

Cystic Adventitial Popliteal Disease: Personal Experience and Literature Review

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Abstract

Background: Cystic adventitial disease (CAD) primarily affects arteries and is described as a rare vascular disorder characterized by fluid accumulation in the adventitial layer of a vessel which may cause luminal narrowing or even complete occlusion. Nowadays different pathogenetic theories exist as well as different treatment solution.

Case Report: We report two cases of cystic adventitial disease of the popliteal artery treated over the last 4 years in healthy men. Both experienced the sudden onset of intermittent calf claudication; one also presented blue toe syndrome. In both cases the diagnose was confirmed after open surgery and histological evaluation. Complete resection and autologous vein bypass were performed in one case; complete resection and direct end to end anastomosis in the others.

Discussion: Cystic adventitial disease is a rare vascular disorder with different interesting theories on its aetiology. Treatment options include percutaneous ultrasound-guided aspiration, endovascular approaches and open surgical cyst excision with or without direct reconstruction. In our Institution, we faced two popliteal CAD cases; open surgery represented the treatment strategy for both: in the first case an autologous vein bypass was performed, while in the second case the artery contiguity was restored with an end-to-end anastomosis. Both patients experienced full relief of symptoms without any recurrence of the disease.

Conclusion: Even if rare, popliteal artery CAD has to be suspected in young healthy patients who refer the sudden onset of acute or sub-acute symptoms related to peripheral arterial occlusive disease. Differential diagnose between popliteal entrapment syndrome and popliteal aneurysms is not always easy. When CAD is suspected or diagnosed a conventional surgical approach is recommended.

Keywords: Cyst, Disease, Popliteal Artery, Lower Leg Ischemia, Bypass

Introduction

Cystic adventitial disease (CAD) is a rare vascular disorder characterized by fluid accumulation in the adventitial layer of a vessel, which may cause narrowing or even complete occlusion of the lumen [1,2]. The cystic fluid has been described as gelatinous or mucoid with a high content of hyaluronic acid and different combinations of mucopolysaccharides and mucoproteins [1,3].

The first case of CAD was described in 1947 by Atkins and Key who reported a case of myxomatous tumour at the level of the external iliac artery [4]. This vascular disease primarily affects arteries; venous CAD is extremely rare and predominantly described at the level of the common femoral vein being an unusual cause of venous thrombosis [5].

Popliteal artery is the site where CAD is most frequently localized although external iliac, femoral and radial arteries are possible sites of presentation [6,7]. Cystic adventitial disease of the popliteal artery is characterized by variety of symptoms similar

to peripheral arterial occlusive disease; main differences are the absence of atherosclerotic risk factors and the age of their onset. Multiple techniques have been described for the treatment, with open surgical repair remaining the most successful one.

We report our experience in the treatment of cystic adventitial disease of popliteal artery describing two cases of surgical resection and different methods of reconstruction, performing a reversed small saphenous vein bypass in one patient and a direct artery reconstruction in the others. We also describe the main pathogenetic theories of CAD, its management and the different suitable treatment options by reviewing articles published between 2010 and March 2021 searched via PUBMED databases.

Case Report

Case 1: A healthy and active 56 years old man was referred to our institution because of 3-months history of intermittent claudication involving the left calf after a walking distance of 300 meters. During last 10 days he experienced the progressive worsening of the symptoms and the sudden appearance of cya-

nosis of the homolateral big toe. The only atherosclerotic risk factor was smoking. The physical examination of the affected leg showed normal femoral pulse, no popliteal and distal pulses and cyanosis of the big toe. All contralateral leg pulses were present.

Color Doppler ultrasonography (DUS) revealed at the site of popliteal artery a bilobed anechoic lesion completely inclosing the vessel wall. Contrast enhanced computed tomography (CT) showed compression of the left popliteal artery by a non-enhancing structure, initially described as Baker's cyst, associated with the complete thrombosis of the lumen of the artery and its re-habitation at the distal segment under the knee Figure 1.

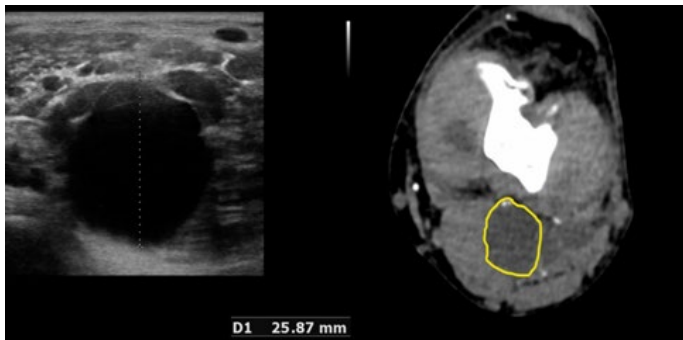


Figure 1: Ultrasound imaging showing a bilobed anechoic lesion compressing the left popliteal artery. Angio CT scan showing complete occlusion of the left popliteal artery and the cystic lesion in its place.

After a collegial consultation with radiologist and orthopedic colleagues, a suspicious diagnosis of CAD was advanced and surgical treatment was chosen. After obtaining patient's informed consent the popliteal artery was exposed through a posterior approach via an S-shaped incision over the popliteal fossa. The mass was completely mobilized circumferentially revealing an apparent communication between the cyst and knee joint. After systemic heparinization (5000 IU of heparin sulphate) and clamping the popliteal artery was incised through a longitudinal arteriotomy: after the adventitial tunica incision the Authors observed the leaking of a gelatinous material that strongly confirmed the diagnosis of CAD Figure 2.



Figure 2: Mucoïd material leaking out after incision of the adventitial cyst.

The entire cyst was then resected and a segment of patient's homolateral small saphenous vein was interposed in a reverse fashion Figure 3 the blood flow was restored with the recovery of all distal arterial pulses. The patient made an uneventful recovery and was discharged on the third post-operative day with-

out any complication. Single antiplatelet therapy was prescribed (100 mg/die of acetylsalicylic acid). DUS examination before discharge showed normal triphasic Doppler waveform through the left popliteal artery without anastomotic defects; histological examination of artery's segment defined its content as "amorphous material", excluding any oncological disease.

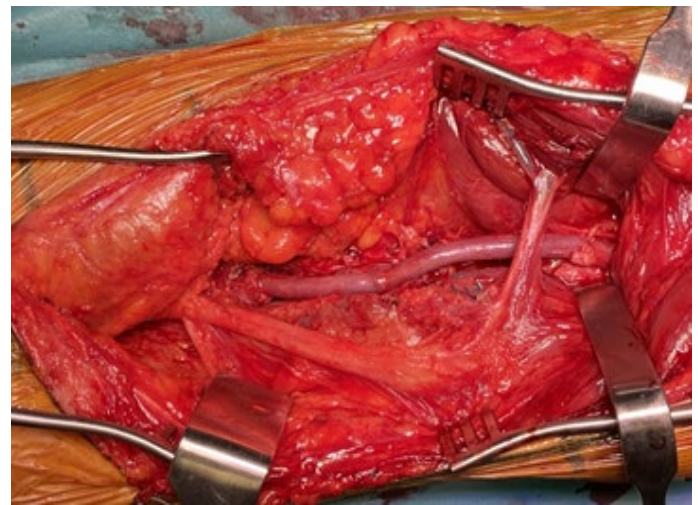


Figure 3: Popliteal artery replaced with reversed small saphenous vein interposition.

The 1-month follow up Duplex ultrasound exam showed good patency of the vein graft and the absence of cyst recurrence. Furthermore, the patient experienced fully relief of symptoms; the big toe recovered normal color, temperature and sensibility few days afterwards.

Case 2: A 64 years old sportsman was admitted to our institution with 1-month history of left calf intermittent claudication. Physical examination revealed only a good left femoral pulse. Color Doppler ultrasonography showed sudden focal occlusion of a dilatated popliteal artery with poorly detectable distal flow. Hypertension was the only atherosclerotic risk factor. Patient underwent digital subtraction angiography (DSA) which showed perfect patency of all below the knee vessels and the "scimitar sign" at the popliteal level Figure 4.



Figure 4: Angiography showing the typical curvilinear scimitar sign due to extrinsic compression of the lumen.

In the suspect of a non-atherosclerotic popliteal disease an MRI was then preoperatively performed. T2 weighted sequences showed a fluid aspect inside popliteal artery confirming CAD diagnosis. An open surgical repair through an S-shaped posterior incision over the popliteal fossa was chosen. The patient was systemically heparinized (5000 IU of heparin sulphate), the diseased popliteal artery segment was resected, and the vessel continuity was restored through an end-to end 6/0 prolene suture Figure 5. Patient totally recovered from the symptoms and was discharged on second post-operative day with single antiplatelet therapy (Clopidogrel 75 mg/die). The 3- months follow-up revealed no limb pain and a normal walking distance. The 24 months follow-up Duplex ultrasound showed no signs of recurrence or restenosis. Histological evaluation showed fibrosclerosis of the vessel wall associated with areas of old and fresh haemorrhage.

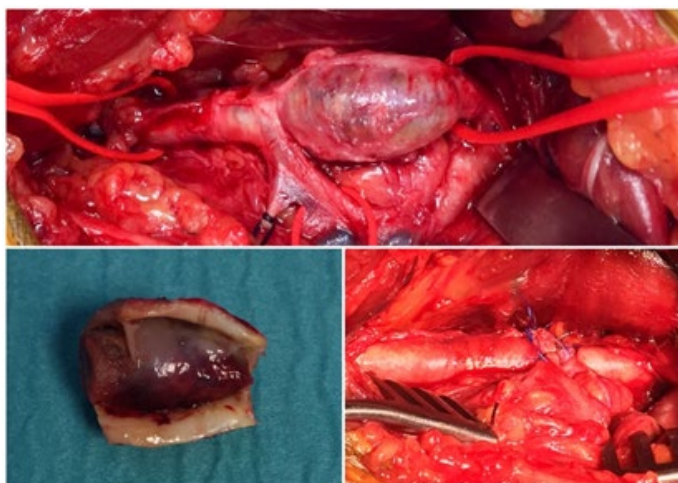


Figure 5: Resected segment of affected popliteal artery with tenacious mucoid material coming from the adventitial cyst. End to end anastomosis to restore the arterial continuity.

Discussion

Adventitial cystic disease is a rare cause of claudication as rare is the pathology itself. It should be suspected in patients with no or minimal cardiovascular risk factors, with a reported male-to-female ratio of approximately 5:1 and mean age of onset between 40 and 50 years [1,2,8]. In our study series mean age at diagnosis was 51 years (Range 5-87) with a prevalence in males (M=90; 76,9%; male-to female ratio 10:3) in Table 1. Because of its rarity, prevalence of CAD is unknown, but it is estimated that CAD accounts for <0.1% of cases of intermittent claudication [1]. A multicentric experience coming from fourteen different institutions, describes an average number of cases per institution of 3 over a period of 10 years, which is in line with our data [3].

Table 1: Patient Demographics

Patients (N, %)	117
• Male	90 (76,9 %)
• Female	27 (23,1%)
Mean age	51 y (Range 5-87)
Presenting symptoms (N, %)	
• Intermittent claudication	104 (89%)
• Burning pain and paresthesia	6 (5%)
• Pain behind the knee	3 (2,6%)
• Rest pain	2 (2%)
• Acute calf pain	1 (0,7%)
• DVT	1 (0,7%)

Aetiology

When it comes to the definition of its origin, different theories can be postulated: the trauma theory, the systemic disorder theory, the synovial theory and the developmental theory. The traumatic theory suggests that multiple (micro) traumas due to anatomical position of the popliteal artery could be responsible for the degeneration of its wall. The systemic disorder theory considers the cystic degeneration of the popliteal artery as part of a generalized disorder involving connective tissue [9]. Because of the histological and biochemical similarity between adventitial cyst and ganglia, the synovial theory suggests a possible origin of the former from the capsular synovial structures that eventually enlarge and involve the adventitia of adjacent vessel [10,11]. Finally, the developmental theory suggests mucin secreting cells becoming incorporated into the vessel wall during embryological development [12,13]. This could support the evidence of an apparent communication between artery and adjacent joint, frequently identified intraoperatively.

The most unifying theory however comes from Levien et al according to whom CAD is caused by the incorporation of mesenchymal cells, that normally are destined to joint tissue, into the developing of non-axial vessel approximately during the 10-22th week of embryological development [14]. These cells would later be responsible of the production of mucoid material.

Clinical Presentation

The most common symptom of popliteal artery CAD is intermittent claudication of the lower extremity: the onset is usually acute and can rapidly progress. Other presenting symptoms can be burning pain and paraesthesia of the affected lower extremity, pain behind the knee, acute calf pain, cyst rupture (rarely), ischemic rest pain and occasionally deep vein thrombosis also [1-3]. In those series the most frequent presenting symptom was calf intermittent claudication (89% of the total), followed by burning pain and paraesthesia (5%), pain behind the knee (2,6%), rest pain (2%), acute limb pain (0,7%) and one case of deep vein thrombosis (0,7%) Table 1.

Diagnosis

No diagnostic criteria have been standardized for CAD: clinical examination, paying attention to absence of risk factors, mean age, sudden as well as rapidly progressive symptoms and duplex ultrasound have to be considered as the first level exams. When popliteal artery appears totally occluded, as described in our cases, it is difficult to differentiate between entrapment syndrome, popliteal artery aneurysms or popliteal cysts using only duplex ultrasound. In these cases, CT and MRI may help: MRI, thanks

to 3D reconstruction, may allow to recognize the cystic nature of the lesion and may show the connection between the adventitial cyst and the adjacent joint capsule appearing the cyst's fluid as a high signal intensity on T2-weighted images [15,16]. Currently angiography is no more considered diagnostic. However, it should be considered a useful completion exam to preoperative planning: for example, the evidence of the "scimitar sign", due to displacement of the artery to one side, provides further support to the diagnosis of CAD.

Treatment

Treatment options of CAD include percutaneous ultrasound-guided aspiration, endovascular approach with percutaneous transluminal balloon angioplasty (PTA) and open surgical cyst excision with patch interposition or bypass grafting; in this case the use of the autologous veins is recommended when available. The surgical approach strongly depends on clinical symptoms and radiological findings; however, it should be recommended even if the diagnosis is not certain. The main goals of the surgical procedure are to remove the mass from popliteal fossa in order to have a certain histological examination and, as in our case, to restore arterial patency preventing progression of symptoms.

The cyst excision is by far the most widely reported surgical treatment: removal of the cyst content allows lumen to return to its regular diameter. Success rates range from 85% to 94% with

failure often due to cyst recurrence [17,18]. The second most reported technique is surgical excision of the pathological artery segment with interposition of venous or prosthetic graft. Success rates between 93.5%-95% are reported for this procedure; among patients treated with resection of the artery and bypass grafting, the use of autologous vein reported better results compared to prosthetic graft [19,20].

The percutaneous cyst aspiration, not always practicable due to viscosity of the cyst content or the presence of multilocular cysts, shows little success; a high recurrence rate is caused by the re-accumulation of fluid or its incomplete aspiration [21,22]. Percutaneous endovascular procedures using angioplasty or stenting initially aroused enthusiasm for the treatment of these lesions. However, considering the poor results mainly due to the nature of the disease that is extraluminal and not atherosclerotic, it has progressively abandoned [23-26].

Table 2 provides an overview of popliteal CAD cases published on PubMed between 2010 and March 2021. In the Study series a total of 117 procedures were described: among surgical techniques cyst resection, with or without bypass interposition, was the most performed technique. Open surgical treatment was performed in 103 of 117 patients (89%); the remaining fourteen patients underwent US guided percutaneous cyst aspiration (n=7; 6%), observation (n=4; 3,4%) or percutaneous angioplasty/stenting (n=3; 2,6%).

Table 2: Published cases between 2010 e 2021 searched via Pubmed databases

	Total Cases	Success	Failure	Comments
Arterial segment resection and vein reconstruction	28	27	1	• Restenosis after 11 months
US-guided percutaneous cyst aspiration	7	4	3	• Cyst's recurrence after 10 months • Cyst's recurrence after 2 weeks • Cyst's recurrence after 3 months
Arterial segment resection and synthetic graft reconstruction	30	29	1	• One bypass loss of patency treated with ballon angioplasty after 6 years
Angioplasty/ Stenting (BMS)	3	3	-	
Arterial segment resection and primary anastomosis	28	15	3	• Restenosis after 6 months • Restenosis after 6 months • Cyst's recurrence after 9 months
Cyst excision	10	8	2	• Cyst's recurrence after 10 months • Cyst's recurrence after 7 months
+ patch closure	6	6	-	
Arterial segment resection and criopreserved arterial graft reconstruction	1	1	-	
Observation	4	3	1	• Acute limb ischemia after 3 months
Total	117	106	11	

The choice to use or not graft interposition largely depended on individual anatomical characteristics. When available, tendency was to use autologous great or small saphenous vein (1/28 restenosis), however good results were achieved also using prosthetic grafts (only one bypass occlusion after 6 years over thirty performed). Recurrence of the cyst was observed in three patients treated with US guided percutaneous cyst aspiration (3/7), in two patients undergoing simple cyst excision (2/10), and in one case of arterial segment resection and primary anastomosis reconstruction (1/10). In this last surgical group also two rest-

enosis occurred (2/10). Three patients treated with angioplasty were asymptomatic after 45, 51, and 117 months respectively, no failures were observed in this group. Among patients treated with simple medical therapy one of them developed acute limb ischemia after three months (1/4).

In our institution we experienced two cases of CAD at popliteal artery; the treatment was surgical for both patients, in fact we performed the complete excision of the diseased segment in both cases. In one case the artery contiguity was restored with

an end-to-end anastomosis thanks to the limited area involved; in the other one an autologous vein bypass was necessary. Both cases experienced fully relief of symptoms without recurrence of cystic adventitial disease.

Highlights

- Cystic adventitial disease is a rare vascular disorder characterized by fluid accumulation in the adventitial layer of an artery.
- Cystic adventitial disease may cause luminal narrowing or even complete arterial occlusion.
- Cystic adventitial disease has to be suspected in young healthy patients who refer to acute or sub-acute symptoms of peripheral arterial occlusive disease.
- A conventional surgical approach through open cyst excision and direct or indirect reconstruction remain the treatments of choice.

Conclusion

Even if rare, CAD has to be suspected in young patients who refer sudden manifestation of progressive claudication or acute limb ischemia with no or few cardiovascular risk factors. When suspected at Duplex ultrasound it should be always mandatory performing a second level exam in order to confirm the diagnosis and planning the strategy of the treatment. Considering the nature of the disease, in our experience conventional surgical approach through arterial segment resection and direct reconstruction or bypass grafting remains the treatment of choice.

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