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A Case of Portal Venous Aneurysm and a Review of the Literature

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Abstract This is a case of a portal vein aneurysm and the presentation of symptoms. There are currently less than 200 cases documented in English literature. The focus of this report is to detail the presentation of symptoms as well as to discuss appropriate work up and management of portal vein aneurysms. Unfortunately, this patient left against medical advice prior to further work up of his aneurysm.

Keywords: *portal vein aneurysm, surgical intervention, aneurysm*

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1. Introduction

Portal vein aneurysm (PVA) is a rare finding with less than 200 cases described in the literature. PVA is defined as a portal vein's maximum diameter exceeding 15 mm in normal livers and 19 mm in cirrhotic patients. Many findings of PVA are incidental, though approximately 50% of patients with PVA present with vague abdominal pain. Potential theories for causes of PVA include the congenital theory and the acquired theory. A clear etiology is still not understood regarding portal vein aneurysm. Thus, there are no clear strategies when it comes to treatment. Both conservative management and surgical procedures can be appropriate. Post-operative mortality rates remain high, so surgical intervention for PVA is currently only recommended for patients with symptomatic or complicated disease.

2. Case Presentation

Patient is a 69-year-old male, poor historian with medical history significant for end-stage renal disease requiring dialysis (Tuesday, Thursday and Saturday), coronary artery disease, CABG x3, hypertension and recent cataract surgery. He presents with complaint of abdominal pain, described as sharp aching pain over the entire abdomen. The pain increases with certain movements and when he eats. There are no alleviating factors. He has decreased appetite and has not eaten in the past two days. He reports non-bloody diarrhea for the past two days, as well as fever, chills, weakness, and diaphoresis. He denied any nausea, vomiting, or melena. All other review of symptoms was negative.

2.1. Physical Exam

On physical exam, the patient was chronically ill appearing, but in no acute distress. He did have scleral icterus. He had a grade III systolic murmur, healed sternotomy incision, and a right tunneled internal jugular dialysis catheter. Patient had a left upper arm AV fistula that had not been fully matured yet. On exam, his abdomen was soft, mildly distended, with tenderness in all quadrants but was not peritoneal.

2.2. Vital Signs and Labs

Patient's vital signs at presentation included temperature of 37 Celsius, blood pressure 183/80, heart rate of 81, respiration rate of 16/min and oxygen saturation of 80% on room air. Which later improved to 94% on 4L nasal cannula. Of note he is not on home oxygen. Abnormal lab values are listed below. He was hyperkalemic at 6.6 and had an elevated creatinine at 13.9. Patient then admitted that he missed his most recent Thursday dialysis. EKG showed very mild ST elevation in V1 and V2 without any reciprocal depressions. Patient denied chest pain but mentioned some shortness of breath which was worse than usual.

Table 1. LAB DATA

WBC	12,600
Alk Phos	168
AST	239
ALT	176
Total bilirubin	2.2
Potassium	6.6
Creatinine	13.9
Troponin I	20.7; repeat 23.1
Pro-BNP	>175,000

2.3. Imaging

CT scan of the abdomen and pelvis showed a possible pancreatic mass, polycystic kidneys, a calcified mass in the left kidney, and diverticulosis. CT is visualized in Figures 1 and 2. The patient was admitted to ICU and given 0.5 inch nitro paste and started on heparin drip. Plan was to perform left heart catherization on the following day. Dialysis was performed overnight. MRCP and right upper quadrant ultrasound were ordered due to elevated LFTs and concern for pancreatic mass.

Right upper quadrant ultrasound demonstrated a 5.8x3.7 cm vascular structure raising concern for aneurysm or pseudoaneurysm. The radiologist recommended CT angiography with venous and arterial phase. CT angiography results returned with concerns from the radiologist for a peripancreatic pseudocyst with portal vein pseudoaneurysm fistulization. After discussing the patient's medical history and clinical exam with the radiologist, he addended his dictation to a portal vein aneurysm due to lack of any history of pancreatitis in the patient.

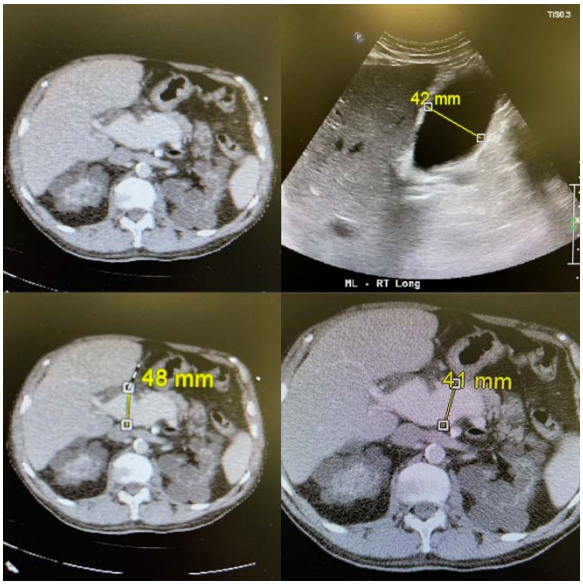

Figure 1. (top left) Axial view, (top right) ultrasound, (bottom) axial measurements

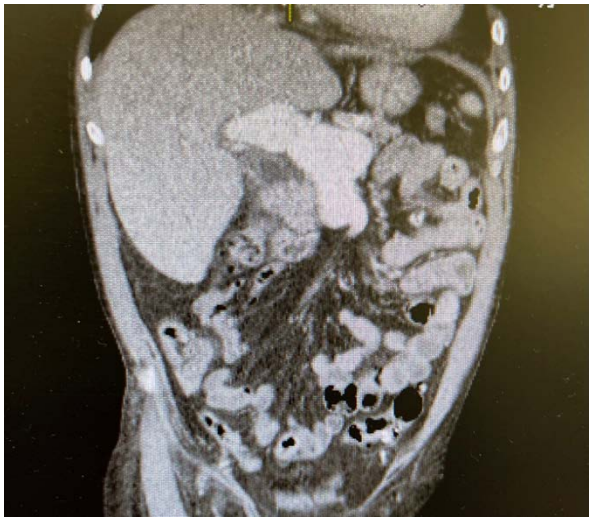

Figure 2. Coronal view

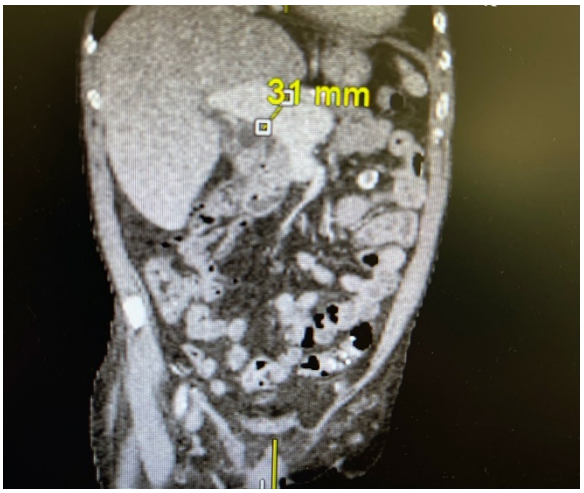

Figure 3. Coronal view measurements

2.4. Care Plan

The following morning the patient went for cardiac catheterization, which showed all grafts were patent. No intervention was performed. At that time, his abdominal pain had resolved, and he stated he was hungry.

At this time, because the patient presented with no known liver problems, further inpatient work up was warranted, including hepatitis panel, trending of LFTs, possible hepatic venous pressure gradient, and transient elastography for workup of possible portal hypertension and cirrhosis, although cirrhosis was not seen on right upper quadrant ultrasound. We were also in search for a potential cause of the portal vein aneurysm, as well as if this was an acute or chronic finding. Given no abnormalities on workup for portal hypertension, this patient's portal vein aneurysm could be attributed to congenital defects in the vessel wall. His portal vein diameter was 58 mm, which is a significant increase from the upper limit of normal for a portal vein (15 mm). The size of this aneurysm raised concern and an MRCP was ordered to check for any underlying obstruction to the biliary system. Suspicion for biliary obstruction was also reinforced by scleral icterus on examination. To our knowledge, there have only been 2 other documented cases of common bile duct compression due to portal vein aneurysm [1]. Unfortunately, we were unable to finish our diagnostic workup as the patient left against medical advice. He stated he would return for outpatient follow-up within the next week. The patient did not return.

With no contributory causes of his elevated liver enzymes on imaging, elevation due to compressive symptoms of the significantly large aneurysm would be a reasonable assumption. This patient had a number of concerning findings including: substantial abdominal pain, aneurysm dilation nearly four times the upper limit of normal portal vein size, and additional signs of surrounding organ compression. Therefore, given the severity of his disease process, it is likely that this patient would have benefited from surgical intervention.

Inpatient work up was warranted, including hepatitis panel, trending of LFTs, possible hepatic venous pressure gradient, and transient elastography for workup of possible portal hypertension and cirrhosis.

3. Discussion

Portal vein aneurysms are a rare vascular finding and were first reported by Barzilai and Kleckner in 1956 [2]. Since then, there have only been 190-200 cases documented in the literature, though these aneurysms are now being detected more frequently as imaging of the abdomen becomes increasingly common. With such a small number of reported cases over the past several decades, a clear understanding of etiology and treatment options has yet to be found. Thus, a literature review was done to elaborate on theories and evolving management strategies in regard to portal vein aneurysms.

This review began with a literature search on the PubMed database, with search terms including: "portal vein aneurysm" OR "visceral venous aneurysm". Only publications in English were retrieved. Additional articles were included after searching through the references section on relevant articles found in the database search.

Portal vein aneurysms (PVA) are defined as a diameter of the portal vein that is 15 mm in non-cirrhotic patients, and 19 mm in cirrhotic patients [3]. This is a largely accepted definition for extrahepatic PVA, although when the PVA in found to be intrahepatic, there is not a universally agreed upon measurement for what constitutes aneurysm [4]. Based on a large scale ultrasonographic study of the portal venous system, it has been suggested that intrahepatic PVA should be defined as greater than 7 mm and 8.5 mm, in non-cirrhotic and cirrhotic patients, respectively [3].

While aneurysms in the venous system do occur, they are more commonly found in superficial veins such as the popliteal, jugular, and saphenous veins [1]. Aneurysm of the visceral veins is much less common. The most common location for an aneurysm of the portal system is in the extrahepatic portion of the portal venous trunk. Between 26%-38% of PVAs occur at this location [1,5]. The second most common location of these aneurysms is at the confluence of the splenic vein and superior mesenteric vein [5]. There is no predisposition for PVA based on gender. The average age of patients diagnosed with PVA is 53, and it has been proposed there may be a bimodal distribution of when portal vein aneurysms occur based on their etiology [6,7]. This bimodal distribution shows that majority of portal vein aneurysms are diagnosed between 13-21 and 47-60 years of age [7].

Because many PVA are found during imaging workup for unrelated issues, it is hard to confidently measure their incidence. For patients presenting with a complaint related to the hepatobiliary system, incidence of portal vein aneurysm is 0.06%. The incidence increases to 0.66% in patients with history of portal hypertension [8]. However, PVA can occur in people who do not have portal hypertension, and who are asymptomatic. Based on a retrospective review of consecutive abdominal CTs, the prevalence of portal vein aneurysms in the general population is estimated to be as high as 0.43% [6].

While the exact causes of portal vein aneurysms are yet to be discovered, the working outline is that there are two general pathways the aneurysm can evolve from; congenital or acquired. The theory of acquired PVAs is that cirrhosis and portal hypertension lead to abnormalities in splanchnic blood flow—including increased flow as well as turbulence—and ultimately cause weaking and dilation of the venous system [4]. There is a known association between patients with liver disease and portal vein aneurysms.

For those patients found to have a PVA in the absence of cirrhosis or portal hypertension, the congenital theory can be applied. Congenital portal vein aneurysms have been defined as presence of PVA in patients without trauma, history of liver biopsy, portal hypertension, or hepatitis [9]. Congenital PVAs are thought to emerge as a result of failure of the distal right vitelline vein to involute during embryologic development. This causes a diverticulum to remain present, predisposing to formation of an aneurysm, especially near the superior mesenteric vein [10]. It is also suggested that interruptions to other developing structures may cause a congenital weakness of the portal venous system, allowing aneurysms to form early.

The congenital theory is further supported by well-documented cases of young children with portal vein aneurysms. One case reports an in-utero portal vein aneurysm. It was first captured by ultrasonography at 37 weeks gestation, showing an intrahepatic left PVA. Presence of a 10 mm diameter aneurysm was confirmed on day 1 of life in this patient with pulsed and color Doppler ultrasound [11]. He was followed for 6 months, remaining asymptomatic and with the aneurysm decreasing to 8 mm in size. Another case demonstrates a five-year-old male with two months of intermittent periumbilical pain found to have a dilation of the portal vein measuring 19 mm. Established normal portal vein measurements for children based on height state that a normal diameter in this patient should have been a maximum of 7.5 mm [7]. This patient's workup was negative for both liver disease and portal hypertension, further validating the congenital variant of portal vein aneurysms.

As one third of patients with portal vein aneurysms are asymptomatic, finding these vascular anomalies commonly occurs during imaging for other diagnostic reasons [5]. 38.2% of aneurysms have been incidentally found by CT or ultrasound imaging [1]. It is a well-established recommendation that diagnosis of these aneurysms is most appropriate by ultrasound with Doppler. Portal vein aneurysms may mimic a cyst on other imaging, so detecting flow through the structure using Doppler ultrasonography is critical in properly diagnosing the dilation properly [4]. Schwope et al. describe the appearance of PVA on ultrasound as an anechoic mass appearing closely located to the portal vein. Use of ultrasound is also helpful to differentiate aneurysm from pancreatic mass, neuroendocrine tumors, or metastasis from a distant malignancy [4]. Furthermore, the rate of thrombosis of portal vein aneurysms is 17%. Doppler ultrasound can detect thrombosis if tubular anechoic structures are visualized around the portal vein, or if calcifications of the portal vein are seen [1,6,9].

Studies estimate that between 50-72% of patients with PVA remain asymptomatic [5,6]. There is a positive association between patients developing symptoms, and increasing size of the aneurysm. The most common presenting symptom is vague abdominal pain. A retrospective study has found that patients who were

symptomatic had significantly larger aneurysms than asymptomatic patients (33.6 ∓ 9.9 mm and 23.1 ∓ 3.3 mm, respectively) [6]. Some patients may also present with GI bleeding or symptoms reflecting compression of surrounding structures, including jaundice and abdominal distension. These are rare, and compression has been reported in only 5 cases: two cases of common bile duct compression, two of duodenal compression, and one PVA imposing on the IVC [1].

In a case review of 190 patients described in the literature, 40 patients underwent operative intervention for repair of their aneurysm. Post-operative mortality was seen to be 17.5% [5]. This can be compared to another systematic review, which found that out of 170 patients with PVA, mortality rate was 10.3%, irrespective of intervention. In this review, five of the nine patients who died had undergone surgical repair, while three of the others died as a result of aneurysm rupture or thrombosis [1]. Because of the high mortality rate following PVA repair, surgical intervention for PVA is considered based on features of the aneurysm itself, and currently only recommended for patients with complicated disease. Complications include symptoms directly related to the aneurysm, thrombosis or increasing size of the aneurysm, which puts patient at risk for rupture [5]. Surgery may be considered appropriate if size of aneurysm is greater than 3 cm [12]. Sfyroeras et al. have recommended that aneurysms of congenital origin are stable and should not be operated on, while acquired PVAs may be considered surgical, though this is not considered a standard approach.

When thrombosis is the only complication, it is recommended that medical therapy be the mainstay of management. Anticoagulation is routinely used in treatment of thrombosed PVA, though thrombosis can also be managed by percutaneous thrombectomy or thrombolysis [6]. Anticoagulation therapy alone has been highly successful in treating thrombosed portal vein aneurysms, with complete or partial patency recovered in 80-90% of patients [6]. Up to 88% of patients who followed up with serial abdominal ultrasounds were found to have stable size aneurysms without need for surgical intervention [1].

When surgery is deemed appropriate, surgical options are divided into two categories, dependent upon if a patient does or does not have portal hypertension. In patients without portal hypertension, aneurysmorrhaphy or aneurysmectomy are performed [5]. Operative approach is chosen based on the location and shape of the aneurysm. Aneurysmorrhaphy tends to be the easiest way to excise an aneurysm and allows the native vessel to be repaired without necessitating a graft [1]. This technique is most appropriate for saccular aneurysms and is associated with low morbidity. Portal venous aneurysmorrhaphy has been documented for 10 cases in the literature. All patients were alive at follow up with only one patient having post-operative complication of internal bleeding [13]. Alternatively, aneurysmectomy can be performed with a cadaveric allograft, or synthetic graft to replace portal vein segment that was removed [1]. Aneurysmectomy is commonly utilized for fusiform aneurysms.

In patients with portal hypertension, the most appropriate surgeries are shunt procedures, splenectomy, or liver transplantation. The rationale behind utilizing these techniques is to treat the potential cause for the PVA by decreasing splanchnic flow and relieving portal hypertension. Five patients with cirrhosis or portal hypertension have undergone surgical treatment. Two of these five patients were deceased within 1 year of the procedure [5,13]. Considering the high mortality rate in patients with liver disease in conjunction with low occurrence of portal venous aneurysm rupture, there is no evidence that patients with these comorbidities should proceed with surgery, as their risk of post-operative complications (e.g. GI bleed) likely outweigh any benefits of trying to remedy the aneurysm [13].

Portal venous system aneurysms remain a relatively rare pathology of the vascular system. Standard of care has yet to be determined , as the majority of research surrounding PVAs occur within isolated cases. Regardless, the most appropriate approach to portal vein aneurysms continues to be conservative with routine follow up. Ultrasonography and anticoagulation are both reasonable management practices as the majority of PVAs are uncomplicated. Surgical intervention is necessary in only a small number of cases, when the benefits from resolution of the aneurysm outweigh the risks of post-operative complications. Patients with portal hypertension and cirrhosis should be treated conservatively as this population has the highest incidence of post-operative mortality.

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