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Clinical Diagnosis of Segmental Arterial Mediolysis: Differentiation from Vasculitis and Other Mimics

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Keywords

segmental arterial mediolysis; vasculitis; diagnosis

Introduction

Segmental arterial mediolysis (SAM) is a rare vasculopathy of unknown etiology characterized by disruption of the arterial medial layer, with resultant susceptibility to vessel dissection, hemorrhage, and ischemia. Since the first case of SAM described by Slavin and Gonzalez-Vitale in 1976 (1), approximately 50 cases have been reported in the literature (2).

Although the abdominal visceral arteries are most frequently affected in SAM (3), any vessel may be involved, including the retroperitoneal (4), intracranial (2,5,6) and coronary arteries (7–9). The histopathologic changes begin with vacuolar degeneration of smooth muscle cells in the arterial media, followed by fibrin deposition at the medial-adventitial junction (7). This in turn predisposes to dissecting aneurysms (3,10). The angiographic appearance of SAM is variable, ranging from arterial dilation to aneurysm formation (single or multiple) to stenoses or occlusion, frequently with dissection (3,11). Correspondingly, symptoms arise both from stenoses and occlusions (e.g., postprandial pain from intestinal ischemia) and from dissections and aneurysms (e.g., sudden and catastrophic intraperitoneal bleeding). In contrast to true vasculitis, inflammatory cells in SAM are present inconsistently and, when present, are thought to be secondary rather than primary to the pathogenesis of the disease (1).

The differential diagnosis of SAM includes atherosclerosis, fibromuscular dysplasia, infection (e.g., mycotic aneurysm and endocarditis), connective tissue diseases (e.g.,

Bechet's disease and polyarteritis nodosa), neurofibromatosis, and inherited defects in vessel wall structural proteins (e.g., type IV Ehlers Danlos and Marfan's syndrome) (Table 1). Herein, we describe two cases of SAM seen at our institution over the past 20 years and review the salient clinical presentation and treatment of SAM. We report characteristics that may be helpful in distinguishing cases of SAM from other entities in the differential diagnosis.

Case 1

The patient was a 25-year-old female who was admitted to the hospital with an eleven-month history of intermittent episodes of anorexia, abdominal pain, and diarrhea. Symptoms had persisted despite discontinuation of oral contraception and initiation of low-dose aspirin therapy.

Her past medical history was unremarkable. Family history was unremarkable except for benign hypermobility syndrome in the patient's mother. On physical examination, the patient was normotensive and had normal height and arm span. She had no carotid, subclavian, abdominal, or femoral bruits. Skin, chest, abdominal, and neurologic examinations were normal. Joint exam was remarkable only for hyperextensibility of the knees, reducible flexion contractures of the fingers, and hammertoe deformities of the feet.

Complete blood count revealed anemia with hemoglobin of 11 g/dl. Serum creatinine, liver enzymes, amylase, and lipase were normal, and urine pregnancy test was negative. Antinuclear antibody assay (ANA) was positive to a low titer of (1:80). The erythrocyte sedimentation rate was 20 mm/hr. The remainder of the serologic, metabolic, immunologic, and hematologic evaluations were within normal limits, including negative hepatitis serologies, negative double-stranded DNA, anti-Smith, and anti-ribonucleoprotein (RNP) antibodies, and normal complement C3 and C4 levels.

Computed tomography (CT) of the abdomen showed thickening of the colonic wall with mucosal enhancement and fat stranding surrounding the splenic flexure. Colonoscopy revealed ischemic colitis of the splenic flexure. Biopsies of the ischemic areas were not obtained due to risk of possible perforation. Biopsies of the non-ischemic areas were normal, as was magnetic resonance angiography (MRA) of the abdomen. Conventional mesenteric angiography revealed focal stenoses of the right and left hepatic arteries, occlusion of the left colic artery near the splenic flexure with collateral vessel formation, and hyperemia of multiple branches of the splenic artery (Figure 1).

Because of persistent ischemic colitis, the patient underwent a partial colectomy of the splenic flexure. Vascular pathology of the colonic arteries showed patchy, isolated destruction of the arterial media involving both the internal and external elastic laminae (Figure 1). In a few sections the media was absent, with direct juxtaposition of the intima and the adventitia. In areas of medial destruction, there was intimal proliferation with marked luminal narrowing. All of the lesions were of a similar age. There was no evidence of inflammation, and giant cells, neutrophils, and cholesterol deposits were absent. After two years of followup, the patient remains asymptomatic.

Case 2

The patient was a 51-year-old Caucasian male who was admitted to the hospital with acute burning and tearing epigastric pain that began while eating. A full cardiac evaluation and computed tomography (CT) of the abdomen were unrevealing. He was discharged the next day with a tentative diagnosis of biliary colic. However, because of unremitting abdominal discomfort he was readmitted 2 days later.

Prior to the initial painful episode, he had no history of epigastric pain. He denied a change in bowel habits, loss of appetite, or weight loss. There was no history of fever, chills, sweats, rashes, oral ulcers, genital lesions, hepatitis, or exposures to sexually transmitted diseases. He did not smoke and only used alcohol occasionally. He admitted to minor chronic headaches but denied visual changes or jaw pain. Other medical history was non-contributory, and he took no chronic medications. Physical examination revealed normal vital signs with full and symmetrical pulses throughout. There were no peripheral thrills or bruits. Abdominal examination was abnormal only for mild guarding, without rebound, and there were no masses, organomegaly, or bruits.

Complete blood count, urinalysis, basic chemistry panel, and liver function tests were all normal except for an albumin of 3.2 gm/dl. C-reactive protein (CRP) and erythrocyte sedimentation rate were normal at 3.9 mg/L and 30 mm/hr, respectively. The remainder of the serologic and immunologic workup was within normal limits.

An abdominal ultrasound was within normal limits, but initial CT angiography a celiac artery aneurysm with possible dissection. Repeat CT angiography a few days later documented extensive aneurysmal dilatation (up to 1.8 cm) of the celiac axis from the proximal celiac artery to the bifurcation, involving the hepatic and splenic arteries with mural thrombus in the splenic artery. Magnetic resonance angiogram (MRA) of the abdomen showed similar findings.

The patient was managed conservatively. A follow-up CT angiogram in 2 weeks revealed enhanced wall thickness of the celiac axis and decreased aneurysmal dilatation of the splenic artery, compatible with evolution and partial improvement of segmental arterial mediolysis (SAM). Despite the encouraging imaging studies, the patient continued to complain of epigastric pain, and it was decided to proceed with resection of the celiac and common hepatic artery aneurysms with aortoceliac artery bypass. Histologic examination of the surgical specimen revealed extensive deterioration of the media with marked intimal hyperplasia. There were no giant cells, granulomas, or signs of vascular inflammation, and there were no significant cholesterol plaques. Two years have passed since the surgical repair and the patient remains asymptomatic.

Discussion

Segmental arterial mediolysis (SAM), also known as segmental medial arteriolysis (SMA), is a rare vasculopathy characterized by non-inflammatory degeneration of the medial layer of muscular arteries (1) and, occasionally, adjacent veins (12). Originally described by Slavin and Gonzalez-Vitale as “segmental mediolytic arteritis” in 1976, Slavin and colleagues later proposed a change in name to “segmental arterial mediolysis” due to lack of consistent evidence of true inflammation in both the clinical presentation and the histologic features of the disease (10).

SAM is a pathologic diagnosis that is defined by characteristic histologic features on surgical specimens obtained from affected anatomic sites. Though it can be difficult to diagnose, clinical clues are usually present that point to the diagnosis of SAM on the basis of history, physical examination, and initial laboratory evaluation. The combination of clinical features and surgical pathology findings usually allows the discrimination of SAM from its mimics (Tables 1 and 2). For example, although atherosclerosis, a common vasculopathy, is usually widespread throughout many regions of the vascular tree, SAM typically is limited to vessels in only one anatomic site (3). In addition, atherosclerosis typically occurs at the branch points of vessels in patients with traditional cardiovascular risk factors, while these

features are absent in SAM (13). Furthermore, although atherosclerosis typically occurs in middle-aged and elderly adults, SAM may present at any age.

It is a particular challenge to distinguish SAM from fibromuscular dysplasia (FMD), especially since SAM is often considered an early lesion of FMD (3,10). Classically, FMD presents in young females and has a predisposition for the renal arteries, causing premature hypertension. SAM, on the other hand, may present at any age, has no gender predisposition, and most commonly affects the celiac artery and its branches. Arterial dissection and hemorrhage are also much more common in SAM than in classic FMD. In addition to FMD, another disease entity classified on the disease spectrum of SAM is cystic medial necrosis (CMN) (1). However, typically, CMN occurs in the aorta and great vessels of patients with Marfan's syndrome (14), whereas these vessels are not typically affected by SAM.

We reviewed all cases of biopsy-confirmed SAM occurring at our institution over the past 20 years. The two cases described here represent the results of our medical record review. Although many more cases of SAM were suspected on the basis of clinical and radiologic features, these were the only two cases with histopathologic confirmation. Of note, the cases identified in our institution lacked physical signs and symptoms and laboratory indicators of systemic inflammation, helping to distinguish SAM from the inflammatory vasculitides.

The most dramatic presentation of SAM is sudden, life-threatening hemorrhage of the abdomen, retroperitoneum, or brain (3,15). Hemorrhage results from either aneurysm rupture or dissections occurring as a result of weakening along the plane separating the outer media from the adventitia (3). Interestingly, each of the cases of SAM identified at our institution presented in a relatively benign fashion, with abdominal pain as the chief complaint. Ischemic colitis, as in Case 1, has been reported as an example of a relatively less acute presentation of SAM (16). More benign presentations of SAM could easily escape clinical diagnosis, and therefore, SAM may be substantially more common than is suggested by the literature (3,17).

Angiography can reveal several patterns that are consistent with SAM, including single or multiple aneurysms, dissections, stenoses, and occlusions (3). As seen in our cases and in another report by Michael and colleagues (18), lesions of SAM may evolve rapidly over the course of weeks on serial angiography. Despite patterns on angiography that are suggestive of SAM, histopathology remains the gold standard for definitive diagnosis (Table 2). This is especially important in the case of polyarteritis nodosa (PAN), which can have an angiographic appearance identical to that of SAM (3). Lack of inflammation on arterial biopsy in SAM allows these two entities to be readily distinguished. Patients with PAN also generally have clinical evidence of systemic inflammation, whereas patients with SAM do not.

The discrimination of SAM from systemic inflammatory vasculitides is particularly important, since corticosteroids and immunosuppressive agents, which are crucial in the treatment of the inflammatory vasculitides, have no proven benefit in SAM (19). Without any evidence of an inflammatory etiology, the use of immunosuppressive regimens in SAM exposes the patient to undue risks, including infection and poor wound healing, and could possibly worsen prognosis (19). Treatment of SAM involves embolization, surgical bypass, or resection of the injured arteries (20). The long-term prognosis of SAM is somewhat uncertain, since its natural history has not been thoroughly characterized despite its initial description over 30 years ago. It is known, however, that while cases of SAM complicated by intra-abdominal hemorrhage have a mortality approaching 50% (21), the most common scenario is of long-term disease-free survival following embolization, bypass, or resection of the affected areas. There have even been reports of complete spontaneous resolution of the

vascular lesions of SAM (18). Our cases were both followed for 2 years with no recurrence of disease.

In summary, SAM is a rare but important cause of unexplained vascular lesions in patients in whom other inflammatory, infectious, or heritable diseases have been ruled out. The diagnosis should be considered when a patient presents with unexplained acute-onset abdominal pain with or without intra-abdominal bleeding. SAM should also be kept in mind when aneurysms, stenoses, and occlusions are identified in medium and large vessels, especially when these lesions are limited to one anatomic location. Conventional angiography is more sensitive than CT or MR angiography and should be used after more conventional methods of imaging are unrevealing. If a diagnosis of SAM is suspected, a multi-disciplinary approach involving consultation with interventional radiology and vascular or general surgery should be promptly pursued.

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References

1. Slavin R, Gonzalez-Vitale J. Segmental mediolytic arteritis. *Lab Invest.* 1976; 35:23–29. [PubMed: 940319]
2. Ro A, Kageyama N, Takatsu A, Fukunaga T. Segmental arterial mediolysis of varying phases affecting both the intra-abdominal and intracranial vertebral arteries: an autopsy case report. *Cardiovasc Pathol.* 2009
3. Slavin RE. Segmental arterial mediolysis: course, sequelae, prognosis, and pathologic-radiologic correlation. *Cardiovasc Pathol.* 2009; 18(6):352–360. [PubMed: 19026575]
4. Phillips CK, Lepor H. Spontaneous retroperitoneal hemorrhage caused by segmental arterial mediolysis. *Rev Urol.* 2006; 8(1):36–40. [PubMed: 16985559]
5. Collins G, Rich N, Clagett G, Spebar M, Salander J. Fibromuscular dysplasia of the internal carotid arteries. Clinical experience and follow-up. *Ann Surg.* 1981; 194:89–96. [PubMed: 7247539]
6. Ohtoh T, Ono Y, Iwasaki Y, Sakurai Y, Nishino A, Arai H, et al. Non-traumatic recurrent dissection and its spontaneous repair in the circle of Willis: report of two autopsy cases. *Neuropathology.* 2003; 23(3):195–198. [PubMed: 14570286]
7. Slavin RE, Cafferty L, Cartwright J Jr. Segmental mediolytic arteritis. A clinicopathologic and ultrastructural study of two cases. *Am J Surg Pathol.* 1989; 13(7):558–568. [PubMed: 2660608]
8. Gruenwald P. Necrosis in the coronary arteries of newborn infants. *Am Heart J.* 1949; 38(6):889–897. illust. [PubMed: 15395920]
9. Lie JT, Berg KK. Isolated fibromuscular dysplasia of the coronary arteries with spontaneous dissection and myocardial infarction. *Hum Pathol.* 1987; 18(6):654–656. [PubMed: 3596585]
10. Slavin RE, Saeki K, Bhagavan B, Maas AE. Segmental arterial mediolysis: a precursor to fibromuscular dysplasia? *Mod Pathol.* 1995; 8(3):287–294. [PubMed: 7617656]
11. Sakano T, Morita K, Imaki M, Ueno H. SAM studied by repeated angiography. *Br J Radiol.* 1997; 70:656–658. [PubMed: 9227264]
12. Slavin RE, Inada K. Segmental arterial mediolysis with accompanying venous angiopathy: a clinical pathologic review, report of 3 new cases, and comments on the role of endothelin-1 in its pathogenesis. *Int J Surg Pathol.* 2007; 15(2):121–134. [PubMed: 17478765]
13. Slovut DP, Olin JW. Fibromuscular dysplasia. *N Engl J Med.* 2004; 350(18):1862–1871. [PubMed: 15115832]

14. Yamada M, Ohno M, Itagaki T, Takaba T, Matsuyama T. Coexistence of cystic medial necrosis and segmental arterial mediolysis in a patient with aneurysms of the abdominal aorta and the iliac artery. *J Vasc Surg.* 2004; 39(1):246–249. [PubMed: 14718847]
15. Inada K, Maeda M, Ikeda T. Segmental arterial mediolysis: unrecognized cases culled from cases of ruptured aneurysm of abdominal visceral arteries reported in the Japanese literature. *Pathol Res Pract.* 2007; 203(11):771–778. [PubMed: 17920781]
16. Wang JJ, Huang TW. Ischemic colitis caused by an isolated dissecting aneurysm of the left colic artery: a presumed case of segmental mediolytic arteriopathy. *J Formos Med Assoc.* 1994; 93(8): 715–720. [PubMed: 7858458]
17. Hirakawa E, Inada K, Tsuji K. Asymptomatic dissecting aneurysm of the coeliac artery: a variant of segmental arterial mediolysis. *Histopathology.* 2005; 47(5):544–546. [PubMed: 16242010]
18. Michael M, Widmer U, Wildermuth S, Barghorn A, Duewell S, Pfammatter T. Segmental arterial mediolysis: CTA findings at presentation and follow-up. *AJR Am J Roentgenol.* 2006; 187(6): 1463–1469. [PubMed: 17114538]
19. Lie JT. Systemic cerebral and pulmonary segmental mediolytic arteriopathy: villainous masqueraders of vasculitis. *Cardiovasc Pathol.* 1996; 5:305–331.
20. Ryan JM, Suhocki PV, Smith TP. Coil embolization of segmental arterial mediolysis of the hepatic artery. *J Vasc Interv Radiol.* 2000; 11(7):865–868. [PubMed: 10928523]
21. Rengstroff DS, Baker EL, Wack J, Yee LF. Intra-abdominal hemorrhage caused by segmental arterial mediolysis of the inferior mesenteric artery: report of a case. *Dis Colon Rectum.* 2004; 47:769–772. [PubMed: 15054678]

Figure 1.

Angiographic and histologic features of segmental arterial mediolysis in Case 1. (a) Focal stenoses of the right and left hepatic arteries (arrows). (b) Hyperemic blush in the left colonic flexure, suggesting formation of small collateral vessels from the splenic artery in response to occlusion of the left colic artery (not shown). (c) Large bowel with thrombosis in multiple small arteries (long arrows) and focal ischemia (short arrow). Hemosiderin and eosin stain (H&E), 10 $\times$. (d, e) Adventitial arteries with thrombosis, minimal inflammatory infiltrate, and granulation tissue disrupting the media (arrow). H&E, 50 $\times$. (f) Fibrosis of several small colonic arteries. H&E, 50 $\times$. (g, h) Foamy deterioration of media with disruption of internal elastic lamina of colonic vessels. Van Gieson stain, 50 $\times$ (g), 100 $\times$ (h).

Table 1

Key features distinguishing Segmental Arterial Mediolysis (SAM) from its mimics*.

Segmental arterial mediolysis (SAM)	Fibromuscular dysplasia	Polyarteritis nodosa(PAN)	ANCA-Associated Vasculitis	Giant Cell arthritis	Takayasu's	Bechet's	Kawasaki	Mycotic aneurysm	Type IV Ehlers-Danlos	Marfan's syndrome	Neuro-fibromatosis	Pseudo-xanthoma elasticum	Atherosclerosis
equal	female (3:1)	equal	equal	female (2:1)	female (8:1)	equal	male (1.5:1)	equal	equal	equal	equal	female (2:1)	
any; 40s-60s most common	20s-30s	40s-60s	any	>50	15-30s	30s-40s	<5	any	<10	any	<18	<18	>60
none	Caucasian	none	Caucasian	Caucasian	Asian	Asian, Middle Eastern	Asian	none	none	none	none	none	none
no	no	yes	yes	yes	yes	yes	yes	yes	no	no	no	no	no
occas.	yes	yes	occas.	occas.	yes	no	occas.	no	no	no	yes	yes	yes
no	no	yes	yes	occas.	occas.	yes	yes	occas.	yes	yes	yes	yes	occas.
no	occas.	yes	yes	occas.	yes ^b	yes	yes	occas.	occas.	occas.	occas, ^d	no	no
no	occas.	occas.	yes	yes	occas.	yes	yes	no	occas.	yes	yes	yes	yes
occas.	occas.	occas.	occas.	yes	occas.	occas.	no	occas.	occas.	occas.	occas.	occas.	yes
no	no	no	yes	no	occas.	no	no	occas.	no	no	no	no	no
no	no	no	yes	no	no	yes ^c	no	no	occas.	no	occas.	no	no
occas.	yes	occas.	occas, ^a	no	occas.	no	yes	occas.	occas.	yes	occas.	yes	yes
yes	occas.	yes	occas.	occas.	occas.	yes	occas.	occas.	yes	occas.	occas.	yes	occas.
yes	occas.	yes	occas.	occas.	occas.	occas.	no	occas.	occas.	occas.	yes	yes	occas.
no	yes	yes	yes	occas.	yes	no	occas.	occas.	occas.	occas.	yes	no	yes
35													
no	no	yes	yes	occas.	occas.	occas.	yes	yes	no	no	no	no	no
occas.	no	yes	yes	yes	yes	occas.	occas.	yes	no	no	occas.	no	occas.
no	no	yes	yes	yes	yes	yes	yes	yes	no	no	no	no	occas.
no	no	occas.	yes	no	no	no	no	occas.	no	no	no	no	no

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	Segmental arterial mediolysis (SAM)	Fibromuscular dysplasia	Polyarteritis nodum(PAN)	ANCA-Associated Vasculitis	Giant Cell arteritis	Takayasu's	Bechet's	Kawasaki	Mycotic aneurysm	Type IV Ehlers-Danlos	Marfan's syndrome	Neuro-fibromatosis	Pseudo-xanthoma elasticum	Atherosclerosis
	nl	nl	low(occas.)	nl	nl	nl	nl	nl	hi or low	nl	nl	nl	nl	nl
n	neg	neg	pos	neg	neg	neg	neg	neg	neg	neg	neg	neg	neg	neg
	no	no	no	no	no	no	no	no	yes	no	no	no	no	no
	no	occas.	yes	yes	no	yes	occas.	occas.	occas.	no	no	occas.	no	no
OTHER THERAPY	no	no	yes	yes	yes	yes	yes	yes	no	no	no	no	no	no

Reagan's syndrome, aortitis associated with seronegative spondylarthritides; CRP = erythrocyte sedimentation rate; ANCA = anti-neutrophil cytoplasmic antibodies; nl = normal; hi = high; neg = negative; pos = positive

and thoracic back pain are relatively common

Coronary arteries

a result of peripheral neuropathy. Also, bone lesions such as pseudoarthrosis and bone dysplasia are common.

Table 2
features of segmental arterial mediolysis (SAM) and its mimics.

	Polyarteritis nodosum (PAN)	Giant cell arteritis (GCA)	Takayasu's	Behcet's	Kawasaki	Mycotic aneurysm	Type IV Ehlers-Danlos	Marfan's syndrome	Neuro fibromatosis	Pseudoxanthoma elasticum	Atherosclerosis
Age	S, M	M, L	L	S, M, or L; veins	M	S, M, or L	L	M, L	S, M, L, veins	M, L, veins	S, M, or L
Sex	yes; often strikingly segmental	yes	yes	yes	yes	yes	no	no	no	no	occas.
Location	intra-abdominal arteries	intracranial arteries	aorta and great vessels	veins	coronary arteries	any	thoracic and intra-abdominal arteries	aorta	any	any	aorta, coronary arteries, intracranial arteries, lower extremity arteries
Pathogenesis											
Pathology	Focal areas of necrotizing panarteritis	Segmental arteritis predominantly affecting intimal-medial junction; concentric intimal hyperplasia; giant cells	Granulomatous inflammation initially involving vasa vasorum, progressing to fibrosis of all vessel layers	Leukocytoclastic vasculitis and perivascular inflammation	Perivascularitis and vasculitis of microvessels with vessel wall edema	Infectious elements present	Thinning of media with disruption of elastin; deficiency of type III collagen fibers	Massive degeneration of elastic fibers (cystic medial necrosis)	Adventitial and intimal neurofibromatous nodules, intimal fibrosis, intimal and medial hyaline deposition, disruption of elastin with medial thinning	Disruption of elastin with medial calcifications and premature atherosclerosis	Characteristic fibrous plaques with calcification and foamy macrophages
Diagnosis											
Management	Characteristic microaneurysms predominantly at vessel branch points	Angiography has no role in detecting temporal artery involvement or in guiding biopsy site	Large aneurysms and stenoses in aorta and great vessels	Deep venous thromboses (DVTs); rarely, pulmonary arterial aneurysms and rupture	Coronary aneurysms in 15–20% of cases	Aneurysms at branch points	Arterial rupture; dissections less common than in Marfan's; arteriovenous fistulae	Dissections of the ascending aorta.	Vessel narrowing, aneurysms, occlusions	Tortuous, narrowed vessels with angiomatous malformations	Shaggy, irregular appearance; predisposition to branch points; widespread distribution

could be noted that extension to other vascular sizes or other areas of the vascular tree is relatively common.

S = small arteries < 0.5 mm in diameter; M = medium arteries 0.5–1.0 mm in diameter; L = large arteries > 1.0 mm in diameter; veins = veins of any diameter
occas. = occasional
nl = normal