J Ped Surg Case Reports 14 (2016) 8–11

Contents lists available at ScienceDirect

Journal of Pediatric Surgery CASE REPORTS

journal homepage: www.jpscasereports.com

Unusual cause of acute lower extremity ischemia in a healthy 15-year-old female: A case report and review of popliteal artery aneurysm management in adolescents

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ARTICLE INFO

Article history:
Received 24 May 2016
Received in revised form
9 August 2016
Accepted 12 August 2016

Key words:
Pediatric
Popliteal aneurysm
Peripheral ischemia

ABSTRACT

Limb ischemia in healthy teenagers is unusual. While traumatic or iatrogenic injury is the most common etiologies of limb ischemia in the pediatric population, anatomic variants such as true aneurysms should be considered [1]. We report the second documented pediatric case of an idiopathic, isolated true popliteal aneurysm resulting in acute limb ischemia in a previously healthy 15-year-old female. We also review the proper evaluation and surgical management of this anatomic anomaly. In this case, surgical management included resection of the aneurysm, reconstruction with reverse saphenous vein grafting, and distal endarterectomies to restore adequate distal blood flow. Ultimately, this patient's limb and function were salvaged with minimal consequences.

Popliteal artery aneurysms are a rare cause of peripheral arterial ischemia in the pediatric population. While they account for 70% of peripheral aneurysms in the adult population, the prevalence within the general population remains at only 1% [2]. Most affected adult patients have a well-described history of atherosclerosis and may have an associated abdominal aortic aneurysm [3]. In the pediatric population, peripheral aneurysms and pseudoaneurysms have been described in patients with inherited connective tissue diseases that weaken the structure of vascular tissue, such as Ehlers-Danlos and Marfan syndrome [4,5]. Aneurysm formation has also been reported in cases of traumatic injury or in disease states such as fibromuscular dysplasia, osteochondroma, infection, and vasculitides [1, 6–11]. Review of the literature revealed 29 cases from 1834 to 2015 of children with multiple congenital aneurysms involving several anatomic locations without a defined etiology or syndrome [11–18]. However, isolated, idiopathic popliteal artery aneurysms are exceptionally rare in the pediatric population. Review of the literature has shown only one previously reported isolated, idiopathic true popliteal artery aneurysm [19].

We present the second case reported of an isolated, true idiopathic popliteal aneurysm in a 15-year-old previously healthy female who developed acute limb ischemia. We also review the evaluation, management and potential sequelae of popliteal aneurysms in the pediatric population.

1. Case report

A 15-year-old previously healthy, female cross-country runner presented with acute onset of severe right lower extremity pain. She reported a 2-month history of severe episodic right foot pain unassociated with physical activity, which was initially diagnosed as foot strain. On the morning of admission, she experienced moderate pain in her foot, which escalated over the course of 5 h to 9 of 10 in severity. She was brought to the emergency department at an outside hospital where initial examination demonstrated a palpable right femoral artery pulse but absent distal pulses. Investigational workup at the outside hospital included arterial duplex scanning which demonstrated normal flow through the right superficial femoral artery. However, blood flow was documented as 9 cm/s (normal 80–108) in the distal femoral artery and 5 cm/s (normal 55–83) at the popliteal artery with absent flow below the popliteal artery. She was diagnosed with acute

Funding: N/A.
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http://dx.doi.org/10.1016/j.epsc.2016.08.002

limb-threatening ischemia and transferred via helicopter to a tertiary facility for further evaluation.

Nine hours after the onset of symptoms, she presented to the tertiary hospital. Physical exam confirmed the findings at the referring hospital. A computed tomography (CT) angiogram was performed to determine the level of occlusion and evaluate for distal emboli. The aorta below the diaphragm and vessels proximal to the superficial femoral artery (SFA) were unremarkable but a complete cutoff at the distal SFA was identified (Fig. 1). The limb-threatening ischemia was presumed to be due to a thrombotic event in the region of the popliteal artery, yet the etiology of thrombosis remained unclear.

The patient was immediately taken to the operating room. A hypercoagulability panel was sent prior to operation but was not immediately available. Coagulation studies including a thromboelastogram were normal. The popliteal artery was explored through a standard medial incision. A large aneurysmal dilation of the popliteal artery with absent distal blood flow was identified (Fig. 2). The intraoperative decision was made to excise the aneurysm and perform a reverse saphenous vein graft reconstruction. The resected aneurysm was 4 cm in diameter and showed concentric dilation with a complete luminal thrombosis (Fig. 3). Microscopic examination of the aneurysm demonstrated vessel wall dilation with evidence of degenerative changes. Catheter thrombectomies were performed in the superficial femoral artery and in all three limbs of the trifurcation prior to saphenous vein grafting. Reverse saphenous vein grafting was performed using interrupted 7-0 Prolene® polypropylene suture (Ethicon LLC, Somerville, NJ, USA) suture.

Following repair, intraoperative Doppler confirmed adequate blood flow through the vein graft, yet there remained absent flow through the distal vessels at the ankle. Surgical exploration and an arteriotomy of the posterior tibial (PT) artery revealed a chronic and

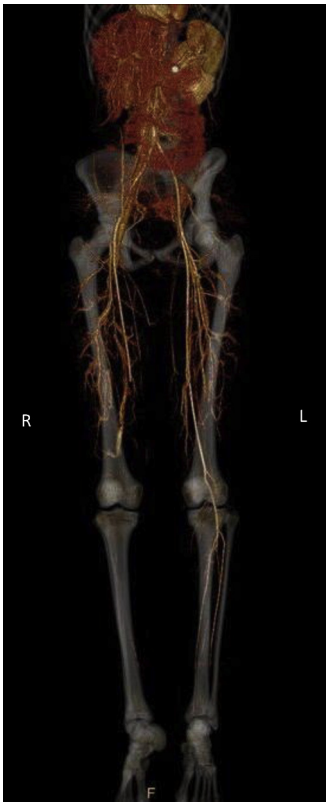

Fig. 1. 3-D reconstruction of CT angiogram demonstrating cutoff of blood flow at the right distal SFA.

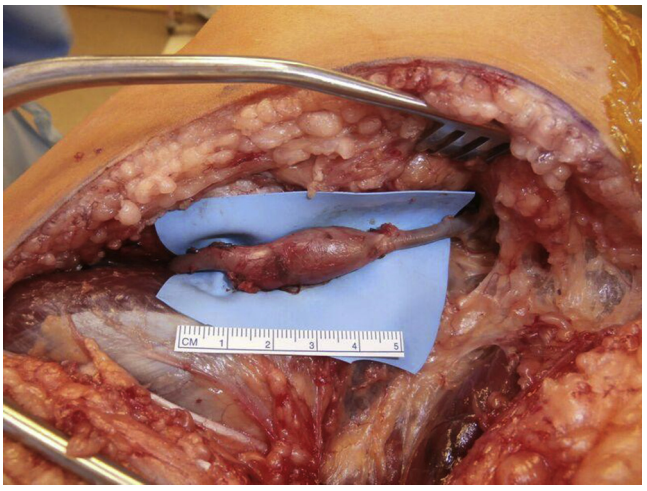

Fig. 2. Intraoperative photo demonstrating aneurysmal sac in the popliteal fossa.

well-organized embolic occlusion that was densely adherent to the vessel wall. Given the extensive thrombosis of the PT artery, exploration of the dorsalis pedis (DP) artery was also performed, revealing a similar chronic distal occlusion. Faced with probable limb loss, the decision was made to perform an endarterectomy of both the PT and DP vessels. A 2 cm long by 2 mm diameter chronic white-tan embolus was extracted from the PT and a 1 cm long by 2 mm diameter embolus was extracted from the DP. Fogarty catheters were passed retrograde from the PT and DP arteries, but no significant clots were removed. The arteriotomies were then closed with standard microvascular techniques with 8-0 Ethilon® nylon sutures (Ethicon LLC, Somerville, NJ, USA).

Despite adequate surgical reconstruction of the popliteal artery and complete distal endarterectomies, intraoperative angiography continued to show absent blood flow to the foot without occlusion. Clinical examination of the foot, however, demonstrated a marked overall improvement in circulation with a capillary refill time of 2–3 s and therefore the patient's inadequate flow on angiography was felt to be due either to vasospasm or chronic microvascular embolic occlusion. A prophylactic four-compartment fasciotomy in the lower leg was performed to prevent compartment syndrome secondary to ischemia-reperfusion injury. The patient was

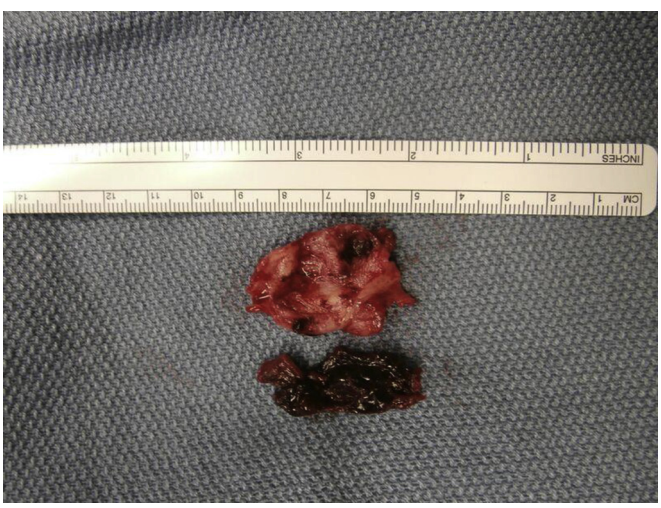

Fig. 3. Resected aneurysm and associated thrombus removed from aneurysm. Microscopic examination confirmed this to be a true aneurysm.

transferred to the pediatric intensive care unit (PICU) where she remained on a heparin infusion.

On arrival to the PICU, the foot remained viable despite lacking a Doppler signal in the foot. However, over the next 24 h, monophasic distal Doppler signals progressively returned to the foot and became biphasic on the second postoperative day. The foot continued to regain color and warmth with the exception of a 2 cm area of ischemia on the distal right great toe. Adjunctive care with air warmers and nitroglycerine paste was employed. The thrombophilia workup returned within normal limits excluding known thrombophilic disease states. She was discharged on hospital day 17 without further complications. The area of ischemic necrosis of the toe was allowed to heal by secondary intention. The pathology report confirmed this to be a true aneurysm.

The patient was placed on a therapeutic dose of low molecular weight heparin for 2 months and aspirin for 6 months. Initial sequelae included ischemic neuropathy of her foot, which was successfully treated with gabapentin. At two-year follow-up, she has a persistent area of distal hypesthesia in the medial forefoot (including the medial 3 toes) which extended proximally to the plantar aspects of the metatarsal heads. The wound at the tip of the great toe demonstrated episodic cutaneous ulcerations related to physical activity over the next 18 months without evidence of osteomyelitis. Despite the sensory loss and occasional skin problem, she successfully returned to long distance running and ambulates without any perceptible gait disturbance or other re-injury.

2. Discussion

Arterial thromboembolic disease is rare in children, occurring in only 8.5 per 10,000 hospital admissions [20]. The most common etiology of arterial thromboembolism in pediatrics is iatrogenic injury during surgery or catheterization leading to mechanical insult to the blood vessel [20,21]. Mechanical, biochemical, and anatomic disturbances underlie the majority of vascular occlusions. In this case, the anatomic disturbance was the aneurysm. No hypercoagulable state was identified.

The initial workup of an ischemic limb should aim to rule out mechanical or anatomic etiologies with imaging and hypercoagulable states with biochemical studies. Arterial aneurysms are one form of anatomic anomaly that predisposes a patient to arterial thrombosis due to stasis of blood flow. An arterial aneurysm is defined as an abnormal dilation of a blood vessel. A true aneurysm involves the symmetric dilation of all three tunics of the blood vessel, preserving histological structure [22]. When an aneurysm is identified, a complete evaluation for abdominal aneurysms or additional extremity aneurysms should be undertaken. CTA (as in this case) gave excellent and rapid images of the abdominal aorta, visceral arteries, and bilateral extremities, but other options include ultrasonography, magnetic resonance angiography, or conventional digital subtraction angiography.

Hereditary thrombophilias encompass several biochemical states that interfere with the inhibition of the coagulation cascade leading to pathologic thrombosis. The most common inherited thrombophilias include antithrombin deficiency, Protein C deficiency, Protein S deficiency and Factor V Leiden. Factor V Leiden is the most common inherited thrombophilia with one allele present in 5% of Caucasians [23]. The prevalence of Protein S deficiency, Protein C deficiency, and antithrombin deficiency within the general population are 0.15%, 0.03–1.3% and 0.2–2% respectively [24–26]. A cohort study of 143 pediatric patients with established thromboembolic disease determined that antithrombin deficiency was present in 6.3% of patients, Protein C deficiency was present in 9.1% of patients, Protein S deficiency was present in 2.1% of patients, and Factor V Leiden was present in 28.7% of patients [27].

True aneurysms in children appear to be extremely rare. Typical causes of true aneurysms in adults include atherosclerosis, hypertension, trauma, vasculitides, infection, and collagen vascular disease [22]. In contrast, a false or pseudoaneurysm is a contained arterial rupture. A pseudoaneurysm is caused by a defect in the vessel wall, which leads to the extravasation of blood into the surrounding connective tissue and is typically seen following iatrogenic injury or trauma [22]. Both true and false aneurysms can affect any blood vessel in the body, although aneurysm formation has a predilection for large, central vessels and intracranial vessels [28,29]. While central and intracranial aneurysms typically rupture, the risk of thrombosis is far greater than the risk of rupture in peripheral aneurysms. In particular, the rupture of a popliteal artery aneurysm is a rare complication with a reported rate ranging from 2 to 4%, while the presence of a thrombus ranges from 39 to 75% [30–32]. No underlying cause for this popliteal artery aneurysm was identified. In adult patients with adequate collateralization, endovascular stenting of popliteal aneurysms have been successfully performed [33]. However, severe acute ischemia caused by complete occlusion of the popliteal artery with limited collateral vascularization threatens limb viability and warrants immediate surgical revascularization. In an otherwise healthy pediatric patient, the additional delay in re-establishing blood flow associated with thrombolytic therapy would contribute further ischemic time in a patient who would also clearly need fasciotomies if flow is re-established. Surgical exploration with resection of the diseased vessel and revascularization with vein grafting has been reported to offer the best chance to prevent limb loss [34].

This case describes a 15-year-old female with distal extremity acute ischemia caused by thrombosis of a popliteal artery aneurysm complicated by chronic distal occlusion of the posterior tibial and dorsalis pedis arteries. Direct exploration with resection of the aneurysm, reconstruction with vein grafting, and endarterectomies of the distal vasculature were required to reestablish flow. This technique of distal endarterectomies has not previously been reported in the pediatric population, but appears to have been effective in this case.

3. Conclusion

Acute limb ischemia in teenagers is rare. While commonly attributable to injury or thrombophilia, anatomic abnormalities such as popliteal aneurysms should be considered as part of the differential diagnosis. In the case of a popliteal artery occlusion due to aneurysmal thrombosis, immediate surgical revascularization with resection of the aneurysm and autologous reverse saphenous vein graft reconstruction is favored. In the setting of chronic occlusion, additional exploration of the distal vessels may be warranted if distal flow is not re-established after successful reconstruction of the popliteal artery. Endarterectomy of multiple chronic emboli in the posterior tibial and dorsalis pedis arteries should be considered and was effective in re-establishing small vessel patency in this rare case.

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