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MINIREVIEWS

Diagnosis and management of fibromuscular dysplasia and segmental arterial mediolysis in gastroenterology field: A mini-review

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Abstract

The vascular diseases including aneurysm, occlusion, and thromboses in the mesenteric lesions could cause severe symptoms and appropriate diagnosis and treatment are essential for managing patients. With the development and improvement of imaging modalities, diagnostic frequency of these vascular diseases in abdominal lesions is increasing even with the small changes in the vasculatures. Among various vascular diseases, fibromuscular dysplasia (FMD) and segmental arterial mediolysis (SAM) are noninflammatory, nonatherosclerotic arterial diseases which need to be diagnosed urgently because these diseases could affect various organs and be lethal if the appropriate management is not provided. However, because FMD and SAM are rare, the cause, prevalence, clinical characteristics including the symptoms, findings in the imaging studies, pathological findings, management, and prognoses have not been systematically summarized. Therefore, there have been neither standard diagnostic criteria nor therapeutic methodologies established, to date. To systematically summarize the information and to compare these disease entities, we have summarized the characteristics of FMD and SAM in the gastroenterological regions by reviewing the cases reported thus far. The information summarized will be helpful for physicians treating these patients in an emergency care unit and for the differential diagnosis of other diseases showing severe abdominal pain.

Key words: Fibromuscular dysplasia; Segmental arterial mediolysis; Mesenteric lesion; diagnosis; Humans

Core tip: The vascular diseases in the abdominal lesions needs to be appropriately diagnosed and treated as it could be lethal if the appropriate management is not provided. Mesenteric ischemia caused by the atherosclerotic changes is rather famous however, fibromuscular dysplasia (FMD) and segmental arterial mediolysis (SAM) which are noninflammatory, nonatherosclerotic arterial diseases are rare and the cause, prevalence, clinical characteristics including the symptoms, findings in the imaging studies, pathological findings, management, and prognoses have not been systematically summarized. Therefore, we have summarized the characteristics of FMD and SAM in the gastroenterological regions and review the cases reported thus far. The information summarized will be helpful for physicians treating these patients in an emergency care unit and for the differential diagnosis of other diseases showing severe abdominal pain.

Ko M, Kamimura K, Ogawa K, Tominaga K, Sakamaki A, Kamimura H, Abe S, Mizuno K, Terai S. Diagnosis and management of fibromuscular dysplasia and segmental arterial mediolysis in gastroenterology field: A mini-review. *World J Gastroenterol* 2018; 24(32): 3637-3649 Available from: URL: http://www.wjgnet.com/1007-9327/full/v24/i32/3637.htm DOI: http://dx.doi.org/10.3748/wjg.v24.i32.3637

INTRODUCTION

A literature search was conducted using PubMed and Ovid, with the term "fibromuscular dysplasia" or "segmental arterial mediolysis" and "mesenteric" to extract studies published in the last 55 years for fibromuscular dysplasia and in the last 21 years for segmental arterial mediolysis. We summarized the available information on demographics, clinical symptoms, image studies, histological findings, treatment, and clinical course.

FIBROMUSCULAR DYSPLASIA

Clinical characteristics

The detailed clinical and pathological classification of fibromuscular dysplasia (FMD) was first reported by Harrison and McCormack in 1971[1]. Since then, several studies regarding clinical course and histological data have been published, and recently, data from the first 447 patients from the United States Registry (US Registry) for FMD have been reported[1]. FMD is a noninflammatory, nonatherosclerotic arterial disease of the medium-sized arteries throughout the body, which could lead to arterial stenosis, occlusion, aneurysm, and dissection[2]. The details of the disease have not yet been clarified; however, it is typically found in the renal, extracranial, carotid, and vertebral arteries[2].

The disease is rare, with a frequency of 0.02%, predominantly occurring in women (91%) with a mean age of 55.7 ± 13.1 years especially in the Caucasian (95.4%)[2].

The mean patient age at first symptom or sign of FMD was 47.2 years (range, 5-83 years)[2]. The mechanisms underlying the pathogenesis of FMD are still poorly understood; however, smoking, hormones, HLA-DRw6 polymorphism, and physiological stimulation have been reported to be risk factors[3]. For example, a significant dose-response relationship between cigarette smoking and the presence of FMD has been reported, with an odds ratio of 8.6 when having smoked more than 10 pack-years of cigarettes[3]. The risk of HLA-DRw6 was reported with an odds ratio of 5.0, adjusted for the level of smoking[3].

FMD can occur in any medium-sized arteries throughout the body, and dissection and aneurysm have been identified in 19.7% and 17.0% of FMD patients, respectively. The three major sites affected with dissection are the carotid arteries (14.8% of all patients enrolled), followed by renal arteries (4.3%), and vertebral arteries (3.4%)[2,4]. The three major sites affected with aneurysm are the renal arteries (5.6% of all patients enrolled), followed by carotid arteries (3.6%), and the aorta (3.4%)[2]. FMD in abdominal lesions, classified as mesenteric FMD, which is caused by the celiac and mesenteric arteries, is a rare condition and often presents as an incidental diagnosis[2]. On the basis of the US Registry data, mesenteric ischemia was reported in only 1.3% of cases, with aneurysm and dissection in these vessels accounting for 6.8% and 22.3% of all cases reported, respectively[2].

Symptoms and imaging

The clinical symptoms depend on the vessels involved. When the renal arteries are affected, renovascular hypertension can be observed. Thus, when the carotid arteries are affected, headache, pulsatile tinnitus, and dizziness are the major symptoms[2]. Mesenteric FMD involves the celiac and mesenteric arteries; therefore, mesenteric ischemic symptoms occur, including unspecific abdominal pain. We reviewed the literature describing the cases and have presented the information in Table 1[5-37]. Our literature review summarized a total of 39 cases of mesenteric FMD, showing predominance in women, as reported, and the median age was 45.2 years (range: 19-78 years). Regarding the risk factors, four patients smoked (10%), two patients had smoking histories (5%), and one patient had taken oral contraceptive pills (2.6%). The most common presenting symptom was abdominal pain (62%), followed by hypertension, diarrhea, nausea or vomiting, and headache. Although approximately 80% of cases showed symptom improvement, eight patients (20%) died because of the severity of the intestinal

Table 1 Summary of mesenteric fibromuscular dysplasia reported to date

Case (*n*)	Ref.	Age (yr)	Gender (Male/ Female)	Risk factors	Symptoms	Vessels Involved	CT	Angiography	Pathology	Treatment	Anti-hypertensive drug	Anti-coagulants	Outcome
1	[5]	62	M	N/A	Upper abdominal pain, hemoperitoneum, shock	Celiac, SMA, IMA, RA	N/A	N/A	Intimal thickening in the branches of the SMA and IMA.	Laparotomy	None	None	Died
2	[6]	45	F	N/A	Abdominal pain	SMA, RA	N/A	N/A	N/A	Ileal resection	N/A	N/A	Improved
3	[6]	50	F	N/A	Hypertension, abdominal pain, diarrhea	SMA, RA, iliac	N/A	Stenosis and string-of-beads like Appearance in the SMA	N/A	SMA revascularization	N/A	N/A	Improved
4	[7]	73	F	N/A	N/A	Celiac, SMA, iliac	N/A	N/A	N/A	N/A	N/A	N/A	Improved
5	[7]	42	F	N/A	N/A	SMA, RA	N/A	N/A	N/A	N/A	N/A	N/A	Improved
6	[7]	50	F	N/A	Hypertension	Celiac, SMA, RAl	N/A	Minimal defects in the SMA and RA; stenosis of celiac artery	N/A	N/A	N/A	N/A	Improved
7	[7]	37	F	N/A	Visceral ischemic symptoms	Celiac, SMA, RA	N/A	N/A	N/A	Revascularization	N/A	N/A	Improved
8	[7]	47	F	N/A	Hypertension, abdominal pain	Celiac, SMA, RA	N/A	Defects in the SMA and RA	Medial hyperplasia	None	N/A	N/A	Died
9	[8]	41	F	N/A	Hypertension	SMA, internal carotid, RA, iliac	N/A	Corkscrew and string-of-beads like appearance in the RA, carotid, iliac artery	Replacement of the normal media with disorganized fibrous and muscular hyperplasia	Thromboendarerectomy on SMA	N/A	N/A	Died
10	[9]	64	F	N/A	Unconsciousness	SMA, circle of Willis	N/A	N/A	Medial hyperplasia	None	None	None	Died
11	[10]	21	M	N/A	Hypertension	Celiac, SMA, RA, carotid	N/A	Stenosis of celiac, SMA, RA	Intimal fibroplasia	Anti-hypertensive drug; revascularization of carotid artery; angioplasty of RA	Yes, N/A	N/A	Improved
12	[10]	20	F	N/A	Hypertension	SMA, IMA, RA, carotid	N/A	Stenosis of carotid, renal, SMA. Total occlusion of the IMA.	Intimal fibroplasia	Anti-hypertensive drug, subclavian-carotid bypass; vascular reconstruction of the kidney	Yes, N/A	N/A	Improved
13	[11]	55	M	N/A	N/A	SMA	N/A	N/A	Intimal hyperplasia in SMA	None	N/A	N/A	N/A
14	[12]	44	F	N/A	Asymptomatic bruit of the aortoiliac system	Celiac, SMA, RA, iliac	N/A	String-of-beads like appearance of iliac artery; aneurysms of SMA, RA	Intimal fibrosis with development of fibrosis	Resection and reconstruction	N/A	N/A	Improved
15	[13]	58	F	N/A	Body weight loss	Celiac, SMA, IMA, RA, iliac, aorta	N/A	Occlusion of celiac, SMA, IMA.	N/A	Open surgery	N/A	N/A	Improved
16	[14]	46	F	None (non-smoker)	Palpitations, headache, hypertension	Celiac, SMA	N/A	Aneurysms in the right RA, celiac; occlusion in the left gastric artery	Muscle hypertrophy and disorganisation of elastic tissue of the media in celiac artery.	Surgical ligation	N/A	N/A	Improved

17	[15]	60	M	None (non-smoker)	Left abdominal pain, diarrhea	SMA	Irregular nodular thickening in transverse colon.	Stenoses of the SMA	Intimal fibrosis and focal replacement of medial smooth-muscle fibers by fibrous tissue .	Splenic flexure resection and angioplasty	N/A	N/A	Improved
18	[16]	54	F	Smoking	Hypertension, headache, abdominal pain	SMA, RA, coronary	Liver cyst	Stenosis of the coronary arteries	Intimal hyperplasia in SMA, RA, coronary, splenic, intrahepatic artery	Anti-hypertensive drug	α,β-blocker → Ca blocker	N/A	Died
19	[17]	39	M	N/A	Melena, lower abdominal pain	Jejunal, Sigmoid	N/A	String-of-beads like appearance in the jejunal and sigmoid arteries.	Adventitia is thickened by fibroplasia	Resection of the jejunum	N/A	N/A	Improved
20	[18]	23	M	N/A	Hypertension	Celiac, SMA, RA, carotid, vertebral, opthalmic, superficial temporal, iliac, lumber, intercostal	Hematomasin the paraduodenal and right superior gluteral lesion and splenic infarction	Multiple saccular aneurysms in the celiac, SMA, RA< splenic, hepatic, iliac, lumber, and intercostal arteries	Mediolytic FMD with segmental dissection and thrombosis	Embolization of the gastroduodenal and right SMA to prevent hemorrhage	N/A	N/A	Improved
21	[19]	33	M	N/A	Abdominal pain	SMA	N/A	String-of-beads like appearance in the SMA	Thickening of the media due to hyperplasia in SMA	Ileal resection	N/A	N/A	Improved
22	[20]	78	F	N/A	Hypertension, abdominal pain, hemoperitoneum.	SMA, RA, colonary	Dilated loop of the small bowel and fluid in the peritoneal cavity.	N/A	Medial and perimedial fibrodysplasia, forms the characteristic petal-like appearance in SMA.	None	None	None	Died
23	[21]	43	M	None (non-smoker)	No symptoms	SMA, iliac	SMA aneurysm	Aneurysms in the SMA, hepatic artery, splenic artery, jejunal artery, iliac arteries.	Medial fibrodysplasia in the arteral walls	Aneurysm resection and arterial reconstruction	N/A	N/A	Improved
24	[22]	48	F	None (non-smoker)	Abdominal pain, hemoperitoneum	Celiac, SMA, RA	N/A	Multiple small aneurysms in celiac, SMA, RA	N/A	Surgical hemostasis and anti-hypertensive drugs	β-blocker	N/A	Improved
25	[23]	57	F	Smoking (40 packs/yr)	Abdominal pain, weight loss, anorexia, nausea, vomiting, diarrhea.	Celiac, SMA	Nothing particular	Stenosis of the celiac artery and SMA	Medial thickening, smooth muscle hyperplasia in SMA and celiac artery	Aortoiliac and aorto-SMA bypass	N/A	N/A	Died
26	[24]	48	F	Smoking (20 packs/yr)	Abdominal pain	Celiac, SMA, IMA	N/A	Occlusion of the celiac, SMA; enlarged hypertrophic IMA	Intimal fibroplasia and an increased deposition of fibrous tissue in the vessel wall media	Reimplantation of the SMA	N/A	N/A	Improved
27	[25]	38	M	Smoking	Gastrointestinal bleeding, anemia	SMA, IMA	N/A	Ectasia in IMA; string-of-beads like appearance in the SMA	Thickening and hyalinization of medium sized vessel walls, with intimal proliferation.	Ileal resection	N/A	N/A	Improved

28	[26]	Not provided	Not provided	None (non-smoker)	Abdominal pain, distension, constipation	SMA	N/A	N/A	Thick cuff (petal like) of smooth muscle proliferation with normal intima and media in mesenteric artery.	Right hemicolectomy	N/A	N/A	Improved
29	[27]	43	F	Smoking (10 cigarettes daily for 20 yr)	Hypertension, headache	SMA, RA	N/A	String-of-beads like appearance in the right RA and SMA; stenosis and multiple irregularities in the left RA	N/A	Angioplasty and anti-hypertensive drugs	Yes, N/A	N/A	Improved
30	[28]	38	M	N/A	N/A	N/A	N/A	N/A	N/A	N/A	N/A	N/A	Improved
31	[29]	43	F	N/A	Hypertension, abdominal pain, headache	SMA, RA	Aneurysms in the left RA	Aneurysms in the left RA with sever fibrodysplastic stenosis; string-of-beads like appearance in the right RA; stenosis in SMA	Intimal fibroplasia, lost of internal elastic lamina, and massive destruction of the media in the aneurysm walls	Aneurysm resection and aortorenal bypass and percutaneous transluminal angioplasty	N/A	N/A	Improved
32	[30]	44	F	Oral contraceptive pills	Hypertension, abdominal pain, diarrhea, vomiting	SMA	Stenosis of SMA and nonspecific colitis	stenosis in SMA	N/A	Angioplasty	N/A	N/A	Improved
33	[31]	30	M	N/A	Abdominal pain, hypertension	Celiac, SMA, RA, iliac	Dissections of the celiac, SMA, left RA, and external iliac artery	stenosis in the right RA	N/A	Anti-platelet and anti-hypertensive therapy and angioplasty for right renal artery.	β-blocker, Ca blocker	warfarin, aspirin (100mg)	Improved
34	[32]	47	F	N/A	Abdominal pain, diarrhea, hypertension	All abdominal arteries	A partial occlusion of the celiac artery and a total occlusion of the SMA	N/A	Intimal and medial proliferation	Anti-hypertensive drug	Yes, N/A	N/A	Died
35	[33]	47	F	None (non-smoker)	Nausea, early satiety, abdominal pain	Celiac, SMA	N/A	Stenosis of the SMA, hypertrophy of the gastroduodenal artery and pancreaticoduodenal arteries	N/A	An aorto-superior mesenteric artery and an aorto-hepatic artery bypass.	N/A	N/A	Improved
36	[34]	19	F	N/A	Abdominal pain, vomiting	SMA, RA	N/A	Stenosis of the origin of the SMA and multiple aneurysms involving the proximal SMA. Right renal artery is mild irregularity.	N/A	Resection of the aneurysmal segment in the SMA; aorto-SMA interposition graft with polytetrafluoroethylene	N/A	N/A	Improved
37	[35]	52	M	Smoking	Abdominal pain	IMA	Stenosis of the IMA	N/A	Necrosis of the mucosa; fibrosis of the inima	Left hemicolectomy	N/A	N/A	Improved

38	[36]	20	F	N/A	Abdominal pain, hemorrhagic shock	SMA, right gastroepiploic, jejunal	Intraperitoneal bleeding in the omental bursa and mesentery of the transverse colon	String-of-beads like appearance in the jejunal artery	N/A	Transcatheter arterial embolization	N/A	N/A	Improved
39	[37]	61	F	N/A	Abdominal pain	SMA, IMA, RA	N/A	Multiple aneurysms and stenoses in SMA, IMA, RA	Multiple tears and dissections of the medial layer and fibrointimal thickening	Anti-coagulation	None	Yes, N/A	Improved

M: Male; F: Female; N/A: Data not applicable; SMA: Superior mesenteric artery; IMA: Inferior mesenteric artery; RA: Renal artery; CT: Computed tomography.

ischemia.

Reflecting the changes in stenosis, dissection, and aneurysm in the medium-sized arteries, FMD leads to the narrowing of the vasculature and shows a beaded appearance[38]. Therefore, catheter-based angiography has been considered to be the gold standard imaging modality; however, recent progress in imaging, such as computed tomography (CT) with high resolution, could support the diagnoses by determining the vessels affected by the disease. With the information obtained from imaging, the disease is classified into four types: multifocal, which comprises 62% of cases, showing multiple stenosis and string-of-beads; tubular, which comprises 14% of cases, showing long concentric stenosis; focal, which comprises 7% of cases, showing short stenosis of less than 1 cm; and mixed[39]. Our summary also showed aneurysms, stenosis, dissection, and occlusions in the cases for which information was available.

Histology

Histopathological findings are characteristics of the disease; thinned media and thickened fibromuscular ridges in which the arterial muscle is replaced by the fibroplasia can be observed. Based on this, the characteristic classification of FMD is essentially based on the arterial layer in which the dysplasia is predominant: intimal fibroplasia, medial fibroplasia, perimedial fibroplasia, medial hyperplasia, and adventitial fibroplasia[40-43]. Intimal fibroplasia, a relatively rare form of the disease, is characterized by focal eccentric or circumferential protuberant intimal proliferation. Medial fibroplasia, the most common type, accounts for more than 70% of this disorder, and angiography shows a typical "string of beads" appearance. Perimedial fibroplasia is the second-most common form of this disorder and is characterized by the accumulation of circumferential aggregations of elastic tissue between the media and the adventitia. Medial hyperplasia is an uncommon form and is characterized by apparent hyperplasia of normal medial smooth muscle with minimal architectural disorganization. Adventitial fibroplasia is characterized by collagenous fibroplasia encircling the adventitia and extending into the surrounding periarterial fibroadipose tissue[20]. In our case summary, fibromuscular change was confirmed histologically in the medial layer in 10 patients (26%), the intimal layer in 9 patients (23%), in both layers in 5 patients (13%), and in the adventitia layer in 1 patient (2.6%) (Table 1).

Treatment

The long-term outcomes of this disease entity have not been clarified to date, and no randomized clinical trials have been conducted to develop a standard treatment for this disease. Therapeutic options have been chosen on the basis of factors such as disease location, symptoms, prior history of symptoms, and the presence and size of aneurysms. Given that FMD often shows ischemic changes causing hypertension and stroke, most patients are treated with anti-platelet, anti-thrombotic, and anti-hypertensive therapy[44]. Anti-hypertensive medications are administered to 71.7% of patients. The median number of medications patients received was one, and 21.5% of patients received three or more anti-hypertensive medications. The most commonly used agents were beta-blockers (40.0%), diuretics (31.3%), and calcium channel antagonists (25.7%). A total of 21.0% of patients received an angiotensin-converting enzyme inhibitor, 21.6% received an angiotensin receptor blocking agent, and 0.8% received both[44]. The use of anti-hypertensive agents is related to the history of hypertension medication, body mass index, and renal function[44]. The use of anti-platelet treatment is associated with cerebrovascular involvement[44]; however, for this entity of medicines, further studies are necessary to determine the clinically meaningful patient outcomes. In our case

summary, insufficient information about medications was provided; thus, the actual number treated with antihypertensive therapy might be lower than 18% (Table 1). Anti-coagulation therapy was attempted for two patients (5%), including one patient each receiving warfarin and aspirin.

Vascular intervention and surgery for revascularization are considered with the appropriate clinical symptoms and are rarely performed other than for the renal artery[44]. For the renal artery, endovascular revascularization using the percutaneous transluminal angioplasty technique or surgical procedures are considered when the patients show hypertension resistant to a regimen of three anti-hypertension drugs, including diuretics, or in cases of renal artery aneurysm or renal artery dissection[2,4,38]. Thus far, no randomized clinical trials of revascularization *vs* medication have been conducted. For other arteries, including the carotid artery, given that FMD is not an atherosclerotic disease, stenting or surgical procedures are not the standard therapy, and medication with anti-platelet, anti-coagulant, and anti-hypertensive agents are the main treatment. However, when symptomatic, interventional radiology using the percutaneous transluminal angioplasty technique can be considered, although it is controversial[38]. In our case summary, open surgery was performed on 23 patients (59%) and endovascular intervention was performed on 9 patients (23%).

Prognosis

Though the prognosis is basically good, when FMD affects the cerebrovascular system, there is a risk of cerebral infarction and rupture. A larger number of cases are necessary to accumulate the information useful to conduct randomized clinical trials.

SEGMENTAL ARTERIAL MEDIOLYSIS

Clinical characteristics

Segmental arterial mediolysis (SAM) was first reported by Slavin and Gonzalez-Vitale in 1976[45] and is a rare disease entity for which 50 cases have been reported to date. It is defined as a nonatherosclerotic, noninflammatory disruption of the arterial medial layer of a medium- to large-sized artery. Histologically, it is characterized as vacuolization and lysis of the outer arterial media[45]. Because of its rarity and difficulty in differential diagnosis from the other vascular diseases, clinical information is insufficient, and little is known to date; however, no significant predominance of sex or age has been reported. The mechanisms underlying the pathogenesis of SAM that have been reported as risk factors are hypoxia, shock, aging, hypertension, circulatory disturbance, arteriospasm, and other vasoconstrictor stimuli[45-47].

Symptoms and imaging

For the abdominal lesion, the most common symptom is nonspecific abdominal and flank pain[46]; diarrhea, nausea, back pain, headache, hypertension, loss of consciousness, and hemiparesis have also been known to be symptoms, although not specific[47]. We reviewed the literature describing the cases and have summarized the information in Table 2[47-71]. The studies reported a total of 26 cases of mesenteric SAM, of which 17 were men and 9 were women, with a slight predominance in men. The median age was 53 years (range: 25-79 years). The most common presenting symptom was abdominal pain (78%), followed by various symptoms, including shock, diarrhea, nausea, back pain, headache, anorexia, hypertension, hemiparesis, and loss of consciousness (Table 2).

With the development of various imaging modalities, it has been reported that, in various combinations, SAM typically affects splenic, celiac, hepatic, mesenteric, and renal arteries in the abdominal lesion[47,72]. Because of the involvement of the celiac artery, splenic arterial aneurysm is frequently found, and its rupture could affect the prognosis. Angiography reveals aneurysms, dissections, occlusions, and stenosis; however, the findings could overlap with those found in collagen vascular diseases and FMD. Therefore, the differential diagnoses between the vascular diseases are based on the histopathological findings. SAM is difficult to distinguish from FMD, although FMD shows predominance in young women and affects renal arteries causing hypertension, whereas SAM commonly affects the celiac arteries. In addition, the clinical course shows ischemic changes in FMD, whereas SAM often causes profuse bleeding from the intestinal arteries. However, these findings often overlap each other; therefore, accumulation of more detailed information is necessary.

Histology

Although the suspicion of SAM is the basis of clinical and radiological features, the gold standard for diagnosis is a pathological finding involving injurious and reparative phases in the arterial lesions of the surgical specimens. These injurious states include mediolysis, separation of the outer media, and formation of arterial gaps; key is that there is no evidence of inflammation. These changes reflect the vascular aneurysms frequently found as angiographic features of this condition. Commonly, the inflammatory markers are negative and genetic diagnosis for collagen vascular disorders shows a normal pattern.

Treatment

The long-term prognosis is unclear, and no standard therapeutic strategy has been proposed, to date; however, given that some SAM cases showed sudden the onset of aneurysm rupture, the condition could be life threatening. Therefore, SAM treatment includes embolization, bypass, and resection of the injured arteries. In addition, anti-hypertensive therapy[28] could prevent further worsening of the arterial lesions. Anti-

Table 2 Summary of mesenteric segmental arterial mediolysis reported to date

Case (*n*)	Ref.	Age (yr)	Gender (Male/ Female)	Risk factors	Symptoms	Vessels Involved	CT	Angiography	Pathology	Treatment	Anti-hypertensive drug	Anti-coagulants	Outcome
1	[48]	65	F	N/A	Abdominal pain	SMA	N/A	Beaded appearance and stenosis of the MCA	Lysis and destruction in the media and intima	Resection of aneurysm in MCA	N/A	N/A	Improved
2	[49]	56	F	N/A	Abdominal pain	IMA	Intraabdominal hemorrhage	Aneurysm in IMA	N/A	Left hemicolectomy	N/A	N/A	Improved
3	[50]	78	M	N/A	Abdominal pain, diarrhea, shock	SMA	N/A	N/A	Destruction of the tunica interna and media in MCA	Emergent surgery (right hemicolectomy); a large hematoma and a ruptured aneurysm upon the surgery	N/A	N/A	Improved
4	[51]	35	F	N/A	Abdominal pain, perforation on transverse colon	SMA	Occlusion of the mesenteric vein and ischemic colitis	Unremarkable	Segmental vacuolar degeneration of smooth muscle with areas of wall thinning	Resection of terminal ileum	N/A	N/A	Died
5	[52]	52	M	N/A	Sudden hemiparesis, hypertension	Celiac, SMA, IMA hepatic artery	Aneurysm in the celiac, hepatic, SMA	Aneurysms in celiac, SMA, ICA, hepatic; stenoses in celiac and SMA	Multiple segmental mediolysis lesions of the muscular and elastic fibers of the media	Reconstruction of hepatic and celiac artery using autologous saphenous vein graft	N/A	N/A	Improved
6	[53]	49	M	N/A	Abdominal pain, shock	SMA	Large hematoma surrounding a high-density aneurysm	Beaded appearance in SMA	Multifocal fragmentation of the elastic fibers of the media	Right hemicolectomy	N/A	N/A	Improved
7	[54]	57	M	N/A	Abdominal pain	SMA, hepatic	Small aneurysm at the middle colic artery and mesenteric hematoma	Aneurysm and stenosis of the celiac, SMA, hepatic artery	N/A	Embolization with N-butyl cyanoacrylate for aneurysm in the SMA	N/A	N/A	Improved
8	[54]	76	F	N/A	Abdominal pain, nausea	IMA	Mesenteric hematoma	Aneurysm in IMA	N/A	Embolization with coil	N/A	N/A	Died
9	[55]	59	M	N/A	Abdominal pain, shock	SMA, RA, gastroepiploic, splenic	SMA dissection, aneurysm in RA, gastroepiploic, splenic artery; rupture of the splenic aneurysm	Saccular aneurysms and multiple stenotic region in gastroepiploic artery	Medial island spared from mediolysis	Emergency embolization of the splenic artery, resection of aneurysm in the gastroepiploic	N/A	N/A	Improved
10	[56]	57	M	N/A	Abdominal pain, diarrhea	SMA	Ascites throughout the abdomen	Aneurysm in SMA	N/A	Transcatheter arterial embolization	N/A	N/A	Improved
11	[57]	60	M	N/A	N/A	SMA	Rupture of the aneurysm of the MCA	Multiple beaded patters and aneurysm in SMA	N/A	Surgical resection	N/A	N/A	Improved
12	[47]	25	F	N/A	Anorexia, abdominal pain, diarrhea	SMA, hepatic	Ischemic colitis of the splenic flexure	Occlusion of IMA; 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	N/A	N/A	Improved
13	[58]	53	M	N/A	None	Celiac, SMA, splenic	Aneurysm in splenic, celiac, SMA; dissection in the celiac.	Aneurysm in the celiac, splenic, and SMA	N/A	Embolization with coil and aortic stent graft	N/A	N/A	Improved

14	[59]	51	M	N/A	Abdominal pain, shock	SMA, IMA	Abdominal hemorrhage	Active bleeding from SMA	N/A	Embolization and ligation of the branches of the SMA	N/A	Warfarin	Improved
15	[60]	29	F	N/A	Hypertension	SMA, RA, hepatic	Renal cortical nephrograms	Scattered microaneurysms in SMA, RA, hepatic artery	Segmental lesions of the media with loss of smooth muscle cells	Anti-coagulants	N/A	Warfarin	Improved
16	[61]	55	F	N/A	Abdominal pain	Celiac, SMA, hepatic, splenic	Unremarkable in vessels	Aneurysms in celiac, SMA, hepatic, splenic artery	N/A	Anti-coagulants	N/A	Warfarin followed by aspirin	Improved
17	[62]	56	M	N/A	Abdominal pain, shock	SMA	Aneurysm in MCA, SMA dissection	Saccular aneurysms in the MCA; dissections in the SMA	N/A	Embolization with coil	N/A	N/A	Improved
18	[63]	64	F	N/A	Abdominal pain, back pain, nausea	SMA, IMA, hepatic	Hematoma in the anterior pararenal space inferior to pancreatic tail; bleeding from aneurysm	Multiple aneurysms in the SMA, IMA, hepatic artery	N/A	Conservative	N/A	N/A	Improved
19	[64]	60	F	Hypoxia	Hypoxia, hypotension, cardiopulmonary arrest	SMA	Large hematoma in the retroperitoneal and intraperitoneal space; SMA aneurysm	Aneurysms and beaded appearance in the SMA	N/A	Conservative	N/A	N/A	Improved
20	[65]	36	M	N/A	Abdominal pain	Celiac, hepatic, anterior inferior pancreaticoduodenal artery	Stenosis and aneurysms in anterior inferior pancreatic duodenal artery	Aneurysms and beaded like appearance in the anterior inferior pancreaticoduodenal artery	N/A	Embolization with coil	N/A	N/A	Improved
21	[66]	47	M	N/A	Loss of consciousness, headache, abdominal pain	SMA	SAH, massive intraperitoneal hematoma	Beaded like appearance in SMA; dissection in 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.	N/A	N/A	Improved
22	[67]	79	M	N/A	Abdominal pain, hypotension	IMA	Active bleeding from IMA and hemorrhage	N/A	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	N/A	N/A	Improved
23	[68]	40	M	N/A	Abdominal pain	Celiac, SMA	Extensive dissection of SMA with the thrombotic occlusion. stenosis and dilation of celiac artery	N/A	N/A	Conservative	N/A	N/A	Improved
24	[69]	32	M	N/A	Abdominal pain	IMA, RA	Aneurysm in renal and IMA, massive amount of hemorrhage	Stenosis and aneurysm in the RA	Media shows myxoid degeneration in the outer one-third adjacent to the adventitia	Surgical hemostasis and left hemicolectomy	Yes, N/A	N/A	Improved
25	[70]	58	M	N/A	Abdominal pain	SMA	Mesenteric hematoma and right inguinal hernia with unremarkable small bowel	Beaded like appearance in SMA	N/A	Immunosuppressive therapy	N/A	N/A	Improved

26	[71]	57	M	N/A	Hypertension, abdominal pain	SMA	Arterial dissection with luminal stenosis and aneurysm formation at the distal portion of the SMA	Segmental dilatation, aneurysm in the SMA	Vacuolization and decrease in the number of vascular smooth muscles	Aneurysmectomy and bowel resection	Ca-blocker	N/A	Improved

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.

coagulation therapy is uncommon, and only a few cases have been reported to date[59-61]. In addition, given it is a noninflammatory disorder, no evidence of efficacy in use of anti-inflammatory agents or immunosuppressive agents has been reported. However, SAM has been treated with these agents when the differential diagnosis from the other arteritis was difficult[70].

For patients presenting acutely with intra-abdominal hemorrhage, patients are treated with emergent catheter angiography, endovascular intervention, or surgical treatment[73].

Shenouda reported that coil embolization was the most common endovascular intervention and was reported as successful in 88% of patients, with no mortality, whereas the open surgical approach was associated with a 9% mortality rate[72]. In our patient summary, open surgery was most commonly performed, and this was performed on 13 patients (50%). Endovascular intervention was performed on eight patients (31%), and anti-coagulation therapy was administered to two patients (7.7%), including warfarin and aspirin administration. Anti-hypertensive therapy was administered to one patient with Ca-blocker.

Prognosis

Although the prognosis of the disease is reported to be good when managed appropriately[72], SAM can be fatal when ruptured[49,73]. Therefore, a careful diagnosis and appropriate management are essential for this disease entity. Our case summary also showed that although 24 (92%) patients improved, 2 (7.7%) patients died, 1 having had a large hematoma and a ruptured aneurysm in the mesenteric lesion that was revealed upon the emergent surgery.

DISCUSSION

The inner wall of a normal artery is smooth and in the normal condition, blood flows through it without difficulty. The major cause of decreasing the blood flow is atherosclerosis which is due to the deposits of fatty materials, such as cholesterol, developing the thickened arterial walls and stenosis of the vasculatures. These changes cause ischemic changes in the organs fed by the vasculatures and if it occurred in the abdominal mesenteric lesions, the symptoms of severe abdominal pain, ischemic changes of the intestine could be observed leading to lethality. For other vascular diseases including aneurysm, occlusion, and thromboses in the mesenteric lesions could cause severe symptoms and appropriate diagnosis and treatment are essential for managing patients. With the development and improvement of imaging modalities, including CT and magnetic resonance imaging, the frequency of diagnosis of vascular disease in abdominal lesions is increasing. Among these, FMD and SAM are known as noninflammatory, nonatherosclerotic arterial diseases, difficult to be differentially diagnosed from each other. Although various arteries are involved in these diseases, we have focused on the mesenteric areas, reviewing cases in this study and summarizing the clinical characteristics of both disease entities (Table 3).

The histologic findings and the imaging findings of FMD and SAM are similar; for example, Lie proposed that SAM can represent a precursor of certain types of FMD[74]. Slavin and colleagues also proposed that SAM could represent a precursor of FMD, although a part of SAM might remain as unspecified aneurysms[46]. Although these similarities in radiological and histological diagnoses have been reported, the two entities exhibit a different clinical profile in terms of age of onset, sex, distribution of affected arteries, and clinical symptoms. Although FMD affects middle-aged women, there is no predilection for age or sex in SAM[2,73].

Considering the mesenteric lesions, as there are no specific symptoms, a greater knowledge and comprehensive understanding of these diseases are important for appropriate diagnosis and treatment. For example, FMD rarely shows significant symptoms and is frequently associated with symptoms of occlusive disease such as renovascular hypertension, headache, and pulsatile tinnitus. Although FMD does not rupture as often, SAM shows hemorrhages resulting from arterial rupture or dissection

Table 3 Clinical characteristic of the fibromuscular dysplasia and segmental arterial mediolysis

	Fibromuscular dysplasia	Segmental arterial mediolysis
Gender	Female (9:1)[2]	No presentation[74]
Age of presentation	Young to middle age[2]	No preference[74]
Laboratory findings	No serological markers[74]	No serological markers[74]
Risk factors	Smoking and extracranial arteries[4]	Hypoxia and shock or other vasoconstrictor stimuli[47]
Vascular distribution	Renal and extracranial arteries[4]	Celiac and mesenteric arteries[48]
CT	Alternating stenosis and aneurysms, less commonly dissections[38]	Dissections with alternating stenosis and aneurysms, dissecting aneurysms[48]
Angiography	Beaded aneurysmal appearance (string-of-beads)[38]	Beaded aneurysmal appearance (string-of-beads)[38]
Pathology	Fibrous or fibromuscular thickening of the arterial wall[38]	Vecuolization and lysis of the outer media[47]
Symptoms	Renovascular hypertension, Headache, Pulsatile tinnitus[4]	Acute abdominal pain, Intraperitoneal bleeding[47]
Treatment	Anti-platelet therapy and anti-hypertensive therapy. Balloon angioplasty and stenting[45]	Anti-hypertensive therapy and endovascular management, surgical management[74]

CT: Computed tomography.

from the weakened arterial wall[4,46] and is therefore symptomatic with acute abdominal and flank pain.

CONCLUSION

Mesenteric vascular diseases are rare compared with other disease entities in lesions; therefore, clinical information is insufficient and clinical trials to develop the standard therapy are lacking. Therefore, an accumulation of cases and a summary of the clinical characteristics of reported cases are important. For this purpose, we have summarized the characteristics of FMD and SAM in abdominal lesions. This information could help physicians to appropriately diagnose and treat cases, including consultation with interventional radiologists and surgeons.

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