

Critical limb ischemia in a young patient with cystic disease of the popliteal artery

Isquemia crítica em membro inferior em paciente jovem com doença cística de artéria poplítea

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Abstract

Cystic adventitial disease of the popliteal artery is a rare vascular disease of unknown etiology in which a mucin-containing cyst may compromise the artery and, because of the compression, develops stenosis or occlusion of affected artery. We report the case of a 45-year-old male with cystic adventitial disease of the left popliteal artery, after admission for an acute limb ischemia in his left leg, diagnosed non-invasively with EcoDoppler vascular dual scan, magnetic resonance angiography, angiography. Performed complete removal of the cyst with arteriectomy and venous replacement. The pathologic result were consistent with the diagnostic of cystic adventitial disease. The patient is in ambulatory follow up and was affected by cystic disease in the contralateral limb. The authors described the best methods of diagnosis and treatment for cystic adventitial disease, discussing if the urgency procedure was the most appropriate and whether there was an alternative treatment for the case. Finally, questioning what would be the best procedure to be performed in the other limb.

Keywords: intermittent claudication; cyst; popliteal artery.

Resumo

Degeneração cística da artéria poplítea é uma doença vascular rara, de etiologia desconhecida, na qual um cisto com conteúdo mucinoide compromete a artéria e desenvolve estenose ou oclusão do vaso. Apresentamos um caso de um paciente do sexo masculino, 45 anos, admitido na emergência hospitalar, que apresentava quadro de isquemia aguda no membro inferior esquerdo. Submetido à EcoDoppler, angiorressonância magnética e angiografia pré-operatória. Realizada arterectomia com enxerto fêmoro-poplíteo infrainguinal de safena invertida. O exame patológico foi consistente com o diagnóstico de doença cística da artéria poplítea. O paciente encontra-se em acompanhamento ambulatorial e foi acometido pela doença cística no membro contralateral. Os autores descreveram os melhores métodos de diagnóstico e tratamento para a doença, discutindo se o procedimento de urgência foi o mais adequado e se haveria opções terapêuticas alternativas para o caso, além de qual seria a melhor conduta a ser realizada no membro contralateral também acometido pela doença.

Palavras-chave: claudicação intermitente; cisto; artéria poplítea.

PART I

Case description

The patient is a 45-year-old male physician, non-diabetic, non hypertensive, non-smoker and athlete, who was admitted to the Emergency Room complaining of severe pain on the left lower limb for 4 hours, decreased temperature, absence of left popliteal and distal pulse, accompanied by decreased sensitivity in the left foot, but

with the motor function preserved. The patient reported bilateral calf intermittent claudication that had began one year earlier and had worsened to approximately 120 m in the last days.

Ankle-brachial index (ABI) measurements showed an, ABI=0.40 at the left and ABI=0.85 at the right lower limb. Color Doppler ultrasound showed a thrombosed left popliteal artery aneurysm and approximately 50% stenosis of the right popliteal artery (Figures 1 and 2). The physician performing the examination raised the suspicion

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of periarterial cyst. Color Doppler imaging showed stenosis and external compression – possibly a cyst – of the right popliteal artery (Figure 3). Knee flexion maneuvers were performed bilaterally, with absence of bilateral distal

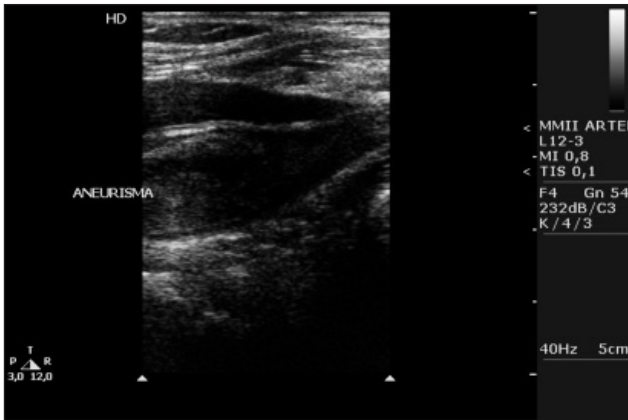


Figure 1. B-mode echography showing thrombosed aneurysm at the left.



Figure 2. B-mode echography showing thrombosed aneurysm at the left. Image indicating adequately the existence of a thrombus.

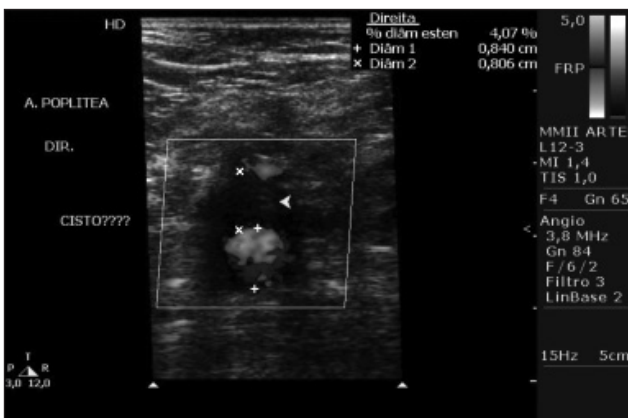


Figure 3. 50% stenosis of the right popliteal artery, suspected periarterial cyst.

pulses. Color Doppler flow showed alterations at maneuvers of hyperextension of the feet.

MR angiography showed left arterial occlusion and no signs of right arterial occlusion. The patient also underwent preoperative catheter angiography (Figure 4), and then was sent to the operating room.

PART II

With acute arterial ischemia of the left lower limb for 6 hours, the patient underwent surgical repair, and tissue induration was observed on the popliteal artery wall, thus occluding the vessel lumen. Tissue samples were collected and sent for analysis.

The patient underwent popliteal artery resection and replacement by a femoropopliteal infragenicular bypass graft with reversed saphenous vein, with good results in the immediate postoperative period. Anatomopathological examination showed the adventitial layer with thickening of the intima in the same proportion and cystic spaces (Figure 5). Multinucleated giant cells, neutrophils, eosinophils, and crystals were not detected, thus excluding other forms of arteritis (Figure 6). The findings were suggestive of cystic disease of the popliteal artery.

Thirty days after operation, the patient presented recurrent claudication for a distance of 70 m on the right lower limb. Ninety days later, the claudication distance was 120 m, so the patient was given vasodilator and antiplatelet treatment. At the day 180, claudication distance was 150 m,

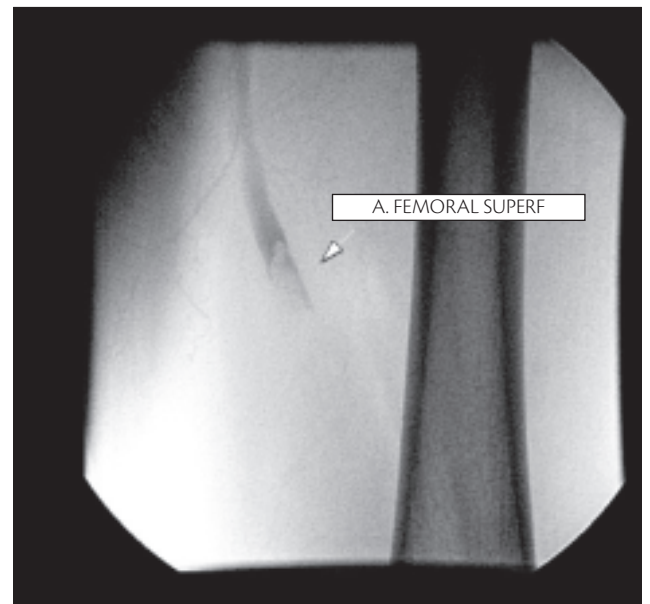


Figure 4. Preoperative angiography.

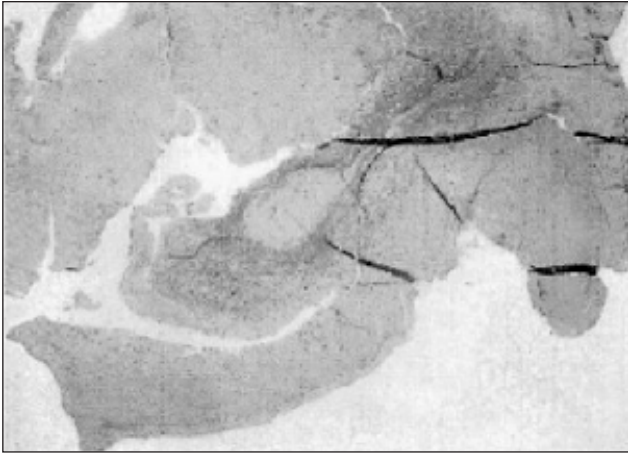


Figure 5. Anatomopathological study (mucicarmim) showing the adventitious layer with tickneing of the intima in the same proportion and few cystic spaces.

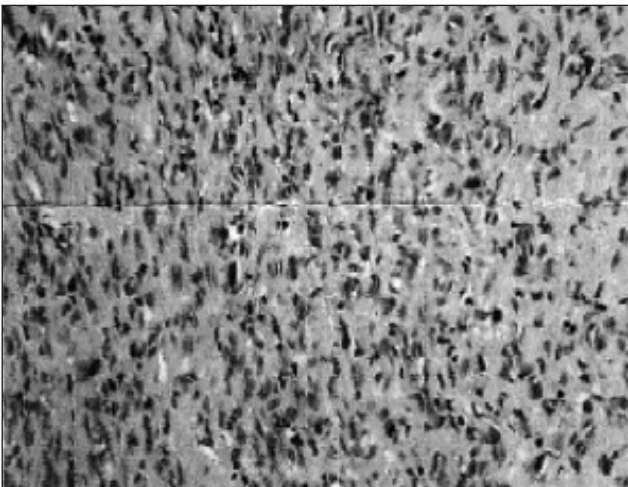


Figure 6. Hematoxylin-eosin staining showing no multinucleated giant cells, neutrophils, eosinophils, and crystals, differing from other arteritis.

so angiography (Figure 7) and CT angiography (Figures 8 and 9) imaging were performed, showing cystic disease affecting the right lower limb.

Was the initial procedure adequate?

Do we have alternative treatment options for this case?

What would be best to perform in the contralateral limb that is also affected by the disease?

Discussion

Arterial cystic disease (ACD) is characterized by the presence of cysts, often multiple, located in the arterial wall. The cysts, which have a gelatinous, viscous content, may compress and distort the adventitia and tunica media,



Figure 7. Angiography of the contralateral limb showing cistos multi-loculados.

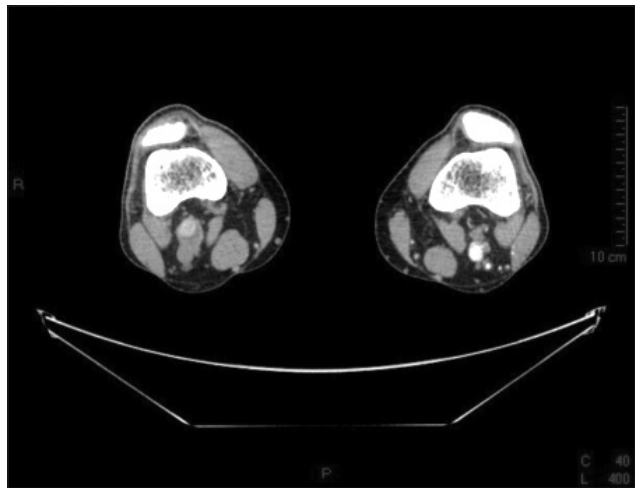


Figure 8. CT Angiography – Cross-sectional view of cysts.

Increased pressure within the cysts can also cause stenosis and occlusion of the affected artery. It is also known as cystic degeneration of adventitia¹, cystic disease of the adventitia^{2,3}, adventitia colloid cyst⁴, mucinous cystic disease⁵, adventitial cyst of the popliteal artery^{6,7}, and other designations⁸⁻¹⁰. Both cystic degeneration of the adventitia and adventitial cyst disease are restricted designations, for they do not encompass all cases, because cysts may be located on the outer layers of the media⁴. More comprehensive names, such as cystic disease, or degeneration of the arterial wall,



Figure 9. CT Angiography – Sagittal view of cysts.

or even degeneration or cystic disease of the popliteal artery, are more appropriate^{11,12}.

The pathogenesis of the disease remains unknown, but four theories have tried to explain it: microtrauma¹³; systemic disease⁴; embryonic origin¹⁴; true synovial cysts^{1,12,15,16}.

The incidence of cystic disease of the popliteal artery is 1 case per 1,200 cases of intermittent claudication, or 1 case in 1,000 femoral angiographies performed¹⁴. This disease affects men more frequently, at a ratio men:women of 4:1¹⁷, and is more commonly observed in individuals in the fourth to the sixth decades of life.

Maffei et al.¹⁸ suggest that non-recognition and lack of report may be the causes of low incidence of this disease, and that the number cases diagnosed is likely to increase with increasing levels of suspicion.

Thus, in younger adults with intermittent claudication and with few or no risk factors for atherosclerosis, one should consider the following differential diagnosis: popliteal artery entrapment syndrome (PAES), cystic adventitial disease (CAD), fibromuscular dysplasia, the external Iliac Artery Endofibrosis (common among cyclists), arteritis, arterial embolism, and pseudoxanthoma elasticum (PXE)-related arterial occlusive lesions.

Currently, there are several imaging methods for diagnosis and surgical treatment planning. It is essential to define the number of cysts, their location, extent and situation. A Doppler ultrasound imaging may be sufficient for diagnosis¹⁹, but a proper surgical planning requires other imaging methods.

Computed tomography (CT) scan²⁰ is still used for diagnosis, but there are better methods such as magnetic

resonance imaging (MRI) or angiography (MRA)²¹. Helical CT angiography associated with three-dimensional reconstruction seems to provide information similar MR or MRA, but it requires contrast injection²².

Maged et al.²³ recently showed good results using High Spatial Resolution Magnetic Resonance Imaging (MR-HSR). They identified connections between the cysts and the adjacent joint, which helped in the ligation of these connections during surgery, avoiding recurrence of the cysts in some cases.

Ortiz et al.²⁴ reported that different therapeutic approaches result in variable recurrence rates. The resection of the affected artery with a vein graft is indicated in cases of complete arterial occlusion and weakening of the arterial wall, or in cases of secondary intimal ulceration.

Some authors^{25,26} show that cyst resection with saphenous vein graft interposition is followed by a recurrence rate of about 5%, and that cyst draining, even with its removal from the wall, is followed by a recurrence rate of 6%. On the other hand, percutaneous aspiration of cyst alone presents a recurrence rate of 34%. Some other methods are rarely used due to high recurrence rates, including endovascular treatment with angioplasty or endovascular stent placement, which have been reported with a recurrence rate of up to 100%.

Maged et al.²⁷ described a case of recurrent popliteal artery cystic disease after surgical enucleation of the cyst. This case was successfully treated with angioplasty, and symptoms were solved in a 24-month follow-up.

Do et al.²⁶ reported the successful treatment of 7 patients (6 men) who had symptomatic cystic adventitial disease (CAD) of the popliteal artery without occlusion by percutaneous image-guided cyst aspiration. In their series, no patient developed complications, and the procedure was technically and clinically successful in all cases. Duplex ultrasonography follow-up lasted 1 to 32 months (mean 14.8 months), and, after the procedure, no patient developed significant stenosis. However, the authors acknowledge that their successful results with the percutaneous aspiration technique have not been reproduced in the literature and that this technique has not gained much recognition worldwide^{24,28}.

Conclusions

Cystic disease of the popliteal artery is a rare entity, but physicians should be attentive to its diagnosis. All young patients complaining of intermittent claudication with no risk factors for atherosclerosis should be managed with the hypothesis of cystic disease of the popliteal artery. Diagnosis must be initially performed using non-invasive methods,

such as color Doppler ultrasound. Bilateral knee flexion maneuvers (Ishikawa), as well as feet hyperextension maneuvers should be part of the clinical examination.

Three-dimensional helical CT scan appears to provide images comparable to those of MRI or MRA, but it requires contrast injection. If possible, high spatial resolution MRI should be used. Although there are several alternative diagnosis methods, one must never forget that angiography is the gold standard before revascularization and it should be, whenever necessary.

Various therapeutic methods can be used for the treatment of popliteal artery cystic disease. Endovascular treatment with angioplasty or intravascular stent placement is not a good choice in such cases, because this disease affects arteries in areas of flexion, and the published data of Ortiz et al.²⁴ showed an occlusion rate of almost 100%.

Image-guided cyst aspiration is a less invasive technique, but it was successful only in the hands of Do et al. Maged et al.²³ and even Do et al.²⁶, in a literature review, showed a recurrence rate close to 34%.

Most studies addressing cyst resection with homologous graft implantation^{20,23,24} and cyst draining with its complete removal from the arterial wall, are the treatments of choice, since they present similar recurrence rates. However, the reports by Melliere et al.²⁹ suggest that the technique of artery resection and homograft implantation provides better short and long-term results, compared to cyst draining alone, even when it is removed from the arterial wall.

Therefore, surgical resection and homograft implantation are the best treatment choices for the popliteal artery cystic disease. Regarding endovascular treatment, it needs need further systematic studies before being indicated; however, it could be considered an adjunctive therapy option in cases of recurrent stenosis after cyst resection and graft placement.

In the current case, since the patient had no ischemia of the right lower limb, we chose to manage him the contralateral limb conservatively, with prescription of exercises aiming at the strengthening of the muscles, use of peripheral vasodilator and anti-platelet aggregation agent and clinical follow-up. In case of ischemia in the future, the best approach would be resection of the affected arterial segment, with autologous vein graft interposition.

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