Endovascular treatment of an isolated iliac artery aneurysm associated with segmental arterial mediolysis

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ABSTRACT

Segmental arterial mediolysis is a noninflammatory nonatherosclerotic vasculopathy of uncertain etiology characterized by dissection and/or aneurysm formation. It affects medium-to-large arteries, primarily the celiac, superior mesenteric, and renal arteries. Iliac involvement is rare, and its specific treatment has not been described. We detail a patient who presented with intrabdominal hemorrhage from a ruptured right colic artery aneurysm. He underwent transcatheter arterial embolization followed by right hemicolectomy. Histopathology confirmed the diagnosis of segmental arterial mediolysis. Endovascular treatment of a 3-cm iliac artery aneurysm was performed 18 months later. There was successful exclusion of the aneurysm demonstrated on computed tomography angiography at 10 years. (J Vasc Surg Cases Innov Tech 2023;9:101224.)

Keywords: Iliac artery aneurysm; Right colic artery aneurysm; Segmental arterial mediolysis

Segmental arterial mediolysis (SAM) is a rare enigmatic noninflammatory nonatherosclerotic nonimmune vasculopathy of uncertain etiology. The disease commonly afflicts patients in the fifth or sixth decades of life, with a slight predisposition for males. It affects medium-to-large arteries, most commonly the splanchnic and renal arteries. The cerebral arteries (younger patients) and coronary arteries (neonates, preterm infants) can be affected to a lesser extent.[1,2] Involvement of the iliac artery is rare.[3]

SAM was first described in 1976 by Slavin et al.[4] The pathogenesis of SAM remains obscure but is thought to be triggered by vasospasm. Lysis of the outer media of the arterial wall occurs, which facilitates dissection, stenosis, occlusion, and aneurysm formation. There is little inflammation. There are no genetic or serological markers. Although there are no proven risk factors, hypertension is commonly associated with SAM.[5] Its presentation can vary from clinically silent disease to hemorrhagic shock from aneurysm rupture, with mortality rates of up to 50%.[6,7] Due to the rarity of SAM, no standard treatment guidelines have been developed.

We describe a patient who presented with sudden abdominal pain and intra-abdominal hemorrhage from a ruptured right colic artery aneurysm. Computed tomographic angiography (CTA) also disclosed beading of the renal artery and a 3-cm isolated iliac artery aneurysm. The patient provided informed consent for the publication of this case and accompanying images.

CASE REPORT

A 56-year-old male with a history only significant for hypertension presented to an outside facility with sudden severe abdominal pain. CTA indicated right-sided hemoperitoneum with no active extravasation. There was focal dissection of the celiac artery with aneurysmal dilatation (16 mm), a "string-of-beads" appearance of the right renal artery, and a 3-cm fusiform aneurysm of the left common iliac artery (Fig 1). Laparotomy was performed with evacuation of hematoma. No bleeding source was identified. The patient continued to require multiple blood transfusions and was transferred to our facility for further care.

On arrival, the patient was afebrile, hypotensive (blood pressure, 90/60 mm Hg) and tachycardic (pulse, 120 bpm). Hemoglobin was 6.3 g/dL. Abdominal examination was significant for tenderness in the upper quadrants without rebound tenderness. He had no history of diabetes, smoking, or intravenous drug use. He had no past surgical history. His only home medications were atenolol and losartan. His family and personal histories were negative for connective tissue disorders, vasculitis, or autoimmune diseases. Laboratory evaluation did not show elevations in leucocyte count, creatinine, liver-associated enzymes, erythrocyte sedimentation rate, or C-reactive protein. Further laboratory results ruled out infectious etiology for his presentation.

Blood transfusion was initiated, and the patient was taken immediately for angiography. Vascular access was achieved through the left common femoral artery under ultrasound guidance and using a 5F sheath. Lateral aortogram was performed. Selective catheterization of the celiac and superior mesenteric arteries was performed with a Levin catheter (Angiodynamics) (Fig 2). There was focal dissection within the celiac artery with aneurysmal dilatation. Multiple tandem areas of aneurysmal dilatation were noted within the branches of the superior mesenteric artery. A site of extravasation was noted in the right colic artery. Renegade STC microcatheter and Transend wire

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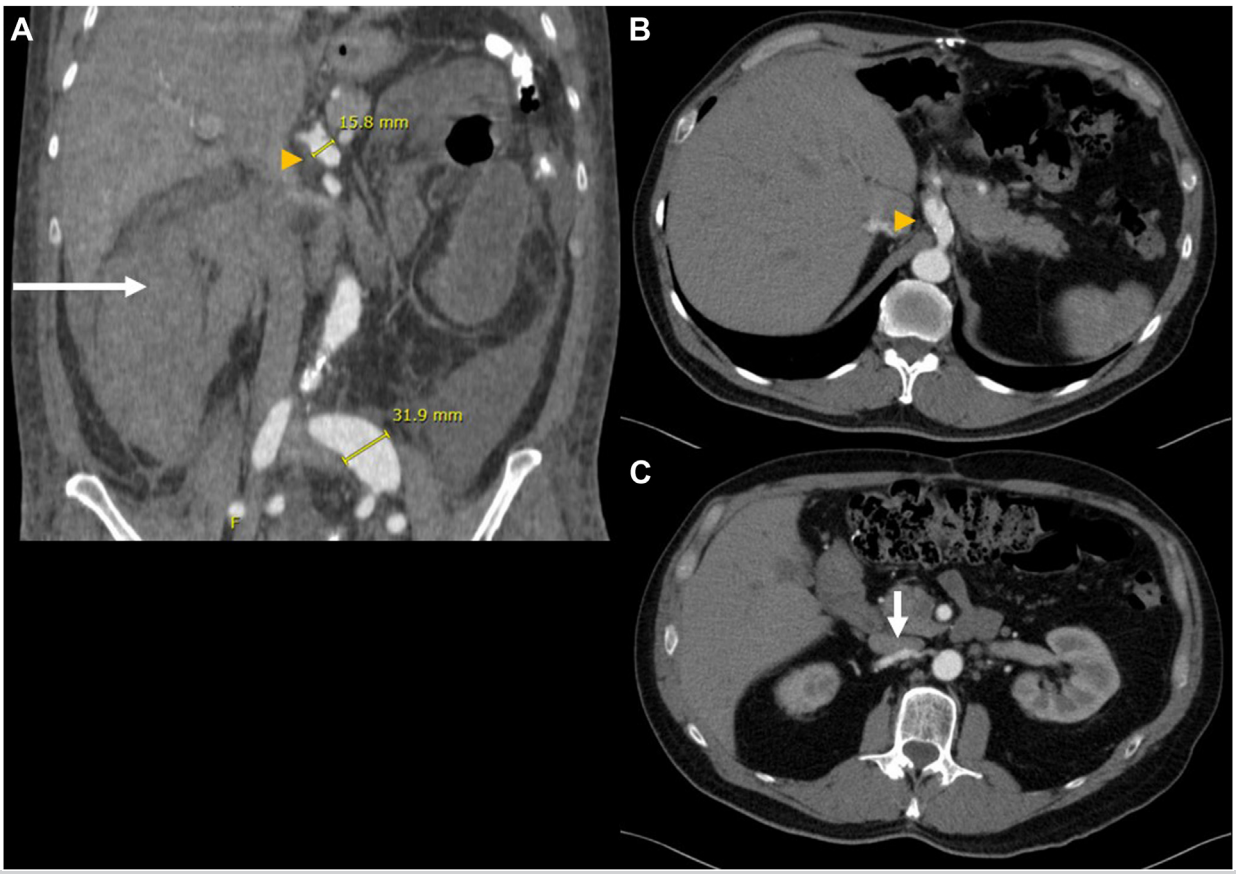

Fig 1. Computed tomography angiography (CTA) of the abdomen and pelvis. **A,** Coronal view indicates right-sided hemoperitoneum (*white arrow*). *Orange arrowhead* indicates celiac artery aneurysm with dissection. A 3-cm left common iliac artery aneurysm is delineated. **B,** Axial view of the celiac axis with focal dissection and aneurysm. **C,** Axial view of the right renal artery with beading.

(Boston Scientific) were advanced coaxially into the right colic artery. Gelfoam, two 3-mm VortX Diamond coils (Boston Scientific), and one 4-mm Nester coil (Cook Medical) were deployed to secure hemostasis. After the endovascular procedure, clinical parameters improved. However, 4 hours later, the patient developed hemodynamic instability and lower gastrointestinal bleeding. The patient was taken emergently for laparotomy. Four liters of blood was evacuated. The proximal transverse colon was dusky, and right hemicolectomy was performed. The patient was transfused a total of 10 units of packed red blood cells. The patient had an uncomplicated postoperative course and was discharged home 10 days later.

Histological examination of the right colon showed marked mediolysis with arterial wall gaps. There was no inflammatory or atherosclerotic changes. Aneurysm formation was evident (Fig 3). These histological findings were consistent with SAM.

Rheumatologic workup was negative for antinuclear antibodies, antineutrophil cytoplasmic antibodies, double-stranded DNA, myeloperoxidase antibodies, anti-proteinase 3 antibodies, rheumatoid factor, abnormalities in complement proteins, cryoglobulins, human immunodeficiency virus, and hepatitis B or C. Antiphospholipid panel including anticardiolipin antibody and lupus anticoagulant was negative. Transthoracic echocardiogram was normal.

Surveillance CTA after 18 months indicated unchanged celiac, right renal, and left iliac artery pathologies. He underwent endovascular repair of the 3-cm left common iliac aneurysm using the Gore Excluder system (Fig 4). Open bilateral femoral artery exposures were performed (ultrasound-guided percutaneous femoral access is our current practice). The left internal iliac artery was embolized with a 20-mm Amplatzer plug II (Abbott Laboratories). The main body (23 mm $\times$ 14.5 mm $\times$ 18 cm) was deployed via the left groin 2 cm below the renal arteries. A 20 mm $\times$ 9.5 cm right contralateral limb was placed. The left side was extended with a 12 mm $\times$ 10 cm iliac extender limb. He had mild left buttock claudication that resolved within 6 months and no sexual dysfunction. Follow-up CTA at 10 years indicated successful exclusion of the left common iliac artery aneurysm and unchanged celiac and right renal artery morphologies (Fig 5). No other stigmata of SAM were identified on follow-up imaging. His blood pressure has been well controlled on two antihypertensive medications.

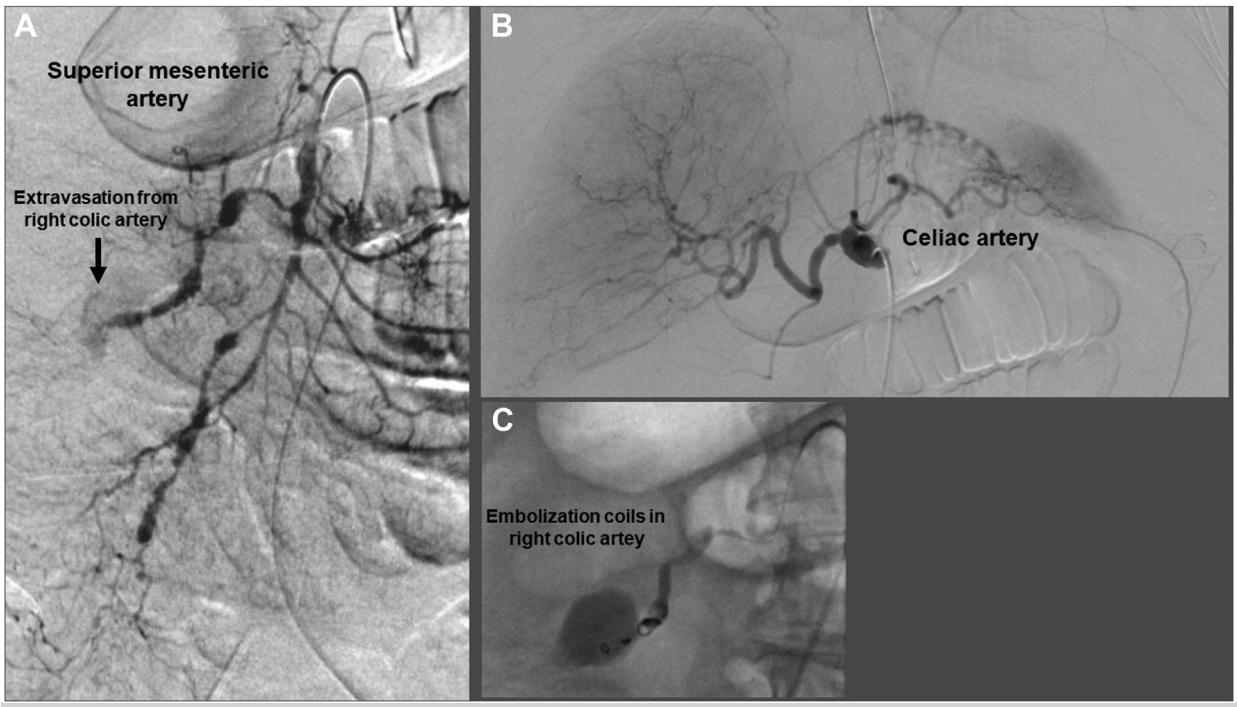

Fig 2. Contrast angiography. **A,** Selective catheterization of the superior mesenteric artery demonstrating multiple tandem areas of aneurysmal dilatation involving the branches of the superior mesenteric artery. There is active extravasation from the right colic artery. **B,** Selective catheterization of the celiac artery demonstrating aneurysmal dilatation. **C,** Magnified view of extravasation from the right colic artery. Embolization coils deployed proximal to the site of active extravasation in the right colic artery.

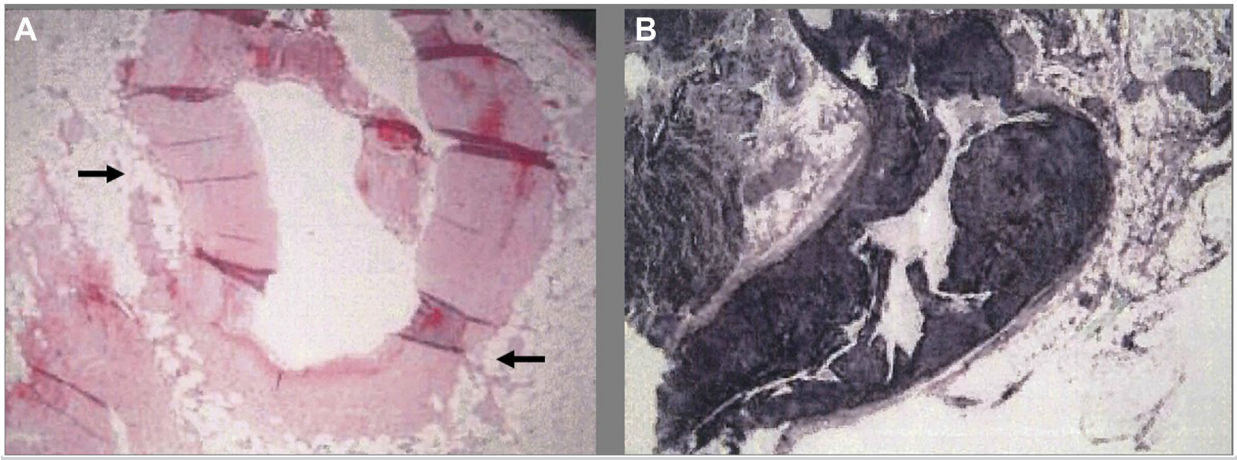

Fig 3. Histolopathological examination of the right colon. **A,** High power view of a medium-sized muscular artery showing changes consistent with segmental arterial mediolysis. There is lack of inflammatory cells and arterial gaps within the media (*black arrows*). **B,** Trichrome stain indicating aneurysmal formation.

DISCUSSION

Because SAM is an uncommon arteriopathy, its diagnosis can be elusive. A high index of suspicion is necessary. The gold standard for diagnosing SAM is histopathologic examination. However, direct arterial sampling is not always performed, as endovascular interventions have now become the first-line treatment. SAM should be suspected in middle-aged to elderly patients presenting with sudden intrabdominal hemorrhage. CTA is the best modality for the radiographic diagnosis of SAM.[8] Naidu et al found that the most common CTA imaging manifestations of SAM were that of dissection (86%), aneurysm (57%), beading (28%), occlusion (19%), and wall thickening (15%).[9] Angiography provides further delineation of the disease and offers the option of endovascular treatment.[10] Transcatheter embolization of aneurysmal disease has reduced the mortality rate for rupture from 50% to 25%.[6] Because pathological evaluation is not possible

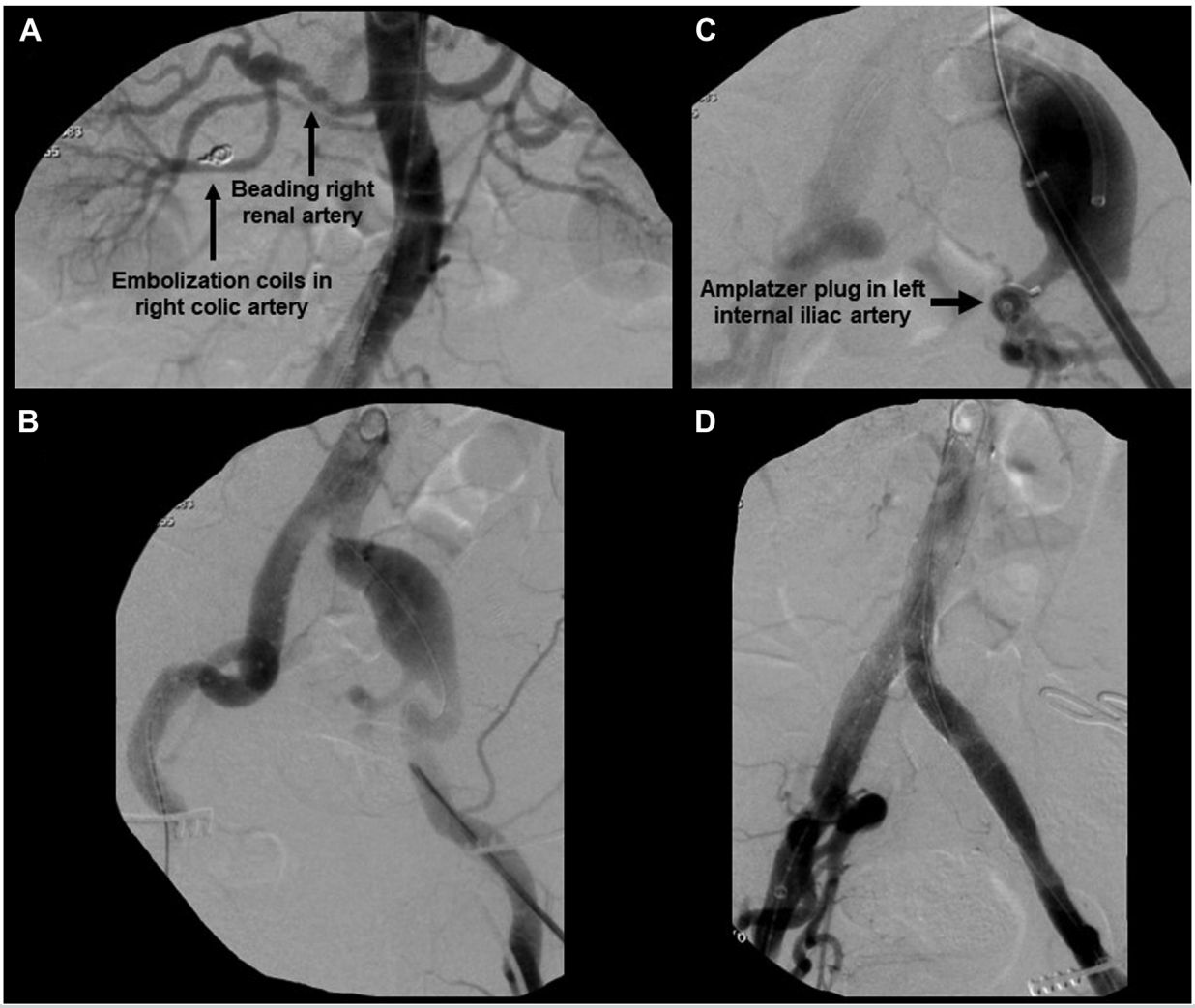

Fig 4. Endovascular repair of left common iliac artery aneurysm. **A,** Abdominal aortogram indicating "string of beads" appearance of the right renal artery. Note embolization coil in the right colic artery. **B,** Large left iliac artery aneurysm prior to repair. **C,** Amplatzer plug deployed in the left internal iliac artery. **D,** Completion angiogram indicating successful exclusion of the left common iliac artery aneurysm.

with endovascular therapy, criteria have been developed for the noninvasive diagnosis of SAM. It incorporates the typical radiographic findings, lack of genetic predisposition, low suspicion of alternative arteriopathies, and absence of inflammatory laboratory markers in order to secure the diagnosis of SAM.[6,11] In our patient, histological evidence was available and provided the definitive diagnosis of SAM. It is unclear whether all renal and visceral artery dissections/aneurysms should be attributed to SAM if there is no evidence of genetic disease, fibromuscular dysplasia, or inflammatory arteritis.

The pathology of SAM consists of two phases: the initial insult, followed by a remodeling and reparative phase. It has been proposed that repetitive vasoconstriction initiates the injurious phase. Vacuolar degeneration occurs within the media. The vacuoles burst and initiate a lytic process in the media (mediolysis). Subsequently, a tear develops, which separates the outer media from the adventitia layer. These arterial gaps weaken the arterial wall, leading to dissection and aneurysm formation. The reparative phase is characterized by exuberant granulation tissue deposition into the arterial gaps, which may cause focal stenosis. There is little inflammatory component to this process, which distinguishes it from vasculitis. The key region of derangement in SAM is the media; in atherosclerosis, the intima is injured. The pathological process can occur in a focal area or within the entire circumference of the artery (segmental skip lesions). The entire panoply of injurious lesions in multiple regions and their asynchronous maturation is the morphological hallmark of SAM. Multiple aneurysms are found in one-third of patients with SAM.[6] Our patient demonstrated the coexistence of celiac artery dissecting aneurysm, right colic artery aneurysm, right renal artery "string-of-beads" morphology, and left iliac artery aneurysm.

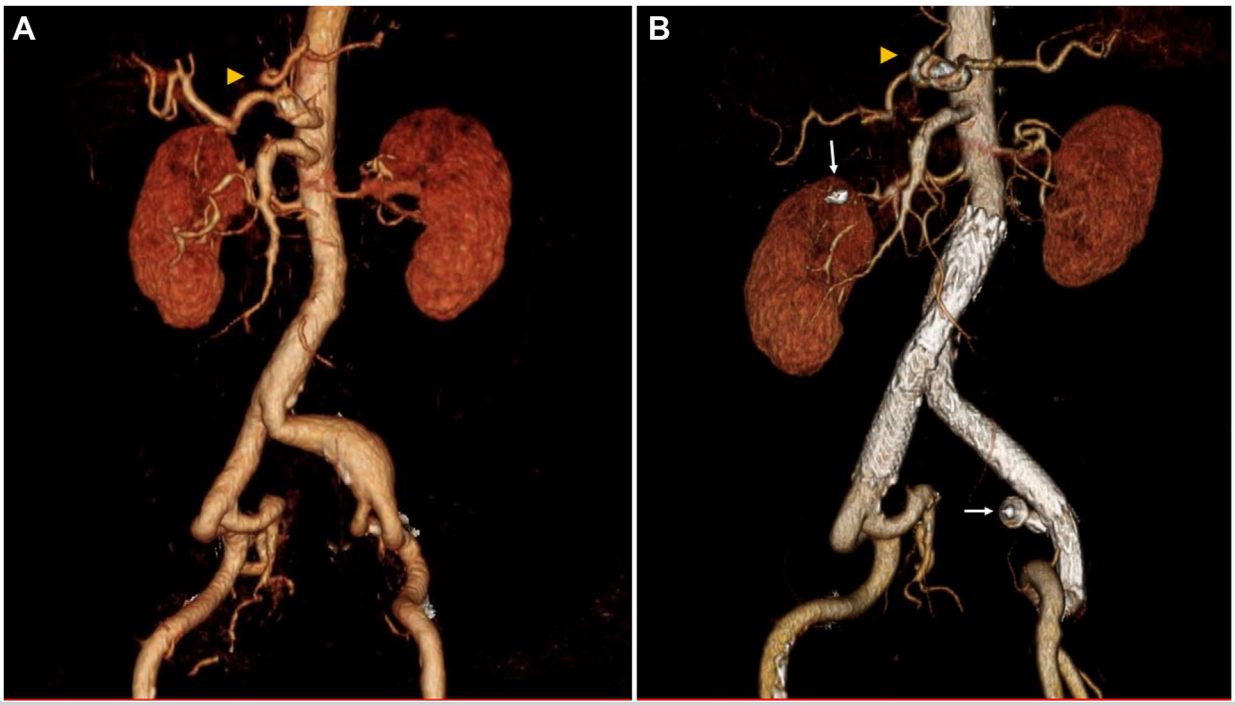

Fig 5. Three-dimensional reconstruction of the computed tomographic angiogram (CTA). **A,** Preintervention view prior to right colic artery embolization and endovascular repair of iliac artery aneurysm. Note the celiac artery aneurysm (*orange arrowhead*) and left iliac artery aneurysm. **B,** Postintervention view 10 years following endovascular repair of the iliac artery aneurysm. Celiac artery aneurysm is unchanged, and the left iliac artery aneurysm has been excluded. Embolization coils are noted in the right colic artery, and Amplatzer plug is noted in the left internal iliac artery (*white arrows*).

The differential diagnosis for SAM is vast and includes inflammatory/immunologic vasculitides (polyarteritis nodosa, antineutrophil cytoplasmic antibody-associated vasculitis, rheumatoid vasculitis, giant cell arteritis, Takayasu's arteritis, Behçet's disease, Kawasaki disease, Henoch-Schonlein purpura), inherited defects in vessel wall structural proteins (Ehlers-Danlos syndrome, Marfan syndrome, Loeys-Dietz syndrome, neurofibromatosis), fibromuscular dysplasia, infection (mycotic aneurysm, endocarditis), and atherosclerotic disease. One disease entity particularly difficult to differentiate is fibromuscular dysplasia (FMD). Some have postulated that SAM is a variant or precursor of FMD.[12,13] FMD is another nonatherosclerotic noninflammatory condition affecting medium-sized arteries with similar radiographic and histological findings. However, the demographics and distribution of affected vessels are distinctly different. FMD occurs mostly in young to middle-aged women; SAM typically affects males. Patients with SAM present with sudden pain and profuse bleeding from the intestinal arteries. FMD may present with ischemic changes in the renal and cerebral beds, causing premature hypertension and stroke and rarely pain or rupture. Vasculitis tends to present as subacute systemic illness with substantial elevation of inflammatory markers (erythrocyte sedimentation rate, C-reactive protein) and/or autoantibodies. There are absent inflammatory markers and autoantibodies in SAM. Our patient did not show systemic features suggestive of a vasculitis process and had negative serology.[14] Congenital diseases such as Marfan, Ehlers-Danlos, and Loeys-Dietz syndromes can present with aneurysm and/or dissection and should be excluded with genetic testing. Our patient did not exhibit clinical features associated with these genetic syndromes. Because there was histopathologic confirmation of SAM, genetic testing was not performed in our patient. Atherosclerotic disease is typically widespread and has a shaggy, irregular appearance favoring branch points and ostia. SAM is randomly distributed along the artery with sparing of bifurcations. In patients with multiple aneurysms or an aneurysm at an arterial bifurcation, a mycotic aneurysm should be considered. Infectious destruction of the arterial wall is often associated with systemic infection. Infectious workup in our patient was negative.

There are no established guidelines for the management of SAM. The natural history is unpredictable, and SAM may evolve over time, remain stable, or suddenly deteriorate. Regression or stability of imaging findings can be seen in 80% of cases.[8,15-18] Management is determined by clinical presentation and can involve medical, endovascular, or surgical intervention. Optimal blood

pressure control is thought to prevent propagation of dissection and further lysis of the arterial wall. Reducing the cardiovascular risk profile with lifestyle modification, tobacco cessation, statins, and antiplatelet medications is advocated.[19] It is important to rule out vasculitis because there is no role for immunosuppression in the management of SAM. Endovascular intervention is recommended as first-line therapy in patients with hemodynamic instability or end-organ ischemia. Our patient had a 3-cm isolated common iliac artery aneurysm. Isolated common iliac artery aneurysms are rare, accounting for approximately 7% of all aortoiliac aneurysms.[20] The natural history of iliac aneurysms is not well-defined, and there is no consensus on the diameter threshold for treatment. A recommended threshold size of 3 to 4 cm has been suggested.[20,21] The European Society for Vascular Surgery recommends that repair of isolated iliac artery aneurysm be considered at a minimum of 3.5 cm in diameter.[22] It is unclear whether SAM alters the natural history of isolated iliac artery aneurysm. Although spontaneous regression of splanchnic artery aneurysms has been observed angiographically, regression of iliac artery aneurysm associated with SAM has not been described.[8,16-18] Our patient preferred treatment over observation and underwent successful repair of the iliac aneurysm with a bifurcated modular endograft. Embolization of the internal iliac artery was required to secure an adequate distal landing zone. The iliac branch endoprosthesis was not available for use in the United States at the time of the procedure in 2013. The patient had mild left buttock claudication, which resolved within 6 months. Surveillance CTA at 10 years has confirmed successful exclusion of the iliac aneurysm.

Endovascular treatment of iliac artery aneurysm associated with SAM is a viable option. There is no consensus regarding surveillance imaging after the diagnosis of SAM. Annual imaging with CTA or magnetic resonance angiography has been suggested.[11,19] Surveillance with yearly CTA and/or color duplex ultrasound will be used in our patient to monitor both the endovascular repair and the celiac artery aneurysm. Prophylactic intervention for aneurysmal disease seems warranted even though the natural history of untreated lesions is not well-defined.

CONCLUSION

Endovascular exclusion of iliac aneurysm associated with segmental arterial mediolysis can be successfully achieved with durable results.

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