A rare cause of acute abdomen: spontaneous dissection of the superior mesenteric artery

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Key Clinical Message
Spontaneous superior mesenteric artery (SMA) dissection is a rare, but potentially fatal disease. Prompt diagnosis and treatment of SMA dissections result in a lower prevalence of intestinal infarction and mortality. In the current era, imaging techniques can promptly diagnose SMA dissection; however, no definitive guidelines have been established to treat this condition.

Keywords
Acute abdomen, anticoagulation, CT angiogram, superior mesenteric artery dissection.

Introduction
Spontaneous superior mesenteric artery (SMA) dissection is a rare disease. In literature, only 168 cases have been reported since 1947 [1]. A study was conducted on 6666 autopsies, which showed an incidence rate of 0.06% [1, 2]. This condition is more prevalent than reported in literature, undetected due to previous limitations in imaging techniques and asymptomatic carriers. Prompt diagnosis and treatment of SMA dissections result in lower incidence of intestinal infarction and a lower mortality rate. In the current era, imaging techniques such as multidetector row computed tomography, multiplanar reconstruction imaging (MPR), and computerized tomography angiography can easily diagnose SMA dissection [2, 3]. The current therapeutic approaches are as follows: conservative management with anticoagulants, endovascular repair with stent placement, and open surgical repair with bypass grafts [2, 3]. Although we are able to treat this condition, no definitive guidelines have been established on how to do so. We present a case of a 57-year-old male with a history of type 2 diabetes mellitus that presented with abdominal pain, nausea, vomiting, and diarrhea originally suspected to have small bowel obstruction based on abdominal X-ray findings. CTA abdomen/pelvis detected a spontaneous dissection of the SMA, without any involvement of the aorta.

Case Presentation
A 58-year-old Hispanic male with a past medical history of type 2 diabetes mellitus presented to the emergency room with the chief complaint of abdominal pain for two days. He reported that the abdominal pain was present in the epigastric area, with no radiation. It was 7/10 on a pain scale and colicky in nature. The patient also complained of nausea, vomiting, and diarrhea. He had four watery bowel movements, without mucus or blood, and vomited four times, which had no blood or bile. His symptoms were aggravated by eating and relieved by fasting. The patient denied any fever, chills, and rigors. He reported that he lost eight pounds in the last one month because he thought he was overweight so he went on a diet of reduced portions of his regular meals. He drank socially, and his last drink was two weeks from the date of admission. He denied tobacco and illicit drug use. The
patient had two episodes of similar symptoms last month. He came to the hospital twice, was given intravenous fluids, felt better, and was discharged within a few hours. At the time of each discharge, he was not told by the emergency room physician his diagnosis. Family history was noncontributory. His vital signs were blood pressure 120/84, pulse rate 73/min, respiratory rate 18/min, and temperature 97.8°F, and oxygen saturation was 96% on room air. An abdominal X-ray obstruction series (Fig. 1) revealed multiple dilated loops of the proximal small bowel and paucity of bowel gas seen within the colon. The findings represented either a partial or early complete small bowel obstruction. Computerized tomography of the abdomen and pelvis, with contrast, (Fig. 2) revealed mild dilation of the thickened bowel, suggestive of a mild ileus pattern without findings of bowel obstruction. There was focal dissection of the superior mesenteric artery. There appeared to be contrast opacification of both the true and the false lumens. There was also contrast opacification of the superior SMA distal to the site of dissection. The SMA at the site of dissection was aneurysmal, measuring up to approximately 1.2 cm in diameter. Computerized tomography angiogram of the abdomen and pelvis (Fig. 3) demonstrated an approximate 2.5 cm dissection shortly beyond the origin of the SMA. Beyond this, the vessels were widely patent with no evidence of compromise of flow to the SMA. The abdominal aorta was normal in caliber. The celiac axis appeared somewhat lobulated in its appearance without evidence of obstruction or dissection. The inferior mesenteric artery was unremarkable. There was no evidence of bowel obstruction. He was started on unfractionated heparin (5000 units/kg bolus and 18 units/kg drip) and bridged to oral warfarin (10 mg on day 1 and day 2, 5 mg on day 3). Our target therapeutic international normalized ratio (INR) was 2–3 and we achieved an INR of 2.3 after 3 days of warfarin therapy. After 6 days, the patient’s symptoms resolved and he was discharged on 5 mg warfarin daily and 81 mg aspirin daily, and advised to follow up as an outpatient with the primary care doctor in 3 days to check his INR and the vascular surgeon in 3 months.

Discussion

Spontaneous dissection of the SMA is a rare, but potentially fatal disease. It is the second most frequent site of isolated spontaneous peripheral arterial dissection after the
Statistically, it is seen mostly in male patients (88% of cases), with an average age of 55 years [6]. It was first described by Bauerfield in 1947 in an 87-year-old female with bowel infarction due to dissection of the SMA [3, 5]. Initially, the prognosis of patients with spontaneous dissection of the SMA was very poor, but modern advancements in imaging, medicine, and surgical techniques have improved the outcome [3, 4, 6].

The etiology of spontaneous SMA dissection has not yet been established, but has been associated with atherosclerosis, cystic medial necrosis, fibromuscular dysplasia, connective tissue disorders, trauma, elastic tissue disorders (Marfan and Ehlers–Danlos syndrome), abnormal curvature of the origin of the SMA, and uncontrolled hypertension [1–3, 5–7]. It has been hypothesized by Solis et al. that the dissection usually begins 1.5–3 cm from the orifice of the SMA, thus sparing the origin of the artery [3, 5]. This hypothesis is based on the fact that this segment of the artery is exposed to high shearing forces because of its anatomical location [3, 5, 7]. It exits the pancreas at its border between the fixed retropancreatic portion and the more distal mobile mesenteric portion [3, 5, 7]. On the microscopic level, there is fragmentation of the elastic fibers, loss of and dispersement of smooth muscle cells, degradation of the internal elastic lamina, areas of cystic degeneration, and atheromatous changes in the arterial wall [5, 6].

The natural history of the disease varies from case to case: (i) no progression or limited progression of the dissection, with false lumen thrombosis; (ii) progressive dissection toward the distal branches of the SMA; (iii) expansion of the false lumen, causing narrowing or obliteration of the true lumen and potentially leading to ischemia and intestinal necrosis; and (iv) artery rupture through the adventitia resulting in hemorrhage [2].

The presentation of the disease is unclear. Most patients will present with acute epigastric pain within 4 weeks of onset of dissection [3, 5]. The pain is caused by inflammation around the dissecting SMA and intestinal ischemia. The pain may be initiated or exacerbated after meals when there is an increased oxygen demand by the intestines [4, 5, 7]. Other common symptoms are nausea, vomiting, diarrhea, melena, and abdominal distension [3, 7]. The mechanism of the acute symptoms after meals is caused by a sudden compression of the true lumen by a rapidly filling false lumen [5]. Laboratory work and plain abdominal radiographs are usually unremarkable, which misleads physicians, to misdiagnoses such as enterocolitis or intestinal obstruction. The patients are kept nil per oral and given intravenous fluids. Their symptoms abate, which prompts physicians to discharge them. Soon thereafter, they return with the same symptoms. This cycle often ensues given the rarity of this condition.

Diagnosis in the acute setting has become possible because of the advances in modern imaging techniques [2–4]. Plain CT shows area of high intensity if there is an acute clot in the false lumen. CTA shows the separated true lumen from the false lumen by identifying the intimal flap. MDCT and MPR although rarely used can be used to create three-dimensional images of the dissection. Abdominal Doppler ultrasound can show the intimal flap and aneurysmal dilation of the SMA. Sakamoto et al. have categorized SMA dissection into four types based on CTA findings: Type 1: patent true and false lumina that show entry and reentry sites; Type 2: patent true lumen but no reentry flow from the false lumen; (A): visible false lumen but no visibly reentry site (blind pouch of lumen); (B): no visible false luminal flow (thrombosed false lumen); and Type 3: SMA dissection with occlusion of SMA [2–4]. However, no one has been able to establish a clear relation between radiological appearance and clinical course.

There are some general principles to treat spontaneous SMA dissection: Conservative management with
anticoagulation therapy inhibits the false lumen from thrombosing and spreading into the SMA [2–4]. These patients need close follow-up to monitor progression of the dissection; however, there is no guideline on how often [2, 3]. There is no consensus on the best medication for anticoagulation. Sparks et al. have suggested that indications for surgery are increasing size of the aneurysmal dilation of the SMA, luminal thrombosis, signs of intestinal ischemia, or persistent symptoms despite anticoagulation [1–3, 6]. Various procedures for surgical intervention have been reported [2, 3]. Minimally invasive techniques include percutaneous endovascular stent placement and intraluminal thrombolytic therapy for patients who are at high risk for surgery [2, 3]. The stent covers the intimal detachment and prevents blood flow through the false lumen [2]. Although this can cause thrombosis of the false lumen due to stasis of blood, the stent keeps the true lumen patent [2]. Problems with this modality of treatment are that it is often difficult to find the site at which tearing of the artery wall starts during dissection of the SMA. Also stents are at risk for reoclusion and possible occlusion of side branches [2, 3]. Intraluminal thrombolytic therapy with urokinase may be useful, but the false lumen often reoccludes [2, 3]. Open surgical techniques used are endoaneurysmorrhaphy, aortomesenteric or iliomesenteric bypass with a vein or prosthesis, thrombectomy, endarterectomy with or without patch angioplasty, ligation, and resection [2, 3, 6]. These techniques have had good short-term results, but not enough data are to establish their long-term efficacy [2, 3].

An algorithm for treatment based on three clinical presentations has been proposed by Buch et al. The first presentation is the asymptomatic patient whose dissection is an incidental finding. The second presentation is a patient with acute transient pain or chronic relapsing pain (noncontinuous pain), and the third presentation is a patient with acute ongoing pain (continuous pain) [4]. In the first type of patient, anticoagulation is suggested with close follow-up with serial CT angiography because an asymptomatic progressive dissection or thrombosis of the true lumen may occur. There is no recommendation for the interval between the scans. For the second type of patient, a stent is recommended, with CT angiography follow-up, with no recommendation on how often [4]. For the third type of patient presentation, surgery is necessary as the dissection has usually spread and is unamenable to more conservative measures [4].

Conclusion
Isolated SMA dissection is a potentially fatal condition, if not diagnosed and treated promptly. Although historically rare, it is imperative that spontaneous SMA dissection be included as a cause of acute abdomen. There are only suggestions on how to treat isolated SMA dissection based on results from individual cases, but there are no established guidelines. Asymptomatic patients can be treated with anticoagulation if there are no ominous signs or symptoms. If the patient has noncontinuous pain, then minimally invasive techniques are necessary. If continuous pain is present, open surgical techniques may come into play. Since this is a rare phenomenon, we believe that data regarding the patients, treatment, and the outcome should be compiled into a central database. Perhaps in the future when there is sufficient research, proper guidelines can be established for the management.

Conflict of Interest
None declared.

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