Isolated dissection of the superior mesenteric artery: a case report and literature review

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ABSTRACT

Isolated dissection of the superior mesenteric artery is rare; it is predominantly observed in men with the highest incidence in those over 40 years old. Contrast-enhanced multi-detector computed tomography is considered essential for the diagnosis, therapeutic management and follow-up. The therapeutic approach ranges from conservative medical treatment to surgery or endovascular stent placement, but there are, to date, no approved guidelines. We report the case of a 68-year old man who entered our Emergency Department just for mild abdominal pain, which later proved to be due to acute dissection of the superior mesenteric artery.

Introduction

Superior mesenteric artery (SMA) dissection is a rare condition, especially if isolated. Given the lack of clinical information on this condition, the therapeutic approach is, to date, far from being standardized. Despite its rarity, SMA dissection may be one of the most important causes of acute intestinal ischemia.1,2 The clinical features of SMA dissection can be extremely variable, ranging from a completely asymptomatic condition, up to forms characterized by real acute abdomen. No laboratory tests help the diagnosis, while the more modern imaging techniques, in particular, contrast-enhanced multidetector computed tomography (MDCT), grant high reliability for a definite diagnosis.3

Here we describe the case of a 68-year-old man who arrived at the Emergency Department of Giovanni Paolo II Hospital of Sciacca (AG) complaining only of mild abdominal pain, which later proved, according to radiological findings, to be acute SMA dissection.

Case Report

A 68-year old man was admitted to the Emergency Department of Giovanni Paolo II Hospital of Sciacca (AG) in August 2011 because of the sudden onset of mild abdominal pain. He reported a medical history of hypertension, under treatment with candesartan, irritable bowel syndrome and recurrent episodes of acute diverticulitis, being treated with periodic cycles of rifaximin; he also reported smoking about 15 cigarettes/day for the previous 30 years.

On physical examination the patient was restless, with a respiratory rate of 70 breaths per minute, a heart rate of 80 beats per minute in sinus rhythm, high blood pressure (160/100 mmHg), diffuse abdominal mild pain (stronger in the right iliac fossa and left hip) and torpid peristalsis, without signs of peritonitis (Blumberg’s sign negative). He underwent routine examinations, which were within normal range, except for the white blood cell count (11.6×10^9/L; reference range, 4.3 to 10.8×10^9/L) and C-reactive protein level (2.60 mg/L; reference range <0.5 mg/L). The chest X-ray was normal, whereas abdominal radiographs revealed small air-fluid levels in lower abdominal quadrants. Urinary tract ultrasound examination showed the presence of cysts in both kidneys (the largest in the right kidney, of about 1.5 cm, the homologous one in the left kidney, of about 9 cm). Finally contrast-enhanced MDCT of the abdomen
was required because of suspected gastrointestinal (cecal) neoplasm. The contrast agent used for intravenous injection was iodixanol, a water-soluble non-ionic contrast medium (containing 320 mg of iodine per milliliter). An 18-gauge needle with cannula was used to administered 120 mL of iodixanol intravenously, followed by 40 mL of normal saline, using a dual headed pressure injector at a flow rate of 3.5 mL/sec. Arterial and venous phase images were obtained using the bolus tracking technique, with the region of interest in the abdominal aorta at the level of the celiac artery (threshold=150 HU). Finally, sagittal, coronal and curved multiplanar reconstructions were made. MDCT showed that the diameter of the SMA was enlarged (12 mm) and identified the vessel’s true and false lumina by the presence of an intimal flap just 15 mm after the origin of the SMA (Figures 1 and 2). The dissection extended anterogradely for 46 mm (Figure 3) and a modest amount of fluid at the origin of SMA was detected. There were no signs of bowel ischemia or ascites.

The patient started therapy with enoxaparin 6000 IU once daily; after 1 month of this therapy, the MDCT showed no meaningful changes, with the exception of reabsorption of the previously reported modest amount of fluid at the origin of the SMA.

Discussion

Isolated dissection of the SMA dissection is an extremely rare condition (Table 1). Bauersfeld et al. documented the first case in 1947. To date, fewer than 100 cases have been reported. Patients with SMA dissection are predominantly men (88% of cases) and over 40 years old (range, 45-87 years). The most frequently reported risk factors include hypertension, atherosclerosis, fibromuscular dysplasia, cystic medial necrosis, and abdominal aortic aneurysm (Table 1); however, some dissections appear to occur spontaneously, without any identifiable etiology.

Solis et al. hypothesized that dissection usually begins 1.5-3 cm from the orifice of the SMA, thus sparing the origin of the artery. This segment of SMA corresponds with the exit of the artery from the pancreas, exactly where it is exposed to shearing forces, because this area represents the border zone between the fixed retropancreatic portion and the more mobile distal mesenteric one.

The natural history of the disease is unclear and differs from case to case. Most patients present with acute epigastric pain, probably due to the dissection itself or subsequent intestinal ischemia, sometimes associated with nausea, vomiting, abdominal distention, and melena. In some cases, patients present with chronic abdominal pain, sometimes related to food intake. Furthermore, patients may, occasionally, be completely asymptomatic.
Acute signs of intestinal ischemia or bleeding are poor prognostic features. Complications of SMA dissection include bowel infarction, intra-abdominal hemorrhage, acute peritonitis, shock, and late complications, such as uremia. Laboratory tests and abdominal radiography are usually unremarkable. Sometimes, laboratory tests show slightly elevated serum amylase levels, due to occlusion of duodeno-pancreatic arcades. Abdominal echo color Doppler can be useful for demonstrating and following up hemodynamic changes within the SMA, bowel movements, and signs of bowel ischemia, such as wall thickening and dilated intestinal loops. Angiography is currently the gold standard imaging technique for diagnosing SMA dissection. Recently, contrast-enhanced computed tomography (CT) has become the most reliable diagnostic modality; indeed, diagnosis in the acute stage has become possible as a result of advanced and increasing use of CT imaging techniques, such as MDCT, leading to multiplanar reconstruction and reconstruction imaging, and computed tomography angiography (CTA). MDCT usually shows that the diameter of the SMA is enlarged and that there is increased attenuation of the fat surrounding the artery itself, together with images of true and false lumina, which can be identified by the presence of an intimal flap after the origin of the SMA (Table 2). In our case, MDCT provided enough information to diagnose spontaneous SMA dissection, and angiography was not necessary. This technique was also useful for our patient’s follow-up. Sakamoto et al. categorized SMA dissection into four types on contrast-enhanced CT scanning. Recently, Yun et al. devised a new classification based on angiographic findings: in this classification, there are three types, with two subtypes (Table 3). However, neither Sakamoto et al. nor Yun et al. found a clear relationship between radiological appearance and clinical course. The differential diagnosis of SMA includes acute SMA embolism, acute SMA thrombosis, non-occlusive mesenteric ischemia, mesenteric venous thrombosis, and aortic dissection (Table 2).

The differential diagnosis of SMA includes acute SMA embolism, acute SMA thrombosis, non-occlusive mesenteric ischemia, mesenteric venous thrombosis, and aortic dissection (Table 2). Acute SMA embolism accounts for approximately 40-50% of all episodes of acute mesenteric ischemia.

Table 1. Summary of superior mesenteric artery dissection.

<table>
<thead>
<tr>
<th>Etiology</th>
<th>According to the hypothesis proposed by Solis et al., the dissection is thought to be caused by stress on the wall of the artery at the inferior pancreatic edge</th>
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<tr>
<td>Incidence</td>
<td>Fewer than 100 cases have been reported in the literature since the first case described by Bauerfeld in 1947</td>
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<td>Gender predilection</td>
<td>Patients are predominantly male (88% of cases)</td>
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<td>Age predilection</td>
<td>Range: 45-87 years old</td>
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<td>Risk factors</td>
<td>Hypertension, atherosclerosis, fibromuscular dysplasia, mycotic infections, trauma, connective tissue disorders, cystic medial necrosis, abdominal aortic aneurysm, vasculitides such as giant cell arteritis, Takayasu’s arteritis, polyarteritis nodosa and iatrogenic dissections due to endovascular interventions; however, some dissections occur spontaneously without any identifiable etiology</td>
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<td>Treatment</td>
<td>Patients should initially receive conservative treatment (anticoagulation, CT follow-up) when the mesenteric blood supply to intestines is not severely compromised by the dissection and the patients have no peritoneal signs. Patients who have recurrent symptoms after conservative treatment should undergo endovascular revascularization if they have no peritonitis, or surgical intervention if peritonitis develops</td>
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<td>Prognosis</td>
<td>The prognosis depends on the extent of involvement of sub-segmental branches. Acute signs of bleeding or ischemia are poor prognostic features</td>
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<td>Findings on imaging</td>
<td>MDCT shows an enlarged SMA diameter, increased attenuation of the fat around the SMA, true and false lumina can be identified by the presence of an intimal flap after the origin of the SMA</td>
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CT, computed tomography; MDCT, multidetector computed tomography; SMA, superior mesenteric artery.
### Table 2. Differential diagnosis of superior mesenteric artery dissection.

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<th>CT</th>
<th>Angiography</th>
<th>Clinical findings</th>
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<tr>
<td>SMA dissection</td>
<td>MDCT shows an enlarged SMA diameter, increased attenuation of the fat around the SMA; true and false lumina can be identified by the presence of an intimal flap after the artery origin</td>
<td>The arteriogram demonstrates a narrowed true lumen and an aneurysmal dilated false lumen. The arteriograms can reveal the intimal flap and a reduced blood flow</td>
<td>The most common symptom is acute epigastric pain (caused by the dissection itself or intestinal ischemia) sometimes associated with nausea, vomiting, melena, and abdominal distention. In some cases, the patient has chronic abdominal pain. Additionally, patients may be asymptomatic</td>
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<td>Acute SMA embolism</td>
<td>CTA can show the acute occlusions as filling defects in the vessel lumen</td>
<td>The angiographic hallmark is abrupt termination of the vessel (cutoff sign)</td>
<td>Most emboli in the SMA lodge just beyond the origin of the middle colic artery</td>
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<td>No or only very few collateral vessels are present</td>
<td>Acute SMA embolism is often characterized by multiple occlusions</td>
<td>Acute embolism is often associated with atrial fibrillation</td>
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<td>In patients with prior embolic events, re-canalized vessels may be seen</td>
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<td>Acute mesenteric artery thrombosis</td>
<td>Filling defect in the vessel lumen, more proximal than embolic occlusion</td>
<td>Angiography can show the acute occlusion as a filling defect in the SMA, more proximal than embolic occlusion</td>
<td>Typically associated with a pre-existing atherosclerotic lesion. In up to 50% of cases, a history of intestinal angina is present. The abdominal pain and other symptoms may be more insidious than those of embolic occlusion because of the development of a collateral circulation</td>
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<td>CTA can show these findings in addition to visualizing collateral vessels</td>
<td>Collateral vessels may be present</td>
<td>The SMA is typically occluded within the first 2 cm from its origin, in contrast to acute embolic occlusions, which occur more distally</td>
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<td>Non-occlusive mesenteric ischemia</td>
<td>The bowel wall of the involved segments may be normal or thickened. Enhancement is variable: absent or diminished enhancement, increased enhancement, or halo or target-type enhancement. Fat stranding of the mesentery and ascites are visible</td>
<td>Angiography does not show vascular obstruction, but diffuse splanchnic vasospasm, narrowing of SMA branches, low flow and poor distal filling</td>
<td>Non-occlusive mesenteric ischemia usually develops during an episode of cardiogenic shock or a state of hypoperfusion in which excessive sympathetic activity results in secondary vasoconstriction of the mesenteric arteries</td>
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<td>Mesenteric venous thrombosis</td>
<td>Persistent, well-defined intraluminal filling defects with central low attenuation, which may be surrounded by well-defined, rim-enhancing venous walls</td>
<td>Mesenteric arteriography with delayed venous phase imaging may demonstrate venous occlusion or an intraluminal thrombus</td>
<td>Associated risk factors are portal hypertension, hypercoagulation, trauma, intra-abdominal inflammatory diseases and recent surgery. Many cases are idiopathic. Acute mesenteric ischemia develops when mesenteric vein thrombosis is associated with a lack of an adequate venous collateral circulation</td>
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<td>Accompanying collateral circulation, engorgement of mesenteric veins, and mesenteric edema may be present</td>
<td>Superior mesenteric arterial spasm is frequently observed, although prolongation of the arterial angiographic phase and other subtle findings may also result from superior mesenteric venous occlusion</td>
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<td>Bowel wall thickening</td>
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<td>Target sign resulting from submucosal edema or hemorrhage</td>
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<td>The thickened bowel wall may appear to be hyperattenuated because of intramural venous engorgement</td>
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<td>Aortic dissection</td>
<td>CTA is an excellent technique for clarifying the dissection, defining entry and re-entry points, differentiating thrombus from slow flow, and evaluating the involvement of branch vessels</td>
<td>The arteriogram demonstrates a narrowed true lumen and an aneurysmal dilated false lumen, the intimal flap and reduced blood flow</td>
<td>The main symptom is the pain</td>
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<td>The most common classification systems of aortic dissection are the Standford (types A and B) and DeBakey (types I, II, III) systems</td>
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CT, computed tomography; SMA, superior mesenteric artery; MDCT, multidetector computed tomography; CTA, CT angiography.
and is often associated with atrial fibrillation. Most SMA emboli lodge just beyond the origin of the middle colic artery. However, acute SMA embolism is often characterized by multiple occlusions. It usually presents with acute and massive abdominal pain, sometimes morphine-resistant, together with vomiting (71%), diarrhea (42%) and hematochezia (21%). The angiographic and CTA hallmark of an embolic occlusion is the abrupt termination of the vessel (cutoff sign) (Table 1).28

Acute superior mesenteric artery thrombosis is typically associated with a preexisting atherosclerotic lesion and it has been estimated to be responsible for 20-30% of all cases of acute mesenteric ischemia. In up to 50% of cases, a history of intestinal angina is present. In contrast to the abrupt, catastrophic onset of symptoms associated with acute SMA embolism, abdominal pain and symptoms associated with acute mesenteric artery thrombosis may be more insidious, because of the development of collateral circulation. Usually this condition is associated with chronic pain spreading after meals, also known as angina abdominis. Angiography and CTA might show occlusion as a filling defect in the SMA, more proximally than in cases of embolism (Table 1).29

Non-occlusive mesenteric ischemia seems to be responsible for approximately 25% of cases of acute mesenteric ischemia, and the mortality rate associated with this condition has been reported to be as high as 70%. It usually develops during cardiogenic shock or hypoperfusion states, in which excessive sympathetic activity results in secondary vasoconstriction of the mesenteric arteries. Hypoperfusion results in increased vascular permeability, which leads to extravascular leakage of plasma, red blood cells, or both into the bowel wall, the mesentery, and the peritoneal cavity. Shock bowel is a variety of non-occlusive mesenteric ischemia, caused by hypotensive shock induced by blunt abdominal trauma. Ischemic colitis and obstructive colitis may be considered similar clinical entities. No defined symptoms or signs can be attributed to non-occlusive mesenteric ischemia. There may be absolutely no early symptoms or symptoms ranging from abdominal discomfort to restlessness. Abdominal pain (reported to be absent in 23% of cases), bloody stools and progressive reduction of consciousness may then appear, leading, finally, to a picture of acute abdomen.30,31 Angiography does not show vascular obstruction, but diffuse splanchnic vasospasm (Table 1). There are few published reports of CTA findings, which usually showed normal mesenteric vessels (both arteries and veins), associated with bowel wall thickening and pneumatosis (i.e. gas in the bowel wall). However, among several forms of mesenteric ischemia, non-occlusive mesenteric ischemia is the most difficult condition to diagnose on CT, and angiography is often required for correct and confident diagnosis.32,33

Mesenteric vein thrombosis accounts for 5-15% of all cases of acute mesenteric ischemia. The most common associated risk factors include portal hypertension, coagulation disorders, and trauma (Table 1). However, many cases are idiopathic. Subsequent acute mesenteric ischemia might develop when mesenteric vein thrombosis is associated with a lack of an adequate venous collateral circulation, which results in the development of intestinal mucosal edema and arterial hypoperfusion. Clinically, at least three different pictures can be differentiated: acute, subacute and chronic. The acute picture usually involves spreading pain, hematochezia and reduction of peristalsis, leading to acute abdomen and shock. Subacute mesenteric vein thrombosis produces the same picture as the acute one, except for a smoother onset. In contrast, the chronic picture may be associated with non-specific symptoms or signs and, sometimes, can even be completely asymptomatic.34 Angiography, with delayed venous phase imaging, may demonstrate venous occlusion or an intraluminal thrombus. CT of mesenteric vein thrombosis shows persistent, well-defined intraluminal filling defects, with central low attenuation (Table 1). Diagnostic angiography is usually reserved for cases in which clinically suspected porto-mesenteric venous thrombosis cannot be established by non-invasive techniques.35

Approximately 5% of patients with aortic dissection develop acute mesenteric ischemia as a complication of the dissection process. Isolated dissections of the visceral arteries usually occur in association with cystic degeneration or as a complication of catheter angiography and are extremely rare. Tearing and ripping pain, which migrates in the direction of dissection, is the most important symptom of aortic

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<th>Table 3. Superior mesenteric artery angiographic classification.</th>
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<td>Type I</td>
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<td>Type II</td>
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<td>Type III</td>
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SMA, superior mesenteric artery. Modified from Yun et al., 2009.19
dissection, associated, usually, with nausea, sweating, shortness of breath, weakness and, sometimes, loss of consciousness. Several others organ-specific signs and symptoms may be connected with occlusion of the arteries which arise from the aorta. Arteriography demonstrates a narrowed true lumen, an aneurysmal dilated false lumen, an intimal flap and reduced blood flow. CTA is an excellent technique for assessing these patients, showing the dissection, defining entry and re-entry points, differentiating thrombus from slow flow, and evaluating branch vessel involvement.

In the context of dissection of the SMA, acute bowel infarction is very rare, and most cases have slow progression to chronic ischemia. The most common CT manifestation of accompanying bowel ischemia is bowel wall thickening. This last, non-specific, finding may be identified from the target sign, with alternating intramural areas of high and low attenuation, resulting from submucosal edema or hemorrhage. The thickened bowel wall may appear to be hyperattenuated: this is assumed to be due to intramural venous engorgement. Other important CT findings include bowel enlargement and, less commonly, intestinal pneumatosis. These last two findings are more specific signs of ischemic bowel disease.

Some treatment algorithms for the management of spontaneous SMA dissection have been reported in literature, but, to date, there is no established, unanimous opinion about conservative medical management, endovascular therapy, or surgical revascularization. Some cases have been successfully treated by conservative therapy, such as anticoagulant drugs, but there is not enough experience to demonstrate that the efficacy and safety of this approach are superior to those of other approaches. However, most of the reports agree about the following therapeutic strategies: i) patients without imminent vessel rupture, symptoms of bowel ischemia or peritoneal signs: conservative treatment; ii) patients with an increased risk of perioperative complications, without severe mesenteric ischemia or peritonitis: percutaneous endovascular stent placement; iii) luminal thrombosis, increasing size of SMA aneurysmal dilatation, peritoneal signs, or persistent symptoms despite anticoagulation: open surgery (aorto-mesenteric or ilio-mesenteric bypass, thrombectomy, intimaectomy, with or without patch angioplasty, ligation and resection). Conservative managements mainly include anticoagulation and blood pressure control. Nevertheless, all studies indicate that a non-operative approach with anticoagulant drugs often does not prevent disease progression, so close follow-up is strongly recommended. Early minimally invasive techniques, such as percutaneous endovascular stent placement, are indicated in cases with short segment dissection without bowel ischemia or signs of peritonitis and in patients not improving on conservative therapy and who have an increased risk of perioperative complications. It is an efficient and less aggressive approach, which provides excellent immediate results and may become a valuable option for mesenteric revascularization in the near future. Finally, surgical methods, such as bypass with venous grafts and thrombectomy, are indicated in all cases with signs of bowel ischemia and perforation; they have been performed with good short-term results.

Conclusions

Isolated SMA dissection is a rare pathology, but it should be considered in the differential diagnosis of abdominal angina and acute abdomen. The clinician’s suspicion can be confirmed by CT studies. MDCT shows that the diameter of the SMA is enlarged SMA and that there is increased attenuation of the fat around the SMA; it may also demonstrate a true lumen and false lumen by the presence of an intimal flap after the origin of the SMA. CT studies, i.e. MDCT, are essential not only for the diagnosis, but also for the therapeutic management and follow-up. There is no consensus on the best treatment of spontaneous isolated dissection of the SMA, and further information is required to establish optimal management guidelines for the disease. However, at present, patients should initially receive conservative treatment when the mesenteric blood supply to the intestines is not severely compromised by the dissection and the patients have no signs of peritonitis, as in our case report. When patients have recurrent symptoms after conservative treatment and no signs of peritonitis, endovascular repair for SMA revascularization is indicated, while surgical intervention is indicated when peritonitis occurs.

References