Common femoral adventitial cystic disease in a young female patient

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ABSTRACT

Adventitial cystic disease (ACD) is a rare condition that typically presents in young healthy men with symptoms of claudication. ACD is characterized by formation of a mucinous cyst within the adventitia of a blood vessel, usually in the popliteal artery, causing compression of the vessel's lumen and leading to reduced flow and symptoms of claudication. We have presented a rare case of ACD of the common femoral artery in a young female patient that was treated successfully with resection and femoral vein graft interposition reconstruction. (J Vasc Surg Cases Innov Tech 2023;9:1-3.)

Keywords: Adventitial cystic disease; Claudication; Femoral artery

Adventitial cystic disease (ACD) is a rare condition that typically presents in young to middle-age men with symptoms of claudication and is often misdiagnosed as atherosclerotic disease.[1,2] The pathophysiology of ACD is characterized by the formation of a mucinous cyst within the adventitia of an artery or vein. The cyst causes swelling within the vessel wall, eventually leading to compression of the vessel's lumen, reduction of flow, and symptoms of claudication.[3] Since the first reported case of ACD in 1947, >700 cases have been reported.[4,5] The popliteal artery was the most common site for cyst formation (90% of cases), followed by the femoral artery (6%), radial artery (3%), and external iliac artery and axillary artery (1%).[6]

ACD should be suspected in patients with intermittent claudication, who do not have any cardiovascular risk factors.[7] The presenting symptoms will mostly depend on the arterial site affected. When the popliteal artery is affected, patients might experience exacerbated symptoms with full flexion of the knee and quick resolution of the symptoms after knee extension.[1] In contrast to atherosclerotic claudication, patients with ACD can experience complete resolution of symptoms, which will usually recur later, with rapid progression at times, depending on cyst fluid production.[8] We have presented the case of a patient with common femoral ACD. She was successfully treated with open vascular reconstruction surgery. Our patient provided written informed consent for the report of her case details and associated imaging studies.

CASE REPORT

A 46-year-old, otherwise healthy, woman with no history of tobacco use had presented to our outpatient vascular clinic with left leg intermittent claudication that had manifested as calf pain after a walking distance of ~200 m. Her symptoms had gradually developed over 1 year, with no history of trauma or strenuous activity.

On physical examination, normal palpable pulses were present in the right leg. Although the left popliteal and pedal pulses were palpable, they were diminished compared with the right pedal and popliteal pulses. The ankle brachial index was normal at rest bilaterally but decreased after exercise on the left side to 0.55, although remaining normal on the right. Duplex ultrasound demonstrated high-grade left common femoral artery (CFA) stenosis. Computed tomography angiography confirmed the presence of high-grade stenosis, with dilation of the left CFA to 1.4 cm. The right CFA and both popliteal arteries were normal (Figs 1 and 2). Although ACD was suspected at that time, the definitive diagnosis remained undetermined, and the differential diagnosis included other etiologies such as an aneurysm with mural thrombus.

Considering the severity of her symptoms and given her acceptable surgical risk, we offered the patient open surgical repair. We chose resection of the affected segment to minimize the risk of recurrence, and an autogenous vein graft was used to reduce the risk of infection. Because the saphenous veins were inadequate for grafting, the ipsilateral femoral vein was chosen as a conduit for arterial reconstruction and revascularization.

The CFA was exposed via a longitudinal incision. A circumferential arterial wall mass extending to the CFA bifurcation was found (Fig 3). Arteriotomy exposed the stenotic segment of the vessel's lumen. The stenosis appeared to be secondary to compression by the cystic structure within the arterial wall. Dissection of the cyst revealed a gelatinous material that was sent to pathology for examination (Fig 4). The affected 5-cm

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Author conflict of interest: none.

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2468-4287

https://doi.org/10.1016/j.jvscit.2022.11.008

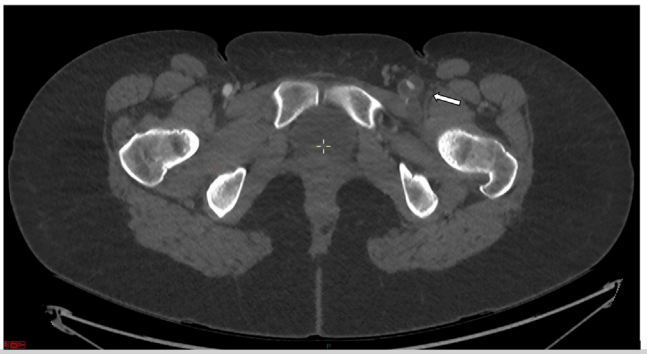

Fig 1. Computed tomography angiogram demonstrating high-grade stenosis of the left common femoral artery (CFA).

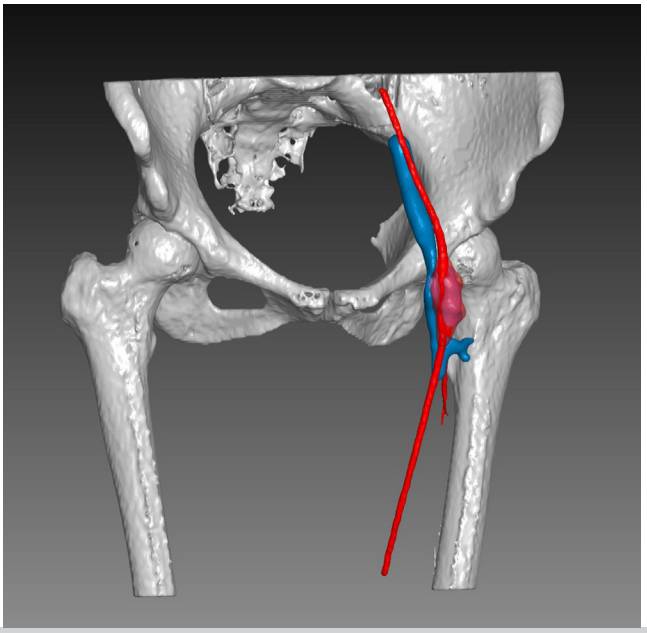

Fig 2. Three-dimensional reconstruction of common femoral adventitial cyst causing severe arterial stenosis.

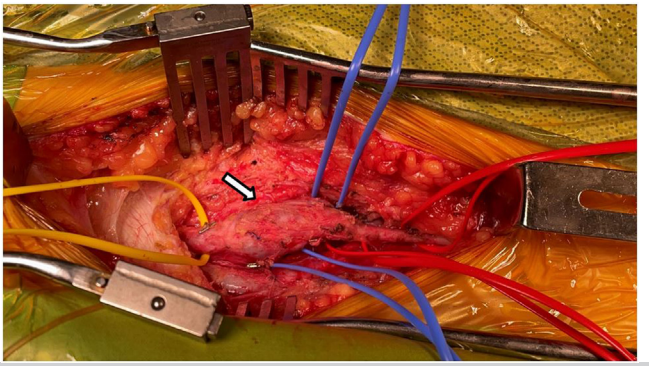

Fig 3. Exposed adventitial cyst in common femoral artery (CFA; *arrow*).

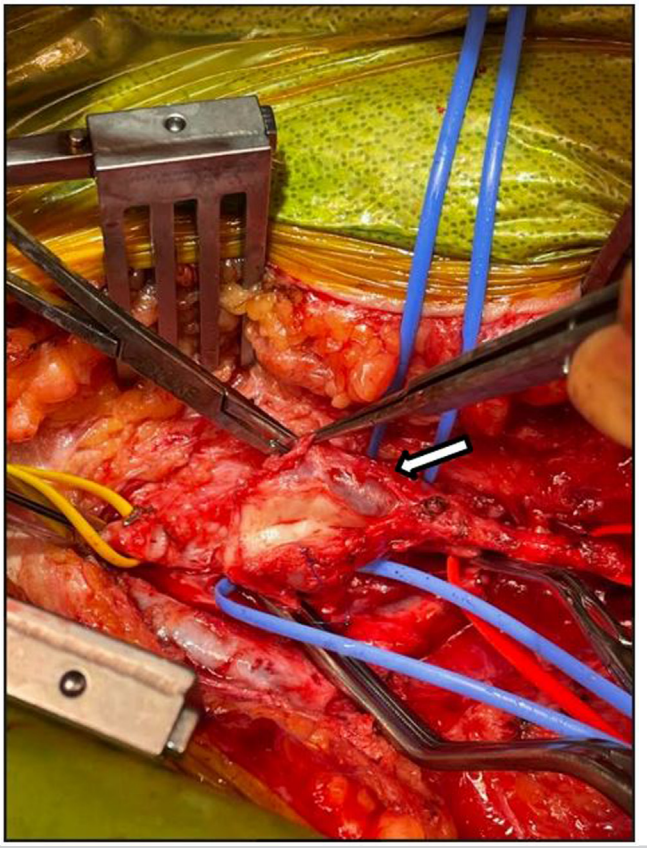

Fig 4. Gelatinous content of adventitial cyst causing severe stenosis (*arrow*).

segment of the CFA was resected, followed by reconstruction with an interposition vein graft. A 6-cm segment of the femoral vein from the proximal thigh was harvested with its branches and fashioned into a reversed "Y"-shaped graft using a large tributary. The reversed vein was anastomosed directly to the origin of the superficial femoral artery, and the tributary was anastomosed directly to the origin of the deep femoral artery. Surgery was performed with no significant blood loss, and no transfusions were needed.

Her postoperative recovery was satisfactory. Deep vein thrombosis (DVT) prophylaxis, including leg elevation, elastic compression therapy, and enoxaparin 40 mg daily, was started. On postoperative day 1, the patient was ambulatory and began double antiplatelet therapy with aspirin and clopidogrel. Her pedal pulses were palpated bilaterally, and an arterial Doppler ultrasound demonstrated normal multiphasic signals. Swelling of the left leg appeared on the fifth postoperative day, and DVT extending from the tibial veins to the proximal femoral vein was diagnosed via duplex ultrasound. The patient's medications were switched to enoxaparin 60 mg twice daily and single antiplatelet (aspirin), and the symptoms had fully resolved by 3 months.

Pathologic examination of the excised tissue showed cystic formation between the media and adventitia of the artery. These findings were confirmed by hematoxylin and eosin and

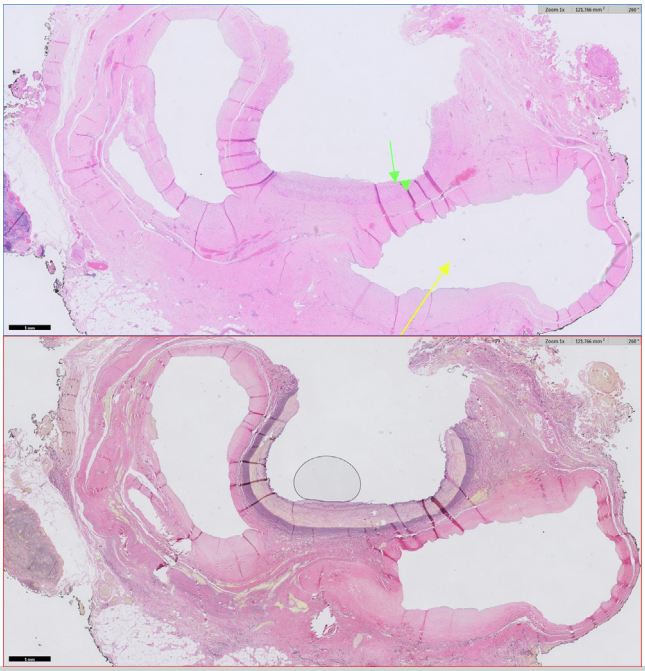

Fig 5. Hematoxylin and eosin and elastic Van Gieson stains. *Green arrows* indicate tunica intima; and *yellow arrow,* the adventitial cyst.

elastic Van Gieson staining and were compatible with ACD (Fig 5).

Serial duplex ultrasound examinations performed 3 and 6 months after discharge demonstrated normal arterial blood flow bilaterally, and the ankle brachial index was normal at rest and after exercise bilaterally. The patient had no complaints of arterial or venous claudication and no lower extremity edema.

DISCUSSION

Claudication complaints in healthy young patients with no cardiovascular risk factors merit consideration of non-atheromatous arterial disease. The differential diagnosis will usually include rare conditions such as popliteal entrapment syndrome, iliac artery endofibrosis, ACD, chronic compartment syndrome, fibromuscular dysplasia, and others.[3] ACD is rarely encountered in patients with intermittent claudication. It occurs mainly in men, with a male/female ratio of 4:1,[1] and usually involves the popliteal artery.[9] AVD was not first in our differential diagnosis list. However, the radiologic appearance on computed tomography angiography and the lack of atherosclerosis risk factors raised a high index of suspicion for femoral artery ACD, which was confirmed by the pathologic examination findings. Although the hypodense lesion within the arterial wall was suggestive of a cyst, the diagnosis can only be suspected from the imaging studies and must be confirmed by pathologic examination.

The natural history of untreated ACD lesions remains unknown, and the optimal treatment has continued to be debated. Treatment is indicated in the presence of symptoms or high-grade stenoses that could potentially progress to thrombosis and embolization. The options for ACD repair include angioplasty, stenting, open or imaging-guided percutaneous aspiration, and cyst resection with or without vascular reconstruction.[1] Our patient was a young and healthy woman. Therefore, we offered her open surgical excision with vascular reconstruction. In our previous experience, femoral vein harvesting for vascular reconstruction has had low rates of complications. In the present patient, the DVT had probably resulted from inadequate prophylaxis and, luckily, had resolved without additional morbidity.

CONCLUSIONS

ACD remains a rare diagnosis, especially in women, but should be suspected in relatively young patients who present with claudication. It is a curative disease with adequate surgical treatment, which will usually include some form of excision, with excellent results.

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Submitted Aug 27, 2022; accepted Nov 10, 2022.