Popliteal artery cystic adventitial disease: A case report and a review of the current literature

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Abstract:
Cystic adventitial disease is a rare condition affecting mostly the popliteal artery, associated to non-atherosclerotic intermittent claudication. We report a case of a 59-year-old male patient, with mild atherosclerotic risk factors, complaining of intermittent claudication in his left calf. Duplex ultrasonography and computed tomography angiography revealed a cystic lesion highly adhered to the popliteal artery, causing a critical stenosis. The patient treated with an excision of the P1 segment of the diseased artery and a venous by-pass grafting. Nowadays, less than 250 cases of popliteal artery cystic adventitial disease are recorded in the literature. Surgical treatment with excision of the cyst and by-pass grafting with autologous vein graft is the treatment of choice.

INTRODUCTION
Cystic adventitial disease (CAD) is a rare condition associated to non-atherosclerotic intermittent claudication in young patients. A cystic lesion could be found in any vessel with a higher predominance at the popliteal artery (PACAD). It concerns a unilocular or multilocular mucin-filled cyst in the vessel’s adventitia which may result in arterial stenosis or thrombosis.1 In the literature, almost 250 cases of cystic adventitial disease of the popliteal artery have been recorded, mostly in case reports and small series.4 Most of the patients underwent a surgical excision of the cyst, accomplished with an artery re-reconstruction, with a low recurrence rate.3 We report a case of a 59-year-old male patient treated in our department for a PACAD and we present a review of the current literature. This case presentation was approved by the Ethical committee of our Hospital.

CASE PRESENTATION
The patient presented to the Vascular Outpatient Service complaining of intermittent claudication in his left calf at 300 meters, during the past 6 months. He was an ex-smoker (tobacco cessation 5 years ago) and hypertensive under treatment. No trauma or other atherosclerotic risk factors were recorded in his medical history. Clinical examination revealed an absent dorsalis pedis and posterior tibial pulse at his left limb. Resting ankle brachial index (ABI) was 1.1 on the right and 0.7 on the left limb. The patient underwent a duplex ultrasonography (DUS) with extremity in complete extension and during knee flexion initially, and subsequently a computed tomography angiography (CTA) of lower limbs which revealed a cyst of 3cm diameter, highly adhered to the popliteal artery, causing a stenosis >90% (Figure 1).

Figure 1. Preoperative CTA image with scimitar sign, the cystic lesion of the popliteal artery (white arrow) and the ligament connecting it to the knee joint (yellow arrow)

Peripheral arteries were all patent with no atheromatosis. The patient was scheduled for surgical treatment. Through a medial approach, the above the knee popliteal artery was dissected, and a cystic formation was revealed at the P1 segment of the popliteal artery. A ligamentous band between the cystic part of the artery and the knee joint was found, ligated and cut (Figure 2).
Post-operatively, the pulses on the left dorsalis pedis and posterior tibial arteries were restored, and ABI was measured at 1.1. The patient was discharged on the 4th post-operative day under a lifelong treatment of aspirin 100mg once daily. The histological report confirmed a cystic lesion firmly attached to the popliteal adventitia. At the 6th month follow-up, the patient was asymptomatic, with complete recovery of his daily routine activities, and the graft was patent on the DUS.

DISCUSSION

PACAD is an uncommon nonatherosclerotic cause of intermittent claudication, with an incidence of 1 in 1200 cases of claudication. It is characterized by unilocular or multilocular myxomatous cysts, situated in the adventia. The majority of CAD occurs in males (ratio 4:1), during the 4th and 5th decade of life, with no or minor atherosclerotic risk factors, as in this case. Almost all patients suffer from intermittent claudication of the calf, with a period of symptoms' recovery longer than in typical atherosclerotic claudication. Nowadays, less than 250 cases of PACAD are recorded in the literature.

CAD pathogenesis has not been clearly defined. Various theories have been suggested. According to the trauma theory, repetitive microtraumatic mechanisms of the nearby joints may provoke the destruction and cystic degeneration of the adventitia. In the developmental theory, stem cells from joints migrate into the adventitia during embryonic development. The systemic disorder theory, which is the most popular, incriminates a systemic connective tissue disease. According to the gaglion theory, adjacent synovial cells of the nearby joints migrate through a low-pressure pathway and form cysts. In many cases, as in this case, a “ligament” between the cyst and the joint is revealed. According to the ligament theory, there is a consistent fluid communication between the knee joint and the cyst.

The typical profile of PACAD is that of a middle-aged male patient with no severe comorbidities or atherosclerotic risk factors who presents with a new onset intermittent calf claudication. Initially, the symptoms could be sudden or insidious. Intermittent claudication tends to wax and wane as the cyst may resolve spontaneously. In the literature, spontaneous resolve of the symptomatology has been associated with spontaneous rupture of the cyst. Peripheral limb pulses are generally palpable in rest but diminished after exercise. Disappearance of the foot pulses during knee flexion could be seen in PACAD, a phenomenon described as the Ishikawa’s sign. This sign facilitates the differential diagnosis between popliteal cystic adventitia disease and the popliteal entrapment syndrome, where foot pulses disappear in ankle plantar flexion. ABI measurement is not pathognomonic for the disease.

DUS is a useful, non-invasive diagnostic media for PACAD. When performed by an experienced radiologist, it is usually the first imaging test that reveals the cystic nature of the lesion and the pre-occlusion stenosis. The hypoechoic characteristics of the cyst facilitates its differential diagnosis over a popliteal aneurysm. An echogenic thin line separating the vessel lumen and the cyst can be imaged. On color DUS, a scintlar sign could be seen at the narrowed lumen, producing high velocity. Intravascular ultrasound, computed tomography (CT) and magnetic resonance imaging have also been advocated in the diagnosis of CAD. In angiography, an eccentric narrow-
ing of the lumen (scimitar sign), hourglass narrowing of the lumen, or complete occlusion with a lack of post-stenotic dilatation may be detected. However, the differential diagnosis between the PACAD and atherosclerotic stenosis occasionally remains quite difficult.8

Surgical treatment with bypass grafting, using an autologous vein graft (great or small saphenous vein), after the cyst excision constitutes the intervention of choice in most of symptomatic patients.2,3,9 Otherwise, cyst evacuation followed by a patch angioplasty can also be performed. Simple resection of the cyst, and CT-guided or ultrasound-guided percutaneous aspiration have also been described and used in monocystic cysts without cyst-artery adhesion. Aspiration of the cyst could be inefficient in multifocal cyst disease and in high viscosity cyst content. Simple cyst excision and ligation of the cyst-joint connections has been proposed in cases where the intima preservation was possible.5 Complete excision of the diseased popliteal artery has shown a low recurrence rate (0-10%) compared to the partial cyst excision with remaining lesions (10-34% recurrence rate).6 Endovascular techniques have no clear place in the treatment of PACAD, because of the compressional nature of the disease. Even if they are described in the literature, they are associated with a high rate of recurrence and a possibility of arterial thrombosis. Conservative treatment is proposed only for asymptomatic patients.10

CONCLUSION

PACAD is an uncommon non-atherosclerotic vascular disease associated with intermittent claudication. A high clinical suspicion and an appropriate imaging investigation are very important to set diagnosis. Excision of the cyst and bypass grafting with autologous vein graft is, generally, the preferred treatment, with a low incidence of recurrence.

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REFERENCES

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