

Portal Vein Aneurysm Complicated by Thrombosis

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Abstract

The current study described a case of portal vein aneurysm at the confluence of the superior mesenteric and splenic veins, complicated by thrombosis in a patient with no liver pathology but with factor V Leiden mutation.

Keywords

Portal vein aneurysm; Thrombosis; Factor V Leiden; Superior mesenteric vein

Introduction

Portal vein aneurysm (PVA) was first described by Barzilai and Kleckner Jr^[1] in 1956 and since then only 190 cases have been reported in the literature^[2-4]. Portal vein aneurysm accounts for 3%^[3-7] of all visceral aneurysms, with an incidence of 0.06%^[5,7], which has increased to 0.43% in recent years^[3,4,6,8] with improved imaging procedures and incidental findings. Portal vein aneurysms usually remain stable in size and do not progress^[6], 63% of them being extrahepatic. Portal vein aneurysms of > 5 cm are extremely rare^[9]. We report here the 191st case of PVA.

CASE REPORT

A 43-year-old female presented to the emergency department on 20 May 2019 reporting sharp crampy epigastric pain for five days associated with constipation and loss of appetite for three days that was relieved by self-induced vomiting. Her medical and surgical history was not significant. She was married with no children and no history of abortions. She was not a smoker and had no history of allergies.

On examination, she was hemodynamically stable, with a temperature of 37°C, heart rate of 80 beats

per minute, and blood pressure of 132/70. She was alert, oriented, and not in distress. Examination of her abdomen showed epigastric fullness with tenderness but otherwise normal findings. Blood work results (complete blood count levels; urea and electrolyte levels; liver function tests; coagulation profile; and amylase, lipase, and lactic acid levels) were all normal, as were the results of an abdominal X-ray.

A computed tomography (CT) scan of the abdomen and pelvis showed a large, homogenous, non-enhancing, well-defined soft tissue density lesion of 8.5 x 6 x 6.5 cm in the porta hepatis with few surrounding collaterals that was inseparable from the pancreas. The lesion was causing thrombosis of the portal vein and its intrahepatic branches, the superior mesenteric vein (SMV) and the right gonadal vein, as well as superior displacement of the caudate lobe of the liver with downward displacement of the duodenum (Fig. 1 and 2).

The patient was admitted to our hospital, given nothing by mouth and kept hydrated, and referred to the vascular, hematology, and gastroenterology teams for endoscopic ultrasound evaluation of a suspected soft tissue lesion. She was initiated on enoxaparin sodium (Clexane) 60 mg twice daily.

Magnetic resonance cholangiopancreatography was performed on May 22, 2019, which showed a 5.6-cm PVA with thrombosis. The thrombus involved the SMV and the splenic vein at the portal venous confluence (Fig. 3).

Screening was done for thrombophilia and antiphospholipid antibodies, and the vascular team was contacted for management, but no further intervention was required. The patient was prescribed therapeutic enoxaparin sodium and discharged home with a follow-up appointment with both the surgical and hematology teams.

The results of her thrombophilia screening turned out to be positive for heterozygous factor V Leiden mutation with a slight elevation in the lupus anticoagulant results (55.7, normal range 0–45). The rheumatology team was consulted but no active treatment was given, and she was discharged. She is still being followed in our clinic and with the hematology team and is doing much better clinically. Her prescription was shifted to warfarin for a follow-up Doppler ultrasound (US) examination this month.

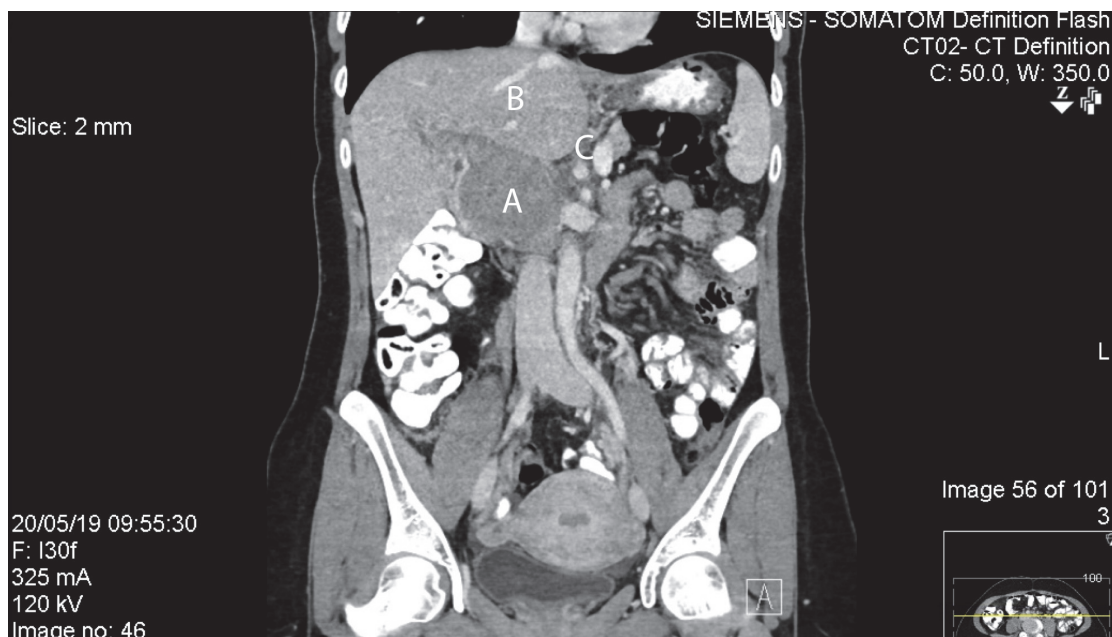


Figure 1. Coronal cut of a computed tomography scan image showing the portal vein aneurysm (A) displacing the liver superiorly (B) and the duodenum downward (C).

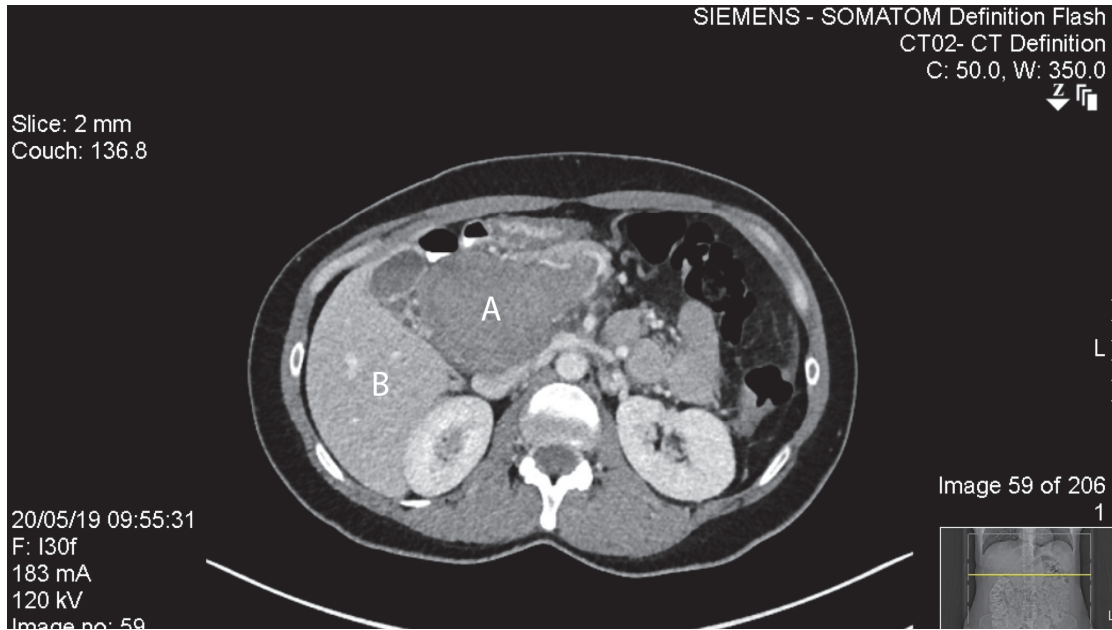


Figure 2. Axial (horizontal) cut of a computed tomography scan image showing the aneurysm (A) pressing on the liver (B).

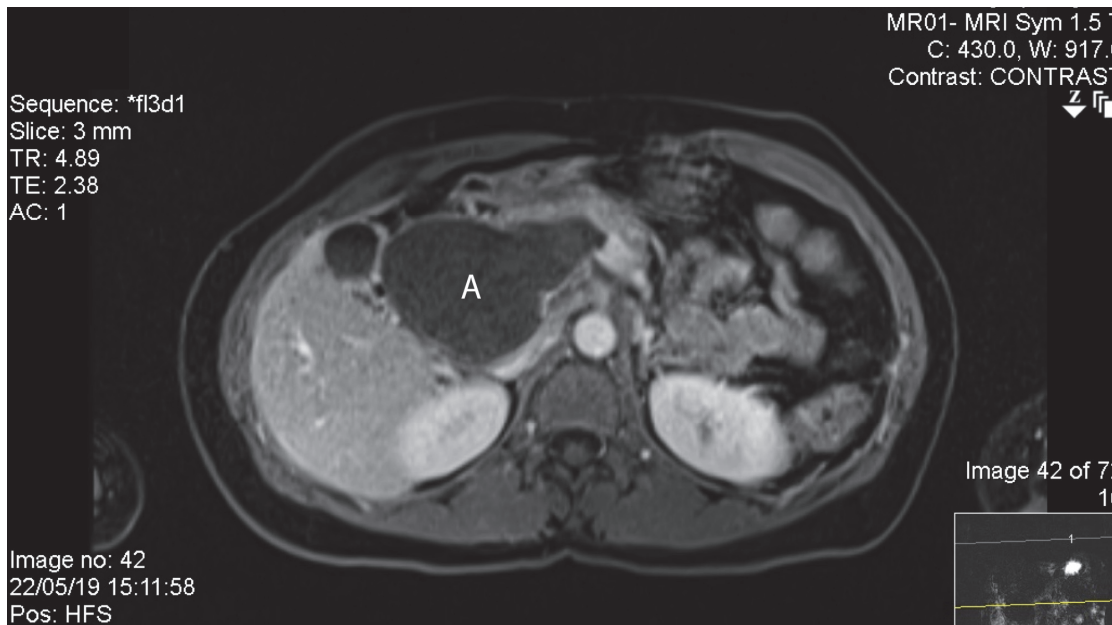


Figure 3. Magnetic resonance image showing the aneurysm with thrombosis (A).

Discussion

Portal vein aneurysms are defined as having a portal vein diameter of > 1.9 cm in patients with cirrhosis and > 1.5 cm in healthy patients, or a portal vein extrahepatic diameter of > 2 cm and an intrahepatic diameter of > 0.9 cm^[3-6,9]. Our patient had a portal vein diameter of 5.6 cm.

Portal vein aneurysms may be congenital or acquired. Congenital causes include either incomplete regression of the right primitive distal vitelline vein or congenital wall defects. Of the 190 patients with PVA reported to date, 25 have been reported to have cutis laxa, or elastolysis, a rare inherited or acquired connective-tissue disorder in which the skin becomes inelastic and hangs loosely in folds. The most common cause of acquired PVA is portal hypertension, the result of a combination of hyperdynamic blood flow and increased portal pressure that induces thickening of the vessel intima, leading to compensatory medial hypertrophy of the vessel. The thickened intima is gradually replaced by fibrous connective tissue that compromises the structural integrity of the venous wall, leading to a reduction in tensile strength and increasing the susceptibility to aneurysmal dilation^[4,9]. Other causes include pancreatitis, trauma, and invasion by malignancy^[2,5,6,9].

Extrahepatic PVAs are more common than intrahepatic PVAs: Of the 190 reported cases of PVA, three were found in the right portal branch, one in the left portal branch, and 19 in the umbilical portion of the left portal vein^[6]. Most cases (72%)^[8] were asymptomatic, but in other cases, the most common symptom was abdominal pain. Gastrointestinal bleeding occurred in 7.3%^[6] of cases, as well as symptoms related to compression of adjacent structures (abdominal distension, jaundice, etc.)^[2,4,6,9] (Table 1).

Table 1. Overview of the data on portal vein aneurysm found in the literature^[3,5,9]

Characteristic	Value
Number of patients	≈ 190
Median age, years	53
Male:female ratio	1:1
Portal hypertension	62 (32%)
Aneurysm localization:	
Main portal trunk	73 (38.4%)
Splenoenteric confluence	45 (23.6%)
Portal bifurcation	72 (38%)

PVA: portal vein aneurysm

The differential diagnosis of PVA includes abdominal soft tissue tumors and liver or pancreatic cysts^[9]. Diagnosis is usually established by Doppler US but a CT or magnetic resonance imaging scan can be done as well. Complications include portal vein thrombosis, which occurred in 20% of cases; compression (duodenal, biliary tract, and inferior vena cava), which occurred in < 10% of cases; and spontaneous rupture, which was reported in four cases^[2,4,6].

The surgical guidelines for management of PVAs are unclear^[6], but treatment is conservative; uncomplicated asymptomatic PVAs of even up to 6 cm can be treated conservatively with long-term surveillance (Doppler US every six months for 18 months). Indications for surgical management include having symptoms or complications (although some cases were successfully treated conservatively, as was the case with our patient) and presenting with an expanding aneurysm^[2,6,9].

Asymptomatic patients are treated conservatively with 6 months of US follow-up for 18 months, whereas symptomatic patients are treated according to whether or not they have portal hypertension. PVAs in patients without portal hypertension are treated with aneurysmorrhaphy or aneurysmectomy, whereas PVAs in patients with portal hypertension are treated with shunt procedures (splenoportal/portocaval shunts) and liver transplantation, which occurred in two patients with end-stage liver disease^[3,5,9]. Among surgically managed cases, the overall postoperative mortality rate was 17.5%. Percutaneous thrombolysis or thrombectomy was also used in cases of widespread thrombus involving the splenic vein and the SMV and unsuccessful anticoagulation^[3]. Spontaneous regression was reported in two cases with cavernous transformation of the main portal vein^[3,5].

Conclusion

Portal vein aneurysm is an uncommon condition that is typically asymptomatic and often discovered incidentally during imaging for other reasons. Thrombosis is just one of the known complications of PVA, which was more prominent in our patient due to the presence of Factor V Leiden mutation, which in itself leads to more thrombus formation. The management of such a condition is usually conservative.

Conflict of Interest

The authors have no conflict of interest.

Disclosure

The authors did not receive any type of commercial support either in forms of compensation or financial for this study. The authors have no financial interest in any of the products or devices, or drugs mentioned in this article.

Ethical Approval

The study design was reviewed and approved by the Unit of Biomedical Ethics Research Committee at King Abdulaziz University.

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المستخلص. في هذه المقالة نصف حالة تمدد الوعاء الوريدي البابي عند التقاء الوريد المساريقي والطحالي مع تخثر في الدم لمريضة تبين أن لديها خلل جيني يعرف بطفرة العامل الخامس ليدين.