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SHORT REPORT

Cystic Adventitial Disease: A Trap for the Unwary

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Cystic adventitial disease is an uncommon condition. A case of cystic adventitial disease of the popliteal artery is reported in a young man who has been followed up for 14 years after surgical treatment. Early recognition and treatment of the condition will prevent progression to popliteal thrombosis and critical ischaemia. However, diagnosis of the condition is difficult.
Characteristic features in the presenting history, such as fluctuation in severity of symptoms, sudden onset after vigorous activity and delayed recovery time after cessation of exercise are identified, which should help the clinician avoid misdiagnosis and delayed diagnosis of the condition. The clinician is also warned of the associated misleading clinical features such as the presence of normal peripheral pulses and normal ankle pressures in some cases of CAD.

Keywords: Cystic adventitial disease; Intermittent claudication.

Introduction

Cystic adventitial disease (CAD) is an uncommon condition with only around 300 cases reported in the literature[1,2] since, the first case was described in the external iliac artery in 1947 by Atkins and Key.[3] CAD consists of a collection of gelatinous material within a cyst adjacent or surrounding a vessel resulting in pressure on that vessel. The reason for the apparent rarity of this condition, however, may be due to the fact that many cases may remain unrecognised. We report a case of cystic adventitial disease in a 36-year-old gentleman whose diagnosis was delayed. He was eventually treated successfully with incision and drainage of the popliteal cyst and ligation of communicating channels to the knee joint and remains asymptomatic 14 years later. We highlight the potential pitfalls in the diagnosis and treatment of this condition.

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Case Report

A 36-year-old man presented with a history of pain in his left calf on prolonged standing and on walking, the symptoms being relieved by rest and elevation. He did not smoke and was very fit. On examination he had gross saphenofemoral incompetence and large varicose veins. All his peripheral pulses were present and there was no objective evidence of ischaemia. He underwent left high saphenous ligation and multiple avulsions. However, his symptoms persisted and he was referred to the Vascular Unit 18 months after first developing his calf pain. He claimed that he needed to rest for at least 10 min before the pain in the calf started to improve. Again his pulses were normal as were ankle pressures. Pressures were measured after exercise and after the pain developed and still there was no drop in pressure. Angiography showed a very slight stenosis of the popliteal artery and angioplasty was offered but the patient refused as his symptoms had improved.

He was readmitted 4 months later with worsening symptoms. This time the ankle pressure was found to drop on contracting his calf muscles. A fresh angiogram showed complete occlusion of the popliteal artery on knee flexion against resistance (Fig. 1) and a decision was taken to proceed to surgery based on a

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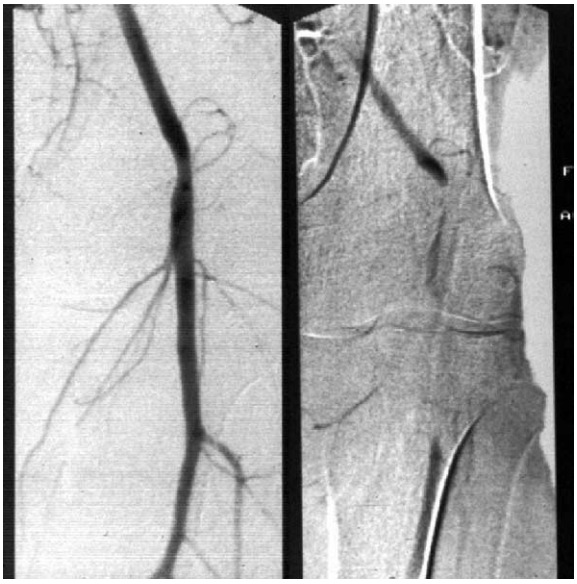

Fig. 1. Angiogram on left: at rest; angiogram on right: on flexion of knee against resistance. Complete occlusion of popliteal artery on knee flexion.

presumed diagnosis of popliteal entrapment syndrome. It was only at surgery that it became clear that cystic adventitial disease was the cause of his popliteal occlusion. Attempts at aspirating the cystic lesion compressing the artery were unsuccessful because of the viscous consistency of the fluid (Fig. 2). The cyst was incised and the thick, clear, mucinous contents evacuated. The evacuated fluid looked like and had the consistency of ganglion fluid. A communicating channel led from the cyst towards the knee joint and this was followed as far as possible and ligated. The adventitia was repaired and the wound closed.

The patient remains entirely asymptomatic 14 years after surgery. His ankle-brachial pressure indices are normal on both sides. An ultrasound scan shows a small remanant of the previous cyst posterior to the popliteal artery measuring 8 mm.

Discussion

Cystic adventitial disease (CAD) is an uncommon condition reported in 85% of cases in the popliteal artery and affecting mainly young males.[2,4] The appearance of claudication in a young non-smoking male and the typical angiographic findings have been claimed to confirm the pathology.[5,6] However, elderly males with the risk factors for peripheral vascular disease have also been reported to suffer from the condition.[7–10] There is, therefore, no age-group that is immune from CAD.

Early recognition of the condition is important because it is often rapidly progressive[8] and because treatment of the condition before it progresses to popliteal occlusion is associated with less morbidity. However, reaching a diagnosis of CAD is fraught with problems.

The symptoms of calf claudication in patients with CAD tend to wax and wane. Severe claudication may suddenly improve spontaneously and completely disappear, only to recur a few months later.[11,12] Direct communication between the cyst and the knee joint is often demonstrated[13] and this could explain the reported waxing and waning of symptoms as well as

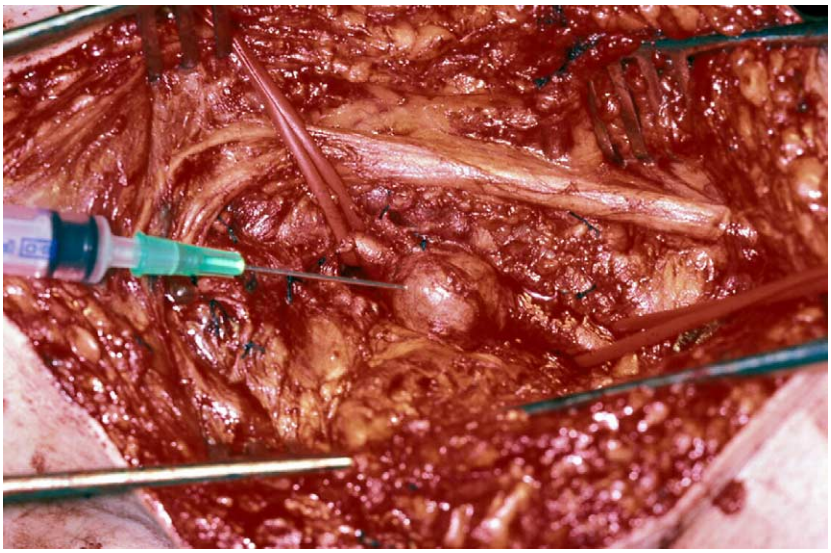

Fig. 2. Cystic adventitial disease of the popliteal artery: attempt at aspiration.

the reported cases of spontaneous resolution of CAD.[14,15] One patient was reported to suffer from periodic distressing and unrelieved right calf claudication and pallor of the foot on walking 100 yards against a background of an extraordinary capacity for exercise exemplified by a hard game of football or running a marathon.[16] Such a history coupled with perfectly normal peripheral pulses and ankle pressures (as in our patient) is enough to make a vascular cause for the presenting symptoms unlikely to all except the most attentive of clinicians. Normal pulses, ankle pressures and even ankle pressures after exercise have been reported in association with CAD.[7,8,17] In patients with a clear history of intermittent claudication and normal peripheral pulses it is worth checking for foot pulses during knee flexion. Ishikawa *et al*. reported disappearance of foot pulses on knee flexion or after exercise in patients with CAD, Ishikawa's sign.[18] To exclude popliteal artery entrapment syndrome as the cause, attempted plantar flexion of the foot against resistance while keeping the knee extended puts the gastrocnemius muscle under tension, and pedal pulses become weaker or disappear if entrapment is the cause.

Abrupt onset of symptoms and commencement of symptoms after vigorous activity seems to be a recurring mode of presentation. Hildreth reported a case of a 42-year-old gentleman who developed right calf claudication suddenly after a wrestling match with his sons after which he was unable to walk more than 100 yards.[19] Another patient was reported to have developed sudden onset of claudication while playing as catcher in a baseball game.[20] The context of pain in the calf in a young male developing soon after vigorous activity could easily be interpreted as musculoskeletal in origin. The unusual finding that pain in patients with CAD takes longer to subside after exercise ceases than in typical claudication could reinforce the clinician's misdiagnosis. A long recovery time was noted in our patient. Similarly Hunt *et al*.[7] reported that symptoms only improved after 20 min in a patient with CAD. They attributed this long recovery period to the fact that pressure in the cyst increased during exertion and that the pressure only fell as fluid was slowly reabsorbed from the cyst, thus gradually reducing the obstruction of the popliteal artery. Another hypothesis for the long recovery time is that during exercise fluid is forced into the cyst from the knee, and a valve mechanism prevents the return of fluid into the knee joint, so that the elevated pressure is maintained in the cyst even when the pressure in the knee has returned to normal.[7,21]

If CAD is suspected further investigation is, therefore, necessary even in cases where peripheral pulses and resting pressures are normal and the history is atypical. Angiography frequently shows a characteristic smooth tapering stenosis, referred to as the scimitar sign, without poststenotic dilatation and with no evidence of atherosclerotic disease.[22] However, in 30% of cases CAD produces a non-specific complete occlusion that can easily be mistaken for an endoluminal lesion[23] while in other cases angiography may be completely normal.[24] While Doppler ultrasound can be helpful in identifying the cystic lesion of CAD around the vessel, low-echoic lesions can sometimes be missed[25] and in other cases ultrasound may suggest a popliteal aneurysm.[26] Computed tomography, magnetic resonance imaging (MRI) and intravascular ultrasound have all been recommended in the diagnosis of CAD, MR imaging probably being the best modality.[17,23]

In view of the difficulties raised by the presentation and investigation of the condition, it is not surprising that many cases are misdiagnosed. Our patient underwent varicose vein surgery before being referred to a vascular unit for assessment. Another case was admitted under the care of orthopaedic surgeons and underwent fasciotomy.[16] Angiographic changes may be interpreted as being due to atherosclerotic disease and the patient may be referred for angioplasty,[11] which has been shown to be unsuccessful in the treatment of CAD.[27] Delay in diagnosis is commonly seen in cases reported, and often the diagnosis is only made once complete occlusion of the popliteal artery develops.[11,16,28]

Several treatment options have been proposed for CAD. Aspiration of the cystic collection has been performed successfully,[29] although recurrence is frequent.[30] This is not surprising considering the fact that many of these cysts communicate with the adjacent joint and, therefore, aspiration will not obliterate the communication. Surgical incision or partial or complete excision without opening the artery has been performed with good results[6,12,19,31] when the vessel is patent. Identifying and ligating any communicating channels into the adjacent joint is claimed to reduce the risk of recurrence following cyst excision.

Most cases of popliteal occlusion secondary to CAD have been treated with resection of the affected segment and autogenous vein graft reconstruction.[1,16,32–36] The complications associated with this procedure are commoner and more serious than with simple cyst excision.[16,32–34] To avoid resection, thrombolytic therapy followed by non-resectional cystostomy has been used to treat CAD which has progressed to vessel occlusion.[37]

In summary, the clinician should be alerted to the possibility of CAD in patients whose symptoms of

claudication wax and wane and in whom claudication seems to develop suddenly, particularly after vigorous activity. The recovery time after ceasing exercise tends to be longer than in the typical claudicant. The presence of normal peripheral pulses and normal ankle pressures does not exclude CAD. It is worth performing Ishikawa's test in such cases. Angiography and Doppler ultrasound may be unhelpful in some cases. Angiography may only demonstrate occlusion of the popliteal artery on knee flexion against resistance. Simple excision of the cystic lesion is probably the treatment of choice where this is possible.

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