Cystic adventitial disease of the popliteal vein and artery in siblings

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ABSTRACT

Cystic adventitial disease (CAD) is an uncommon condition in which mucoid cysts form within the adventitial layer of the arterial or venous wall. We have presented two cases in which two first-degree relatives (brother and sister) had acquired CAD ~6 years apart, one involving the popliteal artery and the other involving the popliteal vein. We have reported a rare case of a possible familial association of CAD. We have discussed the etiology, diagnostic criteria, and imaging modalities between arterial and venous CAD to aid in the management and selection of optimal treatment strategies. (J Vasc Surg Cases and Innovative Techniques 2021;7:545-8.)

Keywords: Cystic adventitial disease; Popliteal artery; Popliteal vein; Siblings; Vascular diseases

Cystic adventitial disease (CAD) is a rare disease caused by mucinous cystic formation in the adventitia of the vessel.[1-4] Arterial CAD (aCAD) most frequently involves the popliteal artery, and venous CAD (vCAD) most frequently affects the popliteal vein.[1,3,4] aCAD has been more frequently reported, with <50 cases of vCAD described.[5] We have presented the case of two first-degree relatives (a brother and sister) diagnosed with aCAD and vCAD and discussed the etiology and treatment modalities. Both patients provided written informed consent for the report of their case details and imaging studies.

CASE REPORT

Patient 1. A 52-year-old man had presented at our clinic because of persistent right calf pain despite multiple interventions at an outside institution. The patient had been treated 6 months previously for acute thrombosis of his distal right popliteal artery, which had required open thrombectomy with a Fogarty catheter and four compartment fasciotomy of the calf. He had had a return of the pedal pulses at the end of the case. However, because of persistent claudication, he had undergone angiography, which showed complete occlusion of the right popliteal artery with reconstitution of the tibioperoneal trunk and anterior tibial artery. Thus, only percutaneous balloon angioplasty (PTA) was performed. On physical examination at our clinic, the right posterior tibial or dorsalis pedis arteries were not palpable. No masses were noted in either popliteal fossa. A review of his angiograms showed external compression of the popliteal artery, which had resulted in significant curvilinear endoluminal stenosis with normal lumina of the proximal and distal arteries (scimitar sign; Fig 1). Duplex ultrasound (US)

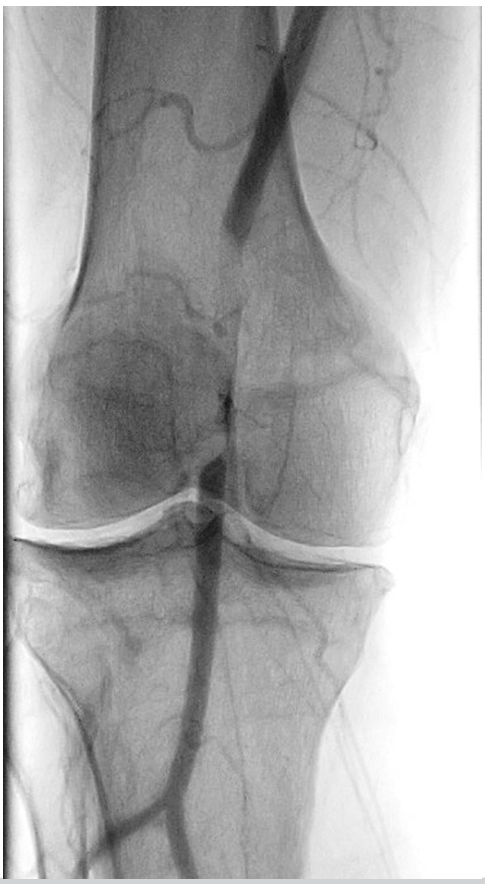

Fig 1. Conventional angiogram demonstrating hourglass-shaped subtotal occlusion from extrinsic compression in the popliteal artery (scimitar sign).

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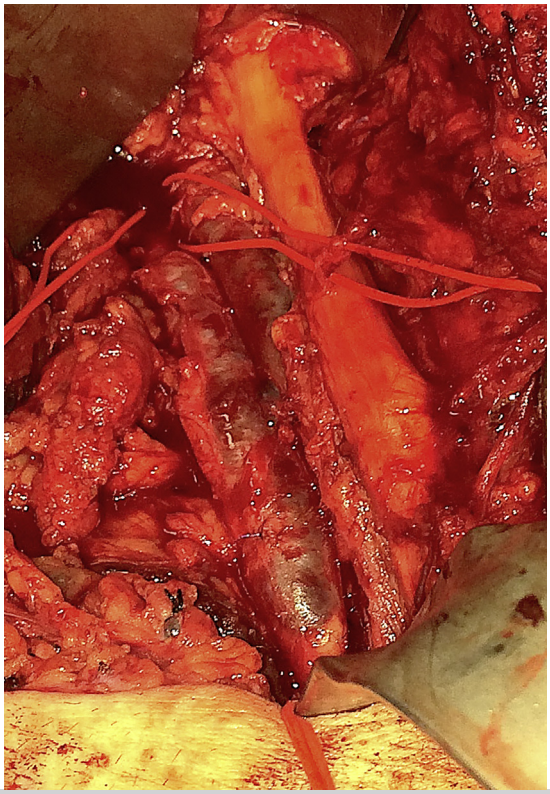

Fig 2. The whole extent of the long cyst surrounding the popliteal artery.

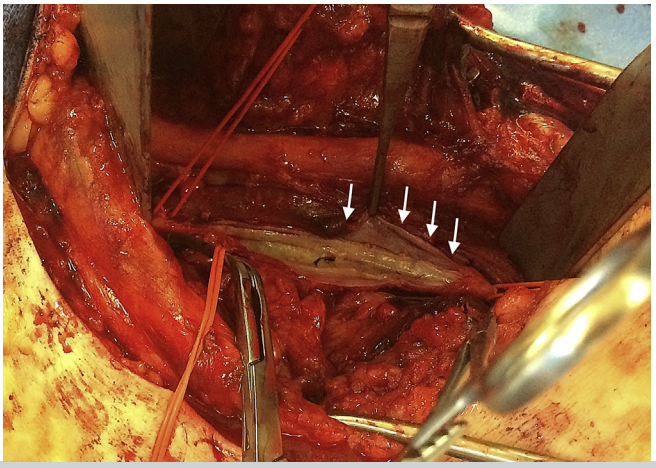

Fig 3. On opening the affected popliteal segment, the cyst wall (*white arrows*) could be seen completely encasing the artery and causing significant stenosis.

showed a cystic structure occluding the popliteal artery above the knee with reconstitution of the flow into the distal aspect. The ankle brachial index of the right leg measured 0.69 compared with 1.27 for the left leg. A presumptive diagnosis of CAD was made at this time, and the patient was taken to the operating room for definitive diagnosis and management. A posterior surgical approach was taken to expose the popliteal vein and artery both (Fig 2). No intra-arterial thrombus was found; however, the arterial wall was notably narrowed with a cystic appearance, resulting in moderate stenosis (Fig 3). The adventitial cyst with the affected artery was completely excised. In situ vascular reconstruction was performed using a great saphenous vein graft. Immediately after the operation, the popliteal and distal pedal pulses were palpable. Histopathologic examination showed a mural myxoid degeneration with focal acute inflammation in the adventitia with mucoid material within the cyst (Fig 4). Follow-up US at 2 weeks, 6 months, and 1 year demonstrated a patent popliteal graft with good perfusion of flow. He had an ankle brachial index of 1.2 bilaterally at 3 years of follow-up. Doppler US did not show any stenosis of the right popliteal artery or a recurring cyst.

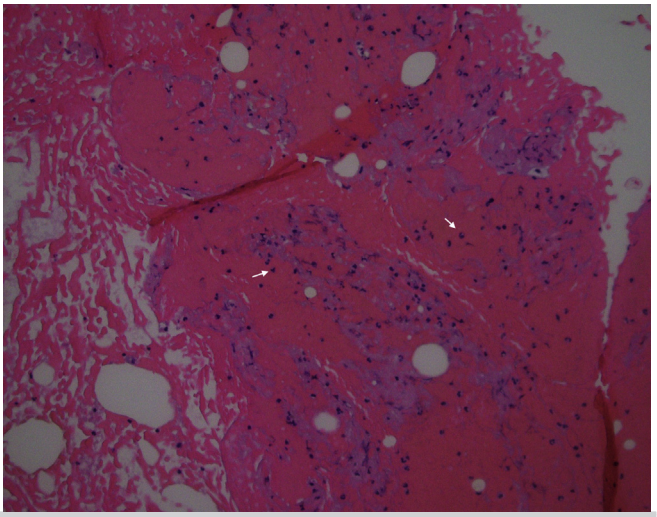

Fig 4. Pathologic examination of the surgical specimen revealed intramural cysts filled with mucoid material and inflammatory cells (*white arrows*; hematoxylin and eosin stain, original magnification ×20).

Patient 2. The 49-year-old female sibling of patient 1 had been referred for evaluation of a 3-month history of right lateral foot and heel numbness. The patient reported that she also had experienced claudication for 1 year with walking, a feeling of fullness in her right knee, and nocturnal cramps. She had initially been seen in the emergency department at an outside institution because of the rapid onset of right lower extremity edema and had undergone right leg venous duplex US, which showed a possible deep vein aneurysm. Magnetic resonance imaging of the right knee showed a multiloculated mass in the popliteal fossa, which the radiologist presumed was a sarcoma from the appearance (Fig 5). Because of its close proximity to the popliteal vessels, the patient was referred to vascular surgery. On physical examination, she had had palpable right posterior tibial and dorsalis pedis arteries but decreased sensation in her right heel. No masses were felt in the popliteal fossa. Arterial duplex US showed evidence of a cystic mass that measured 1.47 × 1.34 cm and involved the right popliteal artery with compression of the vein. Venous duplex US did not show evidence of deep vein thrombosis. In the operating room, a posterior approach was used. Further dissection of the area revealed

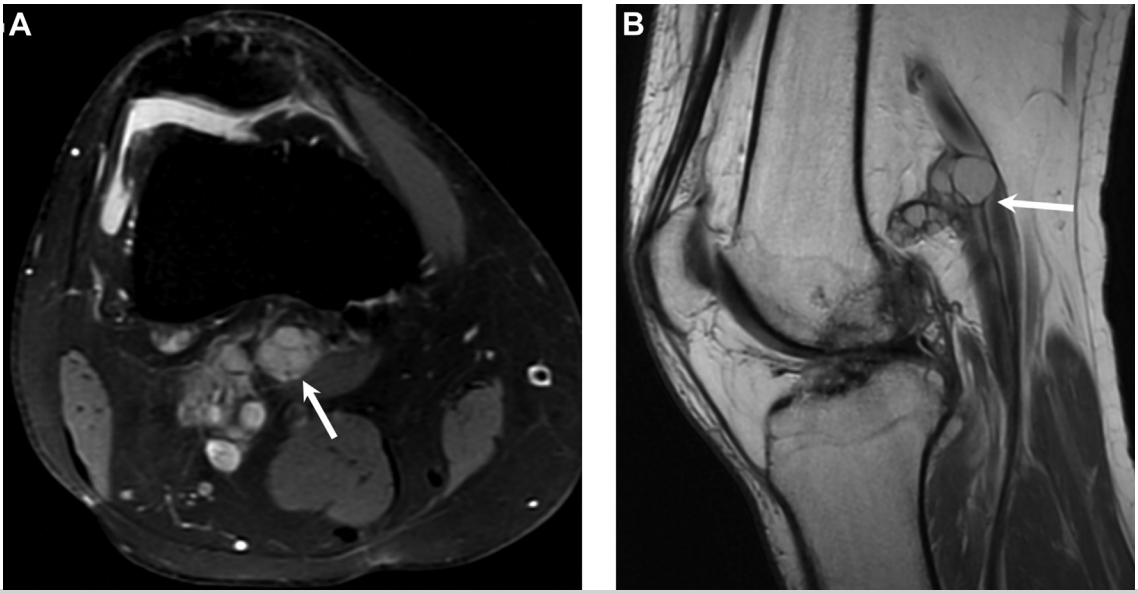

Fig 5. Preoperative magnetic resonance images showing cystic mass (*white arrow*) adjacent to the popliteal artery. **A,** T1-weighted image. **B,** T2-weighted image.

that the cystic lesion had involved the popliteal vein rather than the popliteal artery. On entry into the cyst, communication was found between the compressed venous lumen and cyst, which contained a clear jelly-like substance. The cyst was excised, and the posterior connection was sutured closed. Patch venoplasty of the popliteal vein was completed with an autologous great saphenous vein graft to prevent luminal narrowing. Completion venography showed patent flow through the popliteal vein. The postoperative course was uneventful, and the patient was free of claudication and paresthesia. Pathologic examination showed fragments of fibroconnective and fibrovascular tissue with degenerative changes. Follow-up US at 2 weeks and 6 months postoperatively showed a patent popliteal artery and vein without any hemodynamically significant stenosis.

DISCUSSION

CAD remains a rare condition that is difficult to treat because no unifying consensus has been reached regarding the etiology. Most cases have involved the arterial system, with CAD involving the venous system a rarer entity, with <50 cases reported since 1963.[5] vCAD has most frequently affected the common femoral vein and less commonly the external iliac and popliteal veins.[2,5] Although no single hypothesis has been accepted, four theories have been proposed regarding the etiology of CAD. One theory is the developmental theory in which vessels adjacent to joints have mesenchymal cells that are eccentrically implanted during embryogenesis and secrete mucinous fluid later in life.[1] Another theory proposes the occurrence of microtrauma, which can occur in healthy athletes involving repetitive stretch injuries, causing a separation of the adventitia from the media resulting in adventitial cystic degeneration.[6,7] The de novo systemic disorder theory also suggests degeneration of the adventitial layer and cyst formation due to a general connective tissue disorder. The articular or synovial theory proposes that ectopic synovial ganglions track along vascular branches from an adjacent joint capsule and eventually implant into the adventitia.[1,4,7,8] Both the developmental and the synovial theories have gained support by evidence of a joint connection found intraoperatively between the adventitia and adjacent joints in ≤66.7% of reported cases.[1,2,9] This finding supports that an anatomic connection between the cyst and joint must be excised for complete resolution of the disease.

If the cyst is small, it is possible to monitor for regression or treat it initially with aspiration to improve the symptoms. However, short-term follow-up is warranted owing to the high recurrence rates and the need for secondary intervention. The reference standard for recurrent CAD or patients with total occlusion and severe symptoms is surgery with vessel excision and placement of an interposition or a bypass graft. An attempt to identify and ligate any cyst–joint connections could reduce the potential for recurrence. Endovascular procedures have been associated with high recurrence rates and mixed results after PTA and stenting. Possible explanation for stent occlusion was thought to have been required because of the induction of damage to the healthy intima induced by the radial force from the self-expanding stents and PTA.[10] They are not recommended treatment modalities as demonstrated in this case report.

Our limited experience in a sibling pair is insufficient to support any etiology or pathophysiology for CAD described in our report. However, another report also described a young male with aCAD who had had first- and second-degree relatives with the disease.[11] Although

no evidence has been reported of a genetic predisposition to the disease, perhaps a high degree of suspicion is warranted when family members present with similar symptoms.

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