Repair of popliteal aneurysm and spontaneous arteriovenous fistula in a patient with Marfan syndrome

Patrick Z. McVeigh, PhD,a Ahmed Kayssi, MD,b Aaron Lo, MD,b Dheeraj K. Rajan, MD,c George D. Oreopoulos, MD, MSc,b,c and Graham Roche-Nagle, MD, MBA,b Toronto, Ontario, Canada

Isolated extremity arterial aneurysms remain a rare entity, and the development of a spontaneous arteriovenous fistula from such an aneurysmal segment in a young patient should prompt a search for an underlying genetic predisposition. Endovascular repair of aneurysms or arteriovenous fistulas in the popliteal artery is appropriate in select populations; however, open repair allows for a more durable reconstruction of both the arterial and any involved venous segments in patients who can tolerate the procedure. (J Vasc Surg Cases 2016;2:137-40.)

The development of arteriovenous fistulas (AVFs) in the popliteal fossa are rare and almost always a result of antecedent trauma or medical intervention, with only isolated reports of spontaneous popliteal AVFs.1-3 We describe a case of spontaneous, atraumatic popliteal AVF formation secondary to an isolated popliteal aneurysm in a previously undiagnosed Marfan syndrome patient. Written informed consent was obtained from the patient for publication of this case report and accompanying images.

CASE REPORT

A 29-year-old man presented to the hospital with a 3-week history of right calf pain and worsening knee pain with extension. The physical examination was notable for a relatively tall, thin build, disproportionately long upper extremities and hypermobile joints, but no high-arched palate, chest wall abnormalities, nor any history of ocular pathology. Further note was made of significant tachycardia at rest, right popliteal fossa swelling, and an underlying palpable thrill.

Computed tomography angiography (Fig 1) demonstrated ectatic right common iliac, common femoral, superficial femoral, and popliteal arteries down to the level of a large AVF between a popliteal artery aneurysm and the popliteal vein. Distally, the arteries were normal in caliber and patent. The patient did not endorse any history of trauma or invasive medical interventions in the region. He had no other contributory medical history.

Genetic testing supported a diagnosis of Marfan syndrome with a mutation in the gene encoding fibrillin 1 (FBN1). The patient showed no evidence of aortic aneurysm on imaging. Trans-thoracic echocardiography demonstrated normal ventricular and valvular function and a mildly dilated aortic root at 41 mm.

Consent was obtained for surgical repair. The operative plan was to harvest the contralateral superficial femoral popliteal vein and use a posterior approach with interposition grafting. At operation, the popliteal artery was noted to be segmentally enlarged in keeping with localized aneurysmal dilatation, and the AVF with the massively dilated popliteal vein was immediately identified (Fig 2). Care was taken throughout the dissection to preserve collateral venous branches as best as possible. The AVF did not appear to be acute because numerous venous branches had become arterialized. Vascular control was obtained, and opening the aneurysmal artery revealed the AVF with occlusion of the distal popliteal veins. The only patent segment of the popliteal vein was the proximal high-flow region immediately central to the AVF.

The aneurysmal section of the artery was excised, and the previously harvested superficial femoral popliteal vein was reversed and anastomosed end-to-end using running 6-0 polypropylene suture. There was no excessive suture line bleeding necessitating reinforcement during the repair, and after refusion an excellent Doppler signal was obtained in the extremity. The vein stump was over sewn.

A low-dose heparin infusion was started in the early postoperative period. Anticoagulation was continued for 3 months due to a potentially increased risk of venous thrombosis in the remaining dilated venous system. A popliteal fossa hematoma developed postoperatively, which was treated with percutaneous drainage as an intermediate step before operative drainage; however, the patient was readmitted 3 weeks postoperatively for wound breakdown. The wound was débrided, and a vacuum dressing was applied. Three weeks later, the wound had healed with full return of function. A magnetic resonance imaging study 6 months after the surgery demonstrated a patent graft and ectatic iliac and femoral arteries with normalization of the previous venous dilation (Fig 3).

From the Department of Medical Biophysics, Faculty of Medicine,a Division of Vascular Surgery,b and Joint Department of Medical Imaging,c University Health Network, University of Toronto.

Author conflict of interest: none.

Correspondence: Graham Roche-Nagle, MD, MBA, Toronto General Hospital, 200 Elizabeth St, 6EN-218, Toronto, ON MSG 2C4, Canada (e-mail: graham.roche-nagle@uhn.ca).

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.

2352-667X

© 2016 The Authors. Published by Elsevier Inc. on behalf of Society for Vascular Surgery. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

http://dx.doi.org/10.1016/j.jvsc.2016.03.010
DISCUSSION

Marfan syndrome, a global connective tissue disease with autosomal-dominant inheritance and high rate of sporadic occurrence (25%-30%), is characterized by mutations in \( FBN1 \), the gene on chromosome 15 that encodes fibrillin 1, a large glycoprotein component of extracellular microfibrils.\(^4\)\(^5\) A missense variant in \( FBN1 \) was found in this patient, and expanded genetic testing showed no evidence of Ehlers-Danlos type IV (\( COL3A1 \)). As a result of the role of microfibrils in maintaining elasticity and strength in connective tissue, the resultant disorder has diffuse effects on the cardiovascular, skeletal, central nervous system, and ocular systems\(^6\)\(^7\) and is classically associated with aneurysms of the thoracic aorta.\(^8\) This is largely ascribed to the large diameter and continuous pressure changes in the thoracic segment that leads to dissection and aneurysm formation. Isolated nondissecting abdominal aortic aneurysms are rare in the Marfan population,\(^8\) and there have been only case reports of aneurysmal changes in the extracranial,\(^9\) superior mesenteric,\(^10\) and popliteal\(^11\)\(^12\) arteries.

Repair of popliteal aneurysms may be undertaken with an open surgical or endovascular approach,\(^13\) with
comparable mortality rates. Open repair is associated with superior long-term patency, especially in younger asymptomatic patients and those in whom venous conduits are used, with 1-year primary patency 89.0% vs 67.4%. Unique to this patient, the aneurysmal popliteal artery developed an AVF, without antecedent trauma or medical intervention, which is unusual given the rarity of spontaneous extremity AVFs. Popliteal AVFs may be treated with an endovascular technique in select patients, typically those with suitably sized vessels and landing zones for stent graft placement and who are unable to tolerate open repair. Given the patient’s young age and the relatively large size of the involved vessels, an open surgical approach was used to allow for a durable repair of both the arterial and venous components.

Open repair of popliteal aneurysms may be through a medial or posterior approach, with comparable long-term patency and less aneurysmal re-expansion reported with the posterior approach, which also was necessary in this patient to allow for sufficient exposure of the venous component of the concomitant AVF. The posterior approach carries an increased risk of nerve injury, and wound healing may be poorer, as seen in our patient, with need to use healing by secondary intention with a vacuum dressing after late wound breakdown.

CONCLUSIONS

Isolated extremity arterial aneurysms remain a rare entity, and the development of an unprovoked AVF from an aneurysmal segment, as seen here in a young patient, should prompt a search for an underlying genetic predisposition. Endovascular approaches to popliteal aneurysms and AVFs are appropriate in select populations, but open repair allows for durable reconstruction of both the arterial and venous segments in patients who can tolerate the procedure.

REFERENCES


Fig 3. A follow-up magnetic resonance angiography at 6 months demonstrates no residual popliteal aneurysm and no early venous filling of the popliteal or femoral veins. The right iliac and femoral arteries are ectatic.


