

Cystic adventitial disease

Justin Winn, MD; Charan Singh, MD; Adam Cloud, MD; and Harlan Stock, MD, MBA

CASE SUMMARY

The patient was a healthy, 41-year-old male who presented with a 3-month history of left calf claudication. Physical examination revealed no mass within the popliteal fossa and normal arterial pulses within the femoral, popliteal, dorsalis pedis and posterior tibial arteries. There was no evidence of ischemia, and the ankle brachial index was normal. Color Doppler ultrasonography (US) of the lower extremity was performed, followed by computer tomography angiography (CTA) and magnetic resonance imaging (MRI) of the lower extremity. The patient underwent surgery as the severity of his symptoms interfered with daily activities. At surgery, the multilobulated cystic lesion was found to be incorporated within the adventitia of the popliteal artery. Evacuation was performed, and the popliteal artery was preserved.

IMAGING FINDINGS

Color Doppler ultrasound examination of the left popliteal artery revealed a well-defined, hypoechoic lesion with scant internal echoes and posterior acoustic enhancement abutting the popliteal artery. CTA of the lower extremity revealed a low-density lesion, intimately associated with and compressing the popliteal artery. MRI showed the lesion to be multilobulated with high signal on T2-weighted imaging, and low to intermediate signal on T1-weighted imaging.

DIAGNOSIS

Cystic adventitial disease. Differential diagnoses: Thrombosed aneurysm, Baker's cyst, ganglion cyst or perimeniscal cyst.

DISCUSSION

Cystic adventitial disease (CAD) is a rare vascular disorder. Atkins and Key first described its effect on the external iliac artery in 1947.¹ Since then, more than 350 cases have been reported.² Cystic adventitial disease is located within the popliteal artery 85-90% of the time, and is usually unilateral. The common presentation is a male patient in his 40s who presents with intermittent claudication without risk factors for atherosclerosis. The patient's age can range from 11-70 years, and the presenting symptom may be a palpable mass.³ The incidence of CAD among patients presenting with intermittent claudication is 1 in 1200 cases.³

The etiology of CAD is unknown, although four theories have been proposed. The first theory, proposed by Linquette in 1967, is of a mucinous or myxomatous systemic degenerative process. This theory has gained little traction, as follow-up studies have shown no systemic or contralateral manifestation of this disease. The second theory posits that repeated microtrauma causes degeneration of the arterial adventitia, resulting in cyst formation. However, CAD has been reported to occur in young children and deep vessels (which are presumably more protected) such as the external iliac

artery. A third theory is the ganglion theory, which proposes that ganglion cysts migrate into the arterial adventitia.^{4,5} A fourth theory, proposed by Levien and Benn, states that developing vessels aberrantly incorporate undifferentiated mucoid-secreting mesenchymal tissue which later form the cyst.⁶

Angiography has long been considered the gold standard. However, for diagnosis this modality can be insensitive and nonspecific unless characteristic findings such as the scimitar or hourglass sign is seen in an otherwise normal arteriography.^{5,7} The scimitar sign occurs when there is smooth eccentric vessel tapering, and an hourglass sign occurs with concentric luminal narrowing.^{5,7} Flexion of the knee during arteriography may help reveal the stenosis on angiography.⁷

Ultrasound may show a hypoechoic or anechoic mass arising from the arterial wall in the case of CAD. Internal echoes may correlate with gelatinous intracystic material. In addition, color Doppler ultrasound may be able to estimate the degree of stenosis. However, it may be difficult to differentiate CAD from a thrombosed popliteal aneurysm on ultrasound. A multilobulated lesion would be suggestive of CAD, whereas a thrombosed aneurysm is more likely unilocular.^{3,8}

Computed tomography (CT) or magnetic resonance imaging (MRI) is more informative in detecting CAD because these imaging modalities can better differentiate the anatomy and

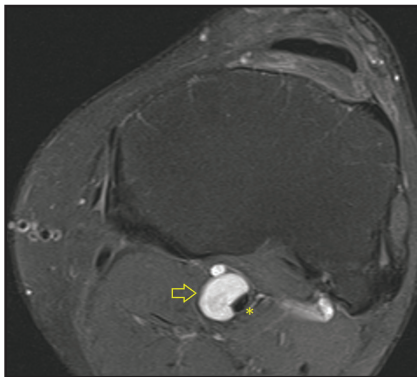


FIGURE 1. Axial proton density weighted image (TR 3050, TE 41) reveals a high signal lesion (arrow) compressing the low signal popliteal artery (asterisk).

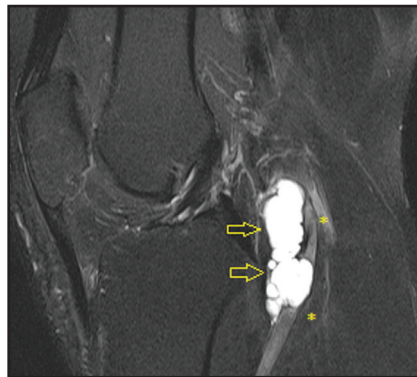


FIGURE 2. Sagittal T2-weighted image (TR 5650, TE 78) reveals a high signal multi-lobulated lesion (arrows) compressing the popliteal artery (asterisk).

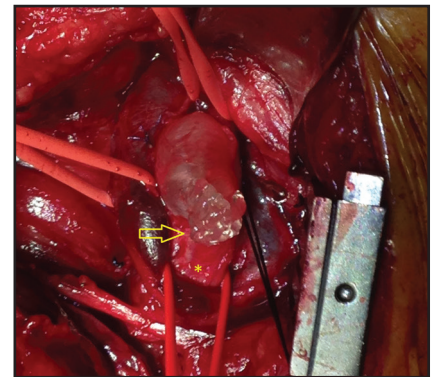


FIGURE 3. During surgery, gelatinous material was found encapsulated by the adventitial layer of the popliteal artery. The cyst material was evacuated, and the artery was preserved.

soft-tissue characteristics of the lesion. Cystic adventitial disease appears as a hypochoic, multi-lobulated lesion arising from the arterial wall on CT. Intravenous contrast would produce mural but not internal enhancement. On MRI, the CAD lesion usually demonstrates low signal on T1-weighted images and hyperintense signal on T2-weighted images. While there have been reports of communication of the CAD lesion with the adjacent knee joint, this finding is usually a surgical one as the connection is typically beyond image resolution.⁸

Differential considerations for CAD on MRI or CT imaging include a Baker's cyst, a ganglion cyst or a peri-meniscal cyst. Location is important. Baker's cysts are situated between the medial head of the gastrocnemius muscle and the insertion of the semimembranous muscle. Ganglion cysts occur along a tendon sheath, such as the semimembranosus tendon, or near the proximal tibiofibular joint. Peri-meniscal cysts are often adjacent to the menisci and seen in the setting of a meniscal tear.⁸ Most importantly, the above lesions rarely cause symptoms of claudication.

Treatment options for CAD include cyst excision with preservation of the native artery, excision of the affected artery with interposition of a bypass graft, CT or ultrasound guided needle aspiration and conservative management.

There have been no controlled studies evaluating the best treatment options for CAD, mostly due to its rarity. Needle aspiration, although relatively noninvasive, may be difficult to perform because of the viscosity of the cyst material; also, the recurrence rate after needle aspiration is 10%.² Spontaneous resolution has been reported in four CAD cases, with no recurrence at 10-year follow-up.⁹ However, if there is degeneration of the medial wall or complete occlusion, complete excision with interposition of a bypass vessel graft is recommended.²

CONCLUSION

CAD is a rare disease affecting mostly healthy male patients in their 40s and classically causes claudication. CT and MRI are the best imaging modalities because each provides pertinent anatomic delineation and soft tissue characterization. Typically a multilobulated cystic structure arising from the popliteal artery causing arterial stenosis is seen. Treatment options include aspiration, surgical excision of the cyst with preservation of the remaining artery, and excision of the affected arterial segment with bypass interposition. The latter is recommended in cases of complete arterial occlusion.

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Prepared by Dr. Winn while a Radiology Resident; Dr. Singh while an Assistant Professor of Radiology; Dr. Cloud while an Assistant Professor and Vascular Surgeon; and Dr. Stock while an Assistant Professor of Radiology and Chief of Musculoskeletal Imaging, at the University of Connecticut Health Center, Farmington, CT.