Operative management of an incidental portal vein aneurysm in the setting of an incarcerated congenital diaphragmatic hernia

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ABSTRACT
Portal vein aneurysms are rare pathologic entities. A 3.7-cm portal vein aneurysm was incidentally discovered in an 80-year-old male patient on imaging for acute abdominal pain secondary to an incarcerated diaphragmatic hernia. The aneurysm was resected, and primary repair of the portal vein was performed during a second-look operation after repair of the incarcerated hernia. Operative intervention was chosen for this patient because of the aneurysm's size and the additional indication for abdominal exploration. (J Vasc Surg Cases and Innovative Techniques 2021;7:64-7.)

Keywords: Aneurysm; Endovascular repair; Portal vein aneurysm; Venous aneurysm

Portal vein aneurysms are rare pathologic entities.1-7 Given their low prevalence, the etiology, pathophysiology, and natural course of these aneurysms are not completely understood.1 Portal vein aneurysms were first described by Barzilai and Kleckner8 in 1956. A systematic literature review of studies related to portal vein aneurysms was performed by Laurenzi et al9 in 2015, which yielded a total of 96 reports with a total of 190 patients. The reported incidence of these aneurysms has been increasing, given the frequent widespread use of abdominal imaging.3,4

In the present case report, we have described a proximal saccular portal venous aneurysm that was surgically managed in the setting of an incarcerated diaphragmatic hernia.

CASE REPORT
An 80-year-old man had initially presented to an outside hospital with a 1-day history of new-onset, left-sided chest pain and severe epigastric pain with associated nausea and vomiting. The patient was taking 81 mg of aspirin daily as his only home medication. He was a previous cigarette smoker, had quit 30 years previously, had consumed alcohol previously, and denied other illicit drug use. He had no history of cirrhosis or chronic liver disease.

The initial workup demonstrated leukocytosis of 16,700 cells/mm3; mild hyperbilirubinemia, and a minimal elevation of serum amylase and lipase levels. A contrast-enhanced computed tomography (CT) scan showed elevation of the left hemidiaphragm and an incidental finding of a saccular aneurysm of the proximal portal vein measuring 3.7 cm × 3.7 cm in the maximum diameter with no thrombus within (Fig 1, A and B).

The patient was transferred to our institution. On presentation, he was hemodynamically stable. However, he had diminished breath sounds on the left side of the chest and a soft, distended abdomen that was diffusely tender to palpation without guarding, rigidity, or rebound tenderness. The patient was initially treated nonoperatively with gastric decompression via a nasogastric tube, bowel rest, and intravenous hydration. However, his symptoms were not relieved through conservative measures. A follow-up CT scan revealed an incarcerated diaphragmatic hernia with displacement of the stomach, left colon, pancreatic tail, and spleen into the left hemithorax. He had a gastric volvulus with an air–fluid level in the stomach and diffuse pneumatisis. The mediastinum was shifted to the right with compression of the left mainstem bronchus and complete atelectasis of the left lower lobe.

The patient was taken for urgent exploratory laparotomy, reduction and repair of the diaphragmatic hernia, partial gastrectomy, and temporary closure of the abdomen with negative pressure wound therapy. Two days later, he returned to the operating room for reexploration of the open abdomen, gastroscopy with gastric feeding tube placement, and formal closure of the abdomen by our acute care surgery team. We performed resection of the portal vein aneurysm with primary repair of the portal vein during this operation. The saccular aneurysm was exposed (Fig 2) and was noted to have a discrete stalk that was 12 mm wide and 24 mm long. The adjacent portal vein was grossly normal, with a maximum diameter of 15 mm.
proximally and distally to the stalk. The patient was fully heparinized, and a side-biting Satinsky clamp was placed along the base of the aneurysm at the confluence with the portal vein (Fig 3). The aneurysm was excised, and the specimen was sent for pathologic examination. The portal vein was repaired by oversewing the stump of the stalk with two layers of 5-0 Prolene suture. The clamp was then removed (Fig 4). No narrowing of the portal vein was found on direct visualization. The abdomen was closed, and the patient was returned to intensive care. He was extubated the next day, and antiplatelet therapy with 81 mg of aspirin was restarted. The patient was discharged to a rehabilitation facility on postoperative day 8 with an oral diet to help him transition back to independent living. At 2 weeks after discharge, the gastrostomy tube was removed without difficulty. A follow-up CT scan of the abdomen and pelvis with intravenous contrast obtained 6 months after discharge demonstrated a widely patent portal vein without stenosis or thrombosis, with a maximal diameter at the site of repair measuring 1.9 cm x 1.1 cm.

DISCUSSION
To the best of our knowledge, <200 cases of portal venous aneurysms have been reported in contemporary literature, highlighting the paucity of evidence about this rare pathologic entity. Portal vein aneurysms do not occur in a predominant age group or gender with a greater incidence. The clinical vignettes reporting the presentation of these aneurysms has also varied. Approximately one half of the patients will present with nonspecific abdominal pain and approximately one third of patients will be asymptomatic, with the aneurysm discovered incidentally, as was the case for the present patient. Different hypotheses pointing toward the etiology of these aneurysms have been proposed. Some lesions appear to be congenital in origin, but others appear to be acquired. In the case of the present patient, it is possible that the aneurysm could have resulted from some occult trauma in the past—especially because our patient did not have chronic liver disease. However, we had no ability to be certain of the etiology.
Multiple investigators have described the normal dimensions of the portal vein via autopsy and ultrasound studies. Douglass et al.\textsuperscript{10} studied 92 autopsies and reported that the diameter of the portal vein was 0.64 to 1.21 cm in patients without cirrhosis and those without portal hypertension. Doust and Pearce\textsuperscript{11} performed a study with ultrasonography and found that the portal vein diameter was never $>1.5$ cm in normal patients and was never $>1.9$ cm in patients with cirrhosis. These studies allowed for any segment of a portal vein with a diameter of $>2$ cm to be defined as aneurysmal.\textsuperscript{6}

The reported complications of portal vein aneurysms include thrombosis, spontaneous rupture resulting in large-volume gastrointestinal bleeding, and compression from a mass effect that results in jaundice or duodenal obstruction.\textsuperscript{2} Most patients, however, remain asymptomatic.

At present, no evidence-based recommendations have been reported regarding the management of portal vein aneurysms, mainly owing to the rarity of the lesion and because the lesion has only been described in case reports and small case series. Laurenzi et al.\textsuperscript{2} reported that as of 2015, of the 190 cases reported, only 40 (21\%) had undergone surgery. According to Laurenzi et al.,\textsuperscript{2} surgical intervention should be considered for patients with complicated portal vein aneurysms, such as those with rupture or thrombosis, symptomatic aneurysms, and enlarging aneurysms with an increased risk of spontaneous rupture. Moreno et al.\textsuperscript{12} suggested that surgical treatment should be offered to all patients presenting with nonthrombotic portal vein aneurysms measuring $>3$ cm in maximum diameter. However, our literature search found no comparative studies or definitive evidence confirming the benefit of surgical intervention for symptomatic patients.\textsuperscript{2}

In the case of the present patient, operative intervention was decided, given the large size of the aneurysm and the additional indication for abdominal exploration (ie, the incarcerated hernia). Observation was considered for this lesion; however, given the extensive abdominal surgery the patient had previously undergone, surgical repair of the aneurysm after it had become symptomatic or had ruptured in the future would have been very difficult. This was likely the best opportunity to perform an open operation of the aneurysm, especially because no known endovascular procedures are available to repair the lesion. Without another indication for laparotomy, the risks of repairing the aneurysm might not have
outweighed the benefits given the patient’s age. In that case, the aneurysm might have been observed with surveillance and serial CT imaging studies of the abdomen.

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