Internal Medicine
The Japanese Society of Internal Medicine
doi: 10.2169/internalmedicine.3094-19
Intern Med 58: 3393-3400, 2019
http://internmed.jp

【 CASE REPORT 】

Rare Mesenteric Arterial Diseases: Fibromuscular Dysplasia and Segmental Arterial Mediolysis and Literature Review

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Abstract:
Fibromuscular dysplasia (FMD) and segmental arterial mediolysis (SAM) are noninflammatory, nonatherosclerotic arterial diseases that cause aneurysm, occlusion, and thromboses. These diseases are rarely seen in mesenteric arterial lesions; however, as they can be lethal if appropriate management is not provided, the accumulation of clinical information from cases is essential. We herein report the cases of a 57-year-old man diagnosed with FMD and a 63-year-old man diagnosed with SAM. We conclude that an early diagnosis with imaging modalities and clinical information followed by the appropriate treatment improves the prognosis of these arterial diseases.

Key words: fibromuscular dysplasia, segmental arterial mediolysis, mesenteric lesion, diagnosis

(Intern Med 58: 3393-3400, 2019)
(DOI: 10.2169/internalmedicine.3094-19)

Introduction

Mesenteric vascular diseases include ischemic changes due to atherosclerotic changes and noninflammatory, nonatherosclerotic arterial diseases of fibromuscular dysplasia (FMD) and segmental arterial mediolysis (SAM). Although atherosclerotic diseases are rather common, and FMD and SAM were originally diagnosed by histological analyses after surgery or upon an autopsy, recent advances in imaging modalities have led to increasing frequencies of FMD and SAM diagnoses. As these diseases can affect various organs and be lethal if not properly managed, an appropriate diagnosis followed by treatment in the early phase is essential (1, 2). However, as the disease entities are rare, we have summarized the characteristics of FMD and SAM in the gastroenterologic regions and reviewed reported cases with a comparison of the imaging findings and histological analyses (3).

To understand these diseases in mesenteric arteries further and diagnose them based on available clinical information and imaging findings, we herein report two recently experienced cases of FMD and SAM with updated information from recently published cases focusing on the mesenteric regions.

Case Reports

Case 1

A 57-year-old man was admitted to our hospital with a chief complaint of acute-onset severe and continuous sharp epigastric pain with no trigger. The pain gradually reduced, but the abdominal discomfort persisted.

He had a history of untreated hypertension, hyperlipidemia, and diabetes. He had been a smoker for 33 years (1 pack/day). Upon admission to our hospital, he had a blood pressure of 142/98 mmHg, heart rate of 92 beats/min, and temperature of 36.5℃. Laboratory findings showed a mild elevation of the white blood cell count (12,800/μL) and C-reactive protein (7.49 mg/dL), blood sugar (152 mg/dL), lactate dehydrogenase (LDH; 243 IU/L), creatinine kinase (538 IU/L), and hemoglobin A1c (6.5%). Computed tomography (CT) and three-dimensional reconstruction showed arterial stenosis, aneurysmal changes, and partial dissection in the superior mesenteric artery (Fig. 1a-c) as well as stenosis and aneurysmal changes in the branch of the right renal artery (Fig. 1d, e). Magnetic resonance angiography (MRA)

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Received: March 26, 2019; Accepted: June 3, 2019; Advance Publication by J-STAGE: July 22, 2019
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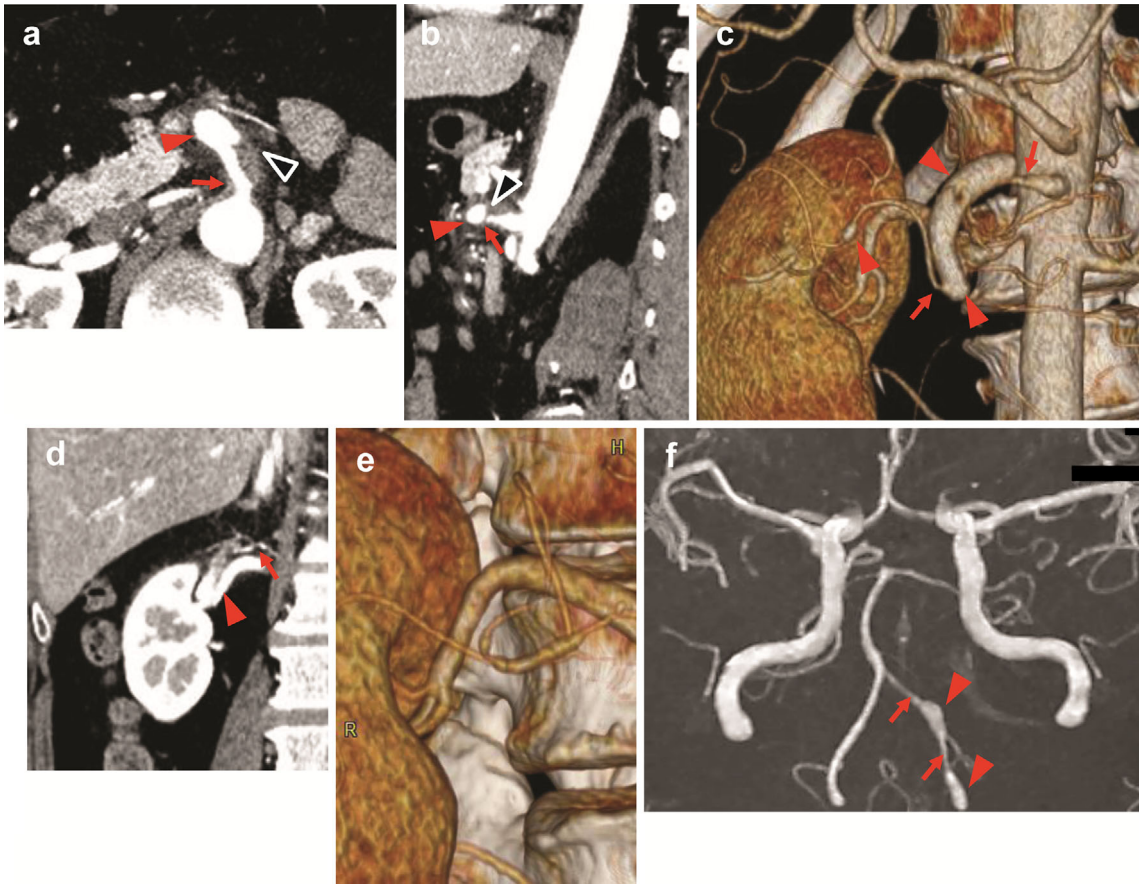

Figure 1. **Imaging studies of FMD in Case 1. (a, b, c) Computed tomography (CT) and three-dimensional reconstruction showed stenosis (red arrows), aneurysmal changes (red arrowheads), and partially dissection (black arrowheads in a and b) in the superior mesenteric artery. (d, e) Stenosis (red arrows) and aneurysmal changes (red arrowheads) in the branch of the right renal artery. Magnetic resonance angiography showed stenosis (red arrows) and aneurysmal changes (red arrowheads) in the left vertebral artery (f). FMD: fibromuscular dysplasia**

showed stenosis (red arrows) and aneurysmal changes (red arrowheads) in the left vertebral artery (Fig. 1f). Because of his noninflammatory, nonatherosclerotic multiple arterial lesions in the midsized arteries, hypertension, and long history of smoking, we diagnosed him with FMD.

Given the diagnosis of FMD and persistently high blood pressure at 160/90 mmHg, antihypertensive therapy was started with continuous intravenous infusion of nicardipine (2 μg/kg/min) followed by the oral administration of valsartan (40 mg/day) starting on hospital day 7, which successfully brought down the blood pressure to 110/60 mmHg. In addition, to prevent cerebral infarction because of aneurysmal lesions in the left vertebral artery, anticoagulant therapy was also started with continuous intravenous infusion of heparin (10,000 IU/day) on hospital day 11, followed by the oral administration of aspirin (81 mg/day). As the epigastric symptoms disappeared soon after the blood pressure came under control, they were considered to have been due to the progression of stenotic changes with vasoconstriction of mesenteric arterial lesions due to the increase in the blood pressure. With treatment of valsartan (40 mg/day) and aspirin (81 mg/day) for 18 months, no recurrence of the symptoms or other complications have been noted to date.

Case 2

A 63-year-old man was admitted to our hospital with a chief complaint of left-sided abdominal pain that showed slow progression from the day before the admission. He was a nonsmoker and had no history of illness.

Upon admission to our hospital, he had a blood pressure of 129/84 mmHg, heart rate of 75 beats/min, and temperature of 36.9℃. Other than the mild elevation of his white blood cell count (12,090/μL) and LDH (283 IU/L), no abnormal findings were noted. CT and three-dimensional reconstruction showed bleeding in the abdominal cavity and dissecting aneurysm in the celiac artery to the splenic artery (Fig. 2a-c). In addition, the bilateral external iliac arteries showed multiple aneurysmal changes and partial dissection (Fig. 2d-f). Because of noninflammatory, nonatherosclerotic multiple arterial diseases, dissecting aneurysm of the celiac and splenic arteries, and bleeding in the abdominal cavity probably from the arterial lesions, we diagnosed him with SAM.

As the symptoms significantly improved after admission

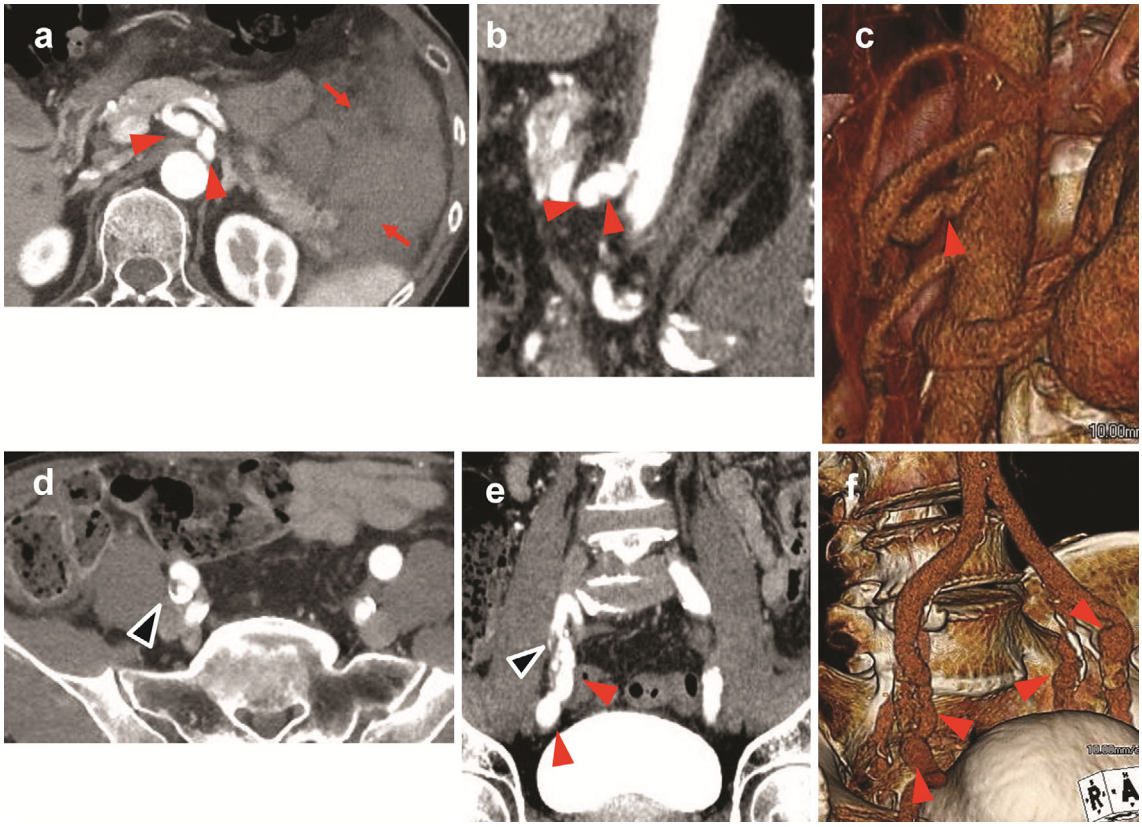

Figure 2. **Imaging studies of SAM in Case 2. (a, b, c) Computed tomography (CT) and three-dimensional reconstruction showed bleeding in the abdominal cavity (red arrows) and dissecting aneurysm (red arrowheads) in the celiac artery to splenic artery. (d, e, f) The bilateral external iliac arteries showed multiple aneurysmal changes (red arrowheads) and partial dissection (black arrowheads in d and e). SAM: segmental arterial mediolysis**

with no increase in the bleeding in the abdominal cavity on day 2 or recurrence of the symptoms, and the hematoma decreased significantly on day 7, no emergent intervention or additional medication was started. However, in order to monitor the size of the aneurysmal changes and dissection, scheduled CT was performed every three to six months, and the blood pressure was carefully monitored. No recurrence of the symptoms or other complications has been noted in 16 months of follow-up.

Discussion

FMD and SAM are noninflammatory, nonatherosclerotic arterial diseases originally diagnosed based on histological findings; however, with marked advances in imaging modalities, including CT and magnetic resonance imaging, opportunities to diagnose these diseases based on imaging findings and clinical information without a pathological examination are increasing (3).

The clinical and pathological classification of FMD was first reported by Harrison and McCormack in 1971 (4), and recently, data belonging to the first 447 patients from the U.S. Registry for FMD were reported (5). FMD is a rare, medium-sized arterial disease occurring throughout the body with a frequency of 0.02%, predominantly in women (5). In addition, smoking, hormones, HLA-DRw6 polymorphism, and physiologic stimulation have been reported to be risk factors (6). The histologic changes in the arterial muscle replaced by fibroplasia can lead to arterial stenosis, occlusion, aneurysm, and dissection, and such events typically occur in the renal, extracranial, carotid, and vertebral arteries. Therefore, although rare, mesenteric FMD can cause unspecific abdominal pain, diarrhea, nausea, and vomiting (2). Imaging studies, including CT and angiography, reveal the narrowing and aneurysmal changes of the vasculature that lead to a beaded appearance (1). Our Case 1 also had a history of smoking, and CT showed a multiple-beaded aneurysmal appearance and partial dissection of the superior mesenteric and right renal arteries. In addition, MRA showed stenosis and dissection of the left vertebral artery, which is rather typical for FMD. Therapeutic options include antiplatelet, antithrombotic, and antihypertensive therapy (7), and our Case 1 was also successfully treated with these approaches with no recurrence.

SAM was first reported by Slavin and Gonzalez-Vitale in 1976 (8) and is a rare disease, with 50 cases reported to date. SAM is caused by the disruption of the arterial medial layer of a medium- to large-sized artery, and its risk factors include hypoxia, shock, hypertension, circulatory disturbance, and other vasoconstrictor stimuli (2, 8, 9). Because of

Table 1. Summary of FMD Casesreported Recently.

Case (No)	Ref	Age (yr)	Gender	Symptoms	Imaging findings	Histological findings	Treatment	Outcome
1	50	81	F	Syncope	Hematoma and hepatic artery rupture. Narrowing and aneurysms in celiac, common hepatic, renal artery. Stenosis in carotid artery.	N/A	Endovascular exclusion of the pseudoaneurysm with a balloon-expandable covered stent. Aspirin and clopidogrel.	Improved
2	49	60	M	Abdominal pain, disturbed consciousness	Dissection in SMA and right vertebral artery.	N/A	Fluid replacement therapy	Improved
3	48	54	F	Diarrhea, abdominal pain, weight loss	Multiple aneurysms in SMA, coeliac, splenic and renal artery. Beaded appearance in both renal arteries.	N/A	TPN, Anticoagulation, open repair of the SMA aneurysms	Improved
4	47	61	F	Abdominal pain	Multiple aneurysms and stenoses in SMA, IMA and renal artery	Multiple tears and dissections of the medial layer and fibrointimal thickening	Anticoagulation	Improved
5	46	20	F	Abdominal pain, hemorrhagic shock	Intraperitoneal omental bursa and mesentery of the transverse colon. "String of beads" appearance in the jejunal and SMA	N/A	Transcatheter arterial embolization	Improved
6	45	52	M	Lower abdominal pain	The inferior mesenteric artery is tortuous and stenosed	Necrosis of the mucosa. Fibrosis of the intima; the media of these vessels was normal.	Left hemicolectomy	Improved
7	44	19	F	Abdominal pain and vomiting	Stenosis of the origin of the SMA and multiple aneurysms involving the proximal SMA. Right renal artery is mild irregularity.	N/A	The aneurysmal segment of the SMA was resected and an aorto-SMA interposition graft with polytetrafluoroethylene was performed.	Improved
8	43	47	F	Nausea, early satiety and upper abdominal pain	Narrowing of the superior mesenteric artery at its origin, with marked hypertrophy of the gastroduodenal artery and pancreaticoduodenal arteries.	N/A	An aorto-superior mesenteric artery and an aorto-hepatic artery bypass.	Improved
9	42	47	F	Abdominal pain, diarrhea and hypertension	A partial occlusion of the celiac trunk and a total occlusion of the superior mesenteric artery.	Intimal and medial proliferation	Antihypertensive drug	Died
10	41	30	M	Abdominal pain and hypertension	Dissections of the celiac, SMA, left renal, and external iliac artery.	N/A	β-blocker, Ca blocker, warfarin, and aspirin. Angioplasty for right renal artery.	Improved
11	40	44	F	Hypertension, abdominal pain, diarrhea and vomiting	SMA stenosis and nonspecific colitis	N/A	Angioplasty	Improved
12	39	43	F	Hypertension, abdominal pain and headache	Aneurysms in the left renal artery with severe fibrodysplastic stenosis. The string-of-beads appearance is shown in the right renal artery. Severe stenosis with post-stenotic dilatation is detected in SMA.	Intimal fibroplasia, loss of internal elastic lamina, and massive destruction of the media are observed in the aneurysm wall.	Aneurysm resection and aortorenal bypass and percutaneous transluminal angioplasty	Improved
13	11	38	M	N/A	N/A	N/A	N/A	Improved
14	38	43	F	Hypertension and headache	String-of-beads appearance in the right renal artery and SMA. Stenosis and multiple irregularities in the left renal artery.	N/A	Angioplasty and antihypertensive drugs	Improved
15	37	N/A	N/A	Abdominal pain, distension and constipation	N/A	A thick cuff (petal like) of smooth muscle proliferation with normal intima and media in mesenteric artery.	Right hemicolectomy	Improved
16	36	38	M	Gastrointestinal bleeding, anemia	Ectasia, bleeding and narrowing in SMA. Ectasia in IMA.	Thickening and hyalinization of medium sized vessel walls, with intimal proliferation.	Ileal resection	Improved
17	35	48	F	Acute abdominal pain	Occlusion of the SMA and celiac trunk, with an enlarged hypertrophic IMA and reinjection of the distal SMA, common hepatic artery and splenic artery.	Intimal fibroplasia and an increased deposition of fibrous tissue in the vessel wall media	Reimplantation of the SMA	Improved
18	34	57	F	Acute abdominal pain, weight loss, anorexia, nausea, vomiting and non-bloody diarrhea.	Long, tubular and narrowing of SMA and celiac artery	Medial thickening, smooth muscle hyperplasia in SMA and celiac artery	Aorto-celiac and aorto-SMA bypass	Died
19	33	48	F	Abdominal pain and hemoperitoneum	Multiple small aneurysms in SMA, Celiac and Renal artery (string-of-beads).	N/A	Surgical hemostasis and antihypertensive drugs(β-blocker).	Improved
20	32	43	M	No symptoms	Aneurysms of the SMA, hepatic artery, splenic artery, jejunal artery and internal iliac arteries.	Medial fibrodysplasia is observed in the artery wall.	Aneurysm resection and arterial reconstruction	Improved
21	31	78	F	Hypertension, abdominal pain and hemoperitoneum.	Dilated loop of the small bowel and a small amount of fluid in the peritoneal cavity.	Medial and perimedial fibro dysplasia, forms the characteristic petal-like appearance in SMA.	None	Died
22	30	33	M	Abdominal pain	Strings-of-beads appearance in SMA	Thickening of the media due to hyperplasia in SMA	Ileal resection	Improved
23	Our Case 1	57	M	Acute epigastric pain, neck stiffness	Multiple beaded aneurysmal appearance (stenosis and aneurysms) in SMA and right renal arteries. Stenosis and dissection in left vertebral artery.	N/A	TPN, heparin, aspirin, Ca-blocker, ACE inhibitor	Improved

FMD: fibromuscular dysplasia, M: male, F: female, N/A: data not applicable, SMA: superior mesenteric artery, IMA: inferior mesenteric artery, RA: renal artery, CT: computed tomography, TPN: Total parenteral nutrition

Intern Med 58: 3393-3400, 2019 DOI: 10.2169/internalmedicine.3094-19

Table 2. Summary of SAM Cases Reported Recently.

Case (No)	Ref	Age (yr)	Gender	Symptoms	Imaging findings	Histological findings	Treatment	Outcome
1	78	49	F	Shock, severe abdominal pain	Massive hematoma, aneurysms in gastroduodenal artery, common hepatic artery, and SMA	N/A	Blood transfusion for shock and coil embolization. Ca antagonist	Improved
2	77	60	M	Acute abdominal pain	Mesenteric hemorrhage, dissection in SMA.	N/A	Embolization with coil	Improved
3	76	65	M	Severe abdominal pain, weight loss, melena, anemia	Ruptured hepatocellular carcinoma with hemoperitoneum and an aneurysm with string-of-beads appearance in SMA.	N/A	Embolization with coil	Improved
4	75	37	M	Abdominal pain	Mesenteric hematoma. Stenosis and aneurysms in celiac. Stenosis in renal artery. Aneurysms in jejunal, left gastric and splenic artery.	N/A	Embolization with coil	Died
5	74	57	M	Hypertension, abdominal pain	Arterial dissection with luminal stenosis and aneurysm formation at the distal portion of the SMA	Vacuolization and decrease in the number of vascular smooth muscles	Aneurysmectomy and bowel resection followed by the administration of Ca-blocker	Improved
6	73	58	M	Abdominal pain	Mesenteric hematoma and right inguinal hernia with unremarkable small bowel. Beading appearance in SMA	N/A	Immunosuppressive therapy and embolization with coil	Improved
7	72	32	M	Abdominal pain	Stenosis and aneurysm in renal and IMA, massive amount of hemorrhage	Media shows myxoid degeneration in the outer one-third adjacent to the adventitia	Surgical hemostasis and left hemicolectomy followed by administration of antihypertensive drugs.	Improved
8	71	40	M	Abdominal pain	Extensive dissection of SMA with the thrombotic occlusion. Narrowing and dilation of celiac artery	N/A	Conservative	Improved
9	70	79	M	Abdominal pain, hypotension	Active bleeding from IMA and hemorrhage	Reduplication of the internal elastic lamina with arterial dissection within the tunica media and thrombus at the site of rupture	Surgical resection of left colic artery	Improved
10	69	47	M	Loss of consciousness, headache, abdominal pain	String-of-beads appearance in SMA. Dissection of VA	Medial islands and medial degenerations in SMA	Embolization with coil for VA and SMA. Surgical resection of part of middle colic artery and descending colon.	Improved
11	68	36	M	Abdominal pain	Stenosis and aneurysm of AIPDA and string-of-beads appearance in a nearby artery	N/A	Embolization with coil	Improved
12	67	60	F	Hypoxia, hypotension, cardiopulmonary arrest	Hematoma in the retroperitoneal and intraperitoneal space. Aneurysm and "bead-like fashion" appearance in SMA	N/A	Conservative	Improved
13	66	64	F	Abdominal pain, back pain, nausea	Hematoma in the anterior pararenal space inferior to pancreatic tail. Aneurysms in SMA, IMA, hepatic artery.	N/A	Conservative	Improved
14	65	56	M	Abdominal pain, shock	Aneurysm in MCA, SMA dissection	N/A	Embolization with coil	Improved
15	64	55	F	Abdominal pain	Aneurysms in SMA, celiac, hepatic, splenic	N/A	Warfarin, aspirin	Improved
16	63	29	F	Hypertension	Scattered microaneurysms in renal, hepatic, SMA. Renal cortical nephrogram.	Segmental lesions of the media with loss of smooth muscle cells	Warfarin	Improved
17	62	51	M	Abdominal pain, shock	Abdominal hemorrhage and active bleeding from a branch of the SMA.	N/A	Embolization and ligation of the branches of the SMA. Warfarin	Improved
18	61	53	M	Unremarkable	Aneurysm in splenic, celiac and SMA. Dissection in origin of the celiac.	N/A	Embolization with coil and aortic stent graft	Improved
19	60	70	M	Unknown	N/A	Massive medial defects and residual medial island with extended adventitia	None	Died
20	9	25	F	Anorexia, abdominal pain, diarrhea	Ischemic colitis of the splenic flexure. Occlusion of the left colic artery. Stenoses of the hepatic artery	Patchy, isolated destruction of the arterial media involving both the internal and external elastic laminae	Partial colectomy of the splenic flexure	Improved
21	59	60	M	N/A	Ruptured aneurysm of the MCA. Multiple wide and narrow and aneurysm in SMA	N/A	Surgical resection	Improved
22	58	57	M	Abdominal pain, diarrhea	Ascites throughout the abdomen. Aneurysm within the left branch of middle-colic artery	N/A	Transcatheter arterial embolization	Improved
23	57	59	M	Abdominal pain, shock	SMA dissection, aneurysm in renal, gastroepiploic, splenic artery. Splenic aneurysm was ruptured	Medial island spared from mediolysis.	Emergency embolization of the splenic artery, resection of the gastroepiploic artery aneurysm.	Improved
24	56	76	F	Abdominal pain, nausea	Mesenteric hematoma and aneurysm in IMA	N/A	Embolization with coil	Died in 3 months
25	56	57	M	Abdominal pain	Mesenteric hematoma, aneurysm and stenosis of the middle colic artery, celiac and hepatic artery	N/A	Embolization with N-butyl cyanoacrylate for SMA aneurysm	Improved
26	55	49	M	Abdominal pain, shock	Large hematoma surrounding a high-density aneurysm. abnormal "beaded" appearance in SMA	Multifocal fragmentation of the elastic fibers of the media	Right hemicolectomy	Improved
27	54	52	M	Sudden hemiparesis, hypertension	Aneurysm in ICA, hepatic, celiac, SMA and narrowing in SMA, celiac	Multiple segmental mediolysis lesions of the muscular and elastic fibers of the media	Reconstruction by using autologous saphenous vein graft in hepatic and celiac	Improved
28	53	35	F	Abdominal pain, perforation on transverse colon	Mesenteric vein occlusion and ischemic colitis	Segmental vacuolar degeneration of smooth muscle with areas of wall thinning	Resection of terminal ileum	Died
29	52	78	M	Abdominal pain, diarrhea, shock,	N/A	Destruction of the tunica interna and media in MCA	Emergency surgery (right hemicolectomy). (At intraoperative findings, a large hematoma and a ruptured aneurysm)	Improved
30	51	56	F	Abdominal pain	Intraabdominal hemorrhage. Aneurysm in IMA	N/A	Left hemicolectomy	Improved
31	Our case 2	63	M	Abdominal pain	Hematoma. Dissection and aneurysms in celiac and right external iliac arteries.	N/A	TPN	Improved

SAM: segmental arterial mediolysis, M: male, F: female, N/A: data not applicable, SMA: superior mesenteric artery, IMA: inferior mesenteric artery, RA: renal artery, CT: computed tomography, MCA: middle cerebral artery, VA: vertebral artery, TPN: Total parenteral nutrition

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the involvement of larger arteries than FMD, the rupture of an aneurysm in these arteries can be life-threatening. Histologically, vacuolization and lysis of the outer arterial media can be seen (8), which can lead to aneurysm, dissection, occlusion, and stenosis. Mesenteric SAM in the splenic, celiac, hepatic, and mesenteric arteries can cause abdominal symptoms, including nonspecific abdominal and flank pain, diarrhea, nausea, and back pain caused by aneurysm and dissection (2, 9, 10). CT and MRA have shown aneurysms, dissection, occlusion, and stenosis. Therapeutic options include antihypertensive therapy (11), embolization, bypass, and resection of the injured arteries. Patients presenting acutely with intraabdominal hemorrhaging are treated with emergent catheter angiography, endovascular intervention, or surgical treatment (12). Our Case 2 also suffered from abdominal pain, which had been caused by the minor rupture of a small aneurysm in the branch of the celiac or splenic artery; however, as the symptoms improved smoothly and the aneurysm was located on the main trunk of the celiac artery, no emergent intervention was performed. Fortunately, no recurrence has been seen to date; however, a careful follow-up of the aneurysm by imaging has been performed once every three to six months. In addition to our Case 2, Cases 12 and 13 improved with conservative therapy, although hemoperitoneum was found in abdominal cavity (Table 2). These three cases showed no progression of hemoperitoneum and no extravasation upon admission, so these signs may be markers supporting the selection of conservative treatment.

Due to difficulty in collecting tissue samples from the arteries in these areas, the importance of imaging studies is increasing, and although some similarities in the radiologic and histologic diagnoses have been reported for FMD and SAM, the two diseases show different clinical profiles in terms of the age of onset, gender, distribution of the affected arteries, imaging, symptoms, and treatment. It is therefore possible to diagnose these diseases clinically and suggest appropriate therapeutic options (Table 1, 2). For example, FMD affects middle-aged women, whereas there is no predilection for age or gender for SAM (3, 5, 13, 14). In addition, while FMD often shows stenosis and aneurysms in medium-sized arteries, including the renal, extracranial, carotid, and vertebral arteries (12), SAM shows changes in larger arteries, such as the celiac and mesenteric arteries (13), leading to a higher risk of arterial rupture and hemorrhaging from the weakened arterial wall in these larger arteries (15).

To improve our understanding of these diseases, we recently reviewed the reported cases of FMD and SAM in the gastroenterologic regions (3, 9, 11, 15-29) and reported the characteristics of imaging studies. For a further understanding of the clinical characteristics, we updated the information, focusing on cases reported within the past 20 years, since imaging modalities have shown significant advances in this time period (30-78). Based on the obtained information, CT revealed stenosis and aneurysmal changes in 33 cases (77%) of FMD and aneurysm, dissection, occlusion, and stenosis in 28 cases (88%) of SAM. In addition, hemorrhaging or hematoma was seen in 15 cases (47%) of SAM. Our cases also showed a similar pattern to the previously reported cases. Regarding the therapeutic options, open surgery was performed in 56%, endovascular intervention in 23%, antihypertensive therapy in 19%, and anticoagulation therapy in 11.6% for FMD. In contrast, open surgery was performed in 41%, endovascular intervention in 42%, antihypertensive therapy in 6.3%, and anticoagulation therapy in 6.3% for SAM (including Case 2). These data clearly demonstrate that early imaging studies and appropriate decision-making are essential for successful management.

Interestingly, 13 cases of FMD (30%) and 19 cases of SAM (59%), mainly recent cases, have been diagnosed without histological examinations and administered appropriate therapies, indicating that the accumulation of the information and results of imaging studies encouraged physicians to be suspicious of and diagnose the cases.

In conclusion, FMD and SAM are rare, and no standard diagnostic criteria or therapeutic methodologies have yet been established. The accumulation of similar cases and the summary of the clinical characteristics of the reported cases are important. In this report, we described two representative recent cases and summarized the findings of cases reported recently in order to improve the understanding and knowledge of these diseases. Further cases and the accumulation of clinical information will help physicians diagnose and treat such cases and facilitate the development of diagnostic criteria and standard therapeutic options.

Written informed consent was obtained from the patient for publication of this case report and accompanying images.

The study was reviewed and approved by the Institutional Review Board of Niigata University.

The authors state that they have no Conflict of Interest (COI).

References

1. Brinza EK, Gornik HL. Fibromuscular dysplasia: advances in understanding and management. Cleve Clin J Med **83** (11 Suppl 2): S45-S51, 2016.
2. Slavin RE, Saeki K, Bhagavan B, Maas AE. Segmental arterial mediolysis: a precursor to fibromuscular dysplasia? Mod Pathol **8**: 287-294, 1995.
3. Ko M, Kamimura K, Ogawa K, et al. Diagnosis and management of fibromuscular dysplasia and segmental arterial mediolysis in gastroenterology field: a mini-review. World J Gastroenterol **28**: 3637-3649, 2018.
4. Harrison EG Jr, McCormack LJ. Parhologic classification of renal arterial diease in renovascular hypertension. Mayo Clin Proc **46**: 161-167, 1971.
5. Olin JW, Froehlich J, Gu X, et al. The United States Registry for Fibromuscular Dysplasia: results in the first 447 patients. Circulation **125**: 3182-3190, 2012.
6. Sang CN, Whelton PK, Hamper UM, et al. Etiologic factors in

renovascular fibromuscular dysplasia. A case-control study. Hypertension **14**: 472-479, 1989.
7. Weinberg I, Gu X, Giri J, et al. Anti-platelet and anti-hypertension medication use in patients with fibromuscular dysplasia: results from the United States Registry for Fibromuscular Dysplasia. Vasc Med **20**: 447-453, 2015.
8. Slavin RE, Gonzalez-Vitale JC. Segmental mediolytic arteritis: a clinical pathologic study. Lab Invest **35**: 23-29, 1976.
9. Baker-LePain JC, Stone DH, Mattis AN, Nakamura MC, Fye KH. Clinical diagnosis of segmental arterial mediolysis: differentiation from vasculitis and other mimics. Arthritis Care Res **62**: 1655-1660, 2010.
10. Shenouda M, Riga C, Naji Y, Renton S. Segmental arterial Mediolysis: a systematic review of 85 cases. Ann Vasc Surg **28**: 269-277, 2014.
11. Veraldi GF, Zecchinelli MP, Furlan F, et al. Mesenteric revascularisation in a young patient with antiphospholipid syndrome and fibromuscular dysplasia: report of a case and review of the literature. Chir Ital **61**: 659-665, 2009.
12. Pillai AK, Iqbal SI, Liu RW, Rachamreddy N, Kalva SP. Segmental arterial mediolysis. Cardiovasc Interv Rad **37**: 604-612, 2014.
13. Olin JW, Gornik HL, Bacharach JM, et al. Fibromuscular dysplasia: state of the science and critical unanswered questions: a scientific statement from the American Heart Association. Circulation **129**: 1048-1078, 2014.
14. Lie JT. Segmental mediolytic arteritis: not an arteritis but a variant of arterial fibromuscular dysplasia. Arch Pathol Lab Med **116**: 238-241, 1992.
15. Sakano T, Morita K, Imaki M, Ueno H. Segmental arterial mediolysis studied by repeated angiography. Br J Rad **70**: 656-658, 1997.
16. Aboumrad MH, Fine G, Horn RC Jr. Intimal hyperplasia of small mesenteric arteries. Occlusive, with infarction of the intestine. Arch Pathol **75**: 196-200, 1963.
17. Ripley HR, Levin SM. Abdominal angina associated with fibromuscular hyperplasia of the celiac and superior mesenteric arteries. Angiology **17**: 297-310, 1966.
18. Wylie EJ, Binkley FM, Palubinskas AJ. Extrarenal fibromuscular hyperplasia. Am J Surg **112**: 149-155, 1966.
19. Claiborne TS. Fibromuscular hyperplasia. Report of a case with involvement of multiple arteries. Am J Med **49**: 103-105, 1966.
20. Lie JT, Kim HS. Fibromuscular dysplasia of the superior mesenteric artery and coexisting cerebral berry aneurysms. Angiology **28**: 256-260, 1977.
21. Rybka SJ, Novick AC. Concomitant carotid, mesenteric and renal artery stenosis due to primary intimal fibroplasia. J Urol **129**: 798-800, 1983.
22. Foissy P, Fabre M, Lebaleur A, et al. Aneurysm of the trunk of the superior mesenteric artery and polyaneurysmal disease of the right paracolic arcade of fibromuscular hyperplasia type. A case. Ann Med Interne (Paris) **135**: 530-532, 1984.
23. den Butter G, van Bockel JH, Aarts JC. Arterial fibrodysplasia: rapid progression complicated by rupture of a visceral aneurysm into the gastrointestinal tract. J Vasc Surg **7**: 449-453, 1988.
24. Salmon PJ, Allan JS. An unusual case of fibromuscular dysplasia. J Cardiovasc Surg **29**: 756-757, 1988.
25. Insall RL, Chamberlain J, Loose HW. Fibromuscular dysplasia of visceral arteries. Eur J Vasc Surg **6**: 668-672, 1992.
26. Moncure AC, Rashid A. Case 9-1995--A 60-year-old man with hypertrophic cardiomyopathy and ischemic colitis. N Engl J Med **332**: 804-810, 1966.
27. Stokes JB, Bonsib SM, McBride JW. Diffuse intimal fibromuscular dysplasia with multiorgan failure. Arch Intern Med **156**: 2611-2614, 1966.
28. Yamaguchi R, Yamaguchi A, Isogai M, et al. Fibromuscular dysplasia of the visceral arteries. Am J Gastroenterol **91**: 1635-1638, 1966.
29. Lee EK, Hecht ST, Lie JT. Multiple intracranial and systemic aneurysms associated with infantile-onset arterial fibromuscular dysplasia. Neurology **50**: 828-829, 1998.
30. Safioleas M, Kakisis J, Manti C. Coexistence of hypertrophic cardiomyopathy and fibromuscular dysplasia of the superior mesenteric artery. N Engl J Med **344**: 1333-1334, 2001.
31. Horie T, Seino Y, Miyauchi Y, et al. Unusual petal-like fibromuscular dysplasia as a cause of acute abdomen and circulatory shock. Jpn Heart J **43**: 301-305, 2002.
32. Kojima A, Shindo S, Kubota K, et al. Successful surgical treatment of a patient with multiple visceral artery aneurysms due to fibromuscular dysplasia. Cardiovasc Surg **10**: 157-160, 2002.
33. Felton TW, Drewe E, Jivan S, et al. A rare case of shock. Ann Rheum Dis **62**: 705-706, 2003.
34. Guill CK, Benavides DC, Rees C, et al. Fatal mesenteric fibromuscular dysplasia: a case report and review of the literature. Arch Intern Med **164**: 1148-1153, 2004.
35. Mertens J, Daenens K, Fourneau I, et al. Fibromuscular dysplasia of the superior mesenteric artery-case report and review of the literature. Acta Chir Belg **105**: 523-527, 2005.
36. Rodriguez Urrego PA, Flanagan M, Tsai WS, et al. Massive gastrointestinal bleeding: an unusual case of asymptomatic extrarenal, visceral, fibromuscular dysplasia. World J Gastroenterol **13**: 5771-5774, 2007.
37. Chaturvedi R, Vaideeswar P, Joshi A, Pandit S. Unusual mesenteric fibromuscular dysplasia a rare cause for chronic intestinal ischaemia. J Clin Pathol **61**: 237, 2008.
38. Malagò R, D'Onofrio M, Mucelli RP. Fibromuscular dysplasia: noninvasive evaluation of unusual case of renal and mesenteric involvement. Urology **71**: 755.e13-e15, 2008.
39. Kimura K, Ohtake H, Kato H, et al. Multivisceral fibromuscular dysplasia: an unusual case of renal and superior mesenteric involvement. Ann Vasc Dis **3**: 152-156, 2010.
40. Senadhi V. A rare cause of chronic mesenteric ischemia from fibromuscular dysplasia: a case report. J Med Case Rep **4**: 373, 2010.
41. Sugiura T, Imoto K, Uchida K, et al. Fibromuscular dysplasia associated with simultaneous spontaneous dissection of four peripheral arteries in a 30-year-old man. Ann Vasc Surg **25**: 838.e9-e11, 2011.
42. Dolak W, Maresch J, Kainberger F, et al. Fibromuscular dysplasia mimicking Crohn's disease over a period of 23years. J Crohns Colitis **6**: 354-357, 2012.
43. Patel NC, Palmer WC, Gill KR, Wallace MB. A case of mesenteric ischemia secondary to Fibromuscular Dysplasia (FMD) with a positive outcome after intervention. J Interv Gastroenterol **2**: 199-201, 2012.
44. Sekar N, Shankar R. Fibromuscular dysplasia with multiple visceral artery involvement. J Vasc Surg **57**: 1401, 2013.
45. Mitchell A, Caty V, Bendavid Y. Massive mesenteric panniculitis due to fibromuscular dysplasia of the inferior mesenteric artery: a case report. BMC Gastroenterol **15**: 71, 2015.
46. Yamada M, Nakada TA, Idoguchi K, Matsuoka T. Fibromuscular dysplasia presenting as hemorrhagic shock due to spontaneous rupture of a right gastroepiploic artery aneurysm. Am J Emerg Med **34**: 677e3-e5, 2016.
47. Erwin PA, Blas JV, Gandhi S, Romero ME, Gray BH. Visceral fibromuscular dysplasia in a patient with chronic abdominal pain. Vasc Med **21**: 170-171, 2016.
48. Shalan A, Hughes M, Nicholls M, et al. A challenging case of fibromuscular dysplasia in a transgender patient: is there a hormonal link? EJVES Short Rep **39**: 16-19, 2018.
49. Sakamoto Y, Hiruta R, Iijima A, et al. Sequential symptomatic arterial dissection in multiple vascular beds in a patient with fibromuscular dysplasia. Intern Med **57**: 2885-2887, 2018.

50. Rothenberg KA, McFarland GE, Stern JR. Endovascular repair of ruptured hepatic artery pseudoaneurysms secondary to fibromuscular dyslasia. Vasc Endovascular Surg **53**: 66-70, 2019.
51. Rengstorff DS, Baker EL, Wack J, et al. Intra-abdominal hemorrhage caused by segmental arterial mediolysis of the inferior mesenteric artery: report of a case. Dis Colon Rectum **47**: 769-772, 2004.
52. Chino O, Kijima H, Shibuya M, et al. A case report: spontaneous rupture of dissecting aneurysm of the middle colic artery. Tokai J Exp Clin Med **29**: 155-158, 2004.
53. Basso MC, Flores PC, De Azevedo, Marques A, et al. Bilateral extensive cerebral infarction and mesenteric ischemia associated with segmental arterial mediolysis in two young women. Pathol Int **55**: 632-638, 2005.
54. Obara H, Matsumoto K, Narimatsu Y, et al. Reconstructive surgery for segmental arterial mediolysis involving both the internal carotid artery and visceral arteries. J Vasc Surg **43**: 623-626, 2006.
55. Abdelrazeq AS, Saleem TB, Nejim A, et al. Massive hemoperitoneum caused by rupture of an aneurysm of the marginal artery of Drummond. Cardiovasc Intervent Radiol **31** (Suppl.2): S108-S110, 2008.
56. Shimohira M, Ogino H, Sasaki S, et al. Transcatheter arterial embolization for segmental arterial mediolysis. J Endovasc Ther **15**: 493-497, 2008.
57. Hashimoto T, Deguchi J, Endo H, Miyata T. Successful treatment tailored to each splanchnic arterial lesion due to segmental arterial mediolysis (SAM): report of a case. J Vasc Surg **48**: 1338-1341, 2008.
58. Hirokawa T, Sawai H, Yamada K, et al. Middle-colic artery aneurysm associated with segmental arterial mediolysis, successfully managed by transcatheter arterial embolization: Report of a case. Surgery Today **39**: 144-147, 2009.
59. Fujiwara Y, Takemura M, Yoshida K, et al. Surgical resection for ruptured aneurysm of middle colic artery caused by segmental arterial mediolysis: a case report. Osaka City Med J **56**: 47-52, 2010.
60. Ro A, Kageyama N, Takatsu A, et al. Segmental arterial mediolysis of varying phases affecting both the intra-abdominal and intracranial vertebral arteries: an autopsy case report. Cardiovasc Pathol **19**: 248-251, 2010.
61. Obara H, Matsubara K, Inoue M, et al. Successful endovascular treatment of hemosuccus pancreaticus due to splenic artery aneurysm associated with segmental arterial mediolysis. J Vasc Surg **54**: 1488-1491, 2011.
62. Tameo MN, Dougherty MJ, Calligaro KD. Spontaneous dissection with rupture of the superior mesenteric artery from segmental arterial mediolysis. J Vasc Surg **53**: 1107-1112, 2011.
63. Filippone EJ, Foy A, Galanis T, et al. Segmental arterial mediolysis: report of 2 cases and review of the literature. Am J Kidney Dis **58**: 981-987, 2011.
64. Taira S, Katori H, Matsuda Y, et al. Case report:a case of segmental arterial mediolysis(SAM) with bilateral renal infarction, superior mesenteric aneurysm and splenic aneurysm. Nihon Naikagakkai Zasshi (J Jpn Soc Intern Med) **100**: 1966-1968, 2011 (in Japanese, Abstract in English).
65. Yoo BR, Han HY, Cho YK, et al. Spontaneous rupture of a middle colic artery aneurysm arising from superior mesenteric artery dissection: diagnosis by color Doppler ultrasonography and CT angiography. J Clin Ultrasound **40**: 255-259, 2012.
66. Horsley-Silva JL, Ngamruengphong S, Frey GT, et al. Segmental arterial mediolysis: a case of mistaken hemorrhagic pancreatitis and review of the literature. JOP **15**: 72-77, 2014.
67. Gulati G, Ware A. Segmental arterial mediolysis: a rare non-inflammatory cause of mesenteric bleeding. BMJ Case Rep **2015**: 210344, 2015.
68. Fujinaga J, Kuriyama A. Segmental arterial mediolysis. J Emerg Med **51**: 732-733, 2016.
69. Shinoda N, Hirai O, Mikami K, et al. Segmental arterial mediolysis involving both vertebral and middle colic arteries leading to subarachnoid and intraperitoneal hemorrhage. World Neurosurg **88**: 694.e5-e10, 2016.
70. Galketiya K, Llewellyn H, Liang X. Spontaneous haemoperitoneum due to segmental arterial mediolysis and rupture of the left colic artery. ANZ J Surg **86**: 201-202, 2016.
71. Kuriyama A. Segmental arterial mediolysis. Am J Emerg Med **35**: 518.e1-e2, 2017.
72. Yoshioka T, Araki M, Ariyoshi Y, et al. Successful microscopic renal autotransplantation for left renal aneurysm associated with segmental arterial mediolysis. J Vasc Surg **66**: 261-264, 2016.
73. Japikse RD, Svenson JE, Pickhardt PJ, et al. Segmental arterial mediolysis: an unusual case mistaken to be a strangulated hernia. WMJ **116**: 173-177, 2017.
74. Akuzawa N, Kurabayashi M, Suzuki T, et al. Spontaneous isolated dissection of the superior mesenteric artery and aneurysm formation resulting from segmental arterial mediolysis: a case report. Diagn Pathol **12**: 74, 2017.
75. Kalfa M, Kocanaogullari H, Karabulut G, et al. Segmental arterial mediolysis mimics systemic vasculitis. Eur J Rheumatol **3**: 136-138, 2016.
76. Liao CY, Kuo WH, Huang EH, et al. A diagnostic dilemma: acute abdomen presenting as segmental arteril mediolysis masked by a ruptured hepatocellular carcinoma. Gastroenterol Rep (Oxf) **5**: 244-246, 2017.
77. Lee J, Ahn HY, I H, et al. Spontaneous intra-abdominal haemorrhage due to segmental arterial mediolysis following oesophagocolojejunostomy. Interact Cardiovasc Thorac Surg **25**: 993-994, 2017.
78. Hayashi S, Hosoda K, Nishimoto Y, et al. Unexpected intraabdominal hemorrhage due to segmental arterial mediolysis following subarachnoid hemorrhage: a case of ruptured intracranial and intraabdominal aneurysms. Surg Neurol Int **9**: 175, 2018.