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Case Report

Segmental Arterial Mediolysis: Report of 2 Cases and Review of the Literature

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Segmental arterial mediolysis (SAM) is an idiopathic noninflammatory vasculopathy involving small to medium arteries, usually in the abdomen, although arteries in the cerebral and coronary circulations also may be affected. Some cases present as abdominal apoplexy due to aneurysmal rupture, but ischemia and infarction also occur. Not uncommonly, SAM may be misdiagnosed as a systemic necrotizing vasculitis. We present 2 patients with bilateral renal infarctions, cerebral arterial dissections, and visceral artery microaneurysms. Both were diagnosed initially as polyarteritis nodosa. The diagnosis was changed to SAM, in one case based on clinical and radiologic features, and in the other, on an open wedge kidney biopsy. We discuss the differential diagnosis and review the literature on SAM.
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Segmental arteriolysis medial (SAM) is an uncommon, usually self-limited, noninflammatory vasculopathy of unknown cause.[1-6] SAM involves predominantly small to medium intra-abdominal arteries, is untreatable, and must be differentiated from classic polyarteritis nodosa (PAN). In the absence of immunosuppressive therapy, PAN usually is fatal.[7,8] The diagnosis of PAN rests on a compatible clinical picture with confirmation by biopsy or angiography.

We report 2 patients presenting with bilateral renal infarctions, visceral artery microaneurysms, and arterial dissections. In both cases, the initial diagnosis was PAN. Therapy was initiated with steroids and cyclophosphamide in one patient. The diagnosis was changed to SAM and treatment was discontinued. The second case was confirmed as SAM by wedge kidney biopsy. We discuss the differential diagnosis and review the literature on SAM.

CASE REPORTS

Case 1

A 43-year-old white woman with no medical history presented to an outside hospital with 1 week of intermittent abdominal pain, nausea, and vomiting. She did not report experiencing fevers, night sweats, arthralgias, or myalgias. There was no anorexia, weight loss, or history of drug abuse. The only medication was daily pseudoephedrine. Enhanced computed tomography (CT) showed a right renal infarct. After several days, she was discharged on enoxaparin therapy. Several days later, she was readmitted with headache, nausea, and vomiting. CT and magnetic resonance angiography showed dissections of the right carotid and both vertebral arteries. The following day, she developed severe abdominal pain, and repeated CT now showed bilateral renal infarctions (Fig 1A). She was transferred to our hospital. Vital signs were normal. Examination findings were remarkable only for abdominal tenderness. Hemoglobin level was 13.6 g/dL (136 g/L), white blood cell count was $10 \times 10^3/\mu L$ (10×10^9/L), platelet count was $264 \times 10^3/\mu L$ (264×10^9/L), serum urea nitrogen level was 6 mg/dL (2.14 mmol/L), serum creatinine level was 1.1 mg/dL (97.24 μmol/L), aspartate aminotransferase level was 165 U/L, alanine aminotransferase level was 259 U/L, alkaline phosphatase level was 128 U/L, bilirubin level was within the reference range, and albumin level was 4.3 g/dL (43 g/L). Urinalysis showed blood (+1), no protein, and no casts. Erythrocyte sedimentation rate (ESR) was 10 mm/h, and C-reactive protein level was 9.47 mg/L (reference, <1.2 mg/L). Findings from a hypercoagulable workup were unremarkable. Antinuclear antibody was positive at 1:320, but C3 and C4 were normal. Antineutrophil cytoplasmic antibody and hepatitis C antibody results were negative. Hepatitis B surface antibody was positive.

A renal arteriogram showed the infarcts, a nonobstructing dissection of the right renal artery (Fig 1B), and microaneurysms in arteries of the kidneys (Fig S1, available as online supplementary material), liver, and spleen. PAN was diagnosed, and treatment with steroids and cyclophosphamide was initiated. Owing to the absence of constitutional symptoms, the normal ESR and hemoglobin values, and the predominance of arterial dissections, the diagnosis was changed to SAM. Positron emission tomography confirmed the absence of vascular inflammation. Immunosuppressive therapy was discontinued. After 12 months, the patient remains well and normotensive with a hemoglobin level of 14 g/dL (140 g/L), ESR of 9 mm/h, and serum creatinine level of 1.0 mg/dL (88 μmol/L).

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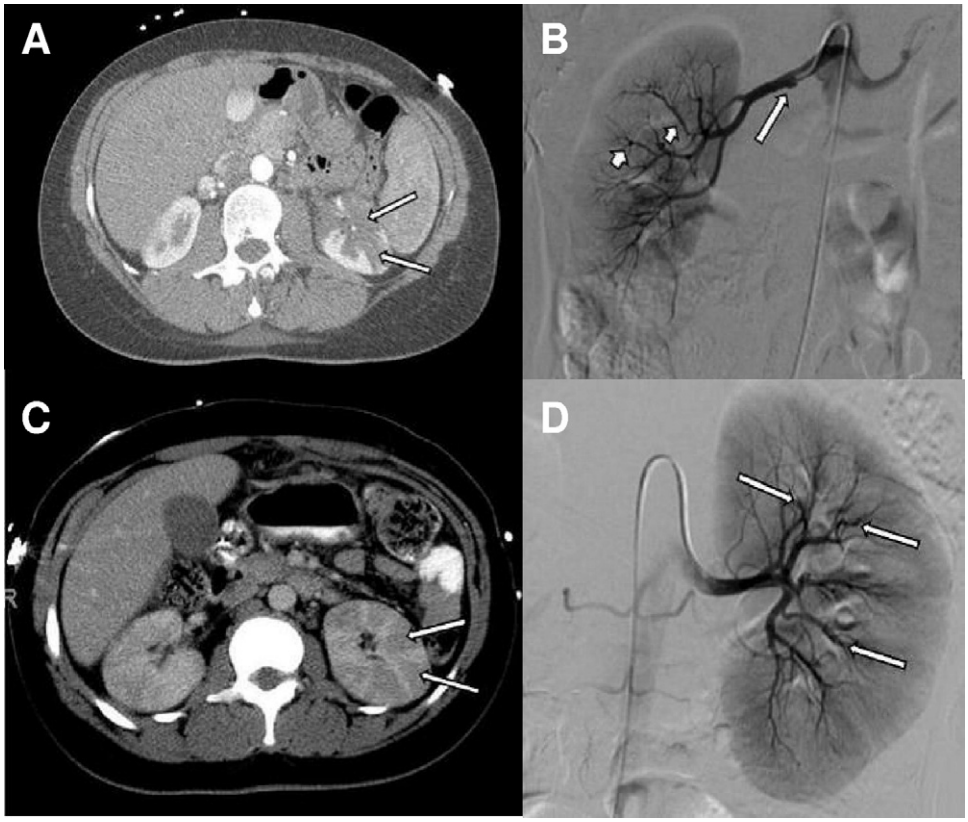

Figure 1. Radiologic images of patients (A, B) 1 and (C, D) 2. Patient 1: (A) contrast-enhanced computed tomographic scan of the abdomen shows wedge-shaped infarcts involving the left kidney (arrows), and (B) selective right renal arteriogram shows focal dissection of the main renal artery (long arrow) and microaneurysms (large arrowhead) and fusiform aneurysms (small arrowhead) of interlobular arteries. Patient 2: (C) bilateral striated renal cortical nephrograms with wedge-shaped areas of hypoattenuation (arrows) compatible with renal infarcts, and (D) selective left renal arteriogram shows abrupt occlusion of multiple interlobular arteries (arrows) compatible with thrombosis.

Case 2

A 29-year-old African American woman with a history of hypertension presented with aphasia and right-sided paresis. CT showed a large infarct in the territory of the left middle cerebral artery. White blood cell count was $13.4 \times 10^3/\mu L$ ($13.4 \times 10^9/L$), hemoglobin level was 8.8 g/dL (88 g/L), and platelet count was $326 \times 10^3/\mu L$ ($326 \times 10^9/L$). Comprehensive metabolic panel and coagulation study results were within the reference ranges.

The patient was taken to surgery for a left hemispheric decompressive craniectomy. On postoperative day 2, magnetic resonance angiography confirmed the infarct, with occlusion of the left internal carotid artery from the base of the skull to the level of the carotid bulb noted, presumably secondary to a dissection.

Hypercoagulable workup results were unremarkable. ESR was 140 mm/h. Serologic study results included negative antinuclear antibody, anti–extractable nuclear antigen, and anti-Ro/SSA. Anti-La/SSB was positive at 292 (reference range, 0-72) U/mL. C3 and C4 levels were within the reference ranges, but serum cryoglobulin results were positive at 1%. Hepatitis B and C serologic tests and antineutrophil cytoplasmic antibody results were negative.

On day 15, contrast-enhanced CT of the abdomen and pelvis showed bilateral striated renal cortical nephrograms (Fig 1C). An angiogram on day 24 showed small wedge-shaped defects in the right renal cortex in addition to scattered microaneurysms and vessel pruning in the distal branches of the right and left renal, hepatic, and possibly superior mesenteric arteries (Fig 1D).

Because of the anemia, elevated ESR, renal infarcts, and visceral microaneurysms, PAN again was suspected. On day 36, a wedge biopsy of the kidney was obtained. Renal cortical infarcts were present (Fig 2A). Segmental lesions of the media with loss of smooth muscle cells (termed "mediolysis" in the literature) were found in the interlobar, arcuate, and proximal intralobular arteries. An accompanying reactive intimal hyperplasia narrowed the lumens to cause the infarctions (Fig 2B-D). The patient was not treated with immunosuppression, but is receiving warfarin as anticoagulation treatment. She continues to be aphasic with residual right-sided paresis.

DISCUSSION

These 2 patients with similar presentations suggesting classic PAN were both eventually given a diagnosis of SAM, one clinically and one confirmed by biopsy. We discuss the differential diagnosis of the first patient in the absence of a tissue diagnosis and review the literature on SAM.

Classic PAN could produce infarctions and microaneurysms. Radiographically, typical findings are 2- to 3-mm aneurysms often associated with areas of narrowing of the arterial lumen. Renal infarction may ensue.[9] The initial radiologic report was PAN, and