Segmental arterial mediolysis

A rare cause of rapidly progressive arterial dissections

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Segmental arterial mediolysis (SAM) is a rare nonatherosclerotic, noninflammatory vasculopathy characterized by lytic degeneration of the arterial media, intramural dissection, and thrombosed/ruptured dissecting aneurysm.¹ We present a putative case of SAM in a patient who presented with rapidly progressive systemic arterial dissections, including the cervical and cerebral vasculature, leading to devastating strokes, subarachnoid hemorrhage (SAH), and death.

Case report

A 49-year-old woman with a history of hypertension presented to a community hospital with abdominal pain. Abdominal ultrasound suggested bilateral renal infarctions and severe renal artery stenosis. Later, she became somnolent, reported a headache and “ripping” back pain, and developed decreased movement in the legs. Pertinent neurologic examination demonstrated mild somnolence, lower extremity paraplegia with hyperreflexia, bilateral extensor responses, and a T12 spinal sensory level. ECG revealed flattened T waves and an associated troponin peak consistent with non-ST elevation myocardial infarction requiring conservative management given clinical instability. Head CT and spinal MRI revealed extensive SAH, with epidural and subdural hemorrhage from C2-T11 with medullary ischemia. CT angiography (CTA) of the head and neck revealed irregular stenoses and luminal contour in the bilateral cervical carotid and right vertebral arteries and multifocal irregularities throughout the intracranial circulation (figure). Thoraco-abdominal CTA demonstrated irregularities in the subclavian, celiac, common hepatic, bilateral renal, and external iliac arteries with intramural hematoma concerning for multifocal systemic dissections.

Laboratory serologic testing showed no supporting evidence of systemic vasculitis, hypercoagulability, or infection given negative antineutrophil cytoplasmic antibodies, extractable nuclear antigen, antinuclear antibodies, double-stranded DNA, Factor II and V Leiden mutations, protein C and S deficiency, antiphospholipid antibodies, hepatitis B/C, and cytomegalovirus antibodies. Temporal artery biopsy demonstrated no evidence of inflammation, but demonstrated prominent intimal hyperplasia compatible with fibromuscular dysplasia. She ultimately became hypotensive, requiring high-dose vasopressors, and her condition rapidly deteriorated to a comatose state with impaired brainstem reflexes. A brain MRI showed large acute infarctions in the bilateral anterior, posterior, left middle cerebral, and cerebellar arterial territories. After further goals of care discussions, she was transitioned to comfort care and died.

Postmortem examination confirmed multifocal arterial dissections of different ages including the cervical, subclavian, coronary, celiac, hepatic, renal, iliac, and small epicardial and intramedullary arteries. Arterial wall histology revealed hematoma between medial and adventitial layers with compression of the true lumen, thinning of the medial layer with degradation of

Practical Implications

Consider segmental arterial mediolysis in patients presenting with a combination of acute abdominal pain and rapidly progressive cerebrovascular vasculopathy.

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Figure Radiographic-histologic correlates

(A) Celiac arterial dissection with hematoma between media and adventitia (Movat pentachrome). (B, C) Focal loss of internal elastic laminae and thinning of media layer in celiac artery (Movat pentachrome, hematoxylin & eosin). (D–F) Right vertebral artery with dissection between outer media and adventitial layers (Movat pentachrome, elastin, Movat pentachrome). (G) CT angiography demonstrates celiac artery dissection. (H) CT angiography head shows left middle cerebral artery stenosis and diffuse irregularity. (I) CT angiography shows right vertebral irregular tapering.

smooth muscle fibers, collagenous loss, and focal elastic breaks with breakdown of the elastic laminae. There was no evidence of vasculitis or atherosclerosis. Presence of foamy or vacuolar degeneration of the smooth muscle in the outer media was considered to be most supportive of segmental arterial mediolysis.

DISCUSSION

SAM typically presents late in the fifth decade, manifesting with abdominal pain due to visceral artery dissections and hemorrhage.² The clinical syndrome is rapidly progressive with a high mortality rate (25%–50%) due to multifocal degeneration of the arterial wall, leading to dissections and aneurysm formation.³ Approximately 18% of cases involve the cerebrovasculature.³ With increases in the use of noninvasive vascular imaging, the diagnosis of SAM has been more widely reported in the last decade.

After proceeding to autopsy, the pathologic differential included SAM, fibromuscular dysplasia (FMD), and cystic medial necrosis (CMN).⁴ SAM is histologically similar to FMD with loss of smooth muscle integrity and intimal proliferation. However, these entities are clinically distinct as FMD often has a favorable prognosis compared to the rapid clinical deterioration in SAM. In addition, FMD shows alternating areas of thinned media and thickened fibromuscular ridges containing increased collagen, not seen in our case. Finally, CMN was unlikely given absence of basophilic ground substance and cystic lesions in the media.

Shared genetic associations in FMD and SAM remain poorly defined and further research is needed to investigate environmental triggers of connective tissue vasculopathies. Of interest, independent genomic analyses have recently discovered a common risk allele for both FMD and cervical artery dissection in the PHACTR1 gene on chromosome 6.^{5,6} In our case, multifocal arterial dissections in the cervical and intracranial vasculature contributed to neurologic decline, with intracranial extension of the right vertebral artery dissection causing SAH. Of note, the intracranial circulation lacks an external elastic lamina, making it prone to arterial rupture in the setting of cerebral vasculopathy.⁷ A remarkable aspect of this patient's disease was the rapid pace with which it progressed from initial symptoms to death over several days. This rapidly progressive phenotype may represent a severe variant along the spectrum of SAM. Neurologists should be aware of this uncommon devastating syndrome in a patient presenting with both cerebrovascular and visceral vasculopathy. Earlier recognition of SAM may lead to prompt management and better outcomes.

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AUTHOR CONTRIBUTIONS

D.A. Lapidès: drafting of manuscript. M.S. Abusamaan: drafting of manuscript. J.J. Davick: pathologic analysis and interpretation. A.M. Sharma: critical revision of manuscript for clinical and intellectual content. J.W. Mandell: pathologic analysis and interpretation. M.B.S. Lopes: pathologic analysis and interpretation. J.J. Provencio: critical revision of manuscript for clinical and intellectual content. B.B. Worrall: critical revision of manuscript for clinical and intellectual content. A.M. Southerland: drafting of manuscript, critical revision of manuscript for clinical and intellectual content.

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