CASE REPORT

A young male runner with a posterior knee mass—not just your typical Baker's cyst

Ayesha Abid,^{1,2} James Fitzhugh Kelley,¹ Donald J Flemming,² Matthew L Silvis^{1,3}

¹Department of Family and Community Medicine, Penn State Milton S Hershey Medical Center, Hershey, Pennsylvania, USA

²Department of Radiology, Penn State Milton S Hershey Medical Center, Hershey, Pennsylvania, USA

³Department of Orthopedics and Rehabilitation, Penn State Milton S Hershey Medical Center, Hershey, Pennsylvania, USA

Correspondence to
Dr Ayesha Abid,
aabid@hmc.psu.edu

Accepted 12 February 2016

SUMMARY

We present the case of a 28-year-old man with a growing mass in his right popliteal fossa causing pain on exertion. The differential diagnosis included Baker's cyst, entrapment syndrome of the popliteal artery, as well as a benign or malignant neoplasm. An ultrasound was non-specific. Follow-up MRI of the knee demonstrated cystic adventitial disease (CAD). With only about 500 cases reported in the literature since its discovery in 1947, CAD is a rare entity. The disease is characterised by mucinous or gelatinous cysts in the arterial or venous adventitia. The disease is predominantly seen in the popliteal artery and typically affects otherwise healthy males in the fourth to fifth decade of life. It presents clinically as intermittent exertional claudication. Examination of our case and a review of the literature will highlight the importance of considering CAD in patients who report of a popliteal mass and intermittent claudication.

BACKGROUND

Cystic adventitial disease (CAD) was first described by Atkins and Key,¹ and accounts for 0.1% of vascular diseases. Of the documented cases, 85% affect the popliteal artery and its branches.² The disease is caused by an accumulation of mucinous fluid in the vessel adventitia, forming cysts. The cysts are composed of mucoproteins, mucopolysaccharides, hyaluronic acid and hydroxyproline that slowly grow in size.³ The presenting symptom, intermittent exertional claudication, carries a broad differential. Diagnosis can be challenging as these patients often have a normal physical examination.⁴ Furthermore, orthopaedic surgeons and primary care practitioners who may not be familiar with the disease process commonly see young athletes. When misdiagnosed, the disease can result in severe vessel stenosis and occlusion over time.^{5 6} Timely diagnosis can help prevent morbidity and allows for early surgical referral, if necessary.

CASE PRESENTATION

A 28-year-old man presented to an outpatient clinic for evaluation of a popliteal fossa mass and a 6-month history of intermittent pain with right knee flexion. The patient had swelling and a soft tissue mass in his popliteal fossa that had been increasing in size over the course of the past 2–3 months. He reported of severe pain localised to the popliteal fossa on exertion. He reported hurting his knee cap several months after the onset of pain but denied any prior trauma. He had neither numbness nor tingling. His symptoms were exacerbated by running and flexing his knee.

Symptoms resolved after 5 min of rest and responded well to ibuprofen.

He was an otherwise healthy male athlete, 172 cm tall and weighing 104 kg, who participated in aerobic and strength training approximately five times a week. His only concerning history was tobacco use of 10 cigarettes per day for approximately 8 years. He had no personal or familial history of cardiac disease, hypertension, diabetes, coronary artery disease or thyroid disease. He claimed a possible elevation in his lipid profile on routine blood analysis 1 year prior to presentation.

Examination of the right knee and leg revealed no significant deformity and no joint abnormality. There was neither oedema nor erythema of the overlying skin. The knee capsule was free of ecchymosis, effusion and dimpling. No tenderness was present over the bony structures. There was a 1 cm×1 cm mass towards the medial aspect of the popliteal fossa, inferior to the biceps femoris. On palpation, the mass was tender, round and hard, and easily moved with finger manipulation. There was full active and passive range of motion of the knee, with only mild calf discomfort with full passive knee flexion. The patient had a negative anterior and posterior drawer test, and there was no laxity and no pain with varus or valgus stress at 0° and 30°. McMurray and Thessaly tests were also negative. Knee flexor and extensor strength was 5/5 bilaterally. Sensation was intact. Pulses were 2+ bilaterally in the dorsalis pedis and posterior tibialis. The popliteal pulse in the right lower extremity was difficult to appreciate in part due to the overlying mass. The popliteal pulse was 2+ in the left lower extremity.

INVESTIGATIONS

Ultrasound imaging of the popliteal fossa showed a non-specific 1 cm×1 cm hypoechoic cyst posterior to the popliteal artery, unrelated to the neurovascular bundle (figure 1). This was not felt to be associated with the patient's symptoms. MRI showed a well-circumscribed high-intensity lesion representing a multiloculated cystic mass associated with the popliteal artery (figures 2 and 3). A fluid signal mass with a 'string of pearls'-like appearance was visible, tracking along the popliteal artery and multiple branch vessels (figure 4). There was no significant major vessel stenosis and the popliteal bursa was not distended. The findings were consistent with a diagnosis of CAD of the popliteal artery.

DIFFERENTIAL DIAGNOSIS

The typical nature of the pain is exertional. This overlaps with the clinical picture of other diseases

CrossMark

To cite: Abid A, Kelley JF, Flemming DJ, et al. *BMJ Case Rep* Published online: [please include Day Month Year] doi:10.1136/bcr-2015-213750

Figure 1 Transverse power Doppler image through the popliteal fossa shows hypoechoic cyst (+ markers) posterior to the popliteal artery (arrow). The association between the cyst and the artery was not appreciated.

such as: Baker's cyst, gradual onset musculoskeletal injuries, chronic exertional compartment syndrome, vascular insufficiency, popliteal artery entrapment syndrome, popliteal artery aneurysm, fibromuscular dysplasia and neuropathies. Of the causes of intermittent claudication, CAD is estimated to be responsible for 1 in 1200.⁷

OUTCOME AND FOLLOW-UP

The patient was presented with risks and benefits of surgical intervention versus conservative management. He declined

surgical intervention and chose observation. At 8-month follow-up, he reported no improvement in symptoms. A follow-up appointment was scheduled to discuss referral to a vascular surgeon.

DISCUSSION

CAD affects otherwise healthy individuals with no history of diabetes, hypertension or coronary artery disease. A case review found 105 reported cases, with a range of 11–70 years of age.⁸ The disease has been documented in children, athletes⁹ and the elderly. A recent review of CAD¹⁰ analysed over 500 cases and described each case by patient demographic, imaging modality, surgical management and recurrence incidence. The average age was 46 years, with 83% of cases affecting men. Five hundred and eighty-seven cases (80.5%) occurred in the popliteal artery, 34 cases in the femoral vein and 28 cases in the common femoral artery. As in this case, CAD often presents with symptoms of calf claudication and pain, which is the primary symptom in 88% of the reported cases.¹⁰ Symptoms fluctuate in intensity^{6 11} and are often related to prolonged postexercise recovery time of 10–20 min.⁵ This is secondary to increased pressure on the cyst during exercise and a gradual opening of the artery secondary to decompression of the cyst postexercise.^{5 12} Decompression of the cyst allows for relief of pain. Neurovascular examination is usually normal.⁵ In rare instances, passive flexion of the knee can cause a diminished distal pulse, known as the Ishikawa sign.¹³ The ankle-brachial index is typically normal but may decrease with exercise.^{9 11}

While still unclear, there are four proposed theories for the aetiology of CAD. One theory proposes a connective tissue disorder intrinsic to the vessel wall.¹⁴ The 'ganglionic theory' promotes the idea that the adventitial cysts are ectopic ganglions that migrated from vascular branches to an adjacent tendon or joint capsule.¹⁵ The microtraumatic theory suggests recurrent trauma that results in separation of the adventitia from the media. This results in bleeding and, eventually, cyst generation.¹⁴ The most widely accepted is the developmental

Figure 2 Axial turbo spin echo proton-weighted sequence with fat suppression through the knee above the joint line shows a multiloculated cystic mass (red arrows) posterior and medial to the popliteal artery (white arrow).

Figure 3 Coronal turbo spin echo proton-weighted sequence with fat suppression through the popliteal fossa shows that the mass (red arrows) is intimately associated with the popliteal artery (white arrow).

Figure 4 Sagittal turbo spin echo T2-weighted image with fat suppression through the popliteal fossa shows that the mass is adjacent to the popliteal artery. Note the 'string of pearls'-like appearance of extension along branch vessels of the posterior knee (red arrows).

hypothesis.¹⁶ In this theory, an embryological error occurs, where undifferentiated mesenchymal cells—targeted to form the knee joint—instead collect in the adventitia of blood vessels, where they secrete mucin, forming cysts.

An appropriate work up should include adequate history, physical examination and imaging.¹¹ Ultrasound imaging offers a good first approach as it is inexpensive and non-invasive.⁴ However, ultrasound is often unremarkable. A colour Doppler can alternatively be used as it can illustrate narrowing of the vessel due to compression by the cystic mass. This appears as reduced or absent blood flow.^{17 18} Better visualisation of anatomy can be achieved with MRI and CT. While CT and ultrasound may be useful in evaluating suspected patients, MRI is preferred due to its ability to define the cyst and its connections to adjacent joints, all of which assist surgical planning.^{10 11} On T2-weighted imaging, a CAD cyst is bright due to the high volume of protein in the cyst. On T1 imaging, cystic lesions have a low-intensity signal. On imaging, CAD consists of multiple, loculated cysts, as demonstrated in figures 1 and 2.¹⁹ It can also aid in differentiating other causes of pathology, including soft tissue tumours.²⁰ MR angiography (MRA) is helpful as it shows sluggish flow in the pathological artery. MRA can also demonstrate networks between the cyst and surrounding joint capsule. If present, involvement of the genicular arteries can also be visualised with MRA.²¹ CT angiography is useful but not frequently utilised due to radiation exposure and low anatomic yield.²² Digital subtraction angiography is not recommended as it is invasive and only diagnostic in a few cases where stenosis is present in a specific conformation.³ A compartment pressure may be obtained and is useful in differentiating CAD from entrapment syndrome of the popliteal artery, which is the cause of approximately 60% of calf claudication in athletes. Of these diagnostic modalities, the current evidence suggests an algorithm of duplex ultrasound, followed by MRA.¹¹

Treatment consists of conservative management and surgical intervention. Conservative management has shown spontaneous

cyst resolution but long-term follow-up for these individuals is unknown.^{10 23} Individuals with symptomatic and non-resolving CAD may choose surgical intervention. For these individuals, there are two approaches, as described by Veraldi *et al.*¹⁵ The first is resection, which involves removal of the vessel and replacement with a graft. This is typically reserved for cases of complete occlusion or involvement of the media where degeneration is present. Non-resection approaches include open cyst evacuation, intraoperative cyst aspiration and radiological interventions such as CT-guided aspiration, and percutaneous ultrasound-guided and percutaneous transluminal angioplasty.⁶ In a study of 724 patients with CAD, Desy and Spinner¹⁰ found 28% required cyst resection and saphenous vein reconstruction; the most commonly used treatment modality. Cyst recurrence occurred in 9% of these cases and was managed by a second surgical intervention.¹⁰

There is a scant amount of data regarding surgical intervention and return to-play times for athletes. Cyst aspiration provides a less-invasive approach, but is coupled with a high recurrence rate.^{2 4 6 10} This option is often unsuccessful as the highly viscous components of the cyst make it difficult to aspirate.^{6 12 14} In general, risks of surgery include recurrence of the cyst even in the grafted vessel,^{6 10} vessel damage, aneurysmal dilation⁶ and anaesthesia risks. These risks and possible prolonged recovery time may make surgical intervention an unattractive approach for an eager athlete. Nevertheless, long-term follow-up is necessary in patients with CAD to ensure that recurrence or graft occlusion does not occur.⁴

Patient's perspective

The pain actually first started about 7 years ago but I did not pay attention because it didn't bother me. It was hurting a lot more for about 6 months before I came into the office. The mass was there for about 6 months or so. I did actually hit my knee several months after the mass appeared. The mass really swelled after that. I started getting more pain. Usually, when I get the pain, it's when I am walking for 2–3 hours, doing exercise or sitting for a long time. I personally thought it was strain or overexertion. And then I thought I should probably get this checked out.

Learning points

- ▶ Although rare, a cystic mass and claudication should raise suspicion of cystic adventitial disease.
- ▶ The disease has a predilection for males and typically presents as intermittent exertional claudication with waxing and waning of symptoms.
- ▶ Misdiagnosis may result in vessel stenosis or occlusion over time.
- ▶ Duplex ultrasound and MR angiography are the preferred imaging studies for diagnosis.
- ▶ Definitive management requires surgical intervention.

Acknowledgements The authors would like to thank the Penn State Department of Family and Community Medicine for its ongoing support of resident research.

Competing interests None declared.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

REFERENCES

- Atkins HJ, Key JA. A case of myxomatous tumour arising in the adventitia of the left external iliac artery: case report. *Br J Surg* 1947;34:426–7.
- Kim SH, Lee CE, Park HO, *et al.* Adventitial cystic disease of the common femoral artery: a case report and literature review. *Korean J Thorac Cardiovas Surg* 2013;46:150–2.
- Peterson JJ, Kransdorf MJ, Bancroft LW, *et al.* Imaging characteristics of cystic adventitial disease of the peripheral arteries: presentation as soft-tissue mass. *AJR Am J Roentgenol* 2003;180:621–5.
- Kawarai S, Fukasawa M, Kawahara Y. Adventitial cystic disease of the popliteal artery. *Ann Vasc Dis* 2012;5:190–3.
- Cassar K, Engeset J. Cystic adventitial disease: a trap for the unwary. *Eur J Vasc Endovasc Surg* 2005;29:93–6.
- Drac P, Kocher M, Utikal P, *et al.* Cystic adventitial disease of the popliteal artery: report on three cases and review of the literature. *Biomed Pap Med Fac Univ Palacky Olomouc Czech Repub* 2011;155:309–21.
- Lewis GJ, Douglas DM, Reid W, *et al.* Cystic adventitial disease of the popliteal artery. *BMJ* 1967;3:411–15.
- Flanigan DP, Burnham SJ, Goodreau JJ, *et al.* Summary of cases of adventitial cystic disease of the popliteal artery. *Ann Surg* 1979;189:165–75.
- Sharma P, Padhiar N, Kyriakides C. Popliteal cystic adventitial disease causing intermittent claudication in a young athlete: a case report. *South Med J* 2008;101:1154–6.
- Desy NM, Spinner RJ. The etiology and management of cystic adventitial disease. *J Vasc Surg* 2014;60:235–45.
- Franca M, Pinto J, Machado R, *et al.* Case 157: bilateral adventitial cystic disease of the popliteal artery. *Radiology* 2010;255:655–60.
- Maged IM, Turba UC, Housseini AM, *et al.* High spatial resolution magnetic resonance imaging of cystic adventitial disease of the popliteal artery. *J Vasc Surg* 2010;52:534–5.
- van Rutte PW, Rouwet EV, Belgers EHJ, *et al.* In treatment of popliteal artery cystic adventitial disease, primary bypass graft not always choice: two case reports and a review of the literature. *Eur J Vasc Endovasc Surg* 2011;42:347–54.
- Tsilimparis N, Hanack U, Yousefi S, *et al.* Cystic adventitial disease of the popliteal artery: an argument for the developmental theory. *J Vasc Surg* 2007;45:1249–52.
- Veraldi GF, Scudo G, Scorsone L, *et al.* Cystic adventitial disease of the popliteal artery: report of two cases and review of literature. *G Chir* 2014;35:229–34. <http://www.ncbi.nlm.nih.gov/pmc/articles/PMC4321496/>
- Levien LJ, Benn CA. Adventitial cystic disease: a unifying hypothesis. *J Vasc Surg* 1998;28:193–205.
- Zhang H, Zhang Y, Wang Q, *et al.* Cystic adventitial disease of the popliteal artery: report of two cases. *Surg Today* 2014;44:1760–3.
- Taurino M, Rizzo L, Stella N, *et al.* Doppler ultrasonography and exercise testing in diagnosing a popliteal artery adventitial cyst. *Cardiovasc Ultrasound* 2009;7:23.
- Wright LB, Matchet WJ, Cruz CP, *et al.* Popliteal artery disease: management and treatment. *Radiographics* 2004;24:467–79.
- Tomasian A, Lai C, Finn JP, *et al.* Cystic adventitial disease of the popliteal artery: features on 3 T cardiovascular magnetic resonance. *J Cardiovasc Magn Reson* 2008;10:38.
- Ypsilantis EA, Tisi PV. Involvement of the genicular branches in cystic adventitial disease of the popliteal artery as a possible marker of unfavorable early clinical outcome: a case report. *J Med Case Rep* 2010;4:91.
- Ortiz M WR, Lopera JE, Giménex CR, *et al.* Bilateral adventitial cystic disease of the popliteal artery: a case report. *Cardiovasc Intervent Radiol* 2006;29:306–10.
- Pursell R, Torrie EP, Gibson M, *et al.* Spontaneous and permanent resolution of cystic adventitial disease of the popliteal artery. *J R Soc Med* 2004;97:77–8.

Copyright 2016 BMJ Publishing Group. All rights reserved. For permission to reuse any of this content visit <http://group.bmj.com/group/rights-licensing/permissions>.

BMJ Case Report Fellows may re-use this article for personal use and teaching without any further permission.

Become a Fellow of BMJ Case Reports today and you can:

- Submit as many cases as you like
- Enjoy fast sympathetic peer review and rapid publication of accepted articles
- Access all the published articles
- Re-use any of the published material for personal use and teaching without further permission

For information on Institutional Fellowships contact consortiasales@bmjgroup.com

Visit casereports.bmj.com for more articles like this and to become a Fellow