Splenic Artery Aneurysms: Two Cases of Varied Etiology, Clinical Presentation and Treatment Outcome

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SUMMARY
Introduction Splenic artery aneurysms are potentially lethal lesions. We report two illustrative cases and discuss etiology, diagnosis and treatment of these aneurysms.

Outline of Cases Both patients, age 31 and 80 years, were biparous women. The younger woman, otherwise healthy, was referred from a local hospital 3 weeks after she underwent a left subcostal laparotomy and exploration for symptomatic abdominal mass diagnosed by CT. Angiography established the diagnosis of a large, non-ruptured splenic artery aneurysm. Elective aneurysmectomy with splenectomy was performed using the approach through the upper median laparotomy and bursa omentalis. Postoperative course was uneventful. Histopathology demonstrated cystic medial necrosis with chronic dissection. The other patient, elderly woman, presented urgently with acute abdominal pain and syncope and was diagnosed by computed tomography with a huge, ruptured splenic artery aneurysm. She underwent immediate aneurysmectomy with splenectomy using the same, above-mentioned approach. External pancreatic fistula and pancreatic pseudocyst complicated the postoperative course, requiring open pseudocyst drainage and cystojejunostomy. After a protracted hospitalization patient eventually recovered. The pathological diagnosis was atherosclerotic aneurysm.

Conclusion Splenic artery aneurysms are infrequent lesions, with varied etiology and clinical presentation. Timely diagnosis and adequate treatment prevent life-threatening rupture and lessen the risk of operative morbidity and mortality.

Keywords: splenic artery aneurysm; aneurysmectomy; splenectomy; pancreatic pseudocyst

INTRODUCTION
Visceral artery aneurysms in general and splenic artery aneurysms (SAA) in particular are infrequent lesions not usually considered in the differential diagnosis of any abdominal complaints, but they are a well recognized and important form of vascular disease. Often clinically silent until late in their course, they show relentless tendency towards progressive enlargement. Increasing use of sophisticated vascular imaging has allowed for earlier diagnosis of these aneurysms and timely therapeutic intervention, thus avoiding the high morbidity and mortality associated with rupture [1-4].

CASE REPORTS
Both patients, age 31 and 80 years, were female and biparous. The younger woman was referred from a local hospital 3 weeks after she underwent a left subcostal laparotomy and exploration for undefined left-upper abdominal mass diagnosed by computed tomography (CT) and presumed to be the cause of her chronic non-specific abdominal complaints. She was apparently healthy, without risk factors for a vascular disease and denied any previous abdominal trauma. Conventional catheter arteriography established the diagnosis of a 5.4 cm, non-ruptured, saccular aneurysm of the distal splenic artery (Figure 1). Elective aneurysm resection with concomitant splenectomy was performed using the upper median laparotomy and direct approach through the bursa omentalis (Figure 2). Histopathological examination of the resected aneurysm wall revealed signs of chronic dissection and of cystic medial necrosis (Figure 3a). After an uneventful recovery, patient was discharged on postoperative day 6. Forty two months later, she was in good health, without signs of postoperative complications or newly developed vascular lesions. The other patient, frail elderly woman with a long-standing history of poorly controlled essential hypertension was admitted urgently a few hours after experiencing sudden severe upper abdominal pain followed by syncope. Urge CT aortography revealed a 9.2 cm saccular SAA with contained rupture (Figure 4). Immediate aneurysmectomy with splenectomy was performed using the same surgical approach as employed in Case 1. Operative findings confirmed the presence of a giant, heavily calcified SAA embedded in the pan-
Figure 1. Preoperative contrast angiogram (right femoral approach with selective catheterization of the coeliac trunk) demonstrates large saccular aneurysm of the distal portion of the splenic artery (maximal transverse diameter of 5.4 cm), encroaching upon the splenic hilum. Nonhomogeneous opacification of the aneurysmal sac suggests the presence of an intraluminal thrombus.

Figure 2. Intraoperative photograph: using the approach through the lesser peritoneal sac, proximal splenic artery was identified, isolated and clamped; large aneurysm of the distal splenic artery was opened, found to be partially thrombosed (arrow) and then partially resected (posterior wall, intimately adherent to the pancreas, left intact); splenectomy was also performed.

Figure 3. a) Histopathological findings in Case 1 included cystic medial necrosis and chronic wall dissection (HE, ×100). b) Case 2: complicated atherosclerotic lesions, with plaque ulceration, intraplaque hemorrhage and heavy calcification (Vb, Vla, Vlb, Vlc intimal lesions, according to the American Heart Association advanced atherosclerosis classification system; HE, ×100).

Figure 4. Contrast-enhanced computed tomogram (a: transverse scan at the level of the upper abdomen; b: coronal reformatted image) showing giant SAA with incipient rupture (discontinuous ring of calcifications at the posterolateral aneurysmal wall with locally contained blood extravasation, arrow).
creatic tissue, with evidence of intramural hematoma and perianeurysmal blood leakage (Figure 5). Aneurysm size and rupture distorted and obscured local anatomy, precluding selective control of the proximal splenic artery and necessitating supraceliac aortic clamping before opening of the aneurysmal sac. Although accidental pancreatic tail injury was evident intraoperatively, it was judged to be insignificant and not to require formal distal pancreatectomy. Instead, local measures were undertaken, including fibrin glue application and omentopexy, to prevent further pancreatic complications. Despite such precautions, quality and duration of postoperative abdominal drainage (drain placed in the splenic bed), as well as elevated serum amylase and lipase levels, indicated development of a low-volume, external pancreatic fistula. Postoperative imaging findings (abdominal ultrasound, abdominal CT, and magnetic resonance cholangiopancreatography [MRCP], performed on postoperative day 5, 14 and 20, respectively) demonstrated progressively larger pancreatic pseudocyst, which eventually completely filled the bursa omentalis (Figure 6). Due to its wide communication with the main pancreatic duct, demonstrated by MRCP, it was not suitable for minimally invasive, interventional treatment (endoscopic or percutaneous drainage). After the period of pseudocyst “maturation”, on the postoperative day 35 the patient was reoperated and internal drainage with Roux-en-Y cystojejunostomy was performed (Figure 6). Postoperatively, protracted abdominal drainage, which seized on conservative treatment, and superficial wound infection required prolonged hospital treatment, but was eventually successful. The pathological examination of the aneurysm was consistent with an atherosclerotic etiology (Figure 3b). At the 6-month follow-up the patient was alive...
and asymptomatic. Both patients also received antipneumococcal vaccine and long-term antiplatelet prophylaxis was introduced (100 mg aspirin daily, indefinitely) to counteract postsplenectomy thrombocytosis.

**DISCUSSION**

Aneurysms of the visceral branches of the abdominal aorta encompass splanchnic and renal arteries aneurysms with splanchnic aneurysms approximately twice as common as renal [1, 2]. They are potentially life-threatening and maybe more common than previously thought [1, 4, 5]. In fact, autopsy studies suggest that visceral aneurysms may be more frequent than abdominal aortic aneurysms [6]. This is in agreement with increased detection of such lesions in the current clinical practice by widespread use of sophisticate diagnostic imaging such as CT angiography [1, 2, 5]. It is important to recognize these lesions timely because up to 25% of them may be complicated by rupture, and the mortality rate after rupture is between 25% and 70% [1, 2, 4]. SAAs are the most common splanchnic artery aneurysms, representing at least 60% of such lesions. They are the third most common site of intra-abdominal aneurysms encountered in the clinical practice, after the aorta and the iliac arteries [4, 5, 7]. The true prevalence of SAAs is difficult to ascertain because most are asymptomatic and diagnosed incidentally. Since the first report of SAA by Beussier at an autopsy in 1770, the estimates of its true prevalence vary widely, ranging from 0.02% to 0.1% in general autopsies to 0.78% in patients undergoing arteriogram for other indications, and up to 7.1% of autopsies in patients with cirrhotic portal hypertension [4, 5, 7, 8]. There is a strong predominance of female sex (female-to-male ratio 4:1). Aneurysms are most commonly found in the distal third of the splenic artery and they are multiple in 20% of patients. They are usually saccular and typically occur at bifurcations. Over 70% of SAAs are true aneurysms [4, 5, 7]. The pathogenesis of SAA is incompletely understood and specific cause remains unknown. Although SAAs often exhibit typical calcific atherosclerotic changes, this is more likely to represent a secondary event than a primary etiology. The major predisposing conditions include medial degeneration with superimposed atherosclerosis, pregnancy and portal hypertension with splenomegaly [4, 5, 7, 8]. Less common causes are pancreatitis, penetrating trauma (false SAAs are most often caused by chronic pancreatitis or by penetrating trauma) [9, 10], septic emboli (leading to infected or “mycotic” SAA) [11], idiopathic dissection, essential hypertension [12], fibromuscular dysplasia, congenital or acquired connective tissue disorders [13] and orthotopic liver transplantation [14]. Dilatation of the splenic artery resulting from increased blood flow during pregnancy and the effects of estrogen on the elastic tissue of the tunica media likely predisposes to aneurysm formation. This is particularly evident with repeated pregnancies, since 40% of women reported to have SAAs have been multiparous (average, 4.5 pregnancies) [4, 5, 7]. Similarly, increased splenic blood flow is considered to be the cause of SAAs in portal hypertension and after liver transplantation; increased estrogen activity associated with cirrhosis may be a contributing factor [4, 5, 12, 14].

Most SAAs are <2.5 cm in diameter and asymptomatic. In the largest series published to date, Abbas et al. [4] (217 patients over an 18-year period) and Trastek et al. [7] (100 patients over a 20-year period) have found a mean diameter for SAAs of 2.2 cm and 2.1 cm, respectively. “Giant” SAAs (≥10 cm) are exceedingly rare [15]. In the past, curvilinear calcifications in the left upper quadrant accidentally detected on the plain abdominal radiograph were often the first sign of SAA [16]. The differential diagnosis includes tortuous splenic artery (common finding in elderly persons), renal artery aneurysm, calcified lymph nodes and calcific cysts of the spleen or adrenal gland. Symptomatic patients manifest with left upper quadrant or epigastric pain that radiates to the left shoulder. Spontaneous rupture is the most common complication and occurs in 2% of patients. However, the risk of rupture is much higher for pregnant women and for aneurysms >2 cm in diameter [4, 5, 7]. Majority of SAAs in pregnant women are diagnosed after rupture and are associated with a 70% maternal and 75–95% fetal mortality rate [17]. Rupture of the aneurysm is manifested initially as upper abdominal pain, but the patient is hemodynamically stable as long as bleeding is contained within the lesser sac by omentum and blood clots that block the foramen of Winslow. Hours or days later, as tension increases and blood overflows into the greater intraperitoneal sac, secondary or free rupture develops with diffuse abdominal pain and hypovolemic shock. It is estimated that this two-stage or “double-rupture” phenomenon, first described by Brockman in 1930 [18], allows time for surgical intervention in 25% of patients [4, 5, 7]. Such sequence of events was present in our Case 2. Arteriovenous fistula formation is a rare complication that leads to portal hypertension [2, 8]. Rarely, the high flow through a splenic arteriovenous fistula causes small-bowel ischemia, the “mesenteric steal syndrome” [19]. Less common complications of SAAs include splenic artery thrombosis, splenic infarction and gastro-intestinal bleeding from erosion and rupture into the bowel lumen [4, 5, 7, 8].

Elective treatment is indicated in patients with SAA >2 cm in diameter, in pregnant women or women of childbearing age with lesions >1 cm, and in symptomatic patients with a lesion of any size. The patient in Case 1 fulfilled several of the abovementioned criteria for (semi) elective treatment (considering the presenting symptoms). An aneurysm between 1 and 2 cm in diameter should be monitored closely with imaging studies every 6 months and treated if enlargement is documented or symptoms ensue [4, 5, 7, 8]. The most appropriate and widely used imaging study for SAA initial diagnosis and follow-up is a duplex ultrasonography. For purposes of surgical or interventional treatment, it is supplemented by computed tomography, magnetic resonance or conventional angiography. Ultrasonography has the advantage of being non-invasive and radiation-free (of particular importance in pregnancy), but,
on the other hand, it is operator-dependant and of limited sensitivity in the presence of small SAAs, obesity and/or intestinal gas [5]. The treatment options for SAAs range from conventional open surgery to minimally invasive laparoscopic ligation or repair and various endovascular techniques using covered stents or embolization materials [4, 5, 7, 8, 20, 21]. Anatomic features and patient selection determine which treatment option would result in the most successful and durable outcome. Until recently, open aneurysmectomy, with or without splenectomy, was recognized to be the best treatment for SAAs. The spleen, which has a good collateral vascular supply via the short gastric arteries, was preserved when possible, preventing potential post splenectomy sepsis. Aneurysms of the proximal splenic artery can be treated with simple ligation [22] but those involving the hilum require splenectomy. Splenectomy is also usually performed for ruptured SAAs [4, 5, 7]. Lesions of the medial third are often adherent to the pancreas; therefore, exclusion by proximal and distal ligation is a safer alternative. Aneurysmectomy with end-to-end anastomosis may be possible for some mid-SAAs in a tortuous and redundant artery [23]. The surgical approach for giant SAAs is challenging because even the celiac trunk may be inaccessible anteriorly due to the size of the mass. Considerable size of the aneurysm in our Case 2 necessitated proximal bleeding control (clamping of the aorta) at the diaphragm level. Long et al. [24] have used a thoracoabdominal incision to gain retroperitoneal access to the subdiaphragmatic aorta and the celiac trunk. In a similar situation, Pescarus et al. [15] used retroperitoneal approach with a chevron incision and medial visceral rotation. Mortality after emergency surgery is as high as 40%, compared with negligible mortality after elective SAA repair [4, 5, 7, 12].

During the past decade, endovascular techniques including transcatheter embolization and stent graft placement were introduced as alternatives to conventional surgery [8, 19, 25, 26]. These techniques can be used to treat most SAAs, except those located at the splenic hilum, especially when the site is difficult to access during conventional surgery or when the operative risk is high [20]. Endovascular treatment is accompanied by high rate of technical success and less morbidity than laparotomy and in many vascular centers is now considered the first choice therapy [5, 8, 19, 25, 26]. Surgical repair is preferred for all symptomatic aneurysms because of the greater likelihood of success. In the presence of portal hypertension, transcatheter embolization or stent-graft placement may be preferred because the extensive collateral circulation that develops as a result of portal hypertension makes surgery more difficult [8, 20, 25, 26]. Tortuosity of the splenic artery, which is frequently present, may preclude stent graft deployment. Giant SAAs may also be unsuitable for endovascular treatment [20, 26]. Transcatheter embolization of the splenic artery pseudoaneurysm is gaining popularity; however, failure does occur, especially when the pseudoaneurysm is associated with a pseudocyst of the pancreas [9, 25, 20, 27]. Splenectomy, with or without distal pancreatectomy is still the standard of treatment. Potential complications of coil embolization include splenic infarction, abscess formation, and higher rates of recurrence [20, 27].

SAAs are clinically significant and increasingly diagnosed vascular lesions, affecting different age groups, with variety of etiologies and clinical manifestations, important to recognize because of the risk for rupture and associated mortality. The diversity of SAAs is matched by the spectrum of treatment options available. Timely diagnosis and individualized treatment approach prevent life-threatening rupture and lessen the risk of operative morbidity and mortality.

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