

Cystic adventitial disease of the popliteal artery presenting with features of entrapment syndrome

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ABSTRACT

Cystic adventitial disease is an uncommon cause of lower extremity claudication resulting from accumulation of mucinous fluid in an arterial subadventitial layer, typically of the popliteal artery. A popliteal bruit and/or reduced distal pulses with knee flexion may be seen on examination. Alternatively, popliteal artery entrapment syndrome triggers claudication via an aberrant arterial pathway or muscular hypertrophy. Decreased distal pressures with plantar or dorsiflexion is a key finding. This report details the case of a middle-aged male with cystic adventitial disease whose diagnosis was complicated by concurrent features of popliteal artery entrapment syndrome. Treatment consisted of venous interposition grafting, which yielded excellent results. (*J Vasc Surg Cases and Innovative Techniques* 2020;6:75-9.)

Cystic adventitial disease (CAD) is a rare vascular disorder characterized by arterial occlusion owing to the development of a cystic mass in the outer (subadventitial) layer. The popliteal artery is most often involved, leading to symptoms of lower extremity claudication. Physical examination is frequently normal, although a popliteal bruit and/or weakened distal pulses with knee flexion may be observed. Diagnosis is supported by imaging, including ultrasound examination, computed tomography (CT) scans, magnetic resonance imaging (MRI), and angiography. Surgical intervention, percutaneous aspiration, and surveillance are among the possible treatment options. Here, we present a case of a 40-year-old man with CAD whose presentation and imaging findings mimicked that of the popliteal artery entrapment syndrome (PAES; [Table](#)). Patient consent for publication was obtained.

CASE REPORT

A 40-year-old Caucasian man with a history of tobacco, alcohol, and illicit drug use, and hepatitis C, presented to the emergency department owing to persistence of a dull ache in his upper left calf for the last month. The discomfort first became apparent after the completion of multihour physical labor that required the patient to be on flexed knees for the duration. Recurrence of the pain occurred with walking short

distances and was relieved with rest. He had no prior symptoms of claudication or associated trauma. There was no skin discoloration or swelling, and his femoral, popliteal, and posterior tibial pulses were palpable bilaterally on physical examination. The ankle-brachial index (ABI) was 1.28 (right) and 1.13 (left). Treadmill stress test was performed, provoking the onset of pain in his left calf with severe drop in the left ankle pressure of greater than 0.15 ([Fig 1](#)).

CT angiography demonstrated an approximate 70% stenosis of the supragenicular popliteal artery owing to extrinsic compression of the artery between the gastrocnemius musculature and what seemed to be a small Baker's cyst. MRI and MR angiography were obtained and showed similar findings as the CT angiography, with no obvious abnormal muscle insertion in relation to the neurovascular bundle and the small Baker's cyst. However, small size of the cyst, for which aspiration was not indicated after evaluation by orthopedic surgery, was inconsistent with continuing symptoms of claudication. An angiogram was then completed, which demonstrated popliteal artery compression with plantar flexion, possibly by the head of the gastrocnemius muscle ([Fig 2, A and B](#)).

Owing to disabling symptoms, surgical intervention was pursued. A preoperative diagnosis of PAES was presumed owing to examination and imaging findings. A posterior approach using a lazy S incision was performed for entry into the left popliteal fossa. Dissection occurred between the heads of the gastrocnemius muscle for exposure of the neurovascular bundle. There was no anatomic abnormality regarding the location of the popliteal artery in relation to the muscles and neurovascular bundle, arguing against PAES. However, the popliteal artery was found to be severely adhered to the vein owing to an inflammatory process ([Fig 3](#)). Further examination of the popliteal artery revealed severe cystic disease of the medial inferior portion of 5 cm in length with significant inflammation and compromise of the lumen. A gelatinous texture of the cystic component was observed ([Fig 4, A and B](#)). No cystic features were seen in relation to the vein. After obtaining proximal and distal control on healthy segments of the popliteal artery, a longitudinal arteriotomy was done over the affected portion. The diseased artery was resected and replaced with an interposition

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Table. Comparison of cystic adventitial disease (CAD) of the popliteal artery and popliteal artery entrapment syndrome (PAES)

	CAD of the popliteal artery	PAES
Pathology	Arterial occlusion owing to the development of a cystic mass in the subadventitial layer of the vessel.	Compression of the popliteal artery via congenital anatomic abnormalities of the artery or muscular hypertrophy (ie, gastrocnemius muscle).
Patient population	Young to middle-aged males; generally healthy, without typical risk factors for peripheral arterial disease.	Active males in second or third decade of life; patients also typically healthy without risk factors for arterial disease. ¹
Presentation	New-onset intermittent claudication of a lower extremity, with a recovery time longer than that seen in peripheral artery disease. ¹⁻⁶	Insidious onset of lower extremity claudication that is not alleviated immediately with rest; commonly bilateral. ^{1,7}
Examination findings	Passive flexion of the knee may elicit diminished distal pulses, termed the Ishikawa sign. ^{3,5} A popliteal bruit may also be present.	May see calf muscle hypertrophy; dorsiflexion and/or plantar flexion of the ankle may elicit diminished, unequal, or even absent pulses.
Characteristics of ABIs	The ABI may be normal in CAD, although a decrease can be observed during exercise. ⁵	Similar findings.
Imaging	Visualization of a thin, echogenic line separating the lumen of the vessel and the cyst, with the narrowed lumen presenting with an ultrasonic scimitar sign on color Doppler imaging and increased velocity. ^{3,5} CT scans and MRIs are often helpful modalities for better demarcation of anatomy. ^{2,4-6} Angiography is another potential imaging tool, with signs including narrowing of the lumen, complete occlusion, and lack of post-stenotic dilatation.	Angiography, CT angiography, Doppler ultrasound examination, MRI may be used to confirm the diagnosis; MRI is the preferred method to visualize the popliteal fossa, but the best method for diagnosis remains unclear. ¹ Advantage of arteriography, unlike CT angiography or MR angiography, is ability to visualize popliteal artery in real-time with use of provocative maneuvers.
Treatment options	Surgical intervention (interposition grafting, cyst excision, circumferential resection of the diseased adventitia), percutaneous drainage of the cyst, and conservative management.	Surgical decompression of the entrapping muscle has been the management of choice; surgery with arterial release allows for definitive reestablishment of normal anatomy and often portends excellent results.

ABI, Ankle-brachial index; *CT*, computed tomography; *MRI*, magnetic resonance imaging.

graft using an autologous reversed great saphenous vein. At the close of the case, posterior tibial pulses were present bilaterally.

Surgical pathology findings confirmed adventitial cystic disease of the popliteal artery, with serial vascular sections demonstrating a 0.9- × 0.5- × 0.4-cm cavitated space exuding a pale gray translucent mucoid-like material.

The postoperative course was uneventful, with no recurrence of symptoms or complications at 6 months. Follow-up duplex ultrasound examination at this time point demonstrated a patent graft (Fig 5). His ABIs were also noted to be normal.

DISCUSSION

First described by Atkins and Key in 1947, CAD is a rare disorder, comprising 0.1% of vascular diseases.²⁻⁵ It is

characterized by the presence of an adventitial cyst resulting in lower extremity arterial stenosis and subsequent ischemic symptoms. The cysts are composed of an accumulation of mucinous fluid formed by proteoglycans, mucoproteins, mucopolysaccharides, hyaluronic acid, and hydroxyproline.^{2,5,6,8} Ninety percent of cases of CAD are unilateral, with involvement of the popliteal artery.²⁻⁶ Less commonly, the external iliac, femoral, radial, ulnar, brachial, and axillary arteries are implicated.^{2,3,5} Rare reports exist of CAD affecting veins.³

Young to middle-aged generally healthy patients, particularly males, are mostly affected.³⁻⁶ The classic presentation consists of new-onset intermittent claudication of a lower extremity, with a recovery time longer than

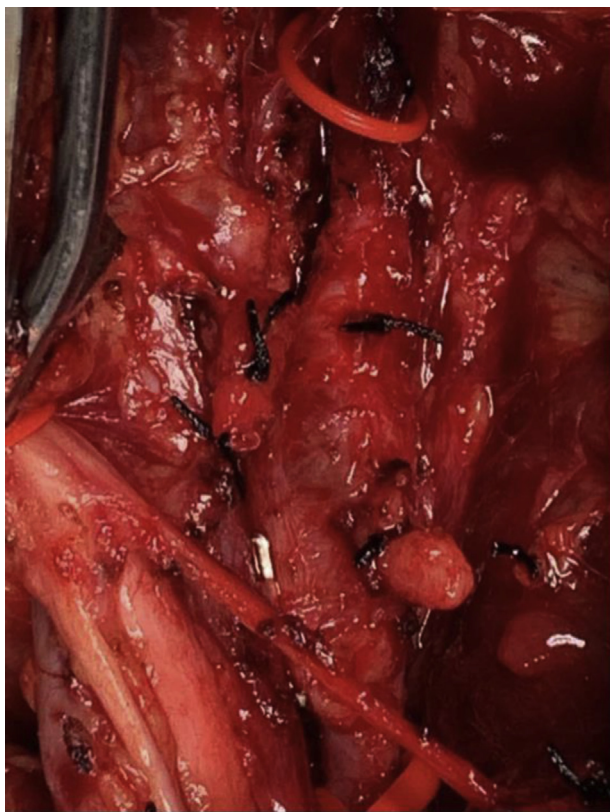


Fig 1. Severe adherence of left popliteal artery to popliteal vein. Exposure of the neurovascular bundle revealed substantial adhesion of the popliteal artery to the vein, requiring careful separation.

that seen in peripheral artery disease (approximately 20 minutes).¹⁻⁶ Recovery after exertion is associated with gradual opening of the artery secondary to decompression of the cyst.⁵ The neurovascular examination is usually normal. However, passive flexion of the knee may elicit diminished distal pulses.^{3,5} This differs from PAES, where maneuvers contracting the gastrocnemius muscle, such as active plantar flexion or passive dorsiflexion, diminish pedal pulses. Interestingly, the angiogram performed in the case of this patient showed compression of the popliteal artery with plantar flexion, supporting a diagnosis of PAES.^{1,7,9} The ABI may also be normal in CAD, although a decrease may be observed during exercise.⁵ Reduction of ABIs with exercise is also seen in PAES. Diminished left ankle pressure was observed in our patient with exertion, again leaving PAES as a potential diagnosis. Differential diagnoses of CAD should also include Baker's cyst.^{2,4,5} Similar to this case, a report exists of a 28-year-old man with CAD masquerading as a Baker's cyst.⁵ The individual had a 1- × 1-cm soft tissue

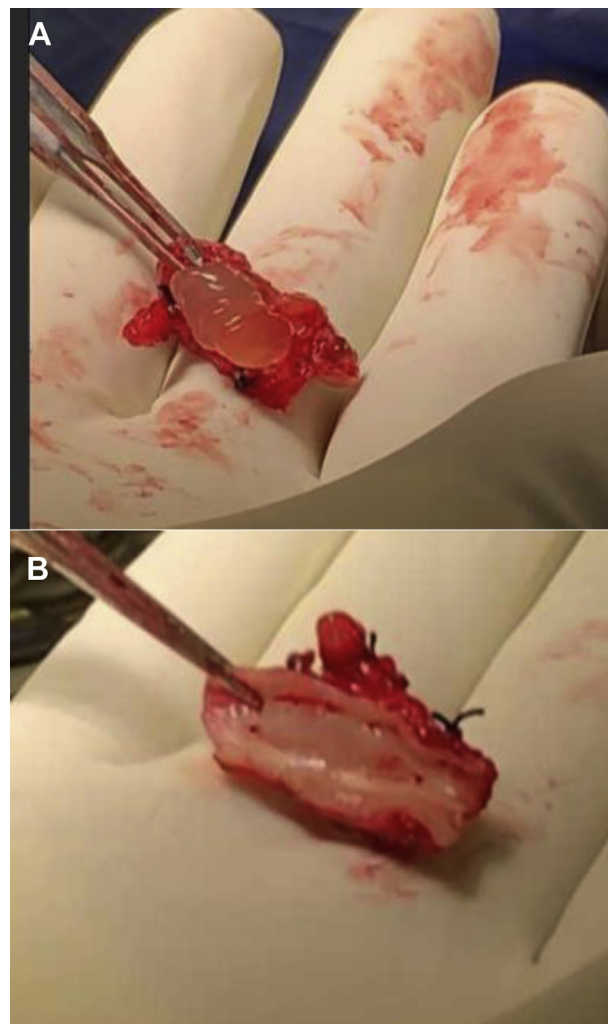


Fig 2. Observable cystic changes with compromise of the lumen. **A**, Dissection of a portion of the diseased popliteal artery exposed plainly visible cystic contents of the vessel wall, which perceptively narrowed the lumen. **B**, The gelatinous texture of the cystic component. The mucoid composition of the cyst is clearly discernable.

mass in the popliteal fossa accompanied by severe local pain with exertion and flexion of the knee.

Various hypotheses exist regarding the etiology of CAD, with no single theory universally accepted. Recurrent trauma of the popliteal artery as the cause of detachment of the adventitia from the media leading to cyst formation^{3,8} was discounted by the lack of associated trauma in the majority of cases of CAD.³ It was hypothesized that the presence of mesenchymal cells in the adventitia during embryogenesis can lead to cyst formation.^{3,5,10} Similarly, the ganglion hypothesis states that CAD may represent ectopic ganglia from adjacent joint

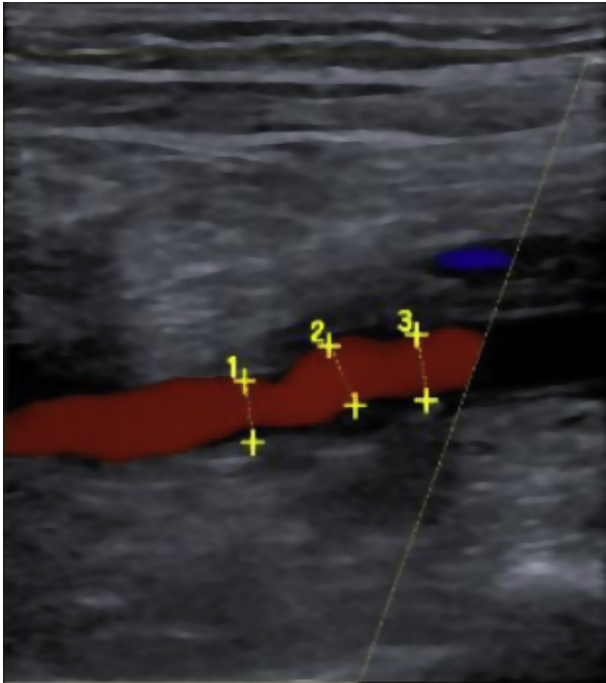
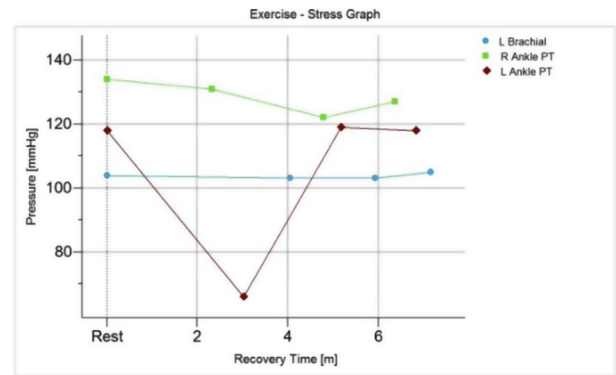


Fig 3. Duplex ultrasound image of venous graft at the 6-month follow-up. The duplex examination displayed patency of the short saphenous venous graft with unidirectional flow. Recurrence of symptoms was denied by the patient.

capsules, with incorporation of the cyst into the vessel at the time of development.^{3,5,9} Finally, the articular, or synovial, theory explains that damage to a joint capsule causes tracking of synovial fluid to the adjacent arterial vasculature, which can subsequently lead to the development of cysts.³

Duplex ultrasound examination is generally accepted as the first-line method for diagnosis.^{3,5,6} Characteristic findings include visualization of a thin, echogenic line separating the lumen of the vessel and the cyst, with the narrowed lumen.^{3,5} Differentiation from a popliteal aneurysm is based on a lack of vascular flow in the cyst and posterior acoustic enhancement.³ CT and MRI allow better demarcation of anatomy.^{2,4-6} MRI is particularly valuable for assistance with surgical planning, owing to its ability to define the cyst and its connection to adjacent joints. Angiography, if used alone, is less helpful, making differentiating the cause of arterial stenosis difficult.²⁻⁶

Treatment options for CAD include surgical intervention, percutaneous drainage of the cyst, and conservative



Exercise Duration: 10:00
 Exercise Type: Treadmill
 Onset of Symptoms: At 01:14, Left Calf, Tightness
 Onset of Symptoms: At 04:32, Left Calf, Pain
 Onset of Symptoms: At 09:39, Left Calf, Pain

Fig 4. Preoperative postexercise ankle-brachial index (ABI).

management. Surgical treatment is recommended owing to the associated low risk of recurrence.^{2-6,8,10} Surgical methods include interposition grafting of the diseased segment, which offers the best long-term patency and was done here, cyst excision, and circumferential resection of the diseased adventitia.^{2-6,8,10} Image-guided cyst aspiration, using ultrasound examination or CT scans, is associated with a higher risk of recurrence.^{5,9} However, reports exist of successful treatment of CAD with aspiration with good 5-year follow-up outcome.⁶ Conservative treatment includes observation for spontaneous resolution. Although the usual trend is for cysts to progress in size and increase the degree of stenosis, dissipation without intervention is possible.^{8,10} Several reports document regression of cysts within 5 weeks after the onset of symptoms, with longer durations reaching 10 and 15 months.¹⁰ Sustained surveillance is mandatory following regression owing to recurrence.¹⁰

CONCLUSIONS

CAD is a rare presentation of popliteal artery stenosis causing symptoms of lower extremity claudication. In the case here, diagnosis was complicated by a presentation encompassing features of both CAD and PAES. A cystic component of the disease was seen on MRI with additional findings suggestive of PAES. Therefore, as in such a scenario, physical examination and diagnostic testing may not always distinguish between CAD and PAES, and operative planning should include the potentiality of encountering either pathology. Surgical exploration owing to disabling symptoms was a successful

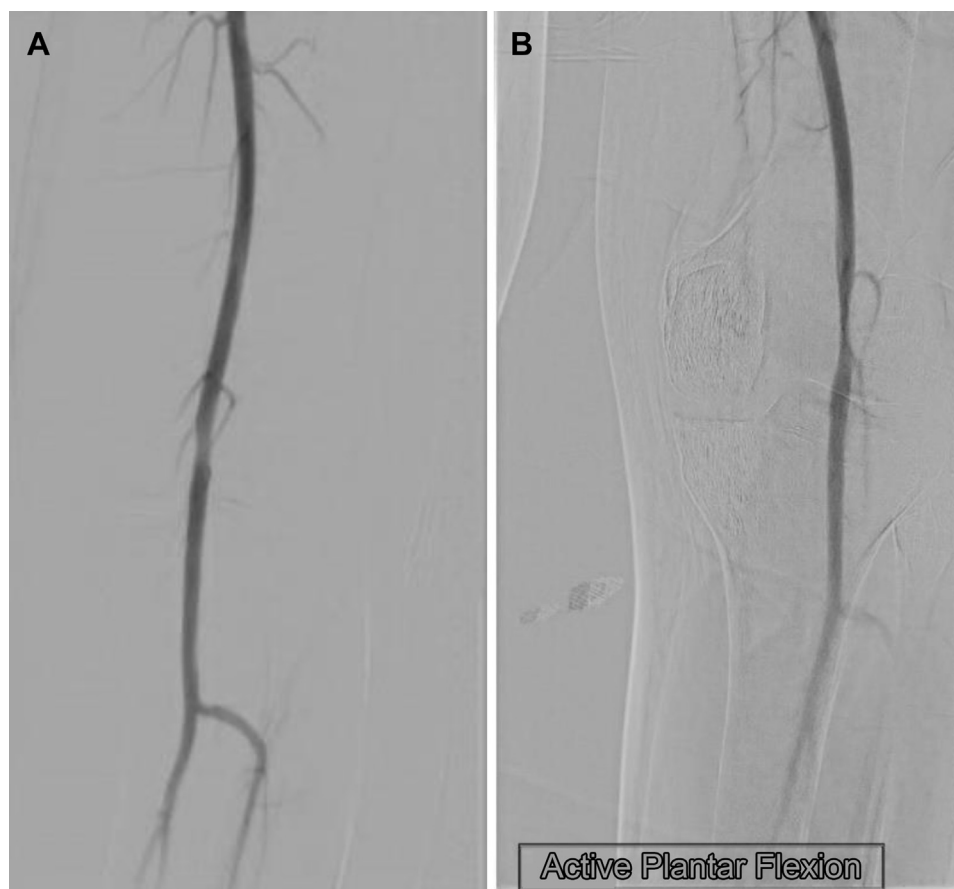


Fig 5. Angiogram of left popliteal artery. **A,** The angiogram at rest shows adequate blood flow through the popliteal artery with three-vessel runoff. **B,** With plantar flexion, blood flow is significantly reduced inferior to the knee via compression of the artery, a sign associated with popliteal artery entrapment syndrome (PAES).

strategy in this case, yielding an excellent long-term outcome.

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