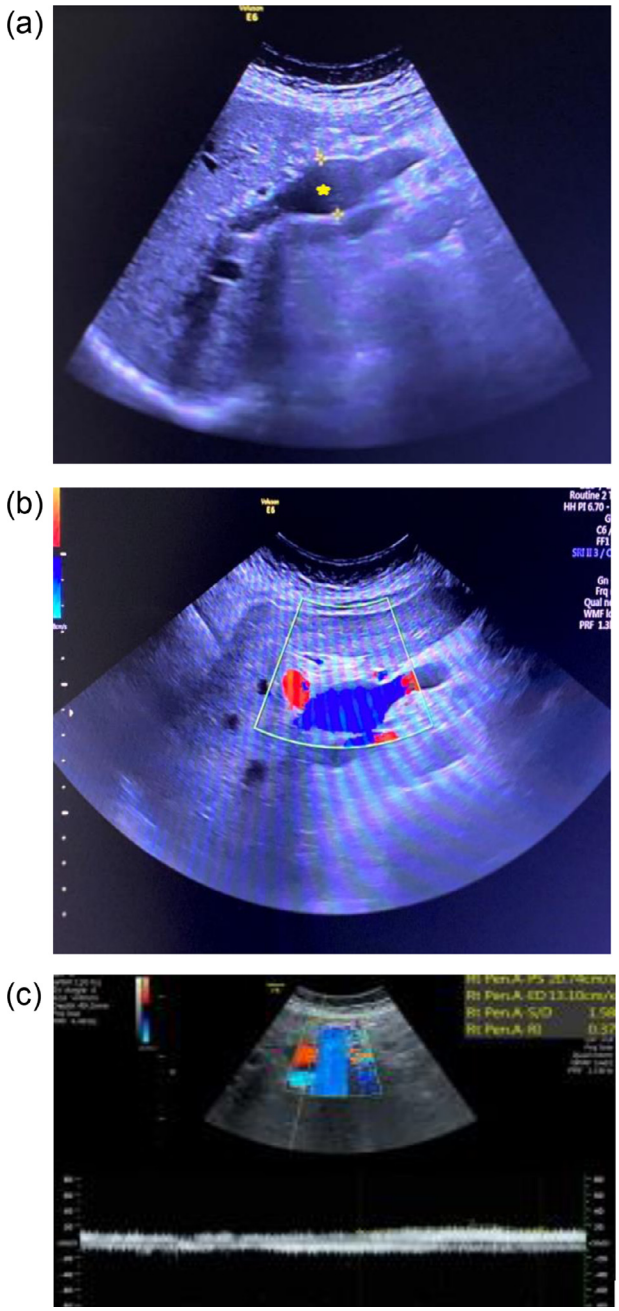

Fig. 1 – (a) Case 1- Ultrasound abdomen, depicting a focal dilatation of portal vein, measuring 2.5 cm (yellow star). (b) Case 1- Blood flow within the PVA on color Doppler. (c) Case 1- Venous waveform of PVA.

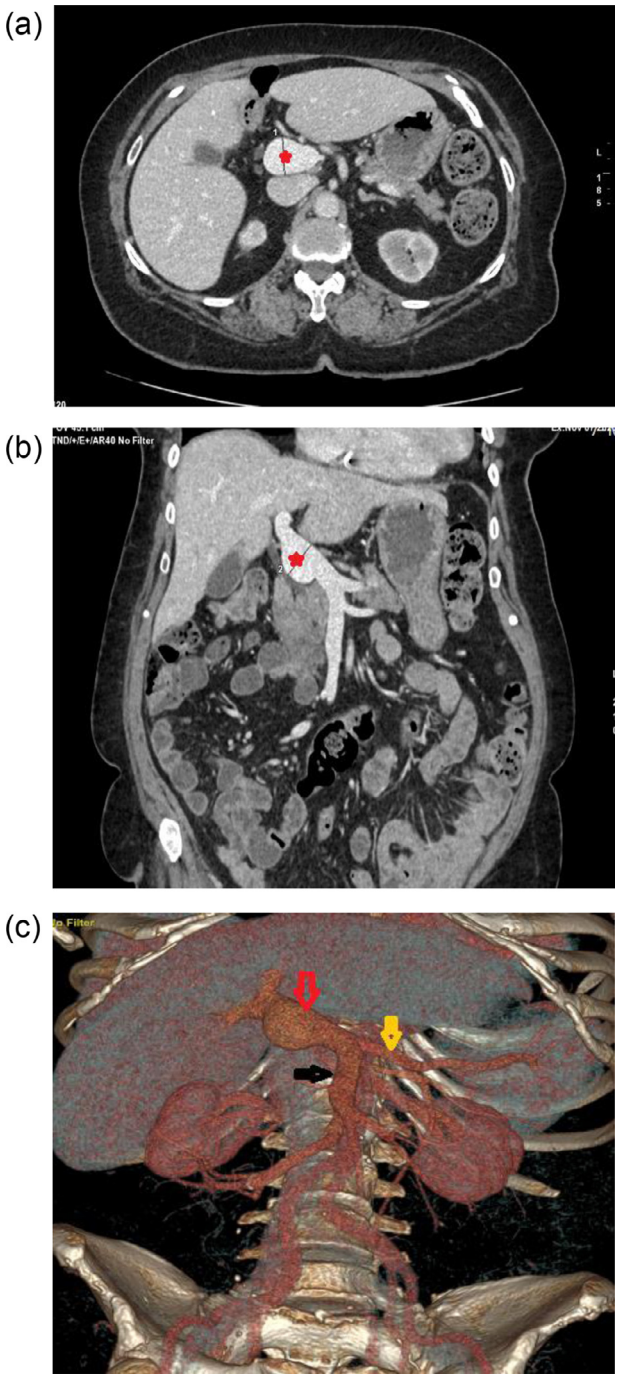

Fig. 2 – (a) Case 1-Portal venous phase CT abdomen (axial section) depicting an extrahepatic portal venous aneurysm (red star). (b) Case 1- Portal venous phase CT abdomen (coronal section) depicting an extrahepatic portal venous aneurysm (red star). (c) Case 1–3 Dimensional VR CT image depicting the formation of portal vein by combination of superior mesentric vein (black arrow) and splenic vein (yellow arrow). PVA (red arrow).

portal hypertension were present. She was a known diabetic for 8 years and was on oral hypoglycemic agents.

Her complete blood count, pancreatic and liver function tests were normal, and she was subsequently advised for radiological investigations.

Her abdominal Ultrasound (USG) revealed an anechoic lesion measuring 2.5 cm in anteroposterior diameter, appearing in continuation with the main portal vein. Color Doppler showed a monophasic, non-pulsatile venous flow pattern

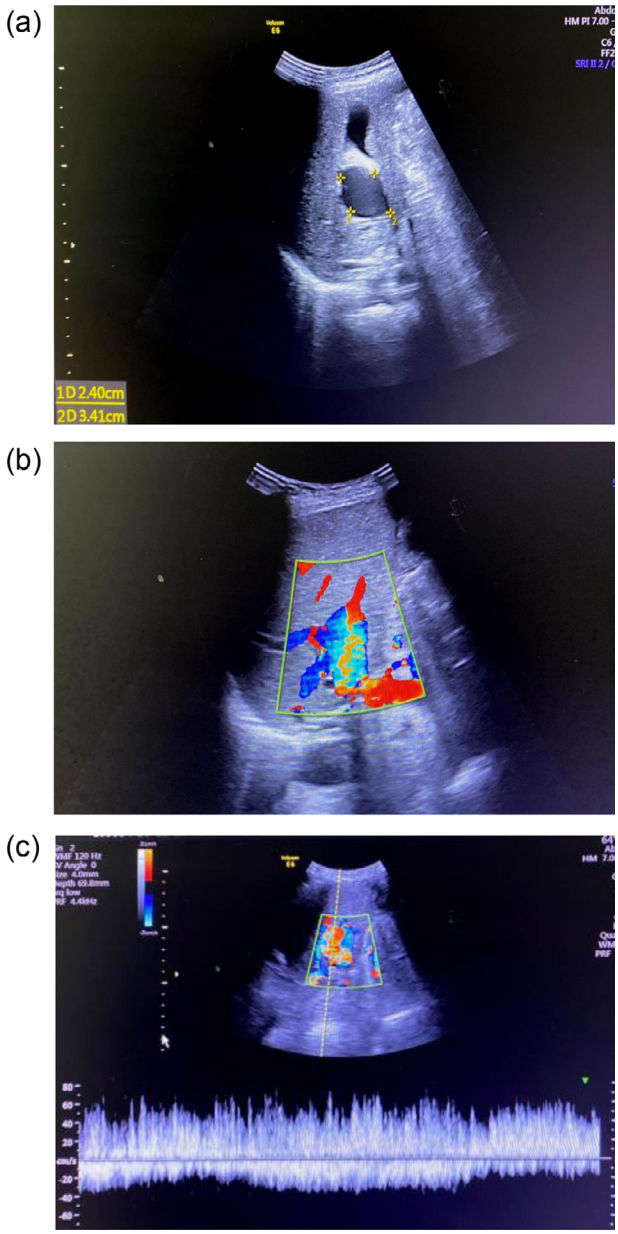

Fig. 3 – (a) Case 2- Ultrasound abdomen, depectic a focal anechoic intrahepatic cystic mass. (b) Case 2- Blood flow within the cystic mass and its branches. (c) Case 2- Monophasic venous waveform of PVA.

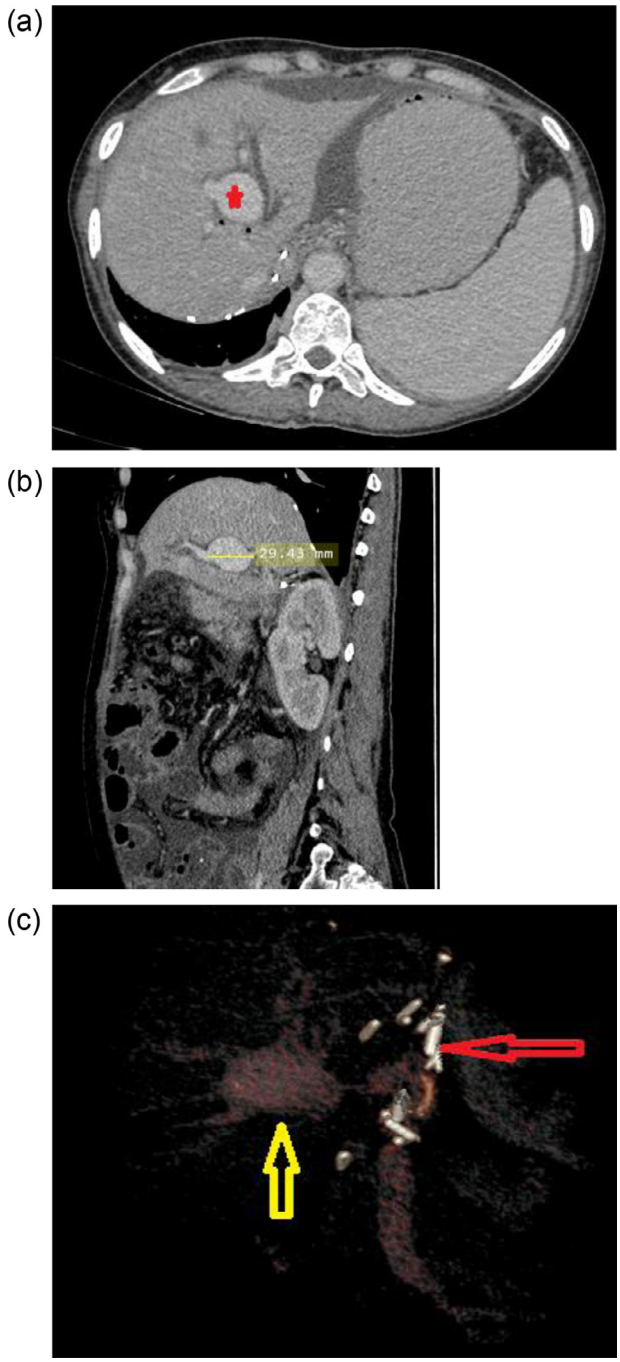

Fig. 4 – (a) Case 2- Portal venous phase CT abdomen (axial section) depicting an intrahepatic portal venous aneurysm (red star). (b) Case 2- Portal venous phase CT abdomen (Sagittal section) depicting an intrahepatic portal venous aneurysm. (c) Case 2–3 Dimensional VR CT image depicting PVA (yellow arrow) and surgical clips (red arrow).

within this lesion and a diagnosis of PVA was made. There was no evidence of any intraluminal thrombosis (Fig. 1). Her other abdominal viscera were grossly normal.

After 1 week of following the out-patient treatment, her pain did not settle and she was subsequently advised for a Triple Phase CT scan (TPCT) of the abdomen firstly, to explore any other potential causes for her symptoms and second, to determine accurate location of the aneurysm, the dimensions and relations with adjacent organs.

The portal venous phase showed a focal, well defined, smoothly marginated, homogenously enhancing mass near the hilum of the liver, appearing in continuation with portal vein and showing contrast enhancement on venous phase similar to that of the portal vein, compatible with an extrahepatic portal vein aneurysm (Fig. 2a and b). There was no evidence of any intraluminal thrombus. No evidence of any liver cirrhosis or portal hypertension was noted.

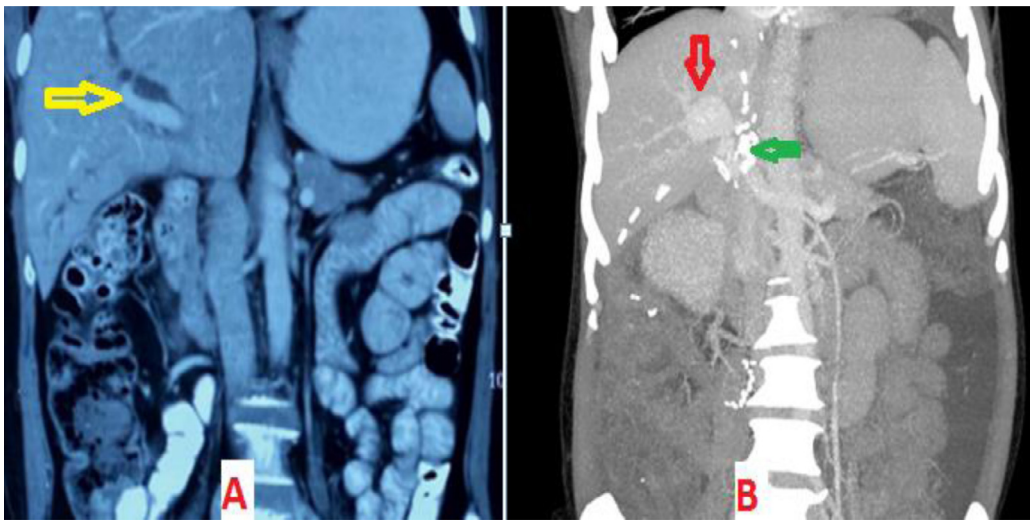

Fig. 5 – Case 2- Comparison of the portal vein status before and after surgery. IMAGE A (Before surgery)- CT abdomen (sagittal section) depicting a normal portal vein (yellow arrow). IMAGE B (postsurgery)- CT abdomen (sagittal MIP section) depicted a portal vein aneurysm (red arrow) and the surgical clips (green arrow).

No other radiological investigations were done for the patient in the past, to help compare the current findings. The patient being asymptomatic was managed conservatively and advised for an annual follow up with USG.

Case 2

A 58-year-old female patient presented with abdominal discomfort. She was an operated case of cholangiocarcinoma (2 years back) and was currently receiving chemotherapy. She had no signs and symptoms of portal hypertension.

Her USG abdomen revealed an intrahepatic, anechoic lesion of size 2.4 ×3.4 cm near the porta, in close proximity to the gall bladder. Few curvilinear anechoic branches were also seen radiating out from the lesion, representing the various vessel branches. Color Doppler showed a venous flow (monophasic waveform) pattern within the lesion. A diagnosis of intrahepatic PVA was made (Fig. 3).

A TPCT was done for her, which depicted a well-defined, intrahepatic lesion. Numerous branches were seen radiating out from the lesion like a pinwheel. The lesion showed enhancement patterns similar to portal vein and a diagnosis of intrahepatic PVA was confirmed (Fig. 4).

On taking a detailed history, her scans prior to the surgery (done in another hospital) were studied, which showed a normal portal vein. Thus, the cause of PVA was attributed to the surgical procedure performed for the patient, in which some portion of the portal vein had been clipped, leading to pressure disturbances within the vessel, eventually causing its dilatation (Fig. 5).

On comparison with postsurgery scans, performed 1 year back, the PVA in the current scan showed an increase in diameter (1.4 cm in the previous scan, 2.4 cm in the current scan). Rest of the findings were grossly the same.

The patient was advised for a 6 monthly close follow-up, as the lesion showed an increase in size, and thus carried a greater risk.

Discussion

PVA represents only 3% of all the venous aneurysms [3] and its etiology is yet to be fully explored. We have come a far way in our diagnostic expertise since the first case of PVA was diagnosed on an autopsy in 1956, after it had ruptured [4]. We are now able to diagnose this entity by radiological investigations, often before it can complicate.

Vascular aneurysms are mainly found in arteries. However, the literature suggests their appearances in certain veins as well, such as popliteal, saphenous, and jugular. Portal vein aneurysms are relatively rare and mainly found in patients with liver cirrhosis or portal hypertension. Although rare, PVA is the most common visceral venous aneurysm.

The portal vein diameter varies from individual to individual depending on age as well as ethnicity. In an ultrasonographic study of the portal vein, Doust and Pearce found that the maximum anterior-posterior diameter never exceeded 1.5 cm in normal patients, and 1.9 cm in cirrhotic patients. The diagnosis of extrahepatic PVA is made when the diameter exceeds 20 mm [5]. And for intrahepatic PVA, a diameter of more than 7 mm in normal subjects and 8.5 mm in cirrhotic patients constitutes the disease [6].

The most common location of PVA is in the extrahepatic portal vein at the confluence of superior mesenteric vein and splenic vein [7]. The other sites are the main portal vein, intrahepatic portal vein, and its branches. The rarest locations are the splenic, mesenteric, and umbilical veins.

Broadly we can divide the causes of PVA into congenital and acquired. The portal venous system develops from

vitelline and umbilical veins. A failure in the regression of the right primitive vitelline vein during embryological development and saccular enlargement of this diverticular remnant may lead to PVA later in life. Other authors describe the development of PVA to an inherent weakness of the vein wall [8]. Case reports of in-utero PVA and its presence in children supports the congenital theory.

Portal hypertension secondary to liver cirrhosis is the foremost acquired cause of PVA. Other causes include pancreatitis, trauma, invasive malignancy and abdominal surgery. Portal hypertension can be both a cause and a complication of PVA [5].

In our first case, the patient did not present any risk factor: no underlying liver disease, no history of pancreatitis, trauma, and surgery. These elements support the congenital cause. In the second patient the cause was acquired (iatrogenic), due to partial clipping of the portal vein (evident by comparison with preoperative scans, which showed a normal portal vein).

Most patients present with vague abdominal pain or are asymptomatic. Its complication can, however, include thrombosis, rupture, and symptoms from pressure on adjacent structures, like the common bile duct leading to jaundice, compression of duodenum leading to obstruction, compression of IVC, gastrointestinal bleed are some of the other presenting symptoms. Acute thrombosis of the portal vein aneurysm can result in severe life-threatening portal hypertension [9].

The diagnosis of PVA in the majority of cases can be made on USG, which is a safe and non-expensive radiological investigation. On B mode scans it appears as an anechoic cystic lesion, in the region of the portal vein. The diagnosis becomes relatively difficult in case of an intra-hepatic PVA (like in our second case), as sometimes we cannot trace the continuation of this cystic lesion with the portal vein, and tend to misdiagnose it as a hepatic cyst. Color Doppler comes to rescue in such cases, where demonstration of blood flow can be made with a non-pulsatile monophasic waveform. Contrast enhanced computed tomography or magnetic resonance angiography can be helpful in patients with equivocal ultrasonographic findings or when surgical intervention is planned.

A focal anechoic (on USG), homogenously enhancing (on CT) mass near the hilum or within the liver might mimic certain other conditions such as a pancreatic or hepatic cyst, duodenal duplication cyst, or a choledochal cyst, which is why Doppler is necessary for demonstrating a non-pulsatile monophasic waveform in the setting of PVA. Hepatic artery aneurysm can be another closely related differential, especially for the fact that it will show color flow on Doppler, but triphasic spectral waveform will help differentiate the two. If thrombosed, the PVA might mimic a solid mass, hypervascular metastasis or a pancreatic mass. Contrast enhanced CT/MRI will be the investigation of choice in such a case.

Due to the rarity of the lesion, the management options often remain controversial. Asymptomatic and small-sized PVAs are often managed conservatively with regular interval follow up. In a case report presented by Shazia A. Rafiq and Michael D. Sitrin, the PVA remained relatively stable and asymptomatic for almost 10 years [9]. Surgical interventions remain the mainstay treatment option, mainly reserved for patients who are either symptomatic or carry a risk of thrombosis or rupture, and range from shunt procedures, aneurysmectomy, aneurysmography to liver transplantations depending on the severity of the illness and also the presence or absence of portal hypertension [10].

The type of procedure is based on the location and size of the aneurysm, co-morbidities and complications. Moreno et al. suggested surgical treatment for patients presenting with a non-thrombotic PVA >3 cm [11].

Aneurysmorraphy is preferred for excision of the aneurysm, mainly when it is saccular. Whereas, in cases of fusiform aneurysms, an aneurysmectomy is performed and the conduit used to replace the portal vein can be an allograft from a cadaveric donor, or a synthetic graft. Both these procedures are considered in patients with a normal liver, that is no cirrhosis/portal hypertension [12]. The presence of portal hypertension associated with chronic liver disease changes the surgical approach. Surgical shunt procedures, with or without splenectomy are the procedures of choice, aimed at decompressing the portal venous system instead of treating the aneurysm itself, to prevent progressive dilatation of the aneurysm [10].

Liver transplantation is reserved for patients with end-stage liver disease and the presence of a PVA at transplantation may require technical innovations [12].

To conclude, PVA is a rare vascular entity, occurring mainly in patients with portal hypertension and liver cirrhosis. It can, however, be congenital. In majority, it remains asymptomatic, but can nonetheless produce life threatening complications such as thrombosis and rupture. Like in our second patient, who depicted an increase in the size of the aneurysm, careful monitoring is inescapable. USG is often the first investigation of choice, with color Doppler offering an excellent diagnostic approach. Management options mainly depend on the patient's symptoms and co-morbidities. With limited cases published so far, there is a need for extensive research in terms of causes and management of this rare entity.

Patient consent

A written, informed consent was taken from the patients, for publication of their case.

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