Three isolated superior mesenteric artery dissections: Update of previous case reports, diagnostics, and treatment options

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Isolated superior mesenteric artery dissection is a relatively rare vascular pathology. However, the number of recent case reports has shown an increasing incidence with the widespread use of computed tomography imaging for abdominal pain. Here we report three cases of isolated superior mesenteric artery dissection. The unique surgical option of small bowel transplantation along with successful medical management is described. A treatment algorithm for isolated superior mesenteric artery dissection is also proposed. (J Vasc Surg 2008;47:649-53.)

Isolated superior mesenteric artery (SMA) dissection without associated aortic dissection is rare. Causes of SMA dissection remain elusive, possibly including cystic medial necrosis, congenital connective tissue disorders, fibromuscular dysplasia, arteriosclerosis, and trauma.[1] No cases of iatrogenic causes have been reported. SMA dissection typically begins 1.5 to 3 cm distal to the origin of the SMA, with differing clinical presentations according to the extent of dissection.[2]

Before 1972, the 11 reports of isolated SMA dissections resulted in death, and the diagnosis was made at the time of autopsy.[3-10] By contrast, since 1975, survival was achieved in all but one reported case by using a variety of treatment options.[11-48] The purpose of this article is to describe three isolated SMA dissections and review the literature for diagnosis and treatment on this unique and relatively rare disease. Currently, there are no established medical or surgical treatment standards.

CASE REPORTS

Patient 1. A 39-year-old woman presented to the emergency department (ED) of Spectrum Health Hospital (affiliated with Grand Rapids Medical Education and Research Center) with a 48-hour history of periumbilical and right lower quadrant pain that developed along with associated symptoms of nausea, vomiting, and diarrhea. Physical examination revealed tenderness in the right upper quadrant without rebound or peritoneal signs. Her medical history included chronic pain secondary to cephalgia and endometriosis, along with asthma, hypothyroidism, and restless leg syndrome.

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Competition of interest: none.

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Results of basic metabolic panel and liver function tests were normal. Stable anemia with a hemoglobin level of 10.7 g/dL was seen on complete blood count. A computed tomography (CT) scan of the abdomen and pelvis showed a dilated duodenum with possible SMA syndrome (nutcracker syndrome) or dissection.

The patient was placed on bowel rest, narcotics for pain control, and intravenous hydration. On hospital day 1 an esophagogastroduodenoscopy (EGD) showed a dilated duodenum but was not suggestive of SMA syndrome. An angiography later on that day showed a markedly abnormal SMA consistent with dissection. The dissection occurred 1.8 cm from the SMA origin, with a false lumen extending into the ileocolic artery and severe focal stenosis (Fig 1). Anticoagulation with heparin was initiated, and the abdominal pain improved. Owing to the extent of dissection to the tertiary branches, the SMA could not be revascularized.

The patient was hospitalized for 15 days at Spectrum Health Hospital. Total parenteral nutrition, narcotic pain control for ongoing abdominal pain, and anticoagulation were used. Repeat imaging, laboratory studies, and serial abdominal examinations did not produce findings of any critical bowel ischemia necessitating immediate resection.

On hospital day 15, the patient was transferred to an outlying university transplant facility for evaluation. One week after transfer, the patient underwent a total enterectomy and right hemicolectomy at the University of Pittsburgh Hospital. The patient was discharged home with total parenteral nutrition after an uneventful stay.

Her next 4 months included three hospital admissions and five ED visits to our facility with multiple complications. She eventually underwent successful small bowel transplantation 1 year after her initial dissection event. Five serial EGDs performed in the year after transplantation showed normal small bowel mucosa, without evidence of rejection. Unfortunately, 22 months after her initial dissection and 10 months after her transplant, the patient attempted suicide with numerous prescription medications. She narrowly survived the attempt and was subsequently lost to follow-up.

Patient 2. A 56-year-old man presented to the ED with a 6-hour history of progressive abdominal pain and nausea. The abdominal pain was diffuse, sharp, and radiated to the back. Physical examination revealed tenderness to palpation in the mid-

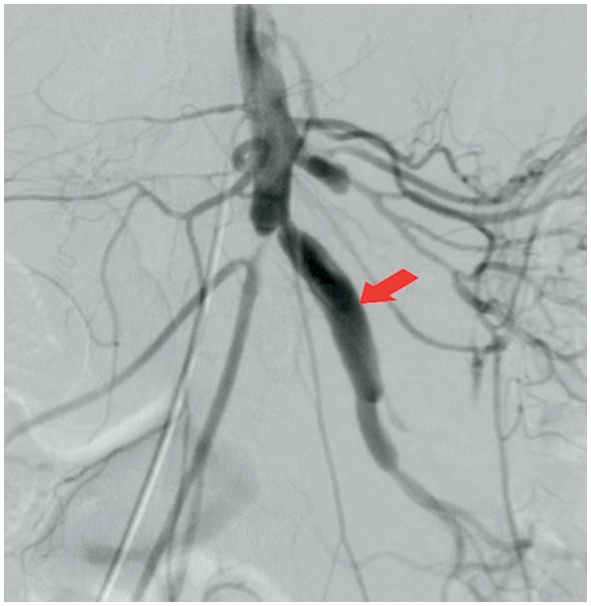

Fig 1. Angiography shows aneurysmal superior mesenteric artery *(arrow)* with distal stenosis.

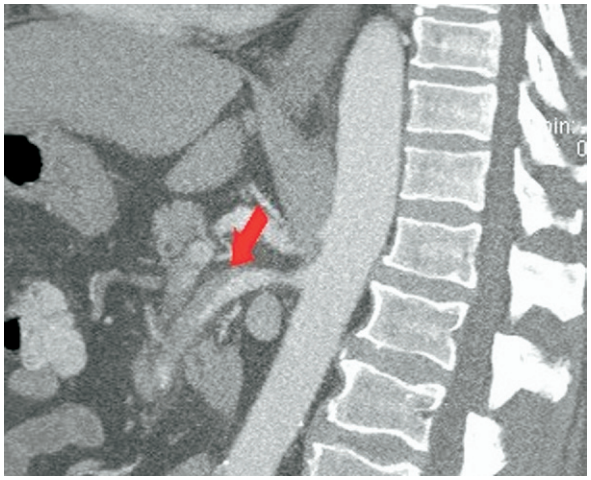

Fig 2. Sagittal computed tomography reconstruction shows proximal superior mesenteric artery dissection and unopacified false lumen *(arrow)*.

dle epigastric region. His medical history was remarkable for hypertension. Results of a complete blood count and basic metabolic panel were within normal limits, with the exception of a leukocytosis of 13,000 cells/mm^3.

An abdominal CT scan showed SMA dissection with distal occlusion. The dissection originated 7 mm from the origin and extended for approximately 6 cm. There were two patent jejunal branches with a completely unopacified SMA distally. No radiologic evidence of bowel ischemia was present (Fig 2). Because of the CT findings, the patient underwent mesenteric angiography, which confirmed long-segment SMA dissection with occlusion of the false lumen and preservation of jejunal branches. Collateral circulation provided flow to the distal SMA distribution.

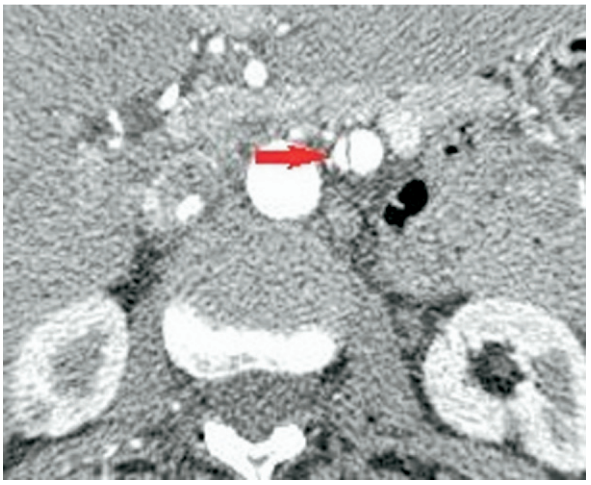

Fig 3. Abdominal computed tomography scan shows the superior mesenteric artery intimal flap or double lumen *(arrow)* with perfusion of the true and false lumens.

Anticoagulation with heparin was administered after angiography. Within 6 hours, the patient's abdominal pain improved significantly. Treatment on hospital day 1 and 2 consisted of bowel rest, heparin anticoagulation, and narcotic pain control. The patient was pain-free by hospital day 3, and a repeat abdominal CT angiogram (CTA) revealed a thread-like true lumen of the middle SMA that filled the distal small bowel branches. Diet was advanced without postprandial pain. Warfarin was initiated, and a therapeutic international normalized ratio (INR) of 2.5 was achieved on hospital day 9. The patient was discharged home. At the 5-month follow-up, the patient was asymptomatic and a CTA of the abdomen showed a very small filling defect in the middle SMA, with complete resolution of the proximal dissection.

Patient 3. A 62-year-old woman underwent a CTA of the abdomen and pelvis, which revealed SMA dissection. The dissection origin was 2.5 cm from the aorta and extended for 10 cm with several patent jejunal branches off of the true lumen (Fig 3). Mesenteric angiography performed 2 weeks later revealed similar findings of the SMA dissection along with fibromuscular disease of the renal arteries.

The patient had been admitted to the hospital 1 month earlier with altered mental status and abdominal discomfort. A CT scan showed evidence of renal infarcts, but a thorough evaluation of this scan failed to show pathologic changes of her SMA. She had been experiencing intermittent nausea and vomiting along with nonspecific abdominal pain for 2 weeks before admission. The patient was somewhat obtunded and had a benign abdominal examination at the initial presentation. Her medical history included carotid artery fibromuscular dysplasia and rib fractures that occurred due to a fall 6 weeks prior. Mild electrolyte abnormalities were evident, but lactic acid and liver functions were within normal limits.

After admission, the patient had continuing decreased mental status with airway compromise and was intubated. The patient was weaned from the ventilator on hospital day 4. Aseptic meningitis

was the presumed diagnosis after exhaustive workup, and the patient was discharged home pain-free on hospital day 9.

The SMA dissection in this case did not correlate with any of the patient's symptoms and was likely an incidental finding. At the 5-month follow-up, the patient continues to be free of abdominal pain, without treatment.

DISCUSSION

A Medline literature search in all languages was performed, and the inclusion of our three cases brings the total of reported isolated SMA dissections to 71 (Table, online only).[1-48] The patient population is 81.7% (58 of 71) male. These patients are relatively young with respect to vascular pathology, with a mean age of 54.6 years (range, 39-87).

The four most common symptoms of SMA dissection have been described, in decreasing order of frequency, as acute isolated abdominal pain, abdominal pain with vomiting, subacute intestinal obstruction, and asymptomatic. Before 2003, these four symptoms encompassed 79.8% of presenting complaints of SMA dissection.[2] In almost all cases, the diagnosis was made by abdominal CT imaging with intravenous contrast administration for evaluation of abdominal pain.

Treatments in the 71 patients included surgery in 34 (47.9%), endovascular stenting in six (8.4%), and medical management in 31 (43.7%). Bowel infarction occurred in eight patients (11.3%). Complications after SMA dissection resulted in 12 deaths, 11 of which occurred before 1972. The diagnosis of SMA dissection was reached at autopsy in all cases.

CT scan is currently the preferred imaging modality for evaluating abdominal pain. It has been a successful tool for identification of SMA dissection in most cases. Suzuki et al[43] described four characteristic CT findings of SMA dissection, which include false lumen or intramural hematoma, intimal flap, increased attenuation of fat around the SMA, and hematoma in the mesentery with hemorrhagic ascites. Catheter-directed SMA angiography is an effective confirmatory study that can also evaluate for collateral blood flow and be used to plan endovascular interventions.

In the present cases, CT scan was used to make the diagnosis in patients 2 and 3. The classic double lumen or intimal flap was identified in all patients, although in retrospect with patient 1. Patient 1 underwent a mesenteric angiography ≤24 hours of a CT scan that clearly identified SMA dissection.

Increase fat attenuation, mesenteric hematoma, or hemorrhagic ascites were not appreciated on any of our radiologic studies. Ultrasonography has been used to specifically evaluate the SMA, but its role for initial evaluation of abdominal pain is minimal. The one patient who died after 1974 was evaluated by abdominal ultrasound imaging, and the diagnosis was missed.[36]

Because SMA dissection is relatively rare, a standard diagnostic and therapeutic approach has not been adopted. Treatments can be separated into medical, surgical, and endovascular. Medical treatments have included reports of anticoagulation and observation. Surgical treatments include thrombectomy, endoaneurysmorrhaphy, intimectomy and patchplasty, ligation, resection, venous graft bypass, and arterial bypass graft. Endovascular intervention involves stent placement or thrombolytics. Indications for surgical repair have been previously described as increasing aneurysmal dilation, thrombosis of the SMA true lumen, persistent symptoms despite anticoagulation,[35] arterial rupture,[43] or bowel infarction. Arterial rupture and bowel infarction are absolute indications for emergency surgical intervention, whereas the others are relative indications for the rare failure of medical management in symptomatic patients.

Yasuhara et al[25] reported successful conservative medical management in two patients without anticoagulation. These cases offer insight into the natural history of SMA dissection. Others have used anticoagulation of indeterminate duration. Nagai et al[38] reported four consecutive cases where anticoagulation was a highly effective approach, with follow-up showing complete normalization of the SMA ≤1 year. One report details the failure of anticoagulation after 1 year. The patient experienced postprandial abdominal pain, and a CT scan revealed enlargement of the false lumen. Endoaneurysmorrhaphy proved effective to alleviate symptoms.[29]

Compared with the carotid artery, the most commonly dissected peripheral vessel, anticoagulation appears to have a beneficial role for treatment of both, but operative and endovascular intervention for SMA dissection has favorable results when indicated.

Endovascular stent placement has also been a successful intervention for treatment of SMA dissection. Leung et al[30] performed the first stent placement in 2000. Six of the past 20 operative interventions have been endovascular stenting. Bare metal stents were used in all cases. Desirable qualities for SMA stents include minimal shortening, good flexibility, and lack of migration despite continuous mesenteric motion.[40] Stents obliterate the false lumen and promote flow through the true lumen. Endovascular stenting shows early promise as a less-invasive alternative to open operative revascularization. Favorable anatomy for stenting includes localized intimal flaps or short segments of dissection. Five of the six stents were placed within the first 2 days of hospitalization and one because of failure of anticoagulation at 1 month. All patients were symptom free at the short-term follow-up. This suggests that timing for stent placement is immediately appropriate when favorable anatomy is encountered, granted no absolute surgical indications are present.

Surveillance for SMA dissection includes an office visit 2 to 4 weeks after hospitalization to assess for symptoms of mesenteric ischemia. A CTA of the abdomen and pelvis should be performed at 6-month intervals to evaluate the SMA for improvement or persistent dissection. Most reports show complete or nearly complete resolution by 6 months. Anticoagulation can be discontinued after resolution. Only two late failures of medical therapy have been reported. These patients presented with symptoms of chronic mesenteric ischemia that occurred after 9 and 12

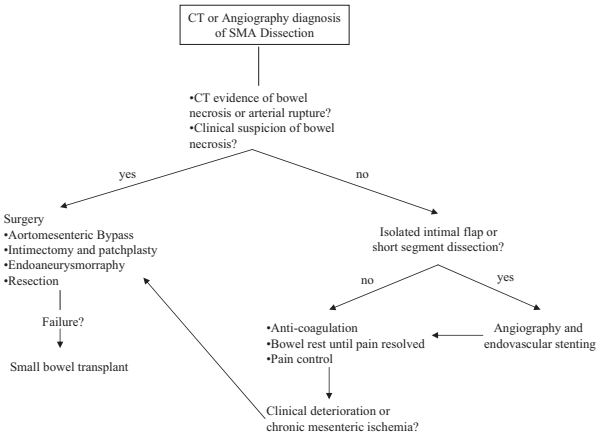

Fig 4. Proposed algorithm for the treatment of superior mesenteric artery *(SMA)* dissection after identification by computed tomography *(CT)* or angiography.

months of anticoagulation with slightly progressive dissection on CT scan.

All surgical options have been performed with good results. Aortomesenteric bypass[14,16,17,21,23,28,33,34] is the most common surgical procedure (8 of 34, 23.5%). Patch angioplasty with or without intimectomy[2,13,17,19,23] and endoaneurysmorraphy[17,29,41] are the most basic surgical revascularization procedures (9 of 34, 26.5%) and have shown good short-term outcomes without failure or reoperation. Long-term follow-up is not available to support any particular approach, however.

Our first case details a unique surgical option. This patient was not a candidate for SMA revascularization. Enterectomy with small bowel transplant was successful, but complications occurred between operations, with post-transplant care and potential ongoing risks that seemed considerably extreme compared with the other previously described successful repairs. Perhaps small bowel transplant should be reserved for failed SMA revascularization with resultant short-gut syndrome. The most recent literature reports a 5-year graft survival rate of 64% after small bowel trantsplant,[49] which is considerably less than the 100% of reported successful SMA revascularizations in this review. A Medline search revealed no results of small bowel transplant for chronic mesenteric ischemia.

The result in patient 2 showed how timely administration of anticoagulation correlated with resolution of acute abdominal pain and recanalization of the SMA. Patient 3 remains asymptomatic 5 months after diagnosis, without anticoagulation. The treatment algorithm we propose is illustrated in Fig 4.

CONCLUSION

Isolated superior mesenteric artery dissection is a rare but increasingly recognized vascular pathology. Documentation of 71 cases to date has provided insight into appropriate treatments and prognosis. The wide spread use of CT scanning for evaluation of abdominal pain and improvement of radiologic images have accounted for the recent increase in SMA dissection identification. CT imaging has proven to be an effective noninvasive tool for accurate diagnosis and surveillance. Medical management with anticoagulation and supportive care has provided good evidence to be the first line of treatment. Options for surgical repair range from endovascular stenting to open revascularization. Immediate operative revascularization is indicated for intestinal infarction or arterial rupture, whereas endovascular intervention can be attempted for patients with favorable pathologic anatomy. Stenting can be used as an adjunct or for failures of medical management. Continued documentation of isolated SMA dissections will provide further support for medical, surgical, and endovascular treatment standards.

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Additional material for this article may be found online at www.jvascsurg.org.

Table. (Online only) Reports of superior mesenteric artery dissection

Case	*First author (year)*	*Age/sex*	*Surgical procedure*	*Bowel infarction*	*Death*
1	Bauersfeld (1947)[3]	87/F	. . .	+	+
2	Foord (1959)[4]	58/M	. . .	−	+
3	Foord (1959)	55/M	. . .	+	+
4	Foord (1959)	73/F	. . .	Unknown	+
5	Ralston (1960)[5]	51/M	. . .	−	+
6	Jean (1961)[6]	53/M	Drainage	−	+
7	Clark (1962)[7]	51/M	. . .	+	+
8	Ramchand (1969)[8]	68/F	. . .	+	+
9	Boquist (1970)[1]	52/F	. . .	−	+
10	Lee (1971)[9]	62/M	. . .	−	+
11	Guthrie (1972)[10]	70/F	. . .	−	+
12	Sisteron (1975)[11]	. . .	Venous graft	−	−
13	Rignault (1967)[12]	50/M	SMA transposition	−	−
14	Krupski (1985)[13]	51/F	Intimectomy and venous patch angioplasty	−	−
15	Takehara (1988)[14]	50/M	Aortomesenteric bypass	−	−
16	Corbetti (1989)[15]	62/M	Resection	−	−
17	Koto (1989)[16]	53/M	Aortomesenteric bypass	−	−
18	Cormier (1992)[17]	60/M	SMA venous bypass	−	−
19	Cormier (1992)	41/M	Endoaneurysmorrhaphy	−	−
20	Cormier (1992)	52/M	Aortomesenteric bypass	−	−
21	Cormier (1992)	50/M	Intimectomy and prosthetic patchplasty	−	−
22	Vignati (1992)[18]	50/M	Right GEA to mesenteric bypass	−	−
23	Solis (1993)[19]	45/F	Intimectomy	−	−
24	Ambo (1994)[20]	56/M	. . .	−	−
25	Ando (1995)[21]	47/M	Aortomesenteric bypass	−	−
26	Hydoh (1996)[22]	66/M	. . .	−	−
27	Murata (1997)[23]	47/M	Aortomesenteric bypass	+	−
28	Murata (1997)	64/M	Resection and prosthetic interpose	−	−
29	Murata (1997)	70/M	Venous patchplasty	−	−
30	Nakamura (1997)[24]	44/M	. . .	−	−
31	Yasuhara (1998)[25]	55/M	. . .	−	−
32	Yasuhara (1998)	45/M	. . .	−	−
33	Barmeir (1998)[26]	48/M	Thrombectomy	−	−
34	Dushnitsky (1998)[27]	58/M	. . .	−	−
35	Iha (2000)[28]	46/M	Aortomesenteric bypass	−	−
36	Sparks (2000)[29]	41/M	Endoaneurysmorrhaphy	−	−
37	Leung (2000)[30]	67/M	Stent	−	−
38	Sheldon (2001)[31]	41/M	Unknown operation	−	−
39	Sheldon (2001)	46/M	. . .	−	−
40	Sagiuchi (2001)[32]	61/M	SMA ligation	−	−
41	Goueffic (2002)[33]	56/M	Aortomesenteric bypass	−	−
42	Hirai (2002)[34]	42/M	Aortomesenteric bypass	−	−
43	Takayama (2002)[35]	63/M	. . .	−	−
44	Javerlait (2003)[2]	51/M	SB resection with celiac stenting	+	−
45	Javerlait (2003)	61/F	Intimectomy and prosthetic patchplasty	−	−
46	Javerlait (2003)	68/M	Intimectomy and prosthetic patchplasty	−	−
47	Sartelet (2003)[36]	44/M	. . .	+	+
48	Yoon (2003)[37]	52/M	Stent	−	−
49	Nagai (2004)[38]	49/M	. . .	−	−
50	Nagai (2004)	56/M	. . .	−	−
51	Nagai (2004)	59/M	. . .	−	−
52	Nagai (2004)	52/M	. . .	−	−
53	Froment (2004)[39]	58/M	Stent	−	−
54	Kim (2004)[40]	48/F	Stent	−	−
55	Kim (2004)	54/M	Stent	−	−
56	Tsuji (2004)[41]	44/M	Endoaneurysmorrhaphy	−	−
57	Nozu (2004)[42]	55/M	. . .	−	−
58	Suzuki (2004)[43]	54/F	. . .	−	−
59	Suzuki (2004)	50/M	. . .	−	−
60	Suzuki (2004)	60/M	. . .	−	−
61	Suzuki (2004)	78/M	SB resection	+	−
62	Suzuki (2004)	57/M	Right GEA to SMA bypass	−	−
63	Suzuki (2004)	50/M	. . .	−	−
64	Miyamoto (2005)[44]	59/M	Stent	−	−
65	Kochi (2005)[45]	43/M	Bypass with right GEA and arterial conduit	−	−
66	Picquet (2005)[46]	53/F	SMA resection; reconstruction used arterial conduit	−	−

Table. Continued

Case	*First author (year)*	*Age/sex*	*Surgical procedure*	*Bowel infarction*	*Death*
67	Chang (2006)[47]	49/M	. . .	–	–
68	Matsushima (2006)[48]	51/M	Exploratory laparotomy	–	–
69	Morris (2007)	39/F	Enterectomy with SB transplant	–	–
70	Morris (2007)	56/M	. . .	–	–
71	Morris (2007)	62/F	. . .	–	–

GEA, Gastroepiploic artery; *SB,* small bowel; *SMA,* superior mesenteric artery.