Intended for healthcare professionals

Endgames Picture Quiz

An unusual case of calf pain

BMJ 2010; 340 doi: https://doi.org/10.1136/bmj.c716 (Published 18 February 2010) Cite this as: BMJ 2010;340:c716

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A healthy 45 year old man with a passion for rock and roll presented to our vascular outpatients clinic. Over the previous few months he had been unable to continue his dancing sessions because of pain in his right calf, which started a few minutes after he started dancing. He had also noticed that the pain would come on after 200 m of walking.

On examination he was well, with a blood pressure of 140/80 mm Hg and a pulse of 80 beats/min, which was regular and of good volume. He had normal heart sounds and normal peripheral pulses in the upper limbs and in the left leg but absent pulses distal to the femoral pulse in the right leg. The ankle brachial pressure index was normal on the left but 0.7 on the right. He had smoked 20 cigarettes a day since he was a teenager but was otherwise fit and well. His fasting blood glucose and haemoglobin were normal, as was his preoperative electrocardiography.

A computed tomography angiogram of his lower limbs had been performed before he was referred to our clinic (fig 1⇓).

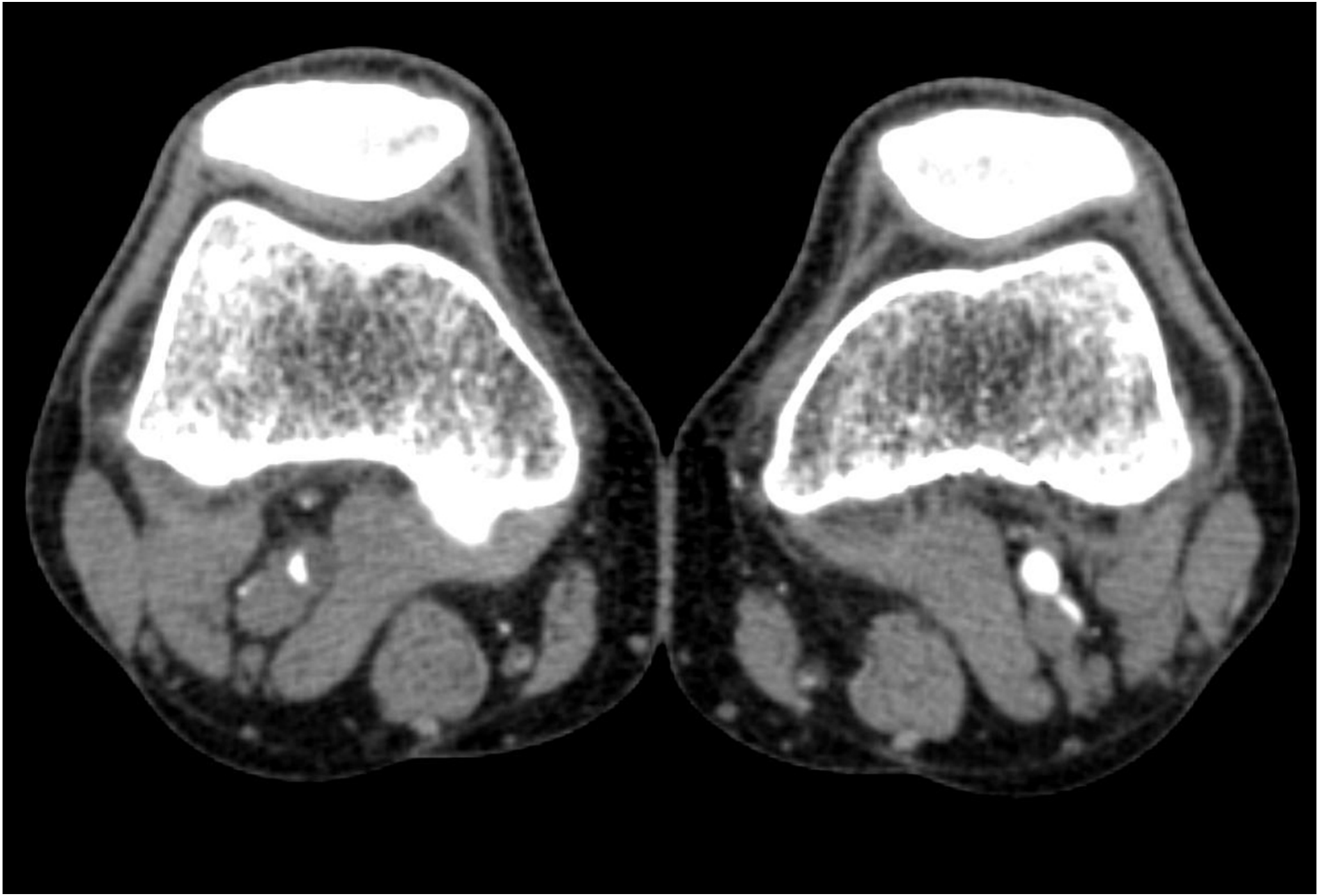

Fig 1 Computed tomography angiogram of the patient's lower limbs

Questions

1 What abnormality is present on the computed tomography angiogram?

2 What is the most likely diagnosis, and what is the differential diagnosis?

3 What further tests can be done to confirm the diagnosis?

4 How should this condition be managed?

Answers

1 What abnormality is present on the computed tomography angiogram?

Short answer

Stenosis of the right popliteal artery is present, as shown by the passage of less contrast through this section of the artery. The lumen is compressed so as to resemble a scimitar (“the scimitar sign”) (fig 2⇓).

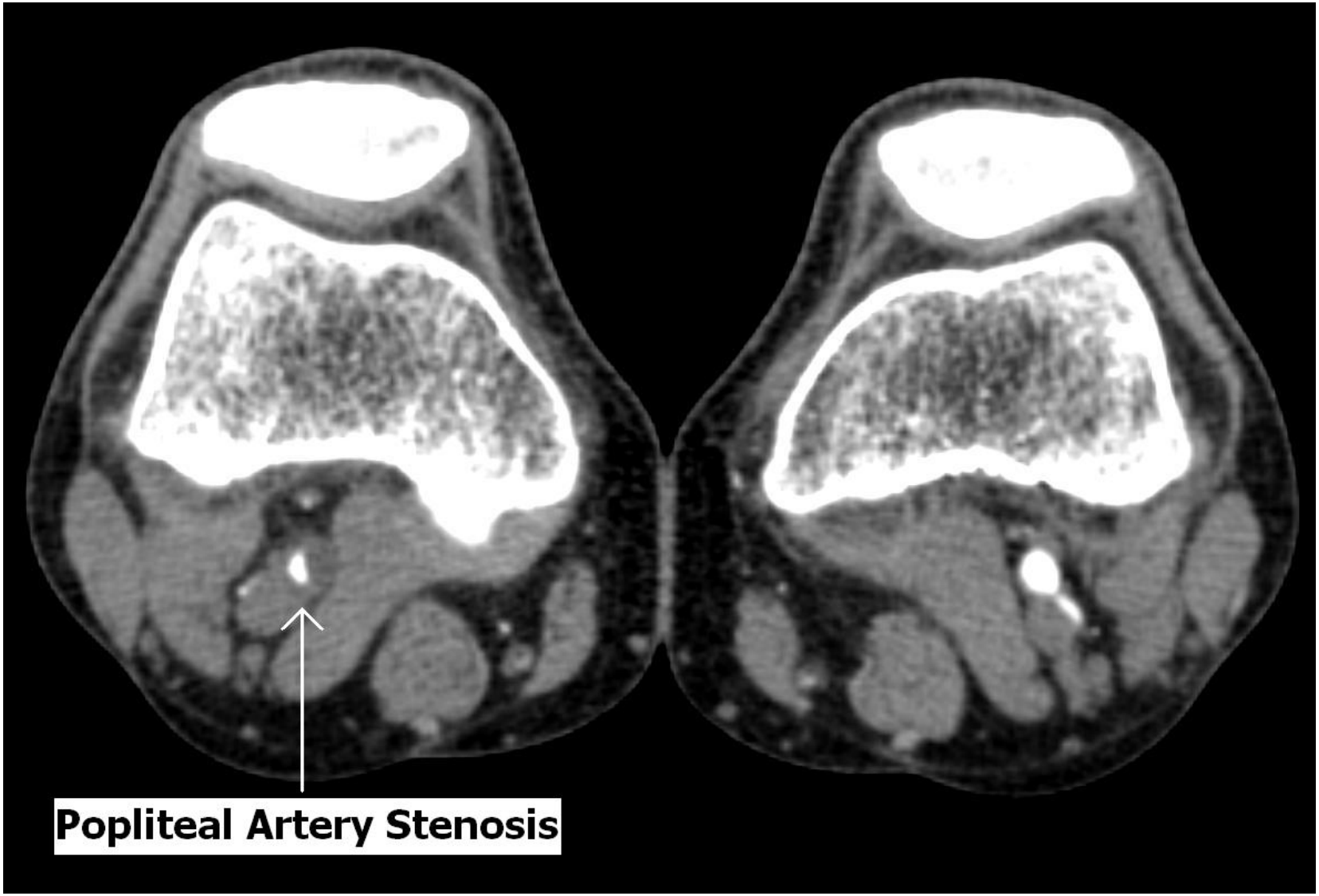

Fig 2 Computed tomography angiogram of the patient's lower limbs showing stenosis of the right popliteal artery (the scimitar sign)

Long answer

Computed tomography angiography is increasingly being used as part of the investigation of disabling intermittent claudication and is replacing traditional digital subtraction angiography.[1] The computed tomography scan showed stenosis in the right popliteal artery, with the characteristic scimitar sign. This implies an eccentrically placed lesion within the wall of the artery. Above and below this section of popliteal artery, the vessel was completely normal with no evidence of any atherosclerotic disease.

2 What is the most likely diagnosis, and what is the differential diagnosis?

Short answer

Considering the young age of this patient and the scimitar sign, the most likely diagnosis is cystic adventitial disease of the popliteal artery, which consists of cystic degeneration in the wall of a vessel. The differential diagnosis includes popliteal entrapment syndrome, which can also occur in this age group, and because the patient smokes, atherosclerotic peripheral arterial disease with an eccentric plaque.

Long answer

Apart from being a smoker, this man had no risk factors for peripheral vascular disease. Considering his young age and the scimitar sign the most likely diagnosis is cystic adventitial disease of the right popliteal artery.[2] Cystic adventitial disease is an uncommon condition of the peripheral vessels arising from cystic degeneration in the adventitia of a vessel. It is thought to account for 0.1% of vascular disease, and features of cystic adventitial disease are seen in one in 1000 angiograms.[3] It has been reported in the femoral, external iliac, radial, brachial, ulnar, and popliteal arteries and veins, and also in the superficial veins of the legs (saphenous veins).[2] The most common site is the popliteal artery, however, with around 85% of cases occurring in this location.[3]

In all cases, the diseased artery or vein overlies a joint. It is thought that adventitial cysts are formed and maintained by communication with a synovial space, with mucin secreting mesenchymal cells from the adjacent joint being included in the adventitia of the artery during development.[4] These adventitial cysts can be single or multiple.

The symptoms of this disease are caused by the cysts compressing the artery and producing stenosis or even complete occlusion. The presentation depends on the degree of compression. Patients can present with intermittent calf claudication if they have compression of the artery or acute ischaemia if they have popliteal artery thrombosis at the site of stenosis.

Cystic adventitial disease most commonly occurs in the fifth decade, with a male to female ratio of 5:1.[5] Cystic adventitial disease should therefore be suspected in healthy young patients (usually with no vascular risk factors) who present with calf claudication.

The differential diagnosis includes popliteal entrapment syndrome, which can also present at a young age and results from abnormal muscular anatomy in the popliteal fossa leading to popliteal artery stenosis.[6] [7] In this condition, the association between the medial head of the gastrocnemius and the popliteal artery is abnormal.[8] The medial head of this muscle may be inserted more laterally than usual, thus compressing the artery, or the artery may take an abnormal course medial to the normally attached medial head of the gastrocnemius muscle. Anomalous muscle and fibrous bands may also occur.[9] Although arterial compression can be shown on conventional angiography or sonography, the underlying anatomical abnormality is best shown using functional magnetic resonance imaging and magnetic resonance angiography.[10] In functional magnetic resonance imaging images are taken when performing manoeuvres, such as forced plantar flexion, that tense the gastrocnemius muscle and thus produce stenosis of the popliteal artery, which may not be evident at rest.

3 What further tests can be done to confirm the diagnosis?

Short answer

Ultrasound duplex scanning would confirm the stenosis and possibly show the cysts in the wall of the artery. Magnetic resonance angiography would also show the popliteal artery stenosis and cysts.

Long answer

Various methods of investigation exist. Duplex ultrasound will show the arterial stenosis and the surrounding cysts, which will contain no flow. The cysts will appear as anechoic or hypoechoic masses in the arterial wall.[11] Magnetic resonance imaging can also help show the cysts,[12] and the image will depend on their shape. If they are concentric, the stenosis will have an hourglass appearance, whereas if they are eccentric, the stenosis will have a scimitar appearance.[13] T2 weighted images will show high signal cysts, whereas T1 weighted images will show variable signal intensity because of the variable amount of mucoid material within the cysts.[12] Angiography (traditional digital subtraction or computed tomography angiography), as used here, can also show the arterial compression.[14] Computed tomography angiography can show the cysts as well, in which case no further tests are needed to confirm the diagnosis. Each imaging modality has its own advantages and disadvantages, and none is universally agreed to be superior.

4 How should this condition be managed?

Short answer

If the cyst is causing stenosis only, it can be dissected off the wall of the artery (cyst enucleation). If however, the cyst has caused arterial occlusion, the diseased segment of the artery should be excised and reconstructed using a vein graft (interposition grafting).

Long answer

The treatment depends on the stage of disease at presentation. If the patient is symptomatic and artery stenosis is present, the cyst should be excised, whereas in the presence of arterial occlusion, it may be necessary to excise the diseased segment of the artery and perform interposition vein grafting.[15] Aspiration of cysts can be successful, but the risk of recurrence is high.[16] Ideally, the patient should be treated before progression to complete arterial occlusion occurs. The small number of reported cases indicate that patients usually have disease in one site only,[17] [18] and most cases affect the popliteal artery only. The risk of recurrence depends on the type of treatment. The risk is highest with simple aspiration, but low with complete excision of the cyst or interposition grafting.[19]

Patient outcome

Our patient was operated on through a posterior popliteal fossa approach. The cyst was enucleated leaving an intact artery (fig 3⇓). The symptoms resolved completely, his ankle brachial pressure index returned to normal, and he was able to resume rock and roll.

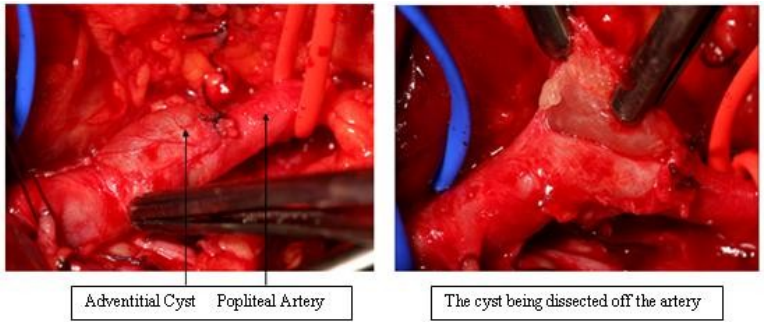

Fig 3 Intraoperative view of the cyst being dissected off the popliteal artery

Notes

Cite this as: *BMJ* 2010;340:c716

Footnotes

- Competing interests: All authors have completed the Unified Competing Interest form at www.icmje.org/coi_disclosure.pdf (available on request from the corresponding author) and declare: (1) No financial support for the submitted work from anyone other than their employer; (2) No financial relationships with commercial entities that might have an interest in the submitted work; (3) No spouses, partners, or children with relationships with commercial entities that might have an interest in the submitted work; (4) No non-financial interests that may be relevant to the submitted work.
- Provenance and peer review: Not commissioned; externally peer reviewed.
- Patient consent obtained.

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