Cystic adventitial disease of the external iliac artery: a rare cause of claudication

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A 40-year-old, healthy female presented complaining of intermittent right calf claudication that occurred after minimal exercise. The symptoms were getting progressively worse over 4 weeks. She denied any rest pain or non-healing wounds. She was unable to do her work as a farm manager due to her troublesome symptoms.

Her past medical history was unremarkable with no previous surgery or trauma.

On examination, her femoral pulse on the right was weak and reduced to 1/3 compared to 2/3 on the left. No bruits were heard. Popliteal, dorsalis pedis and posterior tibial pulses were easily palpable and 2/2 bilaterally. Her resting ankle brachial indices (ABI) were normal at 1.0 bilaterally. This remained normal on the left, but dropped to 0.6 on the right with exercise, using treadmill exercise protocol with speed of 3.2 km/hr and incline of 10 degrees for 5 minutes. Ultrasound duplex scan showed a cyst measuring 3x3x2cm externally compressing the right external iliac artery. Computer Tomography Arteriogram (CTA) showed a non-vascular homogenous cyst compressing the right external iliac artery as shown in Figure 1. The cyst did not have any joint connection.

The patient underwent surgical exploration by a retroperitoneal approach. Intraoperative findings were consistent with a mucinous-filled cyst adjacent to the anterior aspect of the right external iliac artery, as shown in Figure 2. No connection to a joint could be identified intraoperatively. The cyst was resected and the adjacent arterial wall opened, the residual stenosis dealt with by a bovine pericardial patch. Postoperative ABIs returned to 1.0 at rest and post-exercise. The patient recovered well and was discharged home after two days with no residual symptoms. She returned to full-time work after two weeks post-operatively. She remains clinically asymptomatic. Using our standardised follow-up protocol, the patient underwent duplex scanning 3 months after the procedure, showing no signs of recur-

Figure 1: CTA showing a cystic structure compressing the right external iliac artery.
Figure 2: Intra-operative view showing a cyst narrowing the external iliac artery.

Figure 3: Cystic wall surrounding gelatinous material which forms the stroma of the resected cyst. X40 magnification.
rence. The histology of the cyst was consistent with cystic adventitial disease.

Cystic adventitial disease (CAD) causes arterial insufficiency. Atkins and Key reported the first case of CAD in 1947. CAD is defined as a collection of gelatinous material within an aberrant synovial-type cyst in the sub-adventitial plane of a blood vessel which results in external compression. Symptoms of CAD depend on the location of the affected blood vessel. In 85% of reports, CAD affects the popliteal artery. Claudication of a sudden-onset is the most common complaint. The age group affected by CAD is usually 40–60 years old. Nevertheless, CAD has been reported in children.

The pathophysiology of CAD is not well understood. Repeated stretching causes adventitial degeneration and cyst formation, particularly in the popliteal artery. The misplacement of mucin-secreting cells in the adventitial layer of blood vessels during fetal development is thought to cause cyst formation later in life.

Desy et al reported cases in which CAD was caused by a cystic lesion with a direct joint connection, often via small branches of the affected artery. In our case, no joint connection could be identified, nor any arterial side branches in that area.

Macroscopically cysts are often described as "gelatinous, jelly-like material". Microscopically, cysts contain variable amounts of fibrinogen, hydroxyproline, hyaluronic acid and mucin. This explains the differing echogenicity of CAD cysts.

Diagnosis of CAD requires a high index of suspicion. Clinicians should be vigilant of the history, age of the patient and site of the lesion. For popliteal CAD, distal pulses may disappear with maximal flexion of knees due to compression of the artery by the cystic structure. The ankle-brachial index at rest and exercise is useful, however, a drop in pressure post-exercise is not always present.

Imaging is essential in the diagnosis of CAD. Historically, arteriography was used to investigate CAD. The classic appearance of the blood vessel effected by CAD has been described as the ‘Scimitar sign’, a curvilinear narrowing of the vessel. If the cyst itself is concentric, the appearance is more hourglass-like.

Considering the invasiveness of arteriography, imaging modalities such as ultrasound, CTA and MRA are more frequently used. Duplex ultrasound can quantify the stenosis of the blood vessel and identify the cyst.

MRI may provide additional information regarding possible joint connections, but is not as readily available as alternative imaging modalities.

The management of CAD is dependent on the severity of the symptoms. If symptoms are minimal, then conservative management is justified. Surgery is the mainstay of treatment for severe CAD cases. Resection of the cyst and adjacent vessel with a bypass graft, or excision of the cyst with patch repair, are the recommended techniques. Complete resection and bypass/graft interposition is the surgical technique used if the cyst causes occlusion of the blood vessel. The preservation of the native arterial wall with patch repair, rather than excision of arterial wall adjacent to the cyst, is recommended. If joint connection is identified then ligation of such connections in combination with cyst resection is recommended as a treatment strategy.

Percutaneous and endovascular aspiration and angioplasty of CAD have high recurrence rate and are usually not recommended.

As the cyst in our case only affected the anterior aspect of the external iliac artery, a cyst excision and bovine pericardium patch repair was done rather than resection of the whole segment and vein graft interposition, thus saving the patient the risk of vein harvesting morbidities. Potential joint connections were not identified in this case.

Recurrence is low and estimated to be 1 in 40 surgically treated cases. No specific long-term follow up is routinely required other than the usual graft follow-up if the patient remains asymptomatic.

In summary, CAD is a rare cause of arterial insufficiency in otherwise healthy adults. A high index of suspicion combined with clinical examination and US imaging is critical in CAD diagnosis. Surgery is recommended in symptomatic patients.
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