Published online: 2019-12-02

Segmental Arterial Mediolysis with Supra-Aortic Vascular Involvement: Report of Two Cases

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J Clin Interv Radiol ISVIR 2019;3:189–192

Abstract

Segmental arterial mediolysis (SAM) is a rare noninflammatory vasculopathy predominantly involving the visceral arteries of the abdomen. The most common presentation described is massive intra-abdominal hemorrhage secondary to aneurysm rupture. In this report, we describe two cases with characteristic findings of aneurysm, dissection, and beading involving the visceral arteries and the supra-aortic vessels. SAM involving the supra-aortic vessels is extremely rare.

Keywords

- segmental arterial mediolysis
- dissection
- aneurysm

Introduction

Segmental arterial mediolysis (SAM) is a rare noninflammatory and nonatherosclerotic vasculopathy first described by Slavin and Gonzalez-Vitale in 1976.[1] It usually involves the visceral arteries of the abdomen resulting in intra-abdominal hemorrhage. SAM primarily affects the outer layer of the media, leading to smooth muscle cell vacuolar degeneration.[2] Lytic degeneration of medial smooth muscle occurs, culminating in separation from adventitia and adjacent fibrosis.[2] Gaps may be filled with fibrin, or granulation tissue and can lead to saccular aneurysms, dissecting aneurysms, or thrombosis.[3,4] The intima is spared from these lytic changes, and there is minimal inflammation. Also, there is lack of evidence of vasculitis in affected patients.[2-4]

The present article describes clinical and radiological findings of two patients who had angiographic evidence of SAM in the carotid and vertebral arteries in addition to characteristic findings in the visceral arteries.

Case 1

A 37-year-old female presented to an outside hospital with left flank pain. Computed tomography (CT) abdomen revealed left renal infarct. Laboratory findings included an erythrocyte sedimentation rate (ESR) of 24 mm in the first hour, C-reactive protein (CRP) of 5 mg/dL, hemoglobin (Hb) of 11.2 g%, white blood cells (WBCs) of 11,100/cc, and serum creatinine of 0.7 mg/dL. Anticoagulation with enoxaparin was started at 60 mg subcutaneously every 12 hours. She also had a history of ruptured splenic artery aneurysm 4 years prior to the presentation for which aneurysm repair was done. On the third day of admission, she developed sudden onset of severe abdominal pain, hypotension, and tachycardia. Physical examination revealed distended abdomen with epigastric tenderness. Her heart rate was 126/min and blood pressure was 90/60 mm Hg. Hemoglobin decreased from 11.2 to 5.3 g%. After fluid resuscitation, an urgent arterial and venous phase contrast CT of the abdomen was obtained. CT revealed retroperitoneal hematoma, saccular splenic artery aneurysm, and string of beads appearance of the gastric arteries (►**Fig. 1**). Patient was transferred to our hospital for further management. During angiography, the patient had worsening of abdominal pain and splenic aneurysm had reruptured. The aneurysm was embolized with 33% N-butyl-2-cyanoacrylate (NBCA) and lipiodol suspension (►**Fig. 2**). Subsequently, bilateral renal, vertebral, and carotid angiograms were obtained (►**Fig. 3**) to assess the status of vasculature in other territories. It revealed tortuous, ectatic bilateral carotid arteries with beaded appearance of left cervical internal carotid artery. Bilateral vertebral and renal arteries were normal. Considering Hb of 5.3 g% and hypotension, three units of blood was transfused to the patient. She recovered well and was discharged after 10 days.

Laboratory screen for vasculitis (CRP, antinuclear antibodies, antineutrophil cytoplasmic antibodies, antiphospholipid antibody, and rheumatoid factor) was negative. There was no clinical evidence of Marfan's symdrome, Ehlers–Danlos

received
May 27, 2019
accepted after revision
July 1, 2019
published online
December 2, 2019

DOI https://doi.org/10.1055/s-0039-3401352
ISSN 2457-0214.

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syndrome, or neurofibromatosis. The patient remained well at 2 years clinical follow-up.

Case 2

A 39-year-old male was slapped on left side of neck. After 3 hours, he developed sudden onset right hemiparesis. On admission, he was hemodynamically stable. Physical examination revealed normal vital signs and symmetric peripheral pulses. No bruit or thrill was palpable in neck. His past medical and family history was unremarkable. He was a nonsmoker. Complete blood count and serum biochemistry profile were within normal limits. CRP was 4.8 mg/dL. Magnetic resonance imaging (MRI) with MR angiogram of the brain revealed left internal carotid (ICA) occlusion with infarct in left posterior cerebral artery (PCA) territory (►**Fig. 4**). Digital subtraction angiography (DSA) of the cerebral and neck vessels revealed left cervical internal carotid artery dissection with its complete occlusion. Left cerebral anterior circulation was catered by crossflow through the anterior communicating artery. Right internal carotid artery in the cervical segment was tortuous, kinked, and ectatic. There were multiple aneurysms in right vertebral artery and beaded appearance of left vertebral artery in V2 segment. Superior mesenteric artery angiogram demonstrated characteristic dissecting aneurysm (►**Figs. 5** and **6**). Both renal arteries were unremarkable. The patient gradually recovered over 5 days with conservative management. There was no clinical or laboratory evidence of vasculitis. There was no clinical evidence or family history suggestive of neurofibromatosis, Marfan's- or Ehlers–Danlos syndrome. The patient remained well at 2 years clinical follow-up.

Fig. 1 CT angiogram showing splenic artery aneurysm (yellow arrow), beaded left gastric and short gastric arteries (blue arrow), and jejunal branch (green arrow) of superior mesenteric artery (SMA). CT, computed tomography.

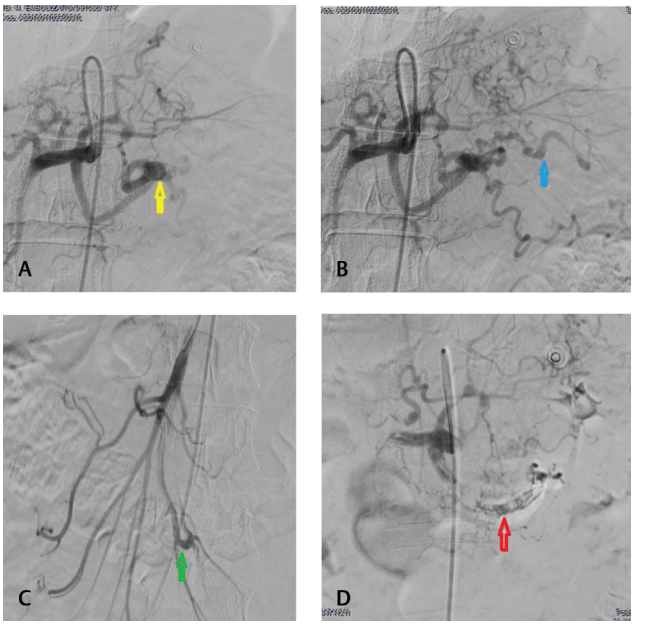

Fig. 2 DSA showing (**A**) splenic artery aneurysm (yellow arrow), (**B** and **C**) beaded gastric arteries (blue arrow) and jejunal branch (green arrow) of SMA, and (**D**) splenic artery aneurysm embolized with NBCA + lipiodol suspension (red arrow). DSA, digital subtraction angiography; SMA, superior mesenteric artery; NBCA, N-butyl-2-cyanoacrylate.

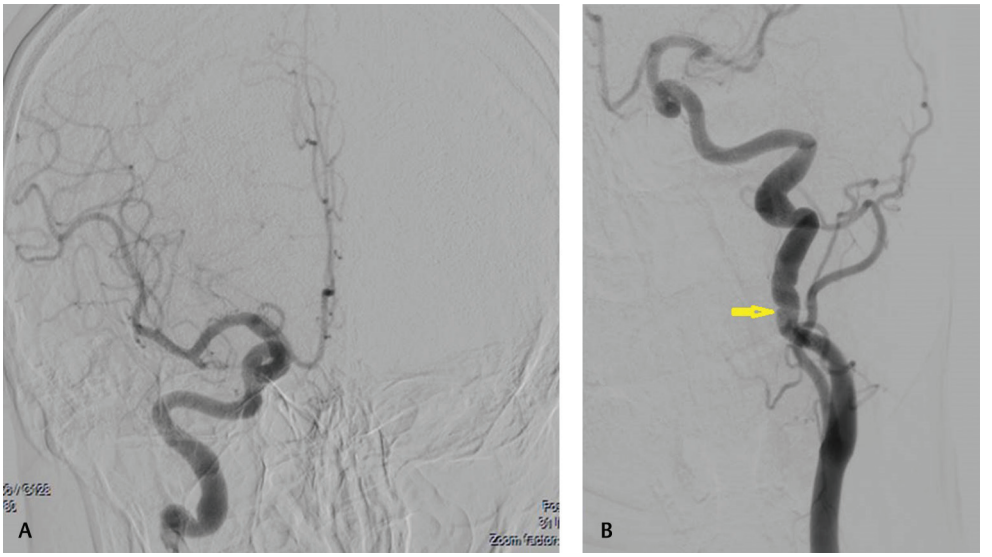

Fig. 3 DSA of both carotids showing tortuousity, ectasia, and kinking. Beaded left carotid (yellow arrow). DSA, digital subtraction angiography.

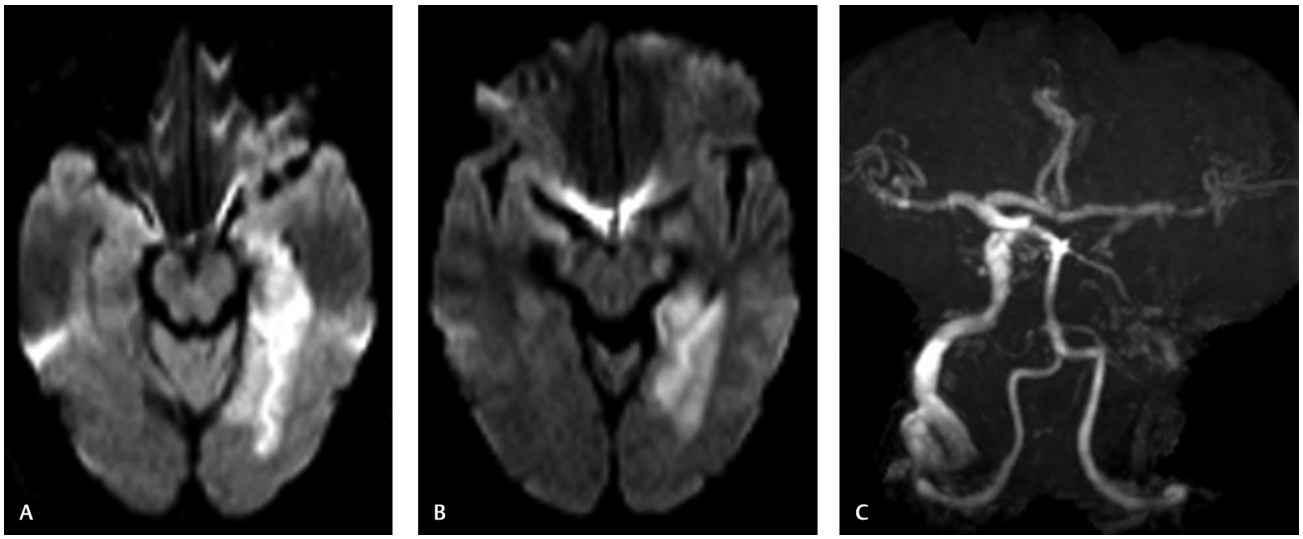

Fig. 4 MRI and MRA showing left PCA territory infarct and left ICA occlusion. MRI, magnetic resonance imaging; MRA, magnetic resonance angiography; PCA, posterior cerebral artery; ICA, internal carotid.

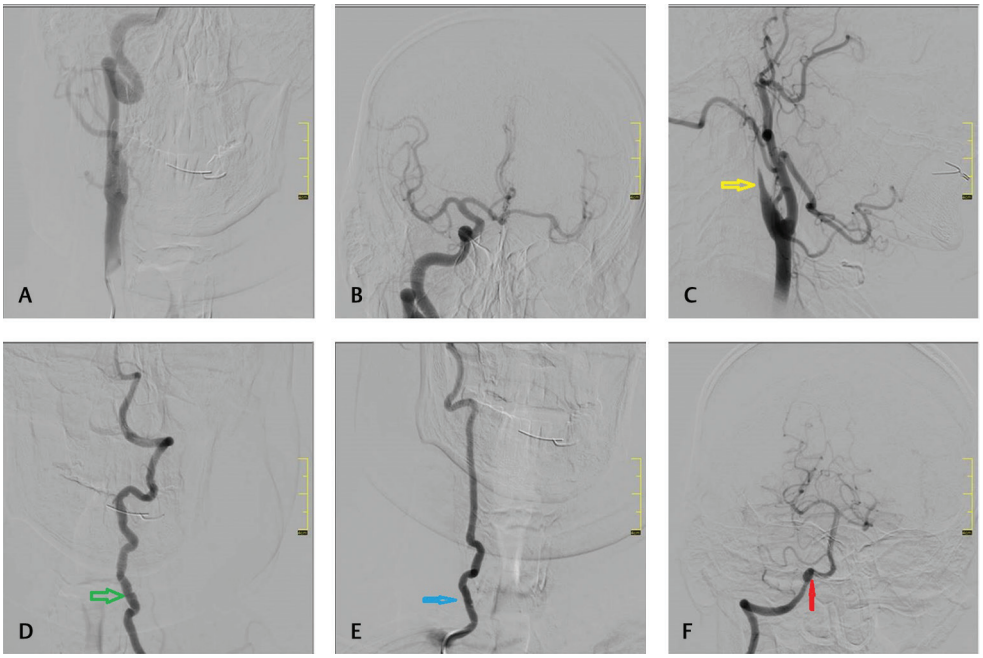

Fig. 5 DSA showing (**A** and **B**) ectatic, tortuous, and kinked right ICA, (**C**) dissection with complete occlusion of left ICA (yellow arrow), (**D**) beaded V2 segment of left vertebral artery (VA; green arrow), (**E**) dissecting aneurysm in V1 segment of right VA (blue arrow), and (**F**) saccular aneurysm in V4 segment of right VA (red arrow). ICA, internal carotid; DSA, digital subtraction angiography;

After excluding vasculitis and congenital vascular condition; angiographic findings of dissecting aneurysms in splanchnic circulation, tortuous ectatic and beaded carotid, and vertebral arteries and normal renal arteries suggested the diagnosis of segmental arterial mediolysis.

Discussion

Segmental arterial mediolysis is a rare entity with a slight male predominance (M/F ratio of 1.5:1). It commonly affects middle-aged and elderly patients.[5] SAM mainly involves the splanchnic vessels.[6,7] The most common sites are celiac axis, splenic, superior mesenteric, renal, and inferior mesenteric arteries. Multiple arteries are commonly involved.[8]

The most common clinical presentation is abdominal apoplexy secondary to ruptured visceral artery aneurysm manifested by abdominal pain, distension, rapidly decreasing hematocrit, and hypovolemic shock.[6,9] Dissection may lead to arterial occlusion which can result in end-organ ischemia, manifested as renal infarct, stroke, and bowel ischemia.[5,10] As in our case where first patient presented with flank pain secondary to renal infarct and second patient presented with stroke.

The principle radiologic hallmark of SAM is the greater prevalence of dissecting aneurysms. Focal areas of vascular

Fig. 6 DSA showing characteristic dissecting aneurysm (yellow arrow) in SMA. DSA, digital subtraction angiography; superior mesenteric artery.

dilation with regions of stenosis proximally may actually represent dissection or dissecting hematoma. Other findings are aneurysm, which may be saccular or fusiform, single or multiple, arterial dilatation or ectasia, arterial stenosis, occlusions and alternating aneurysm, and stenosis giving a beaded appearance. Affected arteries may become elongated and kinked. Unlike mycotic aneurysms, there is no predilection for bifurcation.[4,5,8,9]

Although very rare, SAM has also been reported in the cerebral arteries of young adults.[11] Both of our patients were young and had involvement of carotid as well as vertebral arteries. Many of the cases of young stroke secondary to spontaneous dissection may actually be SAM which has not been thoroughly evaluated.

A combination of clinical features, laboratory and angiographic finding can assist to differentiate SAM from other conditions including various vasculitides, connective tissue disorders, mycotic aneurysms, atherosclerosis, and inherited vascular defects.[12]

Some authors suggested that SAM is a variant of fibromuscular dysplasia (FMD) as the angiographic findings are similar.[3,13] However, FMD occurs more commonly in middle aged women and mostly in carotid and renal arteries whereas SAM affects older age group and involves predominantly the visceral arteries.[14]

Histology is the gold standard for diagnosis; however, vascular tissue is available only in patients undergoing surgery in postmortem cases. Although we could not make a histological diagnosis, the exclusion of all other possibilities and the classical angiographic appearance may suggest the diagnosis.

There are no specific guidelines for the management of SAM. Patient presenting with shock and intra-abdominal hemorrhage are treated by emergency surgery or endovascular therapy. A conservative approach with computed tomography angiography (CTA) follow-up appears most appropriate in patients without initial bleeding complications or with aneurysms of small size and no interval growth.[4] Awareness about this entity among radiologist is important as they may be the one to suggest the diagnosis, treat it by intervention, and follow-up with imaging.

Funding

No source of support in the form of grants, equipment, drugs, or all of these.

Conflicts of Interest

No conflicts of interest.

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