Adventitial cystic disease of the femoral vein in a 5-year-old boy mimicking deep venous thrombosis

Douglas W. Jones, MD,[a] Combiz Rezayat, MD,[a] Patricia Winchester, MD,[b] and **John K. Karwowski, MD,[a]** *New York, NY*

Adventitial cystic disease of the vein is a rare vascular anomaly with 32 reported cases. A 5-year-old boy initially presented with painless leg swelling. He was misdiagnosed with deep vein thrombosis and treated with 3 months of warfarin. When swelling failed to improve, a magnetic resonance venogram showed a mural cystic lesion of the left common femoral vein. In the operating room, the cyst was excised, relieving the obstructive effect and restoring flow. The swelling resolved within days. This is the first reported case of adventitial cystic disease of the vein occurring in a pediatric patient. (J Vasc Surg 2012;55:522-4.)

CASE REPORT

A 5-year-old boy without any medical or surgical history presented with a 5-month history of left lower extremity swelling. He had been previously diagnosed with a deep venous thrombosis (DVT) and treated with hydroxyethylrutoside outside the United States. Upon presentation to our institution, the patient had a swollen but painless left thigh and leg. He had no risk factors for DVT. Left lower extremity venous duplex ultrasound and computed tomography scan were thought to be consistent with a focal DVT of the left common femoral vein. The patient was admitted to the hospital and, after a negative thrombophilia workup, was discharged on warfarin. However, his leg swelling did not improve. Repeat venous ultrasounds 1 and 3 months later showed no change in the focal DVT despite adequate anticoagulation.

He was then referred to vascular surgery for further evaluation. Femoral venous duplex in the vascular laboratory revealed an eccentric mural cyst with significant luminal compromise and an obstructive flow pattern. A magnetic resonance venogram was performed which showed a 1.4-cm round nonenhancing lesion in the wall of the left femoral vein at the junction with the greater saphenous vein consistent with a cyst (Fig 1). Warfarin treatment was discontinued. In the operating room, an oblique incision was made in the groin at the level of the cyst. Dissection was carried down to the femoral vein at which point an eccentric cyst on the posterior wall of the vein was identified (Fig 2). The cyst wall was incised and a dark, thick material drained. After this, the entire abluminal cyst was excised. There was no disruption in the continuity of the vein (Fig 3) and the saphenofemoral junction was preserved. An intraoperative venous ultrasound showed excellent flow throughout the now widely patent vein. Pathology of the cyst wall showed fibromembranous tissue consistent with a cyst. There was no epithelialized lining of the cyst wall.

The patient had an uncomplicated recovery notable for complete resolution of his leg swelling within days without compressive therapy. Follow-up venous duplex ultrasound showed normal flow through the femoral vein with no evidence of cyst recurrence.

DISCUSSION

Adventitial cystic disease (ACD) of the vein is a very rare vascular anomaly. There are only 32 reported cases of ACD of the vein in the world literature, including this case.[1-18] In contrast, adventitial cystic disease of the artery is a well-described, if rare, cause of intermittent lower extremity arteriogenic claudication occurring in 1:1200 cases of claudication.[19,20]

ACD of the artery involves the popliteal artery in 85% of cases.[19,20] In contrast, ACD of the vein affects the popliteal vein relatively infrequently with the most prevalent anatomic sites being the common femoral vein in 16 of 32 cases (50%), the external iliac vein in 8 cases (25%), and the popliteal vein in only 3 cases (9%). Other sites affected have been the small saphenous vein, greater saphenous vein, "wrist" vein, and deep dermal vein in the ankle. Our case represents the 16th case of ACD of the common femoral vein. The lower extremity veins were affected in 28 cases (88%) with right and left sides equally affected (right side = 15 cases; left side = 13 cases).

Many theories as to the etiology of ACD of the artery have been applied in discussions of ACD of the vein,[19,20] but authors have recently argued against the equivalence of the two processes based on epidemiologic factors.[16] In

From the Division of Vascular Surgery[a] and Department of Radiology,[b] New York Presbyterian Hospital, Weill Cornell Medical Center.

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Reprint requests: Douglas W. Jones, MD, New York Presbyterian Hospital – Weill-Cornell Medical Center, Division of Vascular Surgery, 435 East 70th Street, Apt 14L, New York, NY 10021 (e-mail: doj9016@gmail.com).

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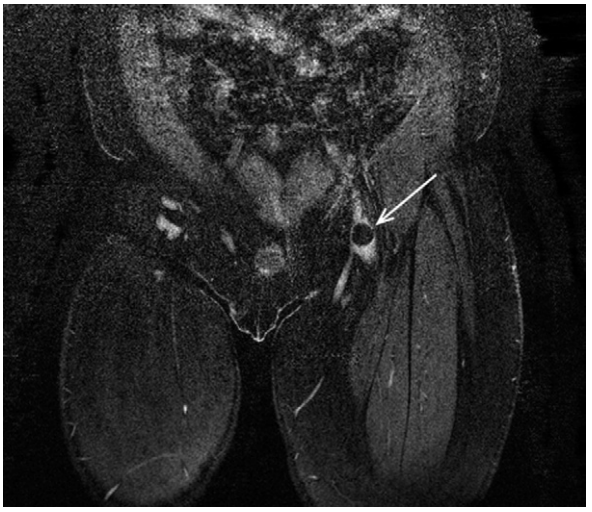

Fig 1. Coronal image from magnetic resonance venography showing cystic dilatation of the left common femoral vein (*arrow*). There is no signal in the cystic structure.

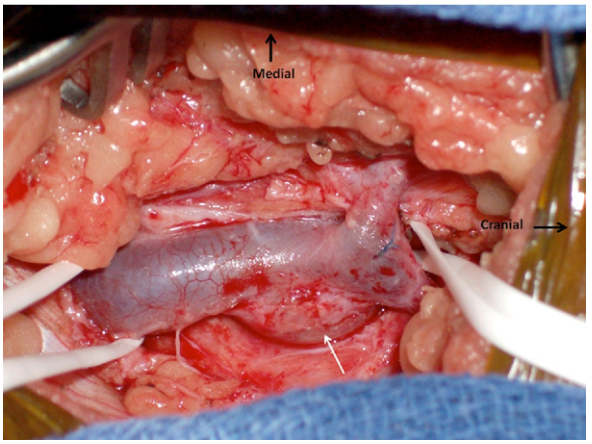

Fig 2. Intraoperative photograph showing cystic dilatation of the posterior wall of the left common femoral vein. *White arrow* indicates cyst.

addition to differences in distribution of affected vessels, distribution of disease between genders also differs when arterial ACD is compared to venous ACD. ACD of the artery affects men much more commonly than women, with a male:female ratio of 15:1.[19,20] Men are also more commonly affected in ACD of the vein, although to a lesser degree, with a male:female ratio of 2.3:1 (21 men, 9 women of 30 reported cases where gender was known). Based on our review of the available literature, ACD of the vein most typically presents in adults, with a mean age of 47 years old (range, 5-75 years old). The youngest age at which ACD of the vein has been reported is 23 years old.[5] At 5 years old, our patient is the youngest reported with venous ACD and is also the first patient in the pediatric population reported to have the disease.

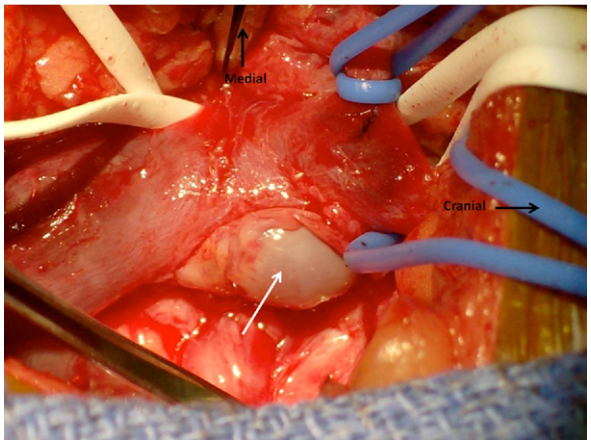

Fig 3. Intraoperative photograph showing the left common femoral vein after cyst wall excision. *White arrow* indicates vessel wall at cyst site after cyst excision.

Bilaterality in ACD of the vein is uncommon. As a result, patients present with asymmetric lower extremity edema. A painless inguinal mass may be palpable on physical examination. Due to compression of the venous system by the cyst, patients may present with DVT[12] or may be misdiagnosed with DVT, as was the case with our patient.

In the setting of unilateral lower extremity edema, venous duplex ultrasonography is often the first diagnostic test performed and can be very reliable at visualizing a cystic mass in the wall of blood vessels with associated narrowing of the vein lumen. As in our case, a magnetic resonance venogram can be helpful in confirming the diagnosis and clarifying anatomy. A computed tomography scan is unlikely to yield additional diagnostic information over venous duplex ultrasonography.

Multiple surgical techniques have been used to treat ACD of the vein. The first option is cyst drainage with or without complete excision of the cyst wall.[2-8,10,13-15,18] This method offers good immediate results with restoration of flow through the native vessel. However, recurrence is common when the cyst wall is not completely excised. Of 6 reported cases where surgical drainage of the cyst was attempted without obvious attempt at cyst excision, there were five recurrences requiring a second operation.[1,5,6,10] In 13 cases, cyst excision was attempted at the initial operation and recurrence occurred in only 3 patients.[4,8,13] Recurrence has been attributed to incomplete excision of cystic tissue; significant portions of the cyst are left in situ and may continue to secrete mucinous material. A more contemporary option is resection of the affected portion of the vein and immediate reconstruction with prosthetic or autologous graft as performed in 5 cases.[8,9,11,12,17] Prosthetic graft was chosen in 4 cases and autologous axillary vein graft in 1 case. Recurrence of a cyst has not yet been reported after vein resection with reconstruction. Percutaneous cyst aspiration and sclerosis has also been reported in 1 case without recurrence.[16]

In our case, complete cyst excision was chosen because it offered the lowest chance of recurrence without exposing the patient to growth-related limitations of a vascular prosthesis. The size of the cystic segment relative to the small caliber of the overall vein in this small child precluded resection and primary anastomosis.

Before current imaging capabilities, the diagnosis of adventitial cystic disease of the vein was made only at the time of surgery and little was known about the most effective treatment of venous ACD. With modern diagnostics, preoperative diagnosis should now be commonplace and proper surgical planning will yield optimal outcomes. Complete excision of the cyst wall is a very effective treatment and should be attempted first if technically feasible. However, vein resection and interposition graft with prosthetic or autologous graft may be necessary if complete cyst excision cannot be accomplished or in cases of recurrence.

REFERENCES

1. Mentha C. [Mucoid degeneration of the veins]. [Article in French] Presse Med 1963;71:2205-6.
2. Gomez-Ferrer F. Cystic degeneration of the wall of the femoral vein. J Cardiovasc Surg 1966;7:162-5.
3. Matsubara J, Ban I, Nakata Y, Hirai M, Miyazaki H, Kawai S, et al. [Cystic adventitia degeneration of the external iliac vein]. [Article in German] Vasa 1978;7:443-6.
4. Frileux C, Le Baleur A, Uzan E. Obstruction of the iliac vein by mucoid cyst: successful result of surgical excision. J Cardiovasc Surg (Torino) 1979;20:517-8.
5. Annetts DL, Graham AR. Cystic degeneration of the femoral vein: a case report. Br J Surg 1980;67:287-8.
6. Fyfe NC, Silcocks PB, Browse NL. Cystic mucoid degeneration in the wall of the femoral vein. J Cardiovasc Surg (Torino) 1980;21:703-6.
7. Ishikawa K. Cystic adventitial disease of the popliteal artery and of other stem vessels in the extremities. Jpn J Surg 1987;17:221-9.
8. Desjardins JF, Turlin B, Kerdiles Y, Ledu J, Clément B. Cystic degeneration of the femoral vein. Lancet 1997;349:1000.
9. Schraverus P, Dulieu J, Mallieux P, Coulier B. Cystic adventitial disease of the popliteal vein: a report of a case. Acta Chir Belg 1997;97:90-2.
10. Yoshii S, Ikeda M, Murakami H. Cystic myxomatous adventitial degeneration of a saphenous vein. J Vasc Surg 1998;27:780-2.
11. Sugimoto T, Yamamoto K, Tanaka S, Saitou N, Kikuchi C, Motohashi S, et al. Adventitial cystic disease of the femoral vein: report of a case. Surg Today 2004;34:286-8.
12. Gasparis AP, Wall P, Ricotta JJ. Adventitial cystic disease of the external iliac vein presenting with deep venous thrombosis. A case report. Vasc Endovascular Surg 2004;38:273-6.
13. Maldonado-Fernández N, Lopez-Espada C, Moreno-Escobar J, Martinez-Gámez J, Rodriguez-Morata A, Garcia-Róspide V. Recurring adventitial cyst in the left external iliac vein. EJVES Extra 2004;8:10-4.
14. Sakamoto A, Tanaka K, Matsuda S, Harimaya K, Nakamura T, Oda Y, et al. Adventitial cystic disease of the popliteal vein: report of a case. Surg Today 2006;36:1098-100.
15. Dix FP, McDonald M, Obomighie J, Chalmers N, Thompson D, Benbow EW, et al. Cystic adventitial disease of the femoral vein presenting as deep vein thrombosis: a case report and review of the literature. J Vasc Surg 2006;44:871-4.
16. 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.
17. Seo JY, Chung DJ, Kim JH. Adventitial cystic disease of the femoral vein: a case report with CT venography. Korean J Radiol 2009;10:89-92.
18. Morizumi S, Suematsu Y, Gon S, Shimizu T, Iwai T. Adventitial cystic disease of the femoral vein. Ann Vasc Surg 2010;24:1135.e5-7.
19. Tsilimparis N, Hanack U, Yousefi S, Alevizakos P, Rückert RI. Cystic adventitial disease of the popliteal artery: an argument for the developmental theory. J Vasc Surg 2007;45:1249-52.
20. Levien LJ, Benn CA. Adventitial cystic disease: a unifying hypothesis. J Vasc Surg 1998;28:193-205.

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