Adventitial cystic disease in the external iliac artery of a 29-year-old man

Jaineet S. Chhabra, MS,[a] Joyce J. Lu, MD,[b] *and* Justin M. Simmons, DO, RPVI,[b] *Huntington, WV; and Grand Rapids, Mich*

ABSTRACT

Adventitial cystic disease (ACD) is an uncommon condition that generally occurs at the popliteal artery but, rarely, can occur in the external iliac artery. To date, only eight cases of ACD occurring in the external iliac artery have been reported. We have reported the case of a 29-year-old man who had presented with new-onset claudication. Despite an extensive imaging workup, ACD was not confirmed until the gross intraoperative examination. We have reported our process of diagnosis and treatment in addition to that reported in previous studies to enhance the historical fund of knowledge for this rare pathology. (J Vasc Surg Cases and Innovative Techniques 2021;7:516-9.)

Keywords: Adventitial cystic disease; External iliac artery; Claudication; Vascular imaging; Popliteal artery

Adventitial cystic disease (ACD) was first documented involving the external iliac artery (EIA) in 1947.[1] However, ~85% of cases will involve the popliteal artery.[2] As of the most recent review in 2015, 746 ACD cysts had been reported in 741 patients.[3] ACD is characterized by unilocular or multilocular cyst formation within the vascular adventitia. Involvement of the medial or intimal layers is believed to be secondary.[4] ACD can occur in both arteries and veins. Arterial occlusion due to ACD can result in intermittent claudication. In contrast, in the venous form, swelling, tenderness, and pain can be common symptoms.[5,6] The incidence of ACD is four to five times more common in males and usually occurs in healthy, active patients aged 40 to 50 years but can affect those aged 11 to 70 years.[3,4,7] Patients will typically present without signs of atherosclerotic degeneration or previous cardiovascular risk factors.[4] Additional approaches to the diagnosis and treatment of ACD in the EIA include mass palpation in the iliac fossa, the detection of low-density wall thickening of the EIA with computed tomography (CT) angiography (CTA) of the lower limbs and confirmation with ultrasound before iliofemoral bypass, magnetic resonance imaging (MRI) confirmation of a cystic lesion, and/or CTA or direct angiography demonstrating a "scimitar" sign.[1,8,9] The patient provided written informed consent for the report of his case.

CASE REPORT

The patient was an otherwise healthy 29-year-old white man, weighing 96 kg with a body mass index of 31 kg/m^2. He had presented with progressive symptoms of right calf claudication and had reported a 1-week history of foot coolness and paresthesia with exertion before visiting the emergency department. On examination, he was noted to have diminished right-sided femoral and dorsalis pedis pulses and the absence of a posterior tibial artery Doppler ultrasound signal. He denied any tobacco or drug use, recent trauma, activities that involved intense hip hyperflexion, and a family history of connective tissue disorders.

Given the palpable pulse on examination in the emergency department and no equipment to perform exercise ankle brachial index (ABI), no ABI was performed. However, arterial duplex ultrasound revealed diminished velocities and abnormal waveforms within all infrainguinal arteries of the right lower extremity and a 4 × 1-cm hypoechoic region along the anterior wall of the right EIA (Fig 1). The CTA findings included a "short segment dissection" beginning in the distal right common iliac artery and extending into the common femoral artery (CFA; Fig 2). The radiologist noted that the differential diagnosis included "arterial dissection with thrombosis of the false lumen, liquefied intramural hematoma, and thromboembolism… [the possibility of an arterial wall mass was] unlikely given the complete lack of vascularity."

Because of the clinical suspicion for potential ACD, magnetic resonance angiography was performed on an outpatient basis. The findings included "this may represent hemosiderin from chronic thrombosed false lumen of a dissection" (Fig 3). The radiologist report did not include the possibility of ACD. The diagnosis was indeterminate despite all the imaging modalities obtained, although ACD was discussed with the patient as a part of the differential diagnosis, in addition to spontaneous dissection, embolic disease, and external iliac artery endofibrosis

From the Joan C. Edwards School of Medicine, Marshall University, Huntington[a]; and the Division of Vascular Surgery, Department of Surgery, Spectrum Health, Michigan State University College of Human Medicine, Grand Rapids.[b]

Author conflict of interest: none.

Correspondence: Jaineet S. Chhabra, MS, Joan C. Edwards School of Medicine, Marshall University, 1130 14th St, Huntington, WV 25701 (e-mail: chhabra@marshall.edu).

The editors and reviewers of this article have no relevant financial relationships to disclose per the Journal policy that requires reviewers to decline review of any manuscript for which they may have a conflict of interest.

2468-4287

https://doi.org/10.1016/j.jvscit.2021.04.008

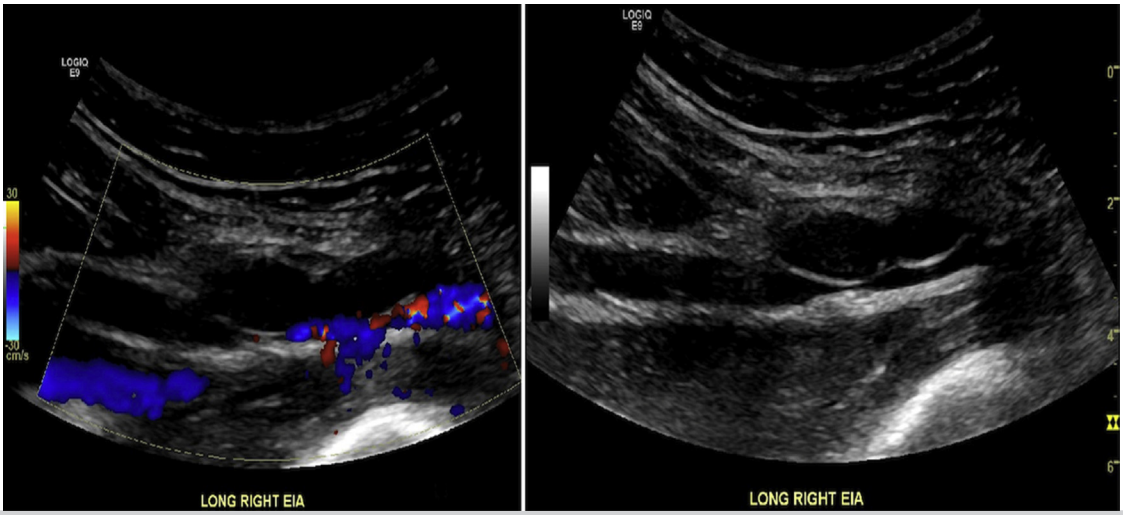

Fig 1. Iliac arterial ultrasound scan in duplex mode (*Left*) and B-mode (*Right*).

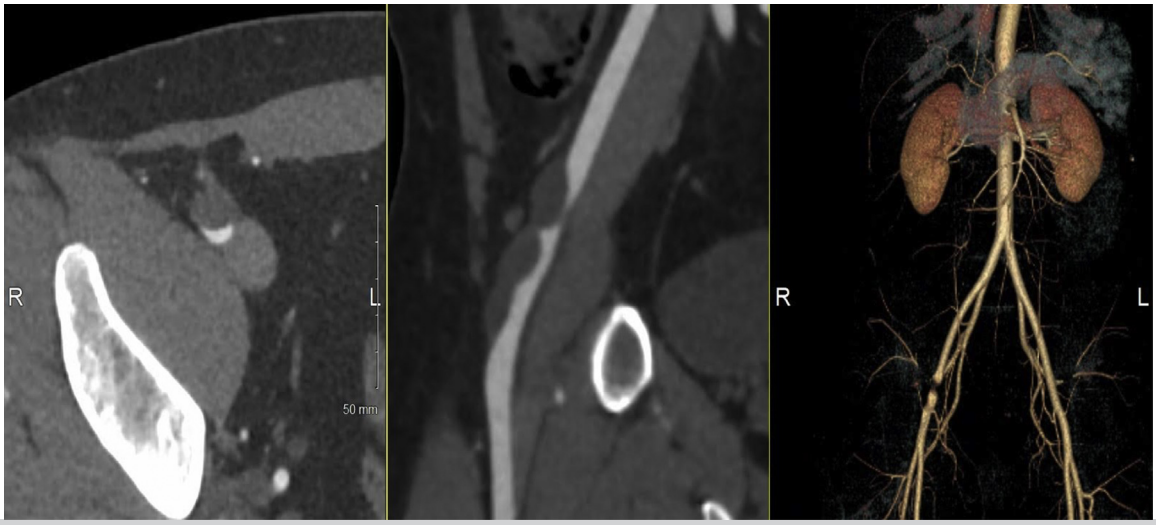

Fig 2. Computed tomography angiography (CTA) on initial presentation. *Left*, Axial view. *Middle*, Rotational coronal/sagittal view. *Right*, Three-dimensional CTA reconstruction.

(although the latter was unlikely because the patient was not a highly active recreational cyclist or athlete). Given the limitations and effects on his lifestyle, open surgical intervention was recommended to the patient in the form of iliofemoral bypass.

For the procedure, a right paramedian incision was made to facilitate entry into the retroperitoneal space, and the EIA was exposed. An oblique incision was made at the groin, and the CFA was also exposed. The epigastric artery and vein and the circumflex arteries were then ligated and divided. The patient was systemically heparinized with 100 U/kg, and atraumatic profunda clamps were applied to the inflow and outflow vessels. The CFA and distal EIA were freed circumferentially, enabling resection of a thick gel-like substance comprising the adventitia of the affected area (Fig 4). Pathologic examination confirmed ACD. An 8-mm Dacron graft was tunneled anatomically beneath the inguinal ligament and the proximal (mid-EIA) and distal (distal CFA) anastomoses were both completed in end-to-end fashion. At the end of the case, the patient's pedal pulses were easily palpable.

The patient experienced no perioperative complications and was doing well at his 1-month follow-up visit. In addition to not experiencing further symptoms of claudication, his ABIs were normal. He was scheduled for annual surveillance visits for outpatient follow-up with ABIs.

DISCUSSION

Fewer than 10 cases of ACD in the EIA have been reported. The etiology of this pathology, regardless of where it presents anatomically, is currently unknown. Proposed theories, including de novo mucinous degeneration due to generalized disease, trauma, articular/synovial theory, and developmental/ganglion theory, are still being debated.[10] Adventitial cysts (in the popliteal artery or otherwise) can be discovered before thrombosis

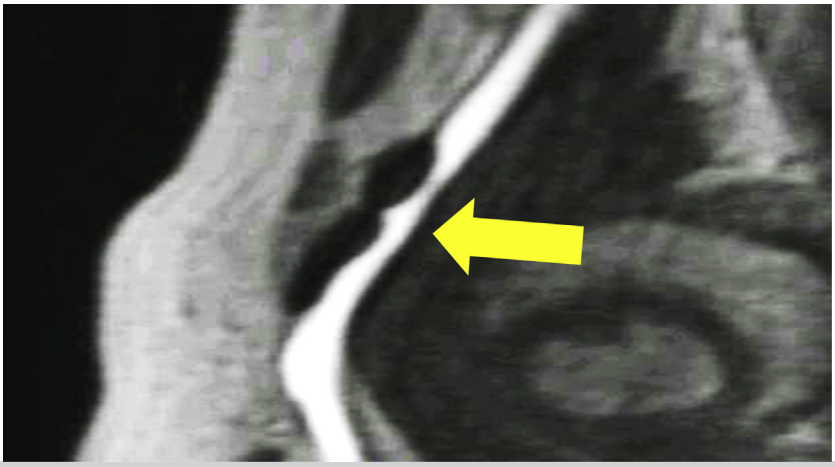

Fig 3. Preprocedural magnetic resonance angiography, sagittal composite image, demonstrating the cyst (*arrow*).

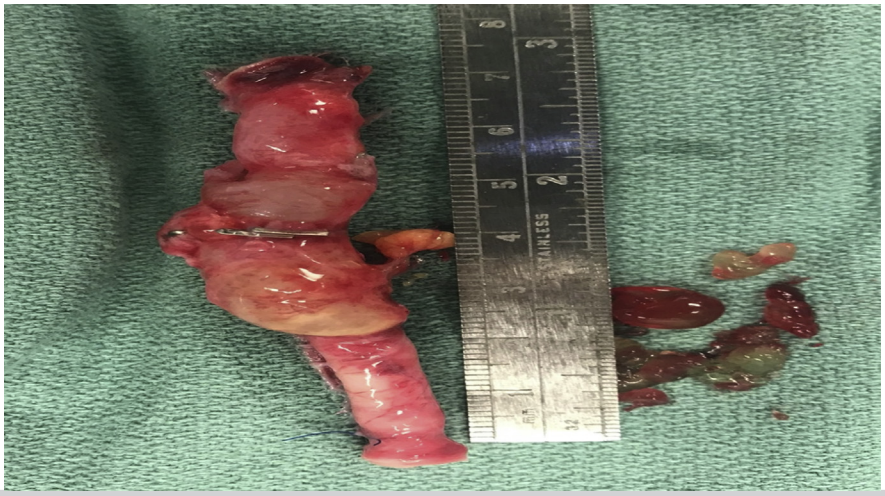

Fig 4. Intraoperative photograph of gross specimen of the right external iliac artery (EIA).

arises if the patient experiences claudication, similar to our patient's presentation. However, patients can initially present with thrombosis and accompanying acute limb ischemia, resembling the symptoms due to a thrombosed popliteal artery aneurysm.[11] This highlights the importance of an early diagnosis, which depends on knowing the target population that ACD affects, choosing the proper imaging modalities, and providing the recommended treatment.

It is important to note that ultrasound, CTA, and MRI were not readily available in 1947 when ACD was first diagnosed.[12] A physical examination finding, the Ishikawa sign, characterized by the disappearance of pedal pulses with knee flexion during a vascular examination due to compression of the popliteal pulses by the adventitial cysts, was reported in 1961.[13] Larger cysts appear to increase the sensitivity of this test. However, radiologic studies are important to best visualize any cystic lesions within the arterial lumen and prevent a misdiagnosis.

Different imaging modalities will demonstrate different physical characteristics of the disease. Ultrasound will reveal hypoechoic lesions within the arterial wall without internal flow and is helpful for identifying cystic changes within an affected vessel. However, ultrasound will usually be supplemented by MRI for better visualization and characterization of the cysts.[14] MRI appears to be a superior image modality for the diagnosis because it permits a thorough cyst assessment and will reveal whether the material extends into an associated joint.[15] Magnetic or conventional angiography can show a characteristic "hourglass sign" or concentric compression of the lumen. If the cyst is large enough to displace the artery to one side, however, a "smooth" occluding and visible contour indicative of a "scimitar" sign could appear. Both CTA and MRI can allow for the estimation of the extent of a diseased segment, aiding in preoperative planning. However, studies have reported that MRI should be used when the CT views are insufficient, further indicating

that MRI is the reference standard imaging study for ACD diagnosis.[16]

It has been reported that endovascular treatment of ACD has mostly been ineffective owing to poor outcomes, including fracture due to exacerbation of the mechanical forces exerted on the stent during movement and restenosis after percutaneous transluminal angioplasty.[17-19] Some patients might benefit from percutaneous ultrasound or CT-guided aspiration of the cyst contents; however, the recurrence rates can be as high as 10% with these procedures.[20] The best treatment of arterial ACD has remained cyst resection and vascular reconstruction with an interposition graft.[3,5]

CONCLUSIONS

A careful assessment of ACD, coupled with knowledge of the affected target population, appropriate imaging techniques, and recommended treatments, can prevent misdiagnosis and allow for an early diagnosis, especially for cases that occur in atypical locations.

We would like to thank Joseph G. Martin, RT (R)(CT)(ARRT), of the Spectrum Health 3-D radiology department, for rendering and processing the magnetic resonance angiography scan allowing it to be included in our report.

REFERENCES

1. Atkins HJ, Key JA. A case of myxomatous tumour arising in the adventitia of the left external iliac artery: case report. Br J Surg 1947;34:426.
2. Torres-Blanco Á, Pepén-Moquete LA, Molina-Nácher V, Gómez-Palonés F. Adventitial cystic disease of radial artery: apropos of a case and review of literature. Cardiovasc Pathol 2017;29:33-6.
3. Lejay A, Ohana M, Delay C, Georg Y, Girsowicz E, Thaveau F, et al. Cystic adventitial pathology as an entity in peripheral arterial disease. J Cardiovasc Surg (Torino) 2016;57:282-91.
4. Veraldi GF, Scudo G, Scorsone L, Mezzetto L, Castellani RL. Cystic adventitial disease of the popliteal artery: report of two cases and review of the literature. G Chir 2014;35:229-34.
5. Desy NM, Spinner RJ. The etiology and management of cystic adventitial disease. J Vasc Surg 2014;60:235-45.e1-245.e11.
6. Mousa AY, Alhalbouni S, Abu-Halimah S, Gill G, Sadek B, Nanjundappa A, et al. Cystic adventitial disease of the common femoral vein: a case report and review of the literature. Vasc Endovascular Surg 2013;47:569-72.
7. Foster DR, Cameron DC. Cystic adventitial disease of the popliteal artery. Australas Radiol 1982;26:161-4.
8. Dharmaraj RB, Griffin J, Ramanathan A, Buckenham T. Case report: cystic adventitial disease of the external iliac artery with imaging features of a complicating proximal dissection. EJVES Extra 2011;21:e36-8.
9. Kim HK, Park S, Hwang D, Huh S. Adventitial cystic disease of the iliac artery with a connection to the hip joint. Vasc Specialist Int 2018;34:10-3.
10. Johnson JM, Kiankhooy A, Bertges DJ, Morris CS. Percutaneous image-guided aspiration and sclerosis of adventitial cystic disease of the femoral vein. Cardiovasc Intervent Radiol 2009;32:812-6.
11. Norgren L, Hiatt WR, Dormandy J, Nehler MR, Harris KH, Fowkes FGR, et al. Inter-society consensus for the management of peripheral arterial disease (TASC II). Eur J Vasc Endovasc Surg 2007;33(Suppl 1):S1-75.
12. Bercovich E, Javitt MC. Medical imaging: from roentgen to the digital revolution, and beyond. Rambam Maimonides Med J 2018;9:e0034.
13. Ishikawa K, Mishima Y, Kobayashi S. Cystic adventitial disease of the popliteal artery. Angiology 1961;12:357-66.
14. Mintz AJ, Weinberg I. Nonatherosclerotic PAD: approach to exertional pain in the lower extremities. Curr Cardiol Rep 2015;17:66.
15. Ali T, Krokidis ME, Winterbottom A. Peripheral non-atherosclerotic arterial disorders: what radiologists need to know. Acad Radiol 2017;24:497-505.
16. Hennessy MM, McGreal G, O'Brien GC. Two cases of popliteal cystic adventitial disease treated with excision and primary bypass graft: a review of the literature. Vasc Endovascular Surg 2017;51:480-4.
17. Patel NJ, Gordon G, Mhatre AU, Verma DR, Pershad A, Heuser RR. Adventitial cystic disease of the popliteal artery contributing to Supera stent fracture. J Invasive Cardiol 2019;31:E160-1.
18. Khoury M. Failed angioplasty of a popliteal artery stenosis secondary to cystic adventitial disease: a case report. Vasc Endovascular Surg 2004;38:277-80.
19. Fox RL, Kahn M, Adler J, Sussman B, Mendes D, Ibrahim IM, et al. Adventitial cystic disease of the popliteal artery: failure of percutaneous transluminal angioplasty as a therapeutic modality. J Vasc Surg 1985;2:464-7.
20. Morbi A, Gohel MS, Hamady M, Cheshire NJ, Bicknell CD. Lower-limb ischemia in the young patient: management strategies in an endovascular era. Ann Vasc Surg 2012;26:591-9.

Submitted Jan 18, 2021; accepted Apr 19, 2021.