Adventitial Cystic Disease of the Popliteal Artery Presenting as an Acute Limb Ischemia in a Female

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Abstract

Adventitial cystic disease (ACD) is a rare vascular disease that involves the arteries and rarely the veins, most commonly, the popliteal artery in male patients. Etiology remains idiopathic. Clinically, the most common presenting symptom is claudication, although they can also present as critical limb ischemia. Diagnosis requires a high index of suspicion especially in patients with limb threatening ischemia, in the absence of other risk factors for atherosclerotic disease. Angiography, ultrasound (US), computed tomography (CT) and magnetic resonance imaging (MRI) can all be used for diagnosis. Treatment of choice of CAD is surgical resection and bypass using an interposition graft.

Keywords: Adventitial cystic disease (ACD), computed tomography angiography (CTA), ultrasound (US), Ishikawa sign, scimitar sign, claudication, limb ischemia.

Introduction

In adventitial cystic disease (ACD)a nonatherosclerotic mucous-like cyst forms in an artery, narrowing its lumen, and eventually obstructing blood flow. The condition usually affects the popliteal artery, which supplies blood to the knee joint, calf muscles, and foot.[1] In rare cases, the condition can cause cysts to form in other arteries. The etiology and optimal treatment of ACD remain controversial.

We report here a rare presentation of a popliteal artery ACD and demonstrate the benefit of an immediate intervention and surgical resection of the cystic segment for limb salvage.[2]

Case Report

A 75 years of age Bahraini Female patient with a past medical history of Hypertension, Hyperlipidemia, type II Diabetes Mellitus, and Osteoporosis, presented to our facility's emergency department in March 2021 complaining of left lower limb pain and coldness up to the left knee of one day duration.

The patient's vital signs were normal. Arterial examination revealed palpable left femoral pulse with absent left popliteal and pedal pulses, and absent pedal arteries doppler signals. All pulses in the right lower limb were palpable.

The patient was admitted and dealt with as acute limb ischemia. On the table angiogram showed focal occlusion of the second segment of the popliteal artery with an "hourglass" appearance. Otherwise, her entire arterial tree was normal with no evidence of atherosclerosis. Below the knee popliteal artery was patent. The upper two third and anterior and posterior tibial and peroneal arteries were patent. Balloon angioplasty of popliteal artery was performed with 4mm and 5mm balloons with good angiographic results and no residual stenosis [Figures 1 and 2]. Catheter directed thrombolysis was initiated to dissolve any distal thrombo-emboli. It is worth noting that the occlusive lesion did not look atherosclerotic in nature. Therefore, uncommon causes of limb ischemia were suspected.

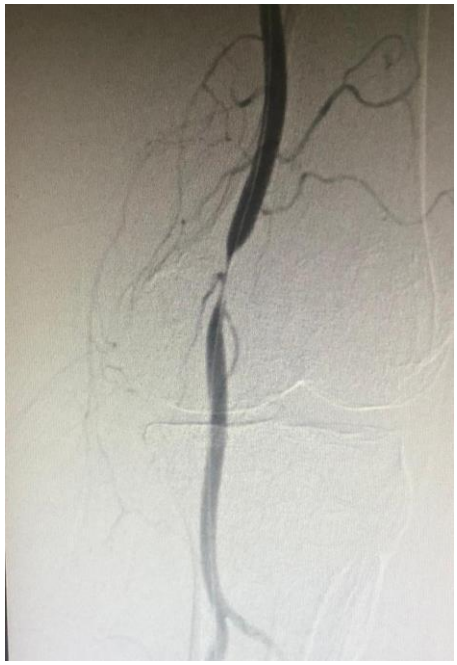

Figure 1: Angiogram shows PA occlusive lesion (hourglass sign).

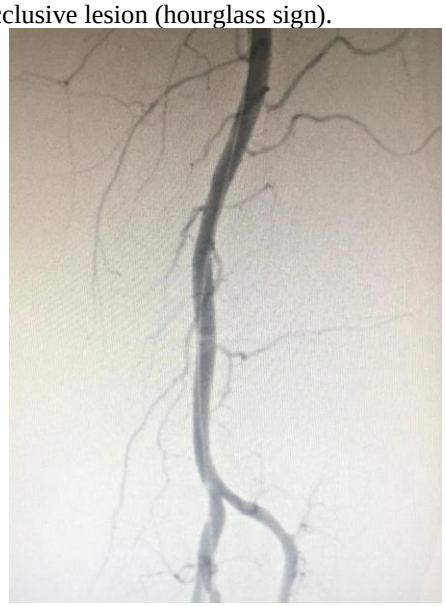

Figure 2: Post balloon angioplasty of PA.

Postoperatively, immediate relief of symptoms was achieved and left dorsalis pedis artery pulse became palpable. However, the next day the patient started to have recurrence of calf pain and weaker distal pulses. An Angiogram was performed the next day as part of the thrombolysis protocol revealed 50% restenosis (recoiling) of the popliteal lesion [Figure 3], with patent anterior tibial & peroneal arteries and good flow to the foot. The posterior tibial artery was patent but diffusely narrowed in its distal half.

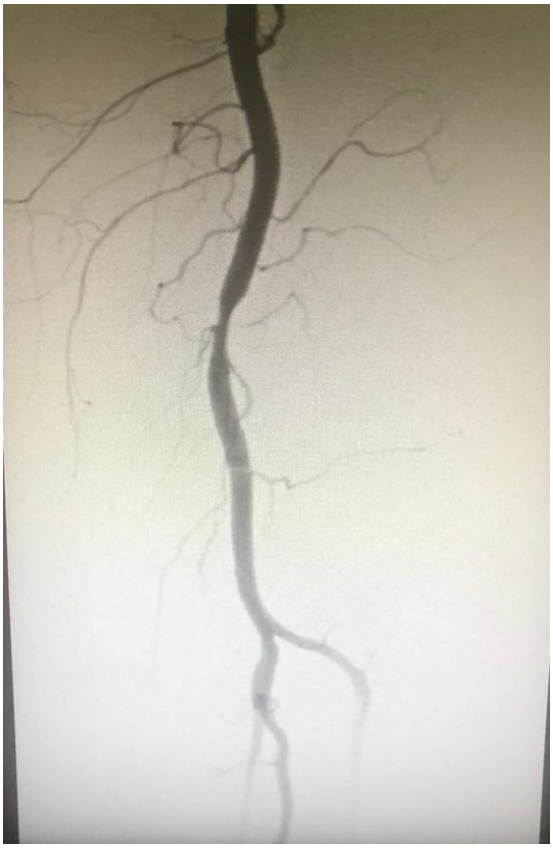

Figure 3: Restenosis of PA after one day of balloon angioplasty.

Cystic adventitial disease of PA was strongly suspected. Arterial duplex of the left lower limb showed 70% focal stenosis of the PA with hypoechoic lesion seen forming hemispherical neural lesion on the same side & bilateral baker's cyst with organized fluid [Figure 4].

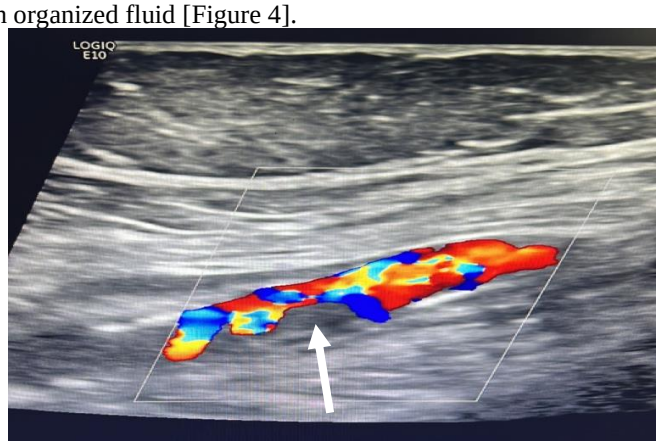

Figure 4: Duplex Ultrasound showing cystic adventitial lesion causing intra-luminal narrowing of popliteal artery (white arrow).

CT angiography of the left lower limb was done and revealed a focal high grade stenosis of the second segment of the popliteal artery. At the same level a mural ovoid shape fluid density lesion was found measuring 5x19 mm both compressing and compromising the arterial lumen. A Backer's cyst measuring 13x22x36 mm was also seen [Figure 5 and 6].

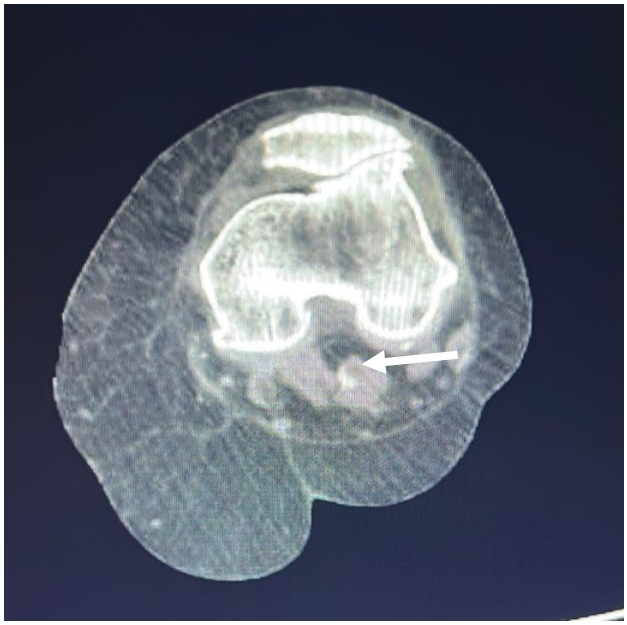

Figure 5: Axial view of CT Angio revealed cystic lesion of the popliteal artery significantly compromising its lumen.

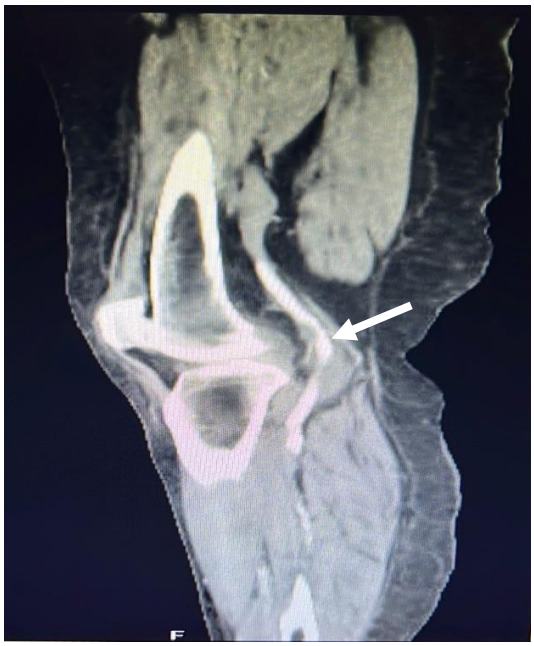

Figure 6: Sagittal view of CT Angio showing luminal narrowing of the 2nd segment of the popliteal artery.

The diagnosis of ACD was entertained. Therefore, the patient was offered a definitive procedure; resection of the popliteal artery and placement of an inter-position vein graft. She consented and prepared for surgery. Venous mapping showed poor superficial veins in the lower limb. Left arm cephalic vein was deemed suitable. Under general anesthesia, the patient was prepped and draped in the usual fashion in prone position. The cephalic vein in the left arm was harvested. A lazy S incision was made in the left popliteal fossa. Dissection was carried down to fascia overlying leg muscles, the sciatic nerve and it branches were identifies, preserved, and gently retracted laterally. The popliteal artery and vein were explored. Adventitial cysts were clearly seen in the diseased segment of the popliteal artery. Interestingly, a communication between the popliteal artery adventitia and the Baker's Cyst was found. After systemic heparinization and control of the popliteal artery, resection of the diseased segment was carried out, and specimen was sent for histopathology. The cephalic vein graft was anastomosed proximally and distally to a healthy segment of the popliteal artery. Arterial clamps were removed, and flow was reinstituted to the lower limb. [Figure 6 and 7] incision was closed in layers. Post operative recovery was uneventful and the patient was discharged from the hospital in good condition.

Histo-Pathology report showed cystic spaces and mucinous changes in the adventitial layer of the wall of the Popliteal artery specimen which is positive for Alcain blue mucin stain. Additionally, there was degeneration of the external elastic layer and focal clacifiation of the wall. Findings consistent with ACD.

During clinic follow up, the patient developed partial wound dehiscence with superficial surgical site infection. This was promptly treated with appropriate antibiotics and negative pressure wound dressing until complete healing was achieved.

She was followed up with regular 4 monthly arterial duplex of the lower limbs, the latest was in March 2023 and showed patent bypass with normal flow velocities and no recurrence of ACD. The contralateral popliteal artery was also free from signs of ACD. A CTA was performed at 1 year post operatively and was normal [Figure 7].

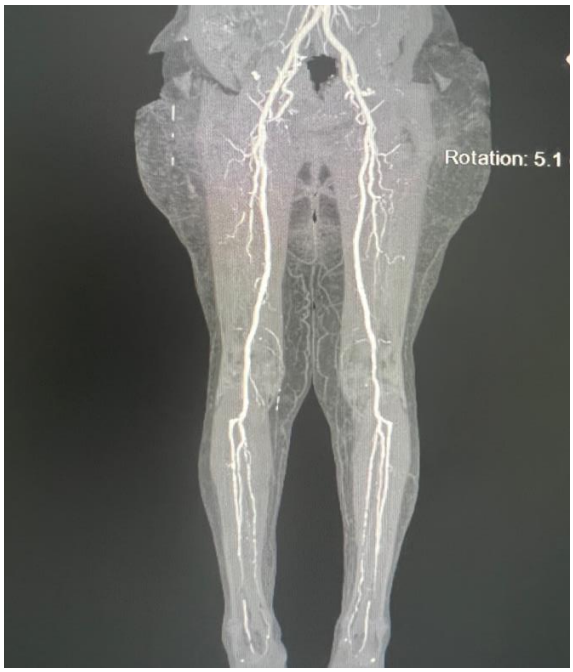

Figure 7: CTA of lower limb showing patent bypass graft after one year of follow up

Discussion

Adventitial cystic disease is a non-atherosclerotic peripheral vascular disease. Functional occlusion of a vessel is caused by compression by a cyst wall.[3,4] It usually occurs unilaterally and most commonly affects the popliteal artery.[1] It is more common in males, with a male to female ratio of 15:1. Affected patients are usually in their mid-40s.[4,5] The disease accounts for 0.1% of lower-extremity claudication in patients with no atherosclerotic risk factors.[6,7]

ACD of the popliteal artery is a rare condition with a poorly understood etiology. Several theories have been proposed to explain the development of ACD, but the exact cause is still not known. Here are some of the most proposed etiological theories for ACD:

1. Inflammation: Some studies have suggested that inflammation in the adventitia of the popliteal artery may lead to the formation of cysts.[8]

2. Arteriosclerosis: Some studies have suggested that arteriosclerosis in the popliteal artery may play a role in the development of ACD.[9]

3. Trauma: It has been suggested that trauma to the popliteal artery could result in the formation of a cyst in the adventitia.[10]

4. Developmental anomaly: Another theory is that ACD may be a congenital or developmental anomaly of the popliteal artery.[11]

In short, the exact cause is still not well understood. Further research is needed to better understand the development of this condition.

CT and magnetic resonance imaging with T1- and T2-weighted imaging have been recommended for the diagnosis of ACD of the popliteal artery,[4] as they are more beneficial for the evaluation of cyst morphology and recognizing the cyst's relationship to surrounding structures. Doppler ultrasonography is a useful non-invasive diagnostic modality. However, it is operator dependent and needs to be performed by an experienced radiologist.

Our patient had signs and symptoms of acute limb ischemia. Two studies of clinical and imaging findings of patients with ACD found that more than half of patients had presented with acute limb ischemia.[8,9] Therefore, it is important for physicians to consider the diagnosis of ACD in patients presenting with acute limb ischemia.

Endovascular management of ACD with balloon angioplasty and, or stenting has been entertained, however, long term results are uncertain.[2,7]]. Complete excision of cysts alone in cases without intimal damage have been described.[12] However, higher incidence of treatment failure or recurrence has been reported for complete cystic excision without arterial reconstruction (10-34%) compared with revascularization surgery (0-10%).[11-13] Accurate incidences of recurrence post each procedure cannot be given due to lack of regular follow up among other factors[14,15]

A literature search (1980-2021, PubMed) shows only one report of successful endovascular treatments for CAD.[15] The same report describes two cases of arterial stenting for popliteal adventitial cystic disease and persistent 4- and 10-year symptomatic patency rate. The authors state that the secret to successful endovascular treatment is stenting for the radial force of the bare-metal self-expanding stent pushes the adventitial cyst apart and dilates the lumen.[15] There have been no current reports of cases of performing endovascular treatments using a drug-coated balloon. The cyst is underneath the adventitia, so it is natural to expect balloon angioplasty to fail. Furthermore, the healthy intima of the artery can be injured during endovascular treatment, inducing a higher risk for arterial thrombosis.[16] On rare occasions, the cyst in the popliteal artery regressed spontaneously.[17] Surgical management is the definitive standard treatment for this disease.[18] Excision of the diseased artery with insertion of autologous venous interposition graft is the main method of surgical treatment. It was mentioned in one study that recurrence developed in 6 cases after either cyst excision alone or patch angioplasty. There was no recurrence after vessel excision with interposition grafting. Vessel excision was a statistically significant factor in recurrence prevention (P=0.026). Based on this result, we can conclude that cyst excision alone and patch angioplasty are not effective, and undergoing vessel excision and interposition should be considered to reduce or prevent disease recurrence.[19]

Our case was unique in the sense that ACD was diagnosed in an elderly female with risk factors for peripheral arterial disease presenting with acute limb ischemia. The hourglass appearance of the occlusive lesion in the popliteal artery on the initial angiogram, and the early recoiling of the lesion raised suspicion for ACD. The diagnosis was confirmed by duplex ultrasound, CT Angiography, and eventually the pathology report. Resection of the diseased segment of the popliteal artery with insertion of an interposition vein graft was undertaken and is the favorable definitive treatment for this condition in our opinion.

Conclusion

Cystic Adventitial disease is a rare, nonatherosclerotic vascular disorder. It must be considered in the differential diagnosis of patients presenting with limb ischemia. Diagnosis is made through careful attention to clinical presentation, radiological signs, behavior of the lesion, use of diagnostic imaging tools, and pathology of the specimen. Treatment is always interventional. Open surgical approach seems to have better outcome than endovascular management for definitive management of this rare condition.

Conflict of interest

The authors declare that there is no conflict of interest regarding the publication of this paper.

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Ethics approval and consent to participate:

Written informed consent was obtained from the Patient. This case report was approved by the Bahrain Defense Force Hospital Research and Ethical Committee.

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