CASE REPORT

Incidental Portal Vein Aneurysm Found in a Patient with a Diaphragmatic Hernia and Evolving Gastric Volvulus

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Rare and poorly described within the literature, a portal vein aneurysm (PVA) is defined as an abnormal vascular dilation of the portal vein exceeding 19 mm in patients with cirrhotic livers and 15 mm in patients with normal livers. Incidence has been estimated to be 0.06% and fewer than 200 cases have been described in the literature[1,4]. We describe an incidental and asymptomatic PVA found in an 80-year-old Caucasian male presenting with evolving gastric volvulus in the context of a large diaphragmatic hernia. Computed tomography (CT) revealed displacement of abdominal contents and 37 mm aneurysm of the main portal vein (image 1,2). Because of the large size of our patients' malformation (37 mm), surgery was pursued in the form of aneurysmorrhaphy. There is a paucity of literature regarding the management of PVA -- however the literature distinguishes between those with and without underlying portal hypertension. Notably, the patient was asymptomatic from this pathology and had no underlying portal hypertension. He was seen in an outpatient clinic two weeks after discharge and had fully recovered without any reported sequela or symptoms related to his vascular surgery.

Keywords: Portal vein aneurysm (PAV), diaphragmatic hernia, volvulus, venous aneurysm, aneurysm

INTRODUCTION

Rare and poorly described within the literature, a portal vein aneurysm (PVA) is defined as an abnormal vascular dilation of the portal vein exceeding 19 mm in patients with cirrhotic livers and 15 mm in patients with normal livers. Incidence has been estimated to be 0.06% and less than 200 cases have been described in the literature[1,4].

We report an exceedingly rare case of an incidental, extrahepatic PVA occurring in the context of a diaphragmatic hernia complicated by gastric volvulus in an otherwise healthy 80 year old Caucasian male. Briefly, we will describe the management of the PVA and its outcome in our patient.

CASE REPORT

Our patient was an 80-year-old Caucasian male who presented from an outside hospital (OSH) with a chief complaint of extreme epigastric pain that radiated to his left chest and back. He also endorsed nausea and nonblood, nonbilious vomiting. Past medical history was significant for appendicitis treated with an uncomplicated laparoscopic appendectomy 7 year prior to presentation. He had no history of gastrointestinal pathology though notably the patient endorsed a 55 pack-year smoking history while denying any alcohol or drug use. Review of symptoms revealed difficulty breathing, chest tightness, chest pain, shortness of breath, abdominal pain, and constipation. The patient denied hematemesis, melena, and hematochezia. On initial presentation, he was hemodynamically stable and afebrile. Physical exam was significant for bowel sounds in the left chest on auscultation and mild tenderness to deep palpation of the epigastric region and left upper quadrant. There were no signs of portal hypertension or liver pathology including ascites, varices, or jaundice though he was noted to have an elevated total bilirubin of 1.5 mg/dL (indirect 1.0 mg/mL, direct 0.5 mg/mL). Lipase, Amylase, AST, ALT, Alkaline phosphatase were all within normal range. He had an elevated WBC of 16,800/mm3.

Computed tomography (CT) of the chest, abdomen, and pelvis was significant for: 1. diaphragmatic hernia with displacement of the stomach, left colon, pancreatic tail and spleen into the left hemithorax with gastric volvulus and diffuse pneumatosis. 2. A 37mm saccular proximal portal vein aneurysm extending from the proximal main portal vein (figure 1,2). Due to the patient's worsening symptoms and radiological signs of incarcerated diaphragmatic hernia, the patient was sent for emergent exploratory laparotomy and hernia repair. During the surgery, there was extensive scar tissue and adhesions around the herniated organs through the 7mm posterior, lateral diaphragmatic hernia which indicated an acute on chronic condition. Based on the clinical evidence, including pathology, it was surmised that the patient had a chronic diaphragmatic hernia that acutely evolved into gastric volvulus requiring gastropexy. During this initial operation, the defect was repaired and he was stabilized in anticipation of another surgery to finalize the repair and remove packing that was placed to stop blood loss. During this second surgery, two days later, the patient was sent for surgery to reinforce the diaphragmatic repair and to look for further necrosis of his viscera (none was found). During his second surgery, attention was brought to the PVA and he was treated with an aneurysmorrhaphy. Surgery was planned over two days because it decided that his volvulus would be treated with two sperate procedures, one to address the immediate threat to life and another to reinforce the operative site and remove packing placed the day prior. Furthermore, this second look operation was necessary to look for further necrosis within the abdomen. Given that his portal vein aneurysm was being repaired semi-electively, we decided to repair it on the second operative day when the patient was more stable and the threat of immediate decompensation had abated. Given that the PVA had no immediate threat to his life, our surgeons opted for more time to plan the procure and wanted the patient to be hemodynamically stable before such an invasive procedure. A surgical specimen was sent to pathology, which confirmed that the sample was a portal vein aneurysm with no signs of malignancy or infiltration. Post-surgical anticoagulation was limited to sequential compression device (SCD).

Patient was discharged on Aspirin 81 MG chew tablet and pain medication. The patient was seen two weeks after being discharged and reported resolution of all symptoms. No further imaging was pursued as the patient had limited access to health care providers in his area and was eventually lost to follow up.

CASE DISCUSSION

PVA is thought to be either an acquired or congenital pathology[2] -- with most acquired cases being the sequela of portal hypertension due to cirrhosis of the liver[3]. Congenital causes have several proposed mechanisms, most notably incomplete regression of the right primitive distal vitelline vein[4]. Other causes have been described in the literature including trauma[7], severe pancreatitis[6], and malignancy of the portal vein[4,8]. At this time, no risk factors have been extensively studied, including age, race, or gender.

Approximately, 33% of PVAs are asymptomatic and are thus found incidentally as in our patient. Furthermore, 50% of patients present with vague and nonspecific abdominal pain. The best imaging modalities for assessing and monitoring PVAs includes computed tomography scan, magnetic resonance angiography, and doppler ultrasonography. Imaging will usually reveal localization of the aneurysm at the level of the main portal trunk (38.4%), portal bifurcation and intrahepatic portal branches (38%), and less commonly at the spleno-mesenteric confluence (23.6%), percentages derived from a review that looked at 96 published reports and 190 patients with portal vein aneurysm[4].

The two feared complications of PVA are thrombosis and spontaneous rupture with subsequent gastrointestinal or intraperitoneal bleeding, though compression resulting in obstructive jaundice or obstruction of the duodenum has also been described[4]. Management of the a PVA remains unsettled[4] but current literature supports a conservative approach (watchful waiting) for asymptomatic cases without portal hypertension[5] while surgery is reserved for cases of rupture, thrombosis, or symptomatic or enlarging aneurysm[4]. Some authors also considered surgical treatment for any non-thrombotic PVA larger than 30 mm[10]. Surgery will entail aneurysmectomy or aneurysmorrhaphy in patients with normal livers while patients with portal hypertension are treated by addressing the portal hypertension directly, be that with a shunt or liver transplantation[4]. Given the rarity of PVAs, surgical management should be reserved to high volume hepato-biliary centers with vascular surgery on staff. Postoperative mortality in the 40 patients in a study that underwent surgical management was 17.5%[4]. Currently the consensus for conservative management is regular 6 to 12 month imaging, ideally with ultrasound. It should be noted that spontaneous regression has been reported within the literature[9].

Still many questions remain about the PVA. First and foremost, natural history has not been fully elucidated nor have the risk factors for its development been described. Furthermore, no study to date has compared outcomes with different surgical approaches or attempted to distinguish management based on location or anatomy of the aneurysm. Finally, the evidence guiding the management of PVAs associated with portal hypertension comes mostly from case reports and there is scant empirical evidence guiding the specific surgical approach indicated.

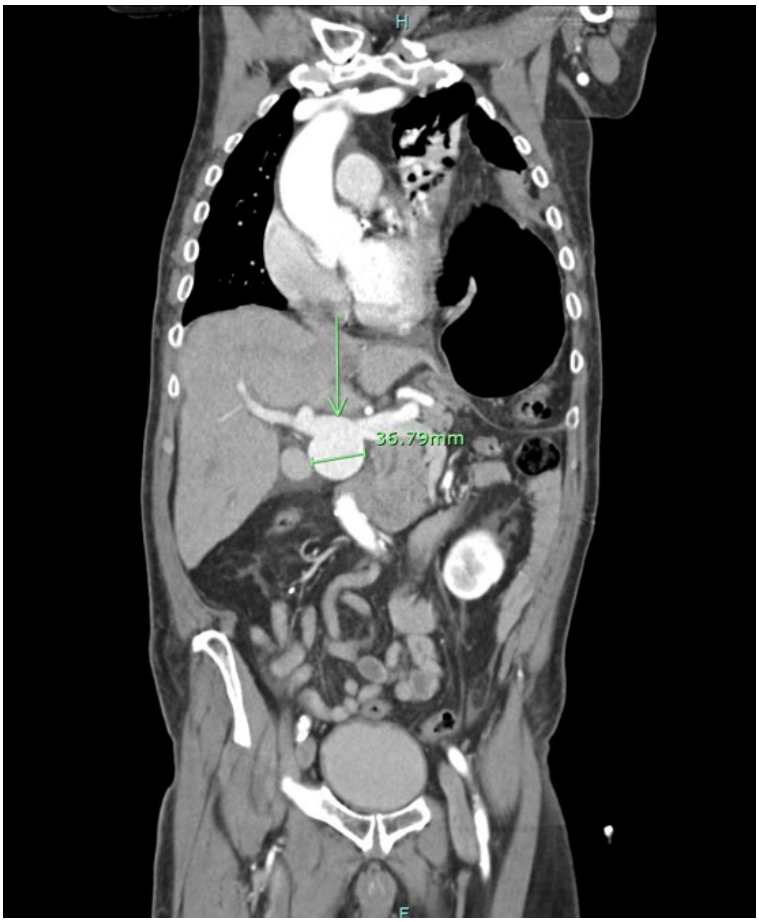

Figure 1. Portal Vein Aneurysm (green arrow) seen in transverse plane. Prominent diagrammatic hernia in left thorax.

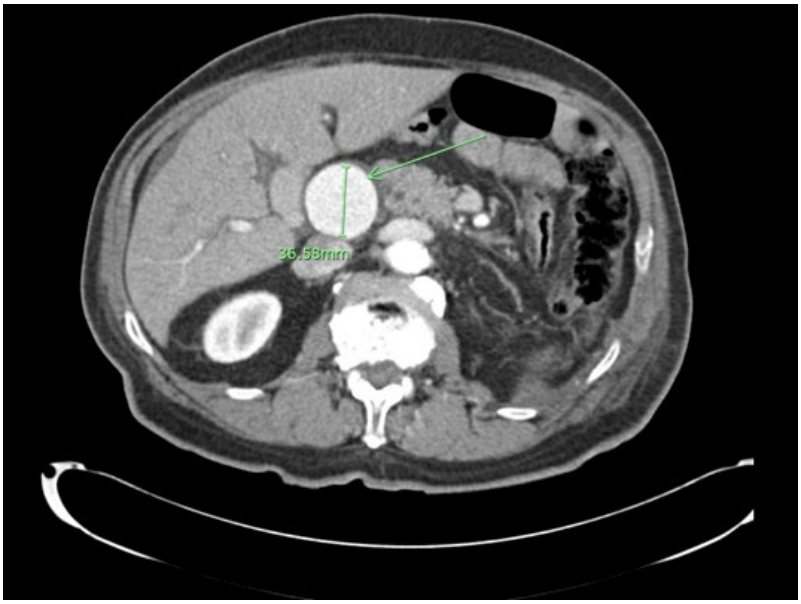

Figure 1. Portal Vein Aneurysm (green arrow) seen in transverse plane.

CONCLUSION

Although exceedingly rare, management of the portal vein aneurysm has become more relevant in recent years because of the increasing incidence of cross-sectional imaging leading to more incidental findings. In our extensive literature review there were no documented cases of PVA in a patient with a diaphragmatic hernia and evolving gastric volvulus. Although our patient had no formal diagnosis of connective tissue disease or familial history of similar symptoms, it remains a possibility that there may be an underlying pathology causing both the PVA and the large diaphragmatic hernia. Whether this may be a connective tissue disease or an underlying congenital malformation that predisposed him to both abnormalities may be elucidated with further research. A diaphragmatic hernia is a protrusion of abdominal contents into the thoracic cavity. Although this defect is usually congenital and present at birth, it is possible to develop an acquired diaphragmatic hernia (ADH). Approximately 80% of ADH cases are a result of blunt trauma to the thoracoabdominal region[11]. The second most common etiology of ADH is iatrogenic, usually a sequela of surgery[12]. Still yet, a smaller subset of patients develop ADH spontaneously. The location of the defect is most commonly at the esophageal hiatus or at other points of failure of diaphragmatic fusion during the embryonic stage[13]. The patient likely developed ADH over the course of years, during which his visceral organs slowly herniated into his thorax. Given that he denied any trauma it's unclear what provoked his gastric volvulus. We scanned the literature for evidence implicating the relationship between portal vein aneurysm and other diseases of connective tissue but there are no other similar case reports represented in the literature today. Currently, there is no data to recommend further imaging or changes to management in patient with hernias of any type. Still, close evaluation of patients with large hernias may reveal aneurysms that can be repaired concurrently with hernia repair.

There is a paucity of literature regarding the management of PVA but the literature distinguishes between those with and without underlying portal hypertension. Given the large size of our patient's malformation (37 mm), surgery was pursued even though he was seemingly asymptomatic from this pathology. Ultimately, surgery was successful and at 2 week follow up he had returned to his baseline health without issues.

The PVA remains a rare and elusive vascular malformation of the abdominal viscera. Further research must be done to clarify etiology, risk factors, management, and outcomes.

Notes

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