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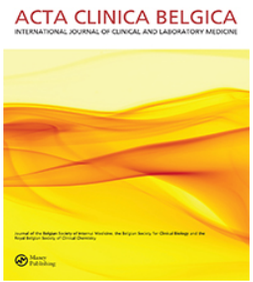

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CASE REPORT

Thrombosis of a portal vein aneurysm: a case report with literature review

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ABSTRACT

Objectives: Portal vein aneurysm is an unusual vascular dilatation of the portal vein. The etiology, diagnosis and management are ill-defined.

Methods: A case of a portal vein aneurysm complicated with complete thrombosis is presented with a literature review providing an overview of the etiology, clinical presentation and management.

Results: Portal venous aneurysms represent approximately 3% of all venous aneurysms with a reported prevalence of 0.06%. The reported incidence is on the rise with increasing use of modern imaging techniques in clinical practice. Usually, portal vein aneurysms are incidental findings and patients are asymptomatic. They can be congenital or acquired and portal hypertension represents the most frequent cause of the acquired version. Various complications such as biliary tract compression, portal vein thrombosis, and rupture can occur. Treatment options are conservative management or surgery. Surgical treatment is currently reserved for symptomatic patients with severe abdominal pain, symptoms of pressure effect or with expanding aneurysms, and/or complications such as thrombosis or rupture.

Conclusion: Conservative management seems the best option in the majority of patients. A multidisciplinary approach discussing the best option on a case-by-case base in light of their individual underlying risk and symptoms is advised.

KEYWORDS
Portal vein aneurysm; portal vein thrombosis

Introduction

Portal vein aneurysm (PVA) is an unusual vascular dilation of the portal vein. Since its first description by Barziali and Kleckner in 1956, less than 200 cases have been reported, mainly as case reports or small case series [1,2].

PVA is defined as a portal vein diameter exceeding 1.9 cm in cirrhotic patients and 1.5 cm in patients without chronic liver disease. For intrahepatic portal vein aneurysms, a size more than 9 mm and any vein significantly larger than the remaining segments of the same vein is considered as aneurysmatic [3].

In this manuscript, we describe a patient with thrombosis of a portal vein aneurysm and review the currently available literature to clarify various aspects of this rare entity and the available treatment options.

Case report

A 67-year-old male was admitted at the emergency department with acute onset abdominal pain, located in the right upper abdomen. His medical history included a metabolic syndrome with diabetes mellitus, obesity and arterial hypertension. The pain was constant, without features of a colic. There were no other concurrent symptoms such as nausea, diarrhea or fever. Clinical abdominal examination was uneventful. He showed no stigmata from chronic liver disease or vascular abnormalities. Biochemical work-up revealed an elevated CRP level (77 mg/L), with normal full blood count, kidney function, and liver function tests. Color – Doppler ultrasonography (US) however documented a hypoechoic mass of 5.2 cm next to the pancreas, extending into the hilum of the liver (Figure 1). Because of the US findings, a computed tomography (CT) scan was performed demonstrating a fusiform dilatation of the portal vein up to 5.5 cm at the level of the portal-splenic venous confluence, complicated with complete thrombosis of the portal vein, extending into to the splenic vein and the superior mesenteric vein (grade 4) (Figure 2). Furthermore a small indeterminate nodule in the corpus of the pancreas was depicted on the CT examination. There were no signs of cirrhosis or chronic pancreatitis. A liver biopsy was compatible with nodular regenerative hyperplasia. Upper endoscopy was negative for varices. Endoscopic ultrasound with fine needle aspiration of the small pancreatic nodule showed a low grade neuroendocrine tumor (NET) with Ki 0.67%. Further diagnostic tests excluded all known

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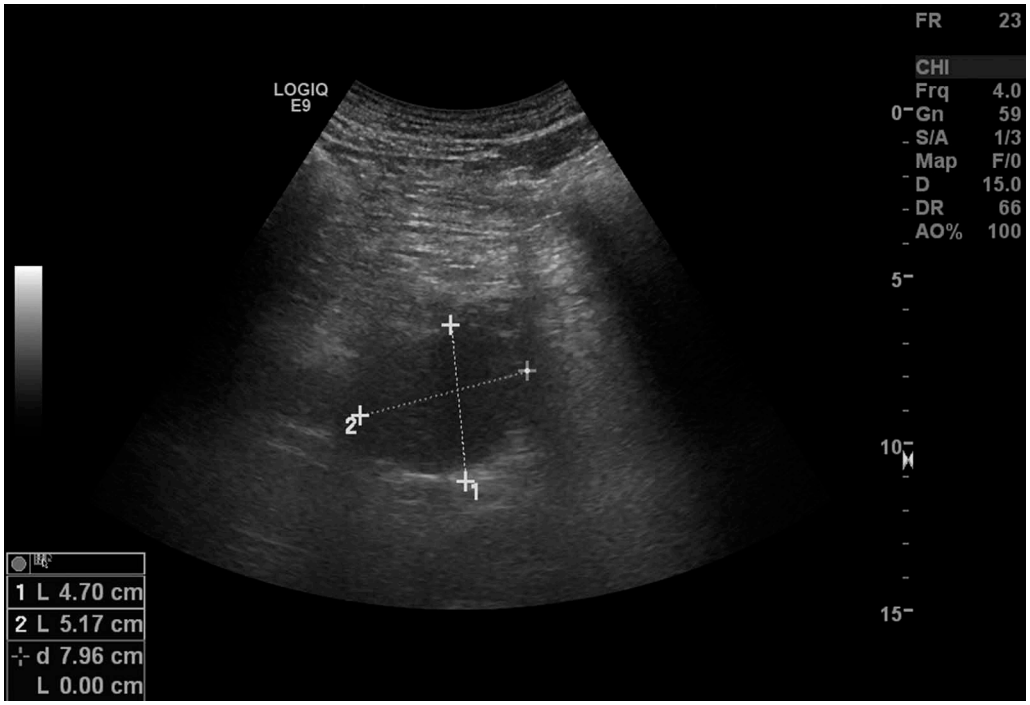

Figure 1. Transverse ultrasound image shows a hypo-echoic mass in the pancreatic region.

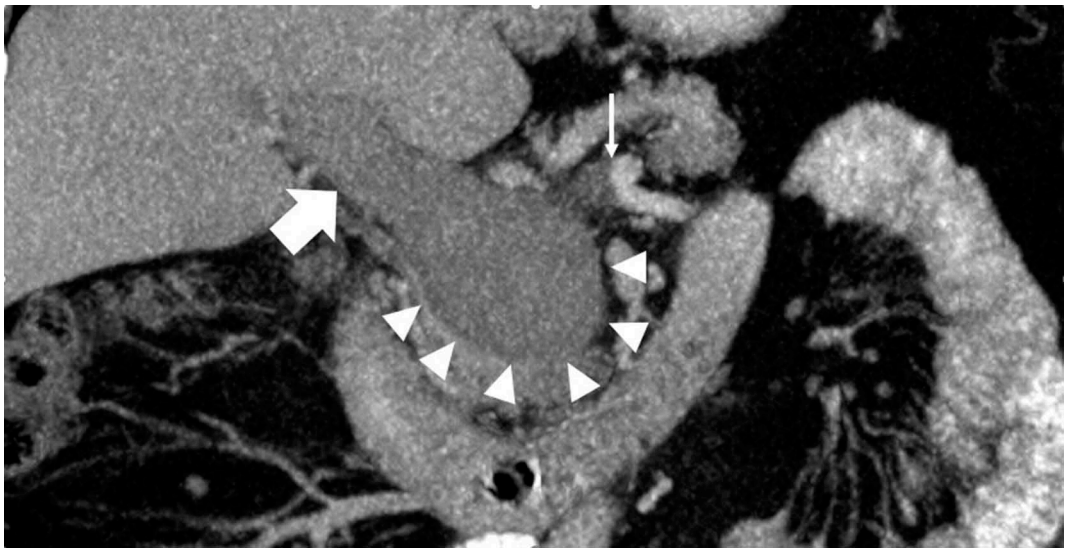

Figure 2. Contrast-enhanced CT image in the portal venous phase. Large thrombosed aneurysm of the splenomesenteric confluence (arrowheads) extending into the main portal trunk (thick arrow) and the splenic vein (thin arrow).

thrombophilic factors (including JAK2-mutation) and a bone marrow aspirate revealed no signs of myeloproliferative diseases. In our case these elements favors a congenital cause for the portal vein aneurysm, there were no risk factors such as an underlying liver disease, history of pancreatitis, trauma or abdominal surgery. Because of the aforementioned work-up and the acute onset of pain, we considered the thrombosis of the portal vein aneurysm as an acute event.

After multidisciplinary discussion conservative management for this thrombosed portal vein aneurysm was adopted and therapy with low molecular weight heparins was started, resulting rapidly in an asymptomatic patient with disappearance of pain. For the NET a conservative approach with watchful waiting was taken. Six months later, the patient was still without symptoms. Follow-up magnetic resonance imaging (MRI) after six month's showed a reduced diameter of the thrombosed confluence aneurysm up to 2 cm (Figure 3). Also, a cavernous transformation of the portal vein was seen, characterized by the formation of a network of dilated collateral veins along the chronic thrombosed and obliterated portal vein. The pancreatic NET remained stable.

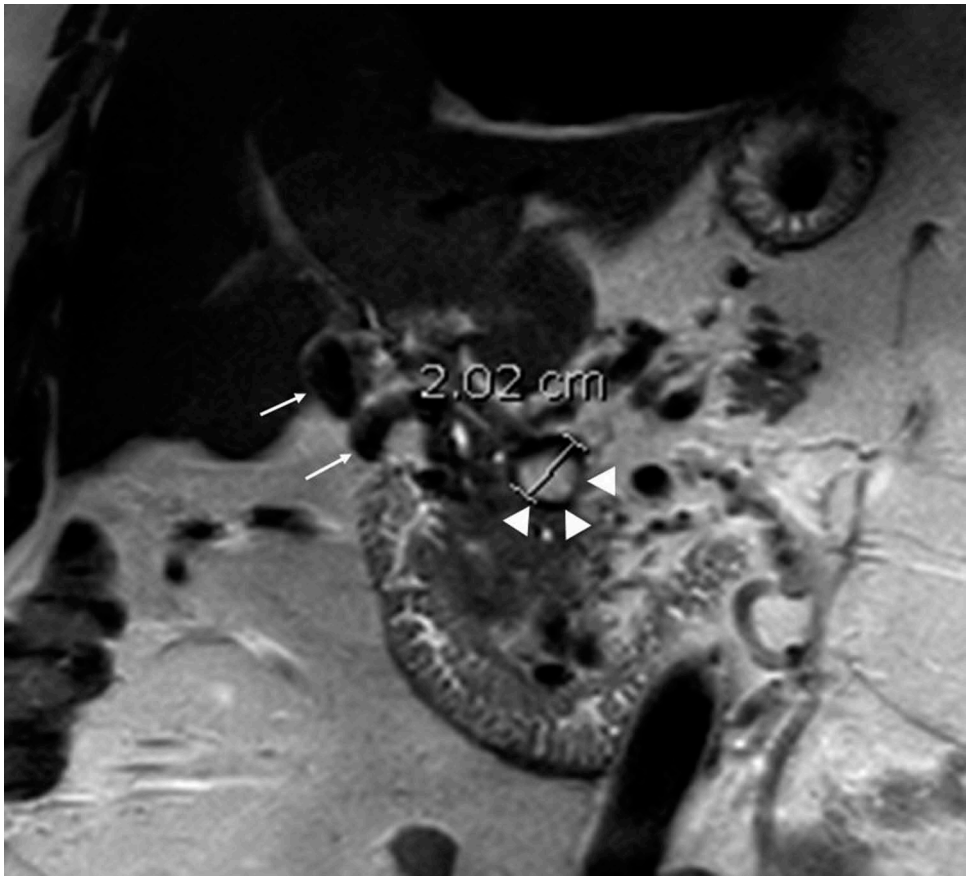

Figure 3. Coronal T2 weighted MR image (TE80 – TR995). Reduced diameter of the thrombosed confluence aneurysm to almost 2 cm (arrowheads), with T2 hyperintens lumen and T2 hypo-intens wall compatible with ageing blood. Portosystemic collaterals in the liver hilum (thin arrows). The pancreatic neuro-endocrine tumors are not depicted on this image.

Discussion

Etiology

PVA is a rare visceral venous aneurysm with an incidence of 0.06% and it represents less than 3% of all visceral aneurysms [1]. Nowadays, with the widened use of modern imaging modalities, the incidence of portal vein aneurysm is markedly on the rise. PVA can occur at any age without gender predilection [4].

The etiology is still unclear, however *congenital* or *acquired* causes have been suggested (Table 1).

More specifically, an incomplete regression of the right primitive distal vitelline vein has been proposed as the underlying mechanism for the *congenital* variant [4–6]. This hypothesis is supported by the in-utero diagnosis of portal vein aneurysms by US [7], as well as the relative frequent occurrence of this entity in children or healthy young adults [8]. In addition, the presence of PVAs in patients with histologically normal livers (without portal hypertension), as well as the frequent stability of such aneurysms over time, supports a congenital etiology [9].

Table 1. Potential etiology of portal vein aneurysm.

Congenital	Acquired
Incomplete regression of the right primitive distal vitelline vein	Portal hypertension = main cause
Presence of vein wall defects	Severe pancreatitis
	Abdominal trauma or surgery
	Invasion of portal vein by malignancy

The *acquired* form of PVA, is thought to be more common. Portal hypertension represents the main cause hereof. In these cases the portal vein dilatation is caused by the high splanchnic flow and hyperdynamic circulation associated with a consequential weakening of the venous wall following increasing shear stress [1,4]. Other acquired causes are necrotizing pancreatitis, abdominal trauma, surgery, or malignancy. Inflammation or local injury in these latter cases could weaken the vessel walls, leading to aneurysmal dilatation. Some PVAs have been described in association with hereditary hemorrhagic telangiectasia [10]. Acquired PVAs, especially when combined with liver disease and portal hypertension, have a more unpredictable course and warrant a closer follow-up and more aggressive intervention when complications occur [11].

Symptoms – complications

Most patients are asymptomatic, but approximately 50% have nonspecific abdominal pain as a major

Table 2. Potential complications of portal vein aneurysm.

Thrombosis (complete or partial) = most frequent
Portal vein rupture
Duodenal compression
Vena cava inferior obstruction
Portal biliopathy causing jaundice, cholelithiasis, cholestasis
Complications of portal hypertension such as gastrointestinal bleeding

symptom. A minority of patients present with a gastrointestinal bleeding [12]. The cause-consequence relationship between these symptoms and a potential PVA remain to be elucidated as we can not exclude mere association and incidental finding as a result of a conventional abdominal work-up.

Complications of PVA include thrombosis, rupture, duodenal compression, vena cava inferior syndrome and portal biliopathy causing jaundice, cholestasis, and cholelithiasis (Table 2). Thrombosis is the most frequent complication, with complete thrombosis and partial thrombus occurring in 13.6% and 6%, respectively [4]. Spontaneous rupture of the portal trunk has only been reported in three cases, ultrarare given the usual low portal pressure [2,4].

Diagnosis

The most common locations for aneurysms of the portal venous system are the main portal trunk (38.4%), the splenomesenteric venous confluence (23.6%), or the intrahepatic portal vein branches at bifurcation sites. The less-frequent locations are the splenic, mesenteric and umbilical vein. Portal venous system aneurysms are more frequently located extrahepatic (63%) compared to intrahepatic, possibly due to the restrictive effects of the liver parenchyma on aneurysm growth [1,4,9]. The main differential diagnoses of portal vein aneurysm are hyperenhancing abdominal tumors or liver or peripancreatic fluid collections [13].

Imaging

Diagnosis of a PVA is frequently an incidental finding on Color Doppler US, CT or MRI [9].

US is the preferred imaging modality for diagnosis and to monitor the evolution of the aneurysms due to lack of radiation and its low cost. On US, the typical appearance of a portal vein aneurysm is an anechoic cyst-like lesion in continuity with the portal venous system. Color Doppler US and spectral Doppler US allows real-time evaluation of portal venous flow with a nonpulsatile monophasic waveform. The imaging appearance of a thrombosed PVA varies, depending on the age of the thrombus and completeness of thrombosis. A completely thrombosed PVA is often hyperechoic but may show variable echogenicity at gray-scale US, thereby it may mimic a solid mass. At Color Doppler US, flow is absent within the thrombosed segment, and spectral measurements cannot be obtained .US may be useful to distinguish a portal vein aneurysm from a hypervascular mass [6].

Contrast-enhanced CT and MRI can be helpful in patients with equivocal sonographic findings, in particular when differentiating slow flow from thrombosis [6]. On CT/MRI scan, portal vein aneurysm is appreciated as a well-defined contrast enhanced focal saccular or fusiform dilatation of the portal venous system during the portal venous phase.

Complications associated with the PVA and planning of surgery/intervention procedures are better evaluated on cross-sectional imaging [4]. At contrast-enhanced CT and MRI, an acute portal venous thrombus appears as a nonenhancing filling defect within the lumen of the vessel with peripheral rim enhancement. Subacute and chronic thrombosis usually manifests as cavernous transformation, characterized by the formation of a network of dilated collateral veins along the chronic thrombosed portal vein. At MR imaging, there is variable signal intensity in the thrombosed PVA on T1-and T2-weighted images due to ageing blood. Given the extreme rarity of portal vein aneurysms, as well as a lack of familiarity with the entity, the diagnosis can be overlooked [9].

Management

The management remains somewhat controversial, due to the lack of scientific evidence as only few cases have been published so far (Table 3). Conservative management with regular follow-up by US is the best option for the majority of patients, especially in patients with small, asymptomatic aneurysms in the absence of cirrhosis or portal hypertension [6]. Most of the portal venous system aneurysms are stable and have low risk of complications with 88% of patients showing no progression of aneurysm size or complications on follow up imaging [2,3]. In the setting of acute portal vein thrombosis, anticoagulation therapy is recommended resulting in complete or partial recanalization in up to 80% of patients [6]. Patients failing anticoagulation therapy, with extensive thrombus involving the splenic and superior mesenteric veins, or with symptoms related to its mass effect may be referred to the interventional radiologist for percutaneous thrombolysis or thrombectomy and restoration of portal vein anatomy [6]. Also endovascular techniques are used for management of PVA with success [4].

Surgical intervention has been proposed, but remains controversial. The management of PVA should be decided on a case-by-case basis, depending on symptoms, presence of cirrhosis, and size and anatomy of PVA [10]. Currently, surgery is considered in some cases of complicated PVA, that is, in severely symptomatic patients, expanding aneurysm, coexistence of mass effect on adjacent structures, thrombosis, or rupture [14]. The surgical treatments differ

Table 3. Suggested management options for portal vein aneurysm, but rather limited evidence (see text).

Asymptomatic	Thrombosed	Symptomatic	
Conservative management and Clinical and radiological surveillance	- LMWH and surveillance	No Portal hypertension: Aneurysmorraphy (restoring normal diameter)	With Portal hypertension Shunt procedure
	Failure of LMWH or symptomatic: - percutaneous thrombolysis - thrombectomy and restoration of portal vein anatomy - endovascular techniques	Aneurysmectomy (aneurysm resection and insertion of graft)	Liver transplantation

according to the presence of portal hypertension. In patients without portal hypertension aneurysmorrhaphy (restores a normal diameter to portal vein) or aneurysmectomy (aneurysm resection, followed by the insertion of a synthetic or cadaveric graft as a replacement conduit), according to the type of aneurysm (saccular or fusiform), has been successfully described [9]. It restores portal vein laminar flow while maintaining normal hepatic flow, and decreases stasis and resultant thrombosis [1,4,6]. In case of portal hypertension associated with chronic liver disease the surgical approach is different and directed towards surgical shunt procedures, with or without splenectomy or liver transplantation [1]. The aim of the shunt procedures (spleno-renal shunt, porto-caval shunt) is to decompress the portal venous system to prevent progressive dilatation of the aneurysm, instead of treating the aneurysm itself [4].

In three pediatric intra-hepatic PVA patients, the use of meso-Rex bypass, which creates a bypass between the mesenteric vein and the Rex recessus has been reported with good clinical outcome [15]. Percutaneous embolization of the PVA is a good alternative treatment option to successfully occlude the aneurysm and prevent further growth or other clinical sequelae. One case report described the resolution of a PVA after TIPS placement [16].

Among the 190 patients with a PVA, described in literature, 40 (21%) underwent surgery, with a postoperative mortality of 17.5% [1].

It can be assumed that a conservative treatment has lower complication rates and reported conservative treatments of thrombosed EPVA have provided good results, as in our case. Subsequently, we would not consider presence of symptoms or thrombosis as strict indications for surgery, and a conservative approach and follow-up in first intent even for aneurysm of great size or extension to SMV/SV is recommended [5]. This approach is also supported by the low risk of aneurysmal rupture, 2.2% [6].

Conclusion

Portal vein aneurysm represents a rare vascular entity, whose management is still not standardized. There are no evidence-based recommendations, mainly due to the fact that the majority of the articles are isolated case reports or small series. Currently, management is largely determined on a case-by-case basis, and each patient with a PVA must be managed in light of their individual underlying risk factors and symptoms, the features of the aneurysm and the growth of the aneurysm over time. Conservative management seems the best option in the majority of patients. Surgical indication is currently reserved to complicated PVA; however, high postoperative mortality has been described. Since treatment decision is complex, we advocate a multidisciplinary approach.

Disclosure statement

No potential conflict of interest was reported by the authors.

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