Popliteal cystic adventitial disease: a rare case of calf claudication in a young man

A J Collingwood, D J Ablett

Abstract

Popliteal cystic adventitial disease is a rare arterial condition characterised by mucinous cyst formation in the adventitia of the popliteal artery producing stenosis or occlusion of the arterial lumen. It predominantly affects males and typically presents in the thirties with disabling intermittent calf claudication. Because of the demographic it affects it is of relevance to military medical practitioners. We present such a case and describe the presentation, clinical features, differential diagnoses and imaging findings as well as the theories surrounding pathogenesis, clinical management and surgical treatment options.

Introduction

Popliteal cystic adventitial disease is defined by mucinous cyst formation within the adventitia of the popliteal artery causing stenosis or occlusion of the vessel resulting in calf claudication. It is a rare condition which accounts for 0.08% of presentations of calf claudication. Cystic adventitial disease (CAD) was first reported by Atkins and Key in 1947 affecting the external iliac artery. The first case affecting the popliteal artery was described in 1954 and the first series was published in 1957 with the condition described as "cystic adventitial degeneration of the popliteal artery". Approximately 85% of CAD cases involve the popliteal artery; it has, however, been documented as affecting not only arteries but also veins throughout the body. The vessels affected are typically adjacent to a joint space. Popliteal CAD (PCAD) affects males more commonly than females, the ratio being 5:1, and patients typically present in their thirties with intermittent claudication of insidious or sudden onset. PCAD normally affects the popliteal artery unilaterally, but cases of bilateral involvement have been reported. The nature of the symptoms of PCAD and the demographic it affects makes it a condition relevant to the military medical community.

Case presentation

A 36-year old male was referred with intermittent claudication symptoms affecting the right calf. These had been present for three months and were progressive, limiting the patient to 100 yards of walking. The patient also reported paraesthesia at the foot associated with the claudication symptoms, but no history of rest pain or ulceration. There was no history of trauma. The patient had undergone vasectomy four months previously; this was complicated by post-operative haematoma and infection, requiring surgical drainage and a seven-day inpatient hospital stay. There was no other significant past medical history. The patient was of normal appearance and build and was on no regular medicines, had no family history of arterial disease and smoked 20 cigarettes per day. Prior to developing intermittent claudication he had jogged regularly.

On examination the affected right lower limb had a normal femoral pulse, a weak popliteal pulse, monophasic Doppler signals at the foot and a reduced ankle brachial pressure index (ABPI) of 0.5 (normal range 0.9-1.2). The asymptomatic left lower limb had easily palpable pulses and a normal ABPI. The patient was advised to stop smoking and was prescribed clopidogrel 75mg once daily. An outpatient computed tomography angiogram (CTA) was organised.

The CTA demonstrated typical appearances of PCAD with near-occlusion of the above-knee popliteal artery and evidence of arterial collateralisation proximal to the lesion (Figure 1). The patient was subsequently seen in clinic. He had stopped smoking and reported that whilst the intermittent claudication symptoms had improved slightly, he was still very limited in his walking distance, with associated severe restriction in quality of life. The case was discussed at the vascular surgery multi-disciplinary team meeting and the patient was subsequently listed for surgery.

Via a posterior surgical approach, the affected 8cm segment of popliteal artery was resected and an interposition bypass performed using ipsilateral reversed great saphenous vein, harvested through the same incision (Figures 2 & 3). The popliteal vein and soft tissue structures were densely adherent to the affected segment of popliteal artery making dissection time-consuming. The operation was performed without complication and the patient was discharged after four days.

Histological assessment confirmed the diagnosis of PCAD. Surveillance Duplex performed at 6 weeks demonstrated triphasic flow within the bypass graft and run-off vessels. At follow up, the patient had a full complement of peripheral pulses, reported no claudication symptoms and had returned
to jogging. The wound had healed satisfactorily (Figure 4). Clopidogrel was discontinued and the patient was discharged.

**Aetiology**

The exact pathogenesis of PCAD is poorly understood and is a source of controversy. Four theories exist as to how PCAD arises.

**Tissue disorder**

PCAD was initially thought to be a consequence of a systemic connective tissue disorder. A lack of evidence of systemic conditions in PCAD patients with long term follow-up and absence of cystic disease in other arteries in patients suffering with PCAD has resulted in a lack of support for this theory.6,7

**Figure 1.** CT angiogram showing PCAD at the right popliteal artery. (A) in lateral view, (B) in posterior view and (C) axial view comparing right and left (normal).

**Figure 2.** Intra-operative photo of the popliteal fossa following dissection showing the segment of popliteal artery with PCAD and adjacent structures.

**Figure 3.** Intra-operative photo showing interposition, reversed great saphenous vein bypass following resection of the 8cm segment of popliteal artery with adventitial cyst.

**Figure 4.** The wound at the right popliteal fossa three months post operation.
Claudication that is notably slower to resolve than claudication due to atherosclerotic peripheral vascular disease. There is rarely a mass felt in the popliteal fossa, and popliteal and pedal pulses may or may not be present. Patients with PCAD may elicit a positive Ishikawa sign with disappearance of the pedal pulses following flexion of the knee. This helps to differentiate PCAD from popliteal entrapment syndrome, where the pulse is present at rest but disappears with contraction of the gastrocnemius during active plantar flexion or passive dorsiflexion of the foot.

Ultrasound is an effective first line investigation method of identifying PCAD. A hypoechoic mass can be seen within the arterial wall if PCAD is present; a multi-lobular lesion is indicative of PCAD, and a uni-lobular lesion could indicate a thrombosed popliteal aneurysm. Colour Doppler may be used to assess the degree of stenosis in the artery.

Angiography was once considered the gold standard, but it lacks sensitivity and specificity owing to its inability to visualise the soft tissue surrounding the vessel lumen. The so-called scimitar sign is pathognomonic of PCAD and occurs when the lesion is large and eccentric, displacing the artery to one side. If the cyst is concentric the stenosis will have an hourglass appearance. Flexion of the knee during imaging aids in eliciting these radiological findings. Computed tomography (CT) or magnetic resonance imaging (MRI) are regarded as more informative in detecting PCAD, as they differentiate the anatomy and soft-tissue characteristics of the vessel more effectively than other modalities. The cystic appearance of PCAD may be confused with other lesions occurring within the popliteal fossa, such as a Baker’s cyst, a ganglion cyst or a peri-meniscal cyst; these conditions rarely cause claudication and occupy subtly different locations within the popliteal fossa.

Clinical management

In a Role 1 environment the patient will probably present with activity-limiting calf pain. The management of the patient is dependent upon the severity of the claudication, the presence of ischaemia and the hostility of the environment. Initial management will require exclusion of more likely conditions. PCAD is a non-emergency and requires a non-urgent evacuation unless acute ischaemia is present. Urgent evacuation should also be considered in a hostile operational environment, as the patient’s lack of exercise capacity could jeopardise their safety.

Several treatment options exist for PCAD, including ultrasound-guided cyst drainage, open cyst excision and interposition or bypass graft, with or without cyst resection. Endovascular intervention (angioplasty +/- stent) has been found to be unsuccessful and has been largely abandoned as a treatment option. Ultrasound guided drainage of the cyst provides a minimally invasive option to manage symptoms over the short-term, but there is a high recurrence of symptoms owing to the cyst refilling with mucoid material and re-occluding the artery; consequently re-intervention is
often required. Open cyst excision can provide long-term symptomatic relief; however, recurrence of symptoms is also common following excision, as a portion of cyst may often be left behind. The management strategy providing the longest symptom-free period and the lowest rates of re-intervention has been shown to be an interposition or bypass graft with cyst resection.

Conclusion

PCAD is a rare cause of intermittent claudication predominantly affecting young males. Outcomes after definitive surgical management involving resection and interposition bypass are excellent. Though rare, PCAD should be considered by clinicians as a potential diagnosis when military personnel present with intermittent calf claudication symptoms.

Acknowledgements

Thanks to Dr Abdel Kader Allouni, consultant interventional radiologist, for reconstructing the CT images.

References


Authors

Alexander J Collingwood
5th year medical student
School of Medicine
Keele University

Surgeon Commander Daniel J Ablett Royal Navy
Consultant Vascular Surgeon
University Hospitals of North Midlands NHS Trust
Royal Stoke University Hospital
Newcastle Road
STOKE-ON-TRENT
ST4 6QG
danielablett@hotmail.com