

## Cystic adventitial disease of the popliteal artery: report of two cases and review of the literature

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**SUMMARY:** Cystic adventitial disease of the popliteal artery: report of two cases and review of the literature.

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*Cystic adventitial disease (CAD) is a rare vascular disease that causes a localized stenosis or occlusion in absence of alterations of blood vessels in other sites of the body. CAD is predominantly located to the popliteal artery, although cases have been described involving other ar-*

*teries. Typically it affects young men with minimal cardiovascular risk factors, presenting a short history of progressive claudication. Imaging is based on US, CTA and MRA. Suspected diagnosis is confirmed at the time of the surgery. We report two cases of CAD involving the popliteal artery. In the first case a 59 year-old man was treated by resection of the popliteal artery and a reversed saphenous vein was used to restore circulation. In the second case a 53 year-old man was treated by resection of the popliteal artery and a cryo-preserved arterial graft was used to restore circulation. We also made a review of the literature on this subject.*

**KEY WORDS:** Cystic adventitial disease - Popliteal artery - Mixomatous degeneration of popliteal artery.

### Introduction

Cystic adventitial disease (CAD) is a rare disease that causes a localized stenosis or occlusion in absence of alterations of blood vessels in other sites of the body. The first case was described by Atkins and Kay in 1947, located to the external iliac artery (1). In 1954 Ejrup and Hiertonn described the first case located to the popliteal artery (2). Until now lesser than 400 total cases have been described in the literature.

CAD is predominantly located to the popliteal artery (approximately 85% of total cases), although cases have been described involving the external iliac artery (1, 3), common femoral artery (4-7), axillary artery (8), distal brachial artery (9), radial and ulnar arteries (10, 11). Rarer cases have been described involving femoral, popliteal or saphenous veins (12-14). The disease is characterized

by the presence of unilocular or multilocular cysts containing mucin, located in the adventitia of the involved vessel. The reported lesions of the media and intima are widely believed to be secondary (15).

CAD is most common in male (m:f ratio 5:1) and it occurs more frequently between the fourth and the fifth decade of life, although several cases have been described in younger or older patients. The patients are usually healthy without atherosclerotic degeneration signs or cardiovascular risk factors, often sportsmen. Nowadays, the origin of this disease still remains unknown.

The presentation is typically characterized by intermittent claudication worsening in the calf in a short time.

In this paper we present two cases of CAD involving the popliteal artery and we make a review of the literature on this subject.

### Case report 1

A 59 year-old male was investigated for a cramp-like pain in the left calf. The intermittent claudication was disabling and the recovery time after the suspension of walking was as much as 5 minutes. The patient was affected by asthma and no other cardiovascular comorbidities were present.

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Physical examination revealed weak left popliteal, posterior tibial and dorsalis pedis pulses.

Colordoppler ultrasound showed an anechoic cyst, located at the popliteal artery, involving adventitia.

Conventional digital subtraction angiography (DSA) showed an hourglass appearance at level of the popliteal artery extended for about 4 cm (Fig. 1 A) and a CT-angiography (CTA) documented near-occlusive stenosis of the popliteal, more than 90% (Fig. 1 B, C, D).

Popliteal fossa was explored by a posterior S-shaped approach. The popliteal artery was swelled with translucent appearance in the middle segment. No connections between cyst and knee joint were found.

After heparinization, the compromised vessel was resected and a segment of reversed saphenous vein was used to restore vascularization. At the end of the procedure limb was well vascularized and distal pulses were present and normal. The postoperative course was regular and complication free.

The patient started walking on the third postoperative day and was discharged on the fourth, treated with ASA 100 mg 1 tablet/day. Histological examination of the surgical specimen confirmed the diagnosis of CAD.

Seven years after surgery, the patient presents a good running autonomy and no episodes of intermittent claudication occurred during the follow-up. The limb is warm, well perfused with normal pulses and colordoppler ultrasound shows regular patency and no aneurysmal degeneration of the graft.

## Case report 2

A 53-year-old male presented with a short history of intermittent claudication in the left calf with a symptom-free walk interval of 10 metres, without rest pain.

He was a smoker until 20 years ago, and no other comorbidities or cardiovascular risk factor were found.

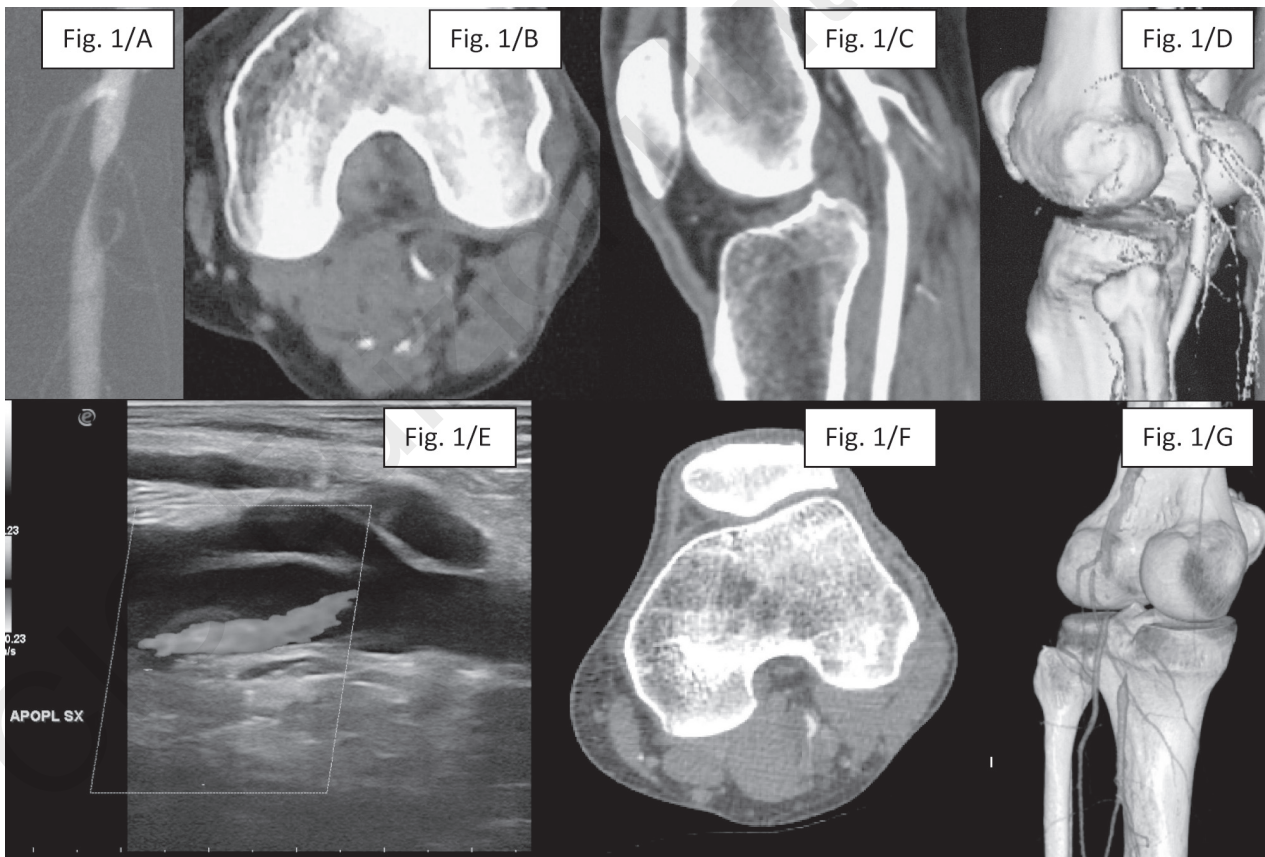
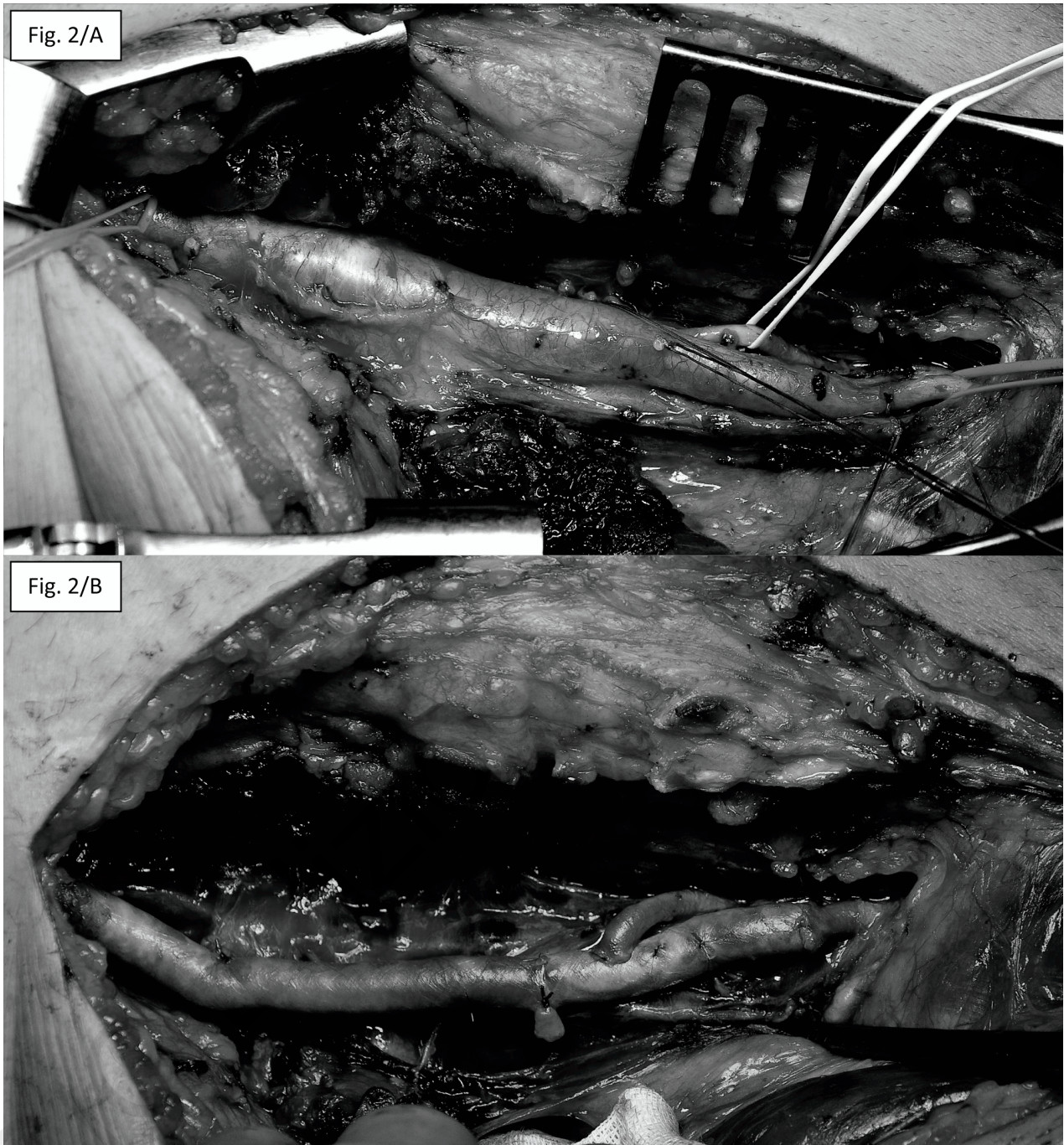


Fig. 1

- A - Case 1: Hourglass appearance at level of the popliteal artery by DSA.
- B - Case 1: Near-occlusive stenosis of the popliteal artery by axial CTA imaging.
- C - Case 1: Near-occlusive stenosis of the popliteal artery by sagittal CTA reconstruction.
- D - Case 1: Near-occlusive stenosis of the popliteal artery by 3D-CTA reconstruction.
- E - Case 2: Colordoppler-US showing on the anterior wall of the popliteal artery a large anechoic cyst.
- F - Case 2: Near-occlusive stenosis of the popliteal artery by axial CTA imaging.
- G - Case 2: Near-occlusive stenosis of the popliteal artery by 3D-CTA reconstruction.



**Fig. 2**  
A - Case 2: Exposure of the affected popliteal artery  
B - Case 2: Complete replacement of the affected artery and reconstruction with tube-homograft and reimplantation of the anterior tibial artery.

Physical examination revealed signs of ischaemia to the left leg. The femoral pulse was normal, the popliteal and posterior tibial pulses were diminished. The left foot was pale and hypothermic compared to the opposite.

The patient performed a color doppler ultrasound of the lower limbs (Fig. 1 E) which showed on the arteri-

or wall of the left popliteal artery a large anechoic area due to the presence of an adventitial cyst, while distally persisting a good flow at the posterior tibial artery and weak at the anterior tibial artery.

A CT-angiography (CTA) of the lower limbs was performed, showing a subocclusive stenosis of the popliteal

artery and of the left tibio-peroneal trunk, more than 90% (Fig. 1 F), extended for about 7 cm, in the suspicion of CAD (Fig. 1 G).

Surgical access was performed through medial route, exploring the popliteal fossa. The popliteal artery showed dilated and with translucent appearance in the middle and distal segments, involving the origin of the tibio-peroneal trunk because of the presence of a cyst (Figure 2 A). No connections with the joint capsule of the knee were found.

After heparinization, compromised vessel was resected, a cryopreserved arterial graft with a diameter of 7 mm was used to restore vascularization, by using end-to-end anastomoses on the popliteal artery and on the tibio-peroneal trunk and the anterior tibial artery was revascularized by using end-to-side anastomosis (Figure 2 B). At the end of the procedure the limb was well vascularized and distal pulses were present and normal.

The postoperative course was uneventful and free of complications. Histological examination of the surgical specimen confirmed the diagnosis of CAD.

The patient started walking on the third postoperative day and he was discharged on the fifth, treated with ASA 100 mg 1 tablet/day.

One year after surgery, the patient is pain free and presenting a good running autonomy. The limb is warm, well perfused with normal pulses.

## Discussion

The etiology of CAD remains unclear. Nowadays four hypotheses have been proposed: microtraumatic, ganglionic, degenerative and developmental. The microtraumatic theory presumes that repetitive traumas, due to the movements of the adjacent joints, cause small detachments of the adventitia from the media and induce intramural bleeding, generating cystic formation because of enzymatic activity (16). The ganglionic theory suggests that adventitial cysts are ectopic synovial ganglions migrated along the vascular branches from an adjacent tendon or joint capsule to the adventitia (17). The degenerative theory suggests a myxomatous systemic degeneration related to a generalized disorder (18). The developmental theory is the most widely accepted and presumes that some undifferentiated mesenchymal cells destined to form the joint are included in the adventitia of the adjacent blood vessel during its development. These cells secrete mucin and form the adventitial cyst later in life (19).

The diagnosis is based on the medical history and the imaging examinations. The typical clinical manifestation is the sudden onset of cramps in the calf, which usually occur in young males with average age of 36 years. The disease can be suspected when a relatively young patient

presents intermittent claudication without evidence of atherosclerosis elsewhere.

Usually, posterior tibial and *dorsalis pedis* pulses are reduced on the limb involved by CAD. Rarely CAD presents as acute limb pain due to bleeding in the cyst or its rupture, alternatively as acute limb ischaemia due to thrombosis of the affected vessel. In some patients passive flexion of the knee, or after exercise, may induce the loss of the *dorsalis pedis* pulses (Ishikawa sign) (20), subsequent to the compression of the popliteal artery by large adventitial cyst.

Diagnostic imaging techniques have improved over the last decades, obtaining more information concerning vascular assessment and have also aided in the management. Nowadays most accessible and detailed techniques are ultrasound (US), CTA, MR-angiography (MRA), DSA.

Standard US can be a useful non-invasive, readily available and cheap screening procedure. Most adventitial cysts appear hypo- or anechoic masses adjacent to the affected vessel. Colordoppler US is slightly more powerful and may show arterial stenosis or occlusion due to the dim or absent blood signal.

CTA is very useful in the preoperative assessment, because it is essential to depict vessel wall and the anatomical surrounding structures. 3D reconstructions can be used to define the aspect and cystic nature of the lesion (21).

MRA has improved and has gained popularity in the last decades, giving reproducible results. It allows demonstrating connections between adventitial cysts and adjacent joint capsule (22). The cyst could not enhance with gadolinium or be suppressed in the fat-saturated sequences. MRA could be much more useful in case of venous CAD.

DSA is commonly performed with limb extension, it does not provide details about adventitial cysts themselves and it could be diagnostic only about typical shaped stenosis ("hourglass appearance" and "scimitar sign") or occlusion caused by the cyst (23).

All articles pertaining to CAD published between 1979 and August 2013 were searched via PUBMED databases, references of the key articles were reviewed. We have supplemented an initial review made by Flanigan (23) in 1978, which collected all the cases since 1953 to 1977 (Table 1).

From the review of the literature we found that multiple treatment options are reported and divided into resectional and non-resectional approaches but surgery with complete removal of the disease is considered the gold standard therapy (23). Some cases of conservative treatment were described (24-26). The group of non-resectional procedures consists of open cyst evacuation including the removal of the cystic wall (7, 27), intraoperative cyst aspiration and interventional radiological tech-

TABLE 1 - SUMMARY OF TOTAL PROCEDURES (INCLUDING FLANIGAN'S REVIEW).

	Total Cases	Cases on follow-up	Mean Time Follow-up (months)
Non-resectional	113	95	18
Resectional	97	67	29

TABLE 2 - SUMMARY OF TOTAL NON-RESECTIONAL PROCEDURES (INCLUDING FLANIGAN'S REVIEW).

Non-Resectional Techniques	Cyst evacuation	Cyst excision	Cyst aspiration	Exarterectomy	Needle aspiration	Percutaneous techniques	Conservative	Others
Cases treated	56	21	6	11	9	5	3	2
Cases Failed	7	3	-	-	1	-	-	-

TABLE 3 - SUMMARY OF TOTAL RESECTIONAL PROCEDURES (INCLUDING FLANIGAN'S REVIEW).

Resectional Techniques	Venous graft	Prosthetic graft	Cryo-preserved homograft	Bypass	End-to-end anastomosis	Others
Cases Treated	69	14	2	4	4	4
Cases Failed	5	2	-	-	-	-

niques, such as percutaneous US-guided (28) or CT-guided aspiration, percutaneous transluminal angioplasty (PTA). Among non-resectional procedures Authors consider cyst evacuation with cyst wall removal as the best technique (29). Non-resectional approaches are indicated in selected cases of arterial CAD, in which is present a stenosis of the involved vessel. Percutaneous techniques, instead of an initial enthusiasm, seem to be inadequate because of extraluminal nature of the disease and the absence of atherosclerosis of the vessels.

Resectional approach consists in the resection of involved vessel and its replacement by use of vein, prosthetic graft or homograft. It is indicated in case of occlusive and near-occlusive disease (23) or in case of media degeneration. This technique consists in the resection of the involved vessel with the complete removal of the cyst. Most Authors use vein reconstruction to restore the arterial segment, if an adequate vein is available. Data demonstrate an initial success of 95% with a failure due to graft thrombosis. However CAD of a vein graft is also possible, probably due to incomplete resection of the cyst (30). The resection of the affected segment or excision of the cyst has been preferred in case of venous CAD (12, 31). Prosthetic graft and homograft showed a good initial success rate ( $\geq 90\%$ ) (32), probably because CAD is characterized by an isolated lesion with a normal inflow and outflow (23).

In the literature review, we collected 210 cases of popliteal artery CAD since the first case described in 1953

(2). These cases are divided in 113 cases treated by non-resectional approaches (Table 2) and 97 cases treated by resectional approaches (Table 3) with, respectively, 95 and 67 cases described on follow up with a mean follow-up of 18 months for the first group and 29 months for the second (Chi Square Test,  $p=.014$ ) (Table 1). We found 11 cases of failure in the non-resectional group ( $11/95 = 11,6\%$ ) and 7 cases of failure for the resectional group ( $7/67 = 10,4\%$ ) ( $p=.8$ ).

## Conclusions

CAD is a rare vascular steno-occlusive disease, usually affecting popliteal artery. Typically it affects young men with minimal cardiovascular risk factors, presenting a short history of progressive claudication. Imaging is based on US and CTA. Suspected diagnosis is confirmed at time of surgery. Percutaneous approaches are adequate in selected cases. Moreover, in our opinion, the treatment of choice for CAD should be the replacement of the compromised arterial segment with autologous grafts that avoids local recurrence and permits long-term patency.

## Conflict of interest

The Authors declare that there is no conflict of interest.

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