

Reconstructive surgery for segmental arterial mediolysis involving both the internal carotid artery and visceral arteries

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Segmental arterial mediolysis (SAM) is a rare, nonatherosclerotic, noninflammatory arteriopathy. A 52-year-old man with sudden hemiparesis of the right side was found to have an aneurysm of the left internal carotid artery and concomitant multiple aneurysms of the extrahepatic, celiac, and superior mesenteric arteries. Reconstructive operations using autologous vein graft were performed to treat the aneurysms. The histopathology analyses of resected arterial and aneurysmal specimens showed characteristics consistent with SAM. To our knowledge, a successfully treated case of SAM affecting both the carotid artery and visceral arteries has not previously been described. (*J Vasc Surg* 2006;43:623-6.)

Segmental arterial mediolysis (SAM) is a rare, nonatherosclerotic, nonvasculitic arteriopathy of unknown cause characterized by development of a dissecting hematoma, aneurysm, occlusion, and hemorrhage following lysis of the arterial media.¹ The disorder affects small- to medium-sized intra-abdominal muscular arteries or, less frequently, intracranial, coronary, or pulmonary arteries. We describe a case of SAM in which preoperative angiography revealed a concomitant left internal carotid artery (ICA) aneurysm and multiple visceral artery aneurysms affecting the celiac to the hepatic artery and the superior mesenteric artery. To our knowledge, a successfully treated case of SAM affecting both the carotid artery and visceral arteries has not previously been described.

CASE REPORT

A 52-year-old man with an 18-year history of well-controlled hypertension had a stroke that resulted in hemiparesis on the right side. The patient's medical history included a hemorrhage of the right thalamus when he was 36 years old and of the left thalamus when he was 43 years old. Clinical examinations indicated a cerebral infarction in the left carotid artery distribution and an aneurysm, with mural thrombus, of the left ICA. A computed tomography (CT) scan incidentally revealed a 3-cm aneurysm of the common hepatic artery. The patient was referred for surgical treatment.

The patient's temperature was 36.3°C; his heart rate, 72 beats/min; and his blood pressure, 135/75 mm Hg. A small 2-cm mass was palpable on the left side of the patient's neck. The results of cardiac, pulmonary, and abdominal examinations were normal, with no bruits observed. Results of laboratory tests included a

white blood cell count of $7.3 \times 10^9/L$, a hematocrit value of 0.40, and a C-reactive protein level of 1 mg/L. An antinuclear antibody test yielded negative results. Except for the aneurysms, the patient had no other clinical characteristics that met the criteria for the Marfan or Ehlers-Danlos syndromes.

Contrast-enhanced CT scanning and angiography showed a 2-cm fusiform left ICA aneurysm with mural thrombus (Fig 1), a fusiform celiac artery aneurysm (Fig 2, A and B), a fusiform distal superior mesenteric artery aneurysm (Fig 2, C), and a large, 6.5- × 5.5-cm saccular aneurysm with mural thrombus of the extrahepatic artery (Fig 2, A and B). The extrahepatic artery aneurysm had grown rapidly since an assessment 5 months earlier. All of the aneurysms contained regions of arterial stenosis in their proximal portions, suggesting a focal spontaneous dissection or primary aneurysmal degeneration of the vessel. The renal arteries appeared normal. The imaging results were considered to be consistent with a diagnosis of SAM.

The hepatic and celiac artery aneurysms were treated with reconstruction by using autologous saphenous vein graft. At operation, the presumptive diagnosis was SAM complicated by an impending rupture of the hepatic artery aneurysm. Because the hepatic artery aneurysm involved both the proper hepatic artery and the bifurcation of the right and left hepatic arteries, the vein graft was therefore interposed between the origin of the celiac trunk and the right hepatic artery, with an end-to-end anastomosis created between the left hepatic artery and a branch of the vein graft. The proximal portion of the splenic artery was involved in the celiac artery aneurysm; therefore, the splenic artery was reanastomosed to the vein graft by using an end-to-side anastomosis. The gastroduodenal, right gastric, and left gastric arteries were ligated. The distal superior mesenteric artery aneurysm was not treated because its diameter was <1.5 cm and no intestinal ischemia was observed.

Two months after treatment of the visceral aneurysms, reconstruction for the left ICA aneurysm was performed with an autologous vein graft, with good immediate results. Postoperative angiography showed good patency of all reconstructed arteries (Fig 3). The patient has been observed for 10 months and is doing well.

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Competition of interest: none.

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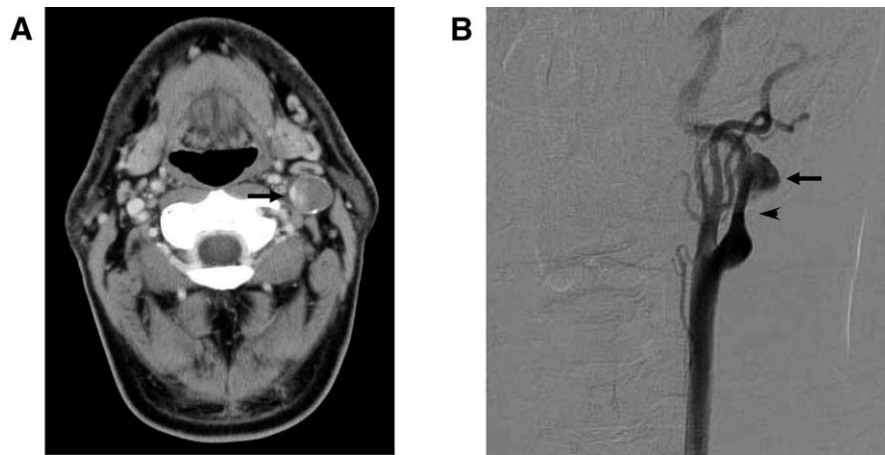


Fig 1. A, Contrast-enhanced computed tomographic (CT) scan shows an aneurysm, with mural thrombus, of the left carotid artery (*arrow*). B, Angiogram of the left carotid artery shows a fusiform aneurysm of the internal carotid artery (*arrow*) with narrowing (*arrowhead*) in the proximal portion.

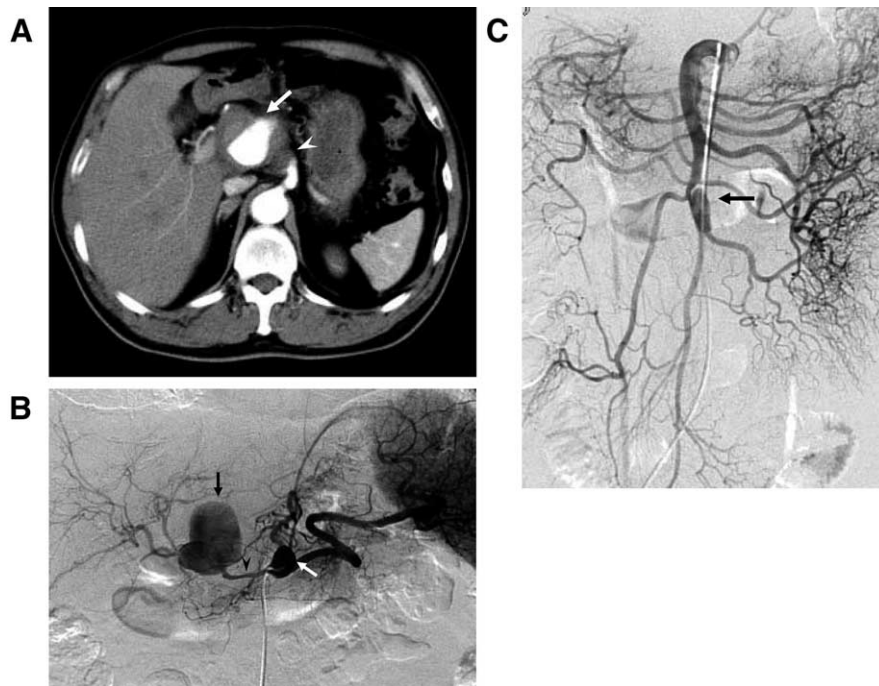


Fig 2. A, Contrast-enhanced computed tomography scan shows a large extrahepatic artery aneurysm (*arrow*), with mural thrombus, and a celiac artery aneurysm (*arrowhead*). B, Selective celiac artery angiogram shows a saccular hepatic artery aneurysm (*black arrow*) with narrowing (*arrowhead*) in the proximal portion, as well as a fusiform celiac artery aneurysm (*white arrow*). C, Selective superior mesenteric artery angiogram shows narrowing of the distal portion of the superior mesenteric artery associated with a poststenotic fusiform dilatation (*arrow*).

Histologic examinations of the resected specimens of the common hepatic, celiac, and left internal carotid arteries (including the aneurysmal portions) revealed marked segmental mediolysis of the muscular and elastic medial fibers at the proximal portion of the resected artery (Fig 4, A and B) and dissection of the media (Fig 4, C), without evidence of vasculitis. These findings were consistent with SAM complicated by aneurysm formation.

DISCUSSION

SAM, first described as a distinct pathological entity by Slavin and Gonzales-Vitale in 1976,² is characterized by necrosis of the outer tunica media-adventitia junction and replacement of the lysed muscle fibers by fibrin, erythrocytes, and granulation tissue.² Transmural me-

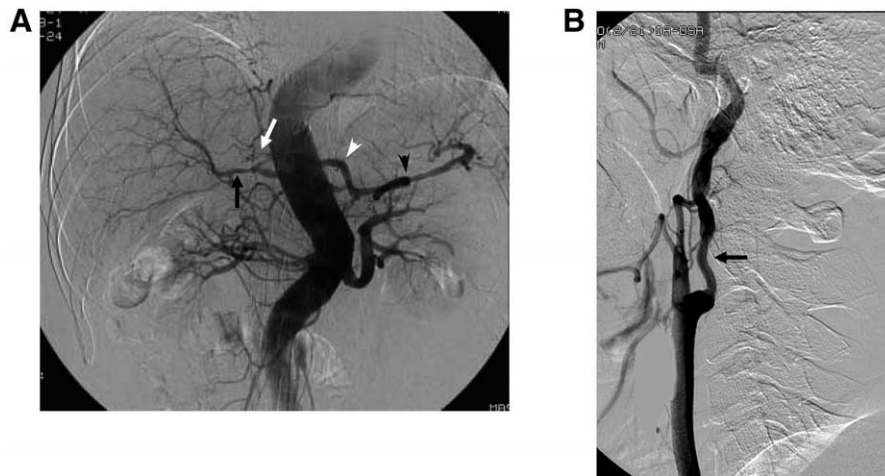


Fig 3. **A**, Abdominal aortogram shows good postoperative patency of the vein graft (white arrowhead), right (black arrow) and left (white arrow) hepatic arteries, and splenic artery (black arrowhead). **B**, Angiogram of the left carotid artery shows good postoperative patency of the vein graft (black arrow).

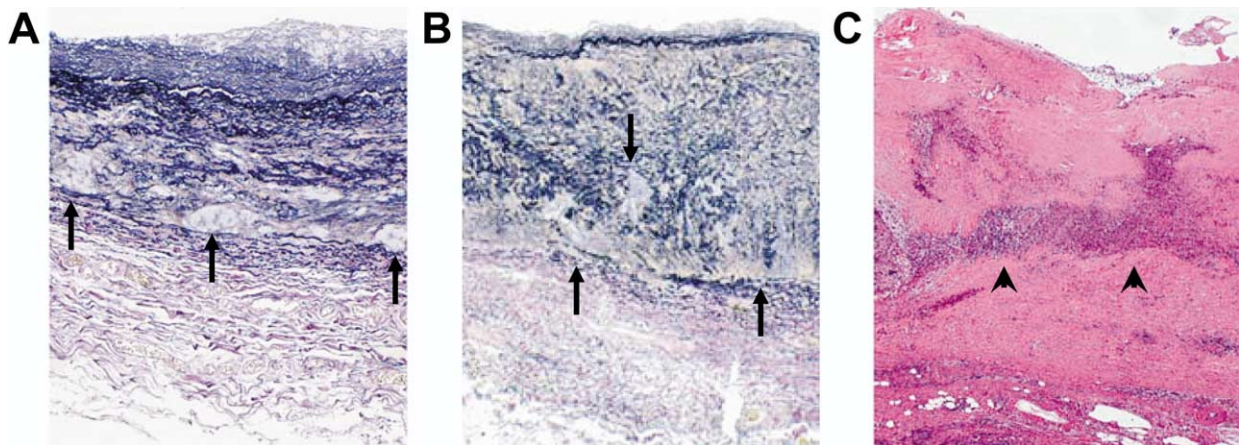


Fig 4. **A** and **B**, Pathology studies of resected arterial wall specimens show multiple segmental mediolysis lesions (arrows) of the muscular and elastic fibers of the media (elastica-van Gieson stain; magnification $\times 20$). **A**, Proximal portion of the resected left internal carotid artery aneurysm. **B**, Proximal portion of the resected common hepatic artery aneurysm. **C**, Pathology study of the resected common hepatic artery aneurysm shows an organized dissection (arrowheads) (hematoxylin and eosin stain; magnification $\times 20$).

diolysis results in arterial wall defects that can lead to focal dissection, aneurysm formation, stenosis, or hemorrhage caused by rupture of the artery. The etiology of SAM is poorly understood, but Slavin *et al*¹ postulated that the arterial lesions develop as a consequence of vasospasm. They also suggested that SAM might be a precursor lesion of fibromuscular dysplasia.¹ The pathologic appearances of fibromuscular dysplasia and SAM suggest an association between the two, and Lie³ proposed that SAM is a variant of fibromuscular dysplasia. The two disorders generally affect different sites, however. Fibromuscular dysplasia, the major cause of renovascular hypertension in young people, most commonly occurs in the renal arteries; visceral artery involvement is uncommon.

Abdominal SAM usually involves more than one visceral artery, and the branches of the celiac artery are much more frequently affected than those derived from the renal artery or the superior and inferior mesenteric arteries.¹ SAM affecting the visceral arteries most commonly occurs in middle-aged or elderly patients, with the typical clinical course consisting of sudden onset of abdominal pain and abdominal distension caused by arterial dissection and rupture with massive hemorrhage.¹ SAM affecting the cerebral arteries is very rare and tends to occur in young adults. The angiographic findings in SAM reflect the histologic changes and may include arterial dilatation, arterial dissection, and vessel occlusion.

In our patient, a left ICA aneurysm was found during the evaluation of a cerebral infarction. Subsequently, con-

comitant multiple visceral aneurysms were detected incidentally. Angiography showed segmental arterial narrowing in the proximal portion of each aneurysm; thus, the arteries may have dissected, resulting in aneurysmal degeneration. Although a definitive diagnosis of SAM requires a histopathology evaluation of the arterial lesions, the preoperative findings in our patient were comparable with SAM,⁴ and the absence of evidence of vasculitis on clinical and laboratory assessments allowed us to determine conclusively that no vasculitic process was present. We therefore assumed preoperatively that our patient had SAM, although fibromuscular dysplasia remained in the differential diagnosis. Later histopathology studies showed segmental mediolysis, wall gaps, and medial dissections in the affected arteries—findings characteristic of SAM.

Extracranial carotid artery aneurysms represent only a small proportion of all aneurysms. The most common causes of these lesions are assumed to be atherosclerosis, surgery, and trauma, although a few cases of SAM affecting the ICA have been observed in young adults who, like our patient, have experienced a cerebrovascular accident or transient ischemia.⁵ Sakata et al⁶ described an autopsy case of SAM involving both intracranial and intra-abdominal arteries. However, to our knowledge, ours is the first reported case of SAM simultaneously affecting both the carotid and visceral arteries that was diagnosed before rupture and treated successfully.

SAM is a noninflammatory process with an acute-phase mortality rate of nearly 50%.⁷ There is no standard treatment for this disorder. In our case, we decided to treat the aneurysms because the large hepatic artery aneurysm was growing rapidly and appeared likely to rupture and because we assumed that the ICA aneurysm had contributed to the

patient's cerebral infarction. Dolapci et al⁸ recommended operative treatment of all diagnosed hepatic aneurysms because of their high incidence of rupture and associated mortality rate of up to 20%. It is also essential to differentiate SAM from vasculitis, which is treated with immunosuppressive agents. So far, only one case in which SAM affecting a hepatic artery aneurysm was treated endovascularly (using coil embolization) has been described,⁴ but endovascular techniques such as coil embolization and stent-grafting may prove to be good treatment options for some patients with SAM.

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