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Cystic adventitial disease of the popliteal artery

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Cystic adventitial disease of the popliteal artery is a relatively rare entity, responsible for approximately 1 in 1,200 cases of claudication. We present a case with both classic history and imaging features. We hope that our experiences may increase radiologists' familiarity with this unusual but treatable entity.

Case report

A 49-year-old healthy, active, nonsmoking male presented with right-lower-extremity claudication and paresthesias. His symptoms began without inciting events approximately eight months prior to presentation and had gradually worsened until presentation. He described a cramping sensation beginning in his posterior thigh and spreading distally, followed by numbness and tingling of the foot and ankle. These symptoms occasionally occurred spontaneously but were typically brought on by walking.

Both initial studies, MRI of the lumbar spine and electrodiagnostic studies of the right leg, were normal. However, postexercise ankle-brachial indices of the right and left lower extremities were 0.28 and 1.26, respectively, raising the possibility for vascular etiology.

Imaging proceeded with contrast-enhanced MR angiography of the lower extremities. While the left side was normal, the right popliteal artery showed a focal narrowing with "hourglass" extrinsic impression (Fig. 1). On the same day, unenhanced MRI of the right knee revealed the source of this focal extrinsic compression to be a multiloculated focus of T2-signal hyperintensity encircling the adjacent popliteal artery. Imaging suggested continuity of this lesion with both the popliteal arterial wall and lateral knee joint (Fig. 2). Ultrasound was immediately performed to exclude vascular etiology for this serpiginous, T2-bright lesion.

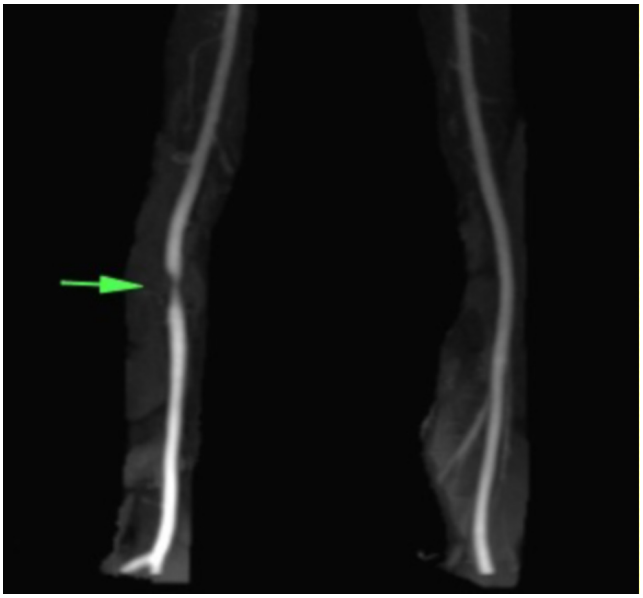

Figure 1. 49-year-old male with cystic adventitial disease. Postcontrast coronal MIP image from lower-extremity contrast-enhanced MR angiography demonstrates focal "hourglass" narrowing of the right popliteal artery (arrow).

Sonographic images confirmed the presence of multilobulated hypoechoic structures without vascular flow, causing focal stenosis of the right popliteal artery (Fig. 3).

Given the rapidity of symptom onset and progression, a diagnosis of cystic adventitial disease was favored. The patient subsequently underwent segmental resection of the diseased popliteal artery with vein graft reconstruction. Surgeons encountered an inflamed cystic structure encasing the popliteal artery. Exploration confirmed a side branch that extended laterally toward, but did not appear to involve, the lateral aspect of the knee joint. Surgical pathology revealed multiloculated cystic structures filled with

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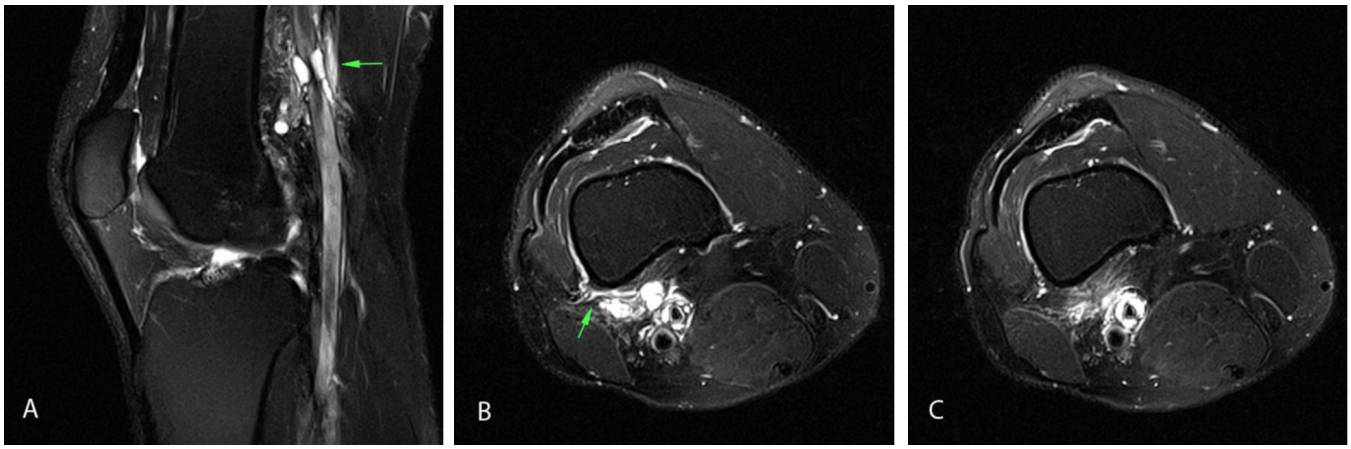

Figure 2. 49-year-old male with cystic adventitial disease. T2-weighted MR. A. Sagittal image demonstrates "hourglass" narrowing of the popliteal artery by an encasing multiloculated, cystic structure. (arrow) B. Axial image suggests continuity of the lesion with both the vessel wall and lateral knee joint (arrow). C. Axial image demonstrates marked circumferential narrowing of the popliteal arterial lumen due to surrounding T2-bright lobulated lesion.

thick, mucoid material. Microscopic findings confirmed cystic adventitial disease (Fig. 4).

Discussion

First described in 1947 by Atkins and Key (1), cystic adventitial disease is a relatively rare entity, responsible for approximately 1 in 1,200 cases of claudication (2). The popliteal artery is most commonly affected, composing approximately 85% of cases with classically unilateral distribution (3). Other reported sites of involvement include external iliac, common femoral, radial, and ulnar arteries (3, 4). Males are affected more often than females, and presentation is usually in the 4th or 5th decade (3, 5, 6). The exact etiology is still unknown; several theories have been proposed, including myxomatous degenerative condition, repetitive trauma causing intramural hemorrhage, synovial/ganglion migration from popliteal artery to the knee joint, and embryologic origin. The embryologic theory is currently the most widely accepted; it postulates aberrant inclusion of mucin-secreting mesenchymal cells from nearby joints into the vessel adventitia during embryogenesis (7).

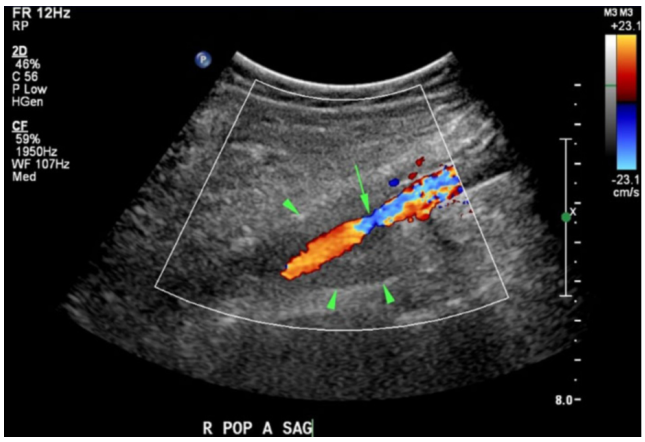

Figure 3. 49-year-old male with cystic adventitial disease. US of the right knee. Sagittal Doppler images confirm a focal stenosis within popliteal artery (arrow) with post-stenotic turbulent flow, seen as multicolor flow on color Doppler. There is absence of Doppler flow within the hypoechoic structure surrounding the popliteal artery, representing the cystic lesions (arrowheads).

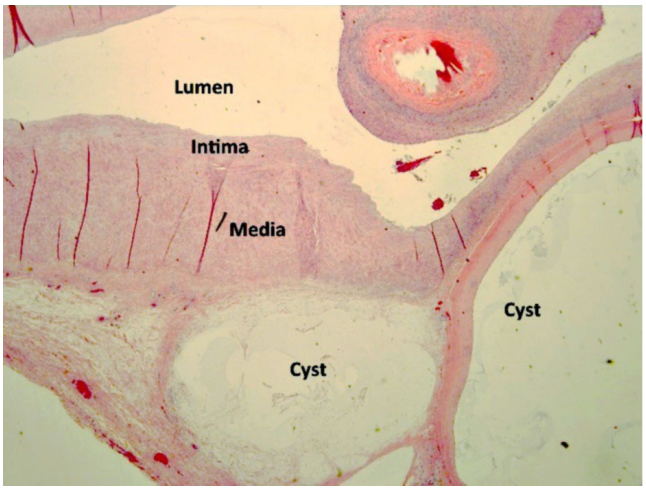

Figure 4. 49-year-old male with cystic adventitial disease. Low-power microscopic image demonstrating a fibrous-walled adventitial cyst with focal, synovium-like lining arising from the popliteal artery wall.

Typical imaging features are those depicted in this case. Ultrasound demonstrates anechoic or hypoechoic cystic

structures eccentric to the artery, often with resultant stenosis (5, 8, 9). Communication between adjacent knee joint and arterial adventitia has been reported (10) but is not required for the diagnosis. The classic arteriographic appearance is "hourglass" narrowing of the vessel when disease is circumferential (11) or a “scimitar” lumen when the lesion is eccentric (12). Ultrasound and angiography may be normal or nonspecific in some cases (6, 12). MRI is most informative, revealing T2 hyperintense cystic lesions surrounding and narrowing the artery. T1 signal within the cystic lesion is variable, depending on the composition of mucoid material (2, 11).

Spontaneous regression of cystic adventitial disease of popliteal artery has been reported; however, the natural history is typically progression toward further occlusion. Therefore, intervention is recommended in nearly all patients. Excision of cysts and artery followed by reconstruction with autologous vein graft is beneficial when the artery is completely occluded or thrombosed (12). In symptomatic patients without thrombotic occlusion, ultrasound-guided percutaneous aspiration has been shown to be safe and efficacious (13). Endovascular treatments have had disappointing results (14-16).

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