Adventitial cystic disease of the popliteal artery

Rachel Rendon, BS^{a,b} , Kristyn Mannoia, MD^c , and William Shutze, MD^{b,d}

^aUniversity of Central Florida College of Medicine, Orlando, Florida; ^bTexas Vascular Associates, Plano, Texas; ^cDepartment of Vascular Surgery, Baylor University Medical Center, Dallas, Texas; ^dDepartment of Vascular Surgery, The Heart Hospital Baylor Plano, Plano, Texas

ABSTRACT

Adventitial cystic disease (ACD) is a vascular disorder most commonly affecting the popliteal artery. ACD is an uncommon and often misdiagnosed cause of lower extremity intermittent claudication that is usually acute in onset and of longer duration than claudication associated with atherosclerosis. We present two cases of ACD affecting the popliteal artery.

KEYWORDS Adventitial cystic disease; arterial repair; claudication; nonatherosclerotic vascular disease; popliteal artery

Adventitial cystic disease (ACD) is a rare vascular disorder characterized by fluid accumulation in the adventitial layer of a vessel, which may cause luminal narrowing or even complete occlusion.^{1,2}

The cystic fluid has been described as gelatinous or mucoid in consistency, with a high content of hyaluronic acid and different combinations of mucopolysaccharides and mucoproteins.^{1,3} Symptoms of ACD depend on the location of the cyst, with the popliteal artery being the most common vessel affected.³ When the popliteal artery is affected, the most common presenting symptom is intermittent claudication that is usually acute in onset and of longer duration than claudication associated with atherosclerosis.^{1–3} We present two cases of cystic adventitial disease of the popliteal artery.

CASE REPORTS

Table 1 presents the clinical information for two patients who recently presented with ACD. Patient 1 was referred for the recent onset of symptoms and a presumed diagnosis of popliteal aneurysm. Ultrasound in our office confirmed a focal cystic mass without any Doppler flow present adjacent to the popliteal artery (*Figure 1a*). Patient 2 was an active runner who suddenly developed claudication symptoms after one block of walking. Computed tomography demonstrated cystic adventitial disease (*Figure 2*).

After their diagnostic workups were complete, both patients were taken to the operating room for exploration under adequate general anesthesia in the prone position. A standard S-shaped curvilinear incision was used in the

popliteal crease. The sciatic nerve and sural nerve were identified and carefully mobilized and protected.

In Patient 1, there was significant venous congestion with some dilated superficial veins. The popliteal vein was densely adherent to the mass, which was carefully dissected out. The mass was completely mobilized circumferentially. The patient was systemically heparinized, and the artery and vein were occluded proximally and distally. The mass was then incised, and some gelatinous material slowly leaked out, confirming that it was a popliteal adventitial cyst (*Figure 1b*). The entire cyst was then segmentally resected, and the artery was skeletonized of any cyst wall. We opened up the artery where the cyst was adherent to it. The artery was healthy both proximally and distally. The interior of the artery was normal. A bovine pericardial patch was cut to the appropriate length and sewn in with running 6.0 Prolene suture (*Figure 3*). Flow was restored. The patient had excellent pulses and a strong palpable posterior tibial pulse at the ankle. The patient's postoperative convalescence was remarkable only for immediate symptom relief, and he was discharged on postoperative day 2. The patient continues to be asymptomatic in follow-up with good pedal pulses after 15 months.

In Patient 2, the popliteal vein was identified and separated from surrounding tissue. The popliteal artery was then identified and dissected from proximal to distal. It was evident that just distal to the knee flexion, in the mid-popliteal space, there was circumferential cystic degeneration around the artery. This cystic structure was dissected off the adventitial surface of the popliteal artery for approximately 6 cm.

Corresponding author: Rachel Rendon, BS, Texas Vascular Associates, 4716 Alliance Blvd., Suite 200, Plano, TX 75093 (e-mail: rachel.rendon@knights.ucf.edu)

Color versions of one or more of the figures in this article can be found online at www.tandfonline.com/ubmc.

Received May 30, 2018; Revised April 18, 2019; Accepted April 22, 2019.

Table 1. Pertinent findings in the two patients

Variable	Patient 1	Patient 2
Age (years)	59	69
Sex	Male	Female
Hypertension	+	–
Hyperlipidemia	+	+
Other past medical and surgical history	Arm surgery, spinal fusion, knee surgery, shoulder surgery, and varicose vein surgery	Hysterectomy
ACD location	Right popliteal artery	Left popliteal artery
Symptoms	Painful popliteal mass	One block claudication
Popliteal pulse	+	+
Pedal pulses	+	+
Ishikawa sign	–	+
Diagnostic imaging	Ultrasound shows cystic mass on popliteal artery	CT shows ACD

ACD indicates adventitial cystic disease.

Figure 1. (a) Ultrasound confirms a focal cystic mass (yellow arrow) without any Doppler flow adjacent to the popliteal artery (green arrow). (b) Intraoperative image showing the proximal popliteal artery (yellow arrow) and the gelatinous material that oozed out of the cyst after incision (green arrow).

The predominant area of stenosis was in the mid-popliteal space, and the cyst in this area was excised in its entirety. Duplex ultrasound confirmed no residual stenosis in the popliteal artery. The wound was then closed. At 1-year follow-up, the patient was symptom free and walking up to 30 minutes at a time. She had palpable bilateral posterior tibial pulses and minimal edema.

DISCUSSION

ACD was first reported in an external iliac artery by Atkins and Key in 1947.¹ Since then, only around 500 cases have been reported, with most (>85%) involving the popliteal artery.^{1,3} However, ACD can also affect veins, albeit much less commonly.¹ Because it is rare, the prevalence of ACD is unknown, but it is estimated that ACD accounts for <0.1% of cases of intermittent claudication.¹ ACD tends to

affect patients with no to minimal cardiovascular risk factors, with a reported male-to-female ratio of approximately 5:1.^{2,4} The peak age of onset is between 40 and 50 years.¹

The most common presenting symptom for ACD affecting the popliteal artery is intermittent claudication of the lower extremity that is usually of acute onset and may progress rapidly.^{1,2} Of note, the claudication associated with ACD takes longer to resolve, which has been thought to be due to an increase in pressure during exercise causing increased fluid secretion and subsequent slow resorption of the secretions.^{1,2} Other presenting symptoms of popliteal artery ACD include burning pain and paresthesia of the affected lower extremity, pain behind the knee, acute calf pain, cyst rupture (rarely), ischemic rest pain, and tissue loss.^{1–3} Among conditions on the differential diagnosis that affect the popliteal artery and may also present with intermittent claudication are popliteal artery entrapment syndrome and popliteal artery aneurysm.⁴

Figure 2. A representative sagittal computed tomography slice of the popliteal fossa showing the proximal popliteal artery (yellow arrow), distal popliteal artery (blue arrow), and cystic structure surrounding the popliteal artery (green arrow).

Figure 3. (a) A normal interior popliteal artery opened after cyst excision. (b) The popliteal artery after closure with a bovine pericardial patch.

Evaluation of ACD is multifaceted. Clinically, patients may present with the Ishikawa sign, which is cessation of a distal lower extremity pulse on the affected side of the body upon hyperflexion of the knee due to occlusion of the popliteal artery during the maneuver.^{1,2} Exercise testing with an ankle-brachial index may be performed to assess for ACD, because it may demonstrate a drop in pressure.² Imaging can be a very helpful tool in diagnosing ACD, with magnetic resonance imaging or magnetic resonance angiography being the generally agreed modalities of choice. They will reveal

the presence of a cystic lesion, the length of the affected segment of the popliteal artery, and the degree of occlusion caused by the cystic lesion.^{1,4,5} These imaging modalities will show either partial or full occlusion of the popliteal artery by a multilocular cystic mass within the arterial wall in an otherwise normal arterial tree.^{2,4} The pathognomonic scimitar sign will appear as a curvilinear smooth narrowing of the vessel in instances of eccentric stenosis, and the arterial stenosis will appear hourglass-shaped in cases of concentric stenosis.^{2,4} Computed tomography findings will show a popliteal artery being compressed by a mass with the same density as fluid but will not show the true multilocular nature of the cyst due to the lower tissue resolution when compared with magnetic resonance imaging.⁴ Ultrasound can also be helpful in diagnosing ACD and will show a well-defined anechoic or hypoechoic mass, depending on the mucin content of the cyst.^{2,4} Doppler ultrasound will also reveal the degree of stenosis caused by the cyst compressing the popliteal artery by showing increased peak systolic velocities.⁴

The standard treatment of ACD of the popliteal artery remains resection of the cyst followed by appropriate arterial repair or reconstruction.^{1,2} However, multiple other modalities have been attempted. Both aspiration of the cyst and balloon angioplasty uniformly fail. Aspiration fails due to recurrence of the cyst. Angioplasty fails due to residual cyst compression of the artery. Stenting of the popliteal artery can overcome cyst compression, but this is not compatible with long-term patency due to the repetitive flexion of the knee in younger, more active patients.¹

ACKNOWLEDGMENT

The authors acknowledge Kathleen Richter for her expertise and assistance in preparing this article.

ORCID

Rachel Rendon <http://orcid.org/0000-0001-9823-9076>

Kristyn Mannoia <http://orcid.org/0000-0002-1630-1874>

William Shutze <https://orcid.org/0000-0002-1627-5434>

1. Del Canto Peruera P, Vazquez MJ, Velasco MB, et al. Cystic adventitial disease of the popliteal artery: two case reports and a review of the literature. *Vascular*. 2015;23:204–210.
2. Paravastu SC, Regi JM, Turner DR, Gaines PA. A contemporary review of cystic adventitial disease. *Vasc Endovascular Surg*. 2012;46:5–14. doi:10.1177/1538574411419377
3. Motaganahalli RL, Smeds MR, Harlander-Locke MP, et al. A multi-institutional experience in adventitial cystic disease. *J Vasc Surg*. 2017; 65:157–161. doi:10.1016/j.jvs.2016.08.079
4. Jaraya M, Simmons S, Farber A, Teytelboym O, Naggara N, Guermazi A. Uncommon diseases of the popliteal artery: a pictorial review. *Insights Imaging*. 2016;7:679–688. doi:10.1007/s13244-016-0513-6.
5. Gwon JG, Kwon YJ, Han YJ, Cho YP, Kwon TW. Chronic nonatherosclerotic occlusive popliteal artery disease. *Ann Vasc Surg*. 2018; 47:128–133. doi:10.1016/j.avsg.2017.07.037