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Popliteal artery entrapment syndrome in a young girl

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Abstract Popliteal artery entrapment syndrome (PAES) is well-described in adults, but is an uncommon cause of lower-limb pain in children. We present an 11.5-year-old girl with thrombosed aneurysm of the right popliteal artery, subsequently diagnosed with bilateral type I PAES. Multimodality illustration of the radiological findings is presented.

Keywords Popliteal entrapment · Popliteal aneurysm · Claudication · Popliteal thrombus

Introduction

Popliteal artery entrapment syndrome (PAES) is well described in adults, but is an uncommon cause of lower-limb pain in children. Accurate and prompt diagnosis is crucial. Imaging modalities play an important role in confirming the diagnosis. We present a case of PAES in a young girl with a multimodality illustration of the radiological findings.

Case report

A previously healthy 11.5-year-old girl presented with an 8-month history of intermittent right-leg pain, mainly with walking and swimming. The pain was relieved by rest. She had noticed coldness and pallor of the right leg for the last 7 months, not associated with numbness. She had a 1-month history of a small wound on the right great toe with progressive swelling. The wound did not improve on systemic antibiotics.

On examination, she was afebrile and had a normal cardiac examination. The right calf was cold with no difference in circumference between the right and left calves. There were redness and bluish discoloration around the right great toe associated with bony tenderness. The right femoral pulse was palpable; however, the popliteal, dorsalis pedis, and tibialis posterior pulses were not palpable. All other peripheral pulses were present.

She had normal complete blood-cell count and coagulation profile. Duplex ultrasound of the lower limbs demonstrated fusiform dilatation of the right popliteal artery, just proximal to the popliteal fossa, with thrombus causing total occlusion of the popliteal artery (Fig. 1). Distally, there was no flow seen in the dorsalis pedis artery; however, some flow was seen within the tibialis posterior artery. The right deep calf veins were patent, in addition to normal arterial and venous flow on the left side. MRI of the lower limbs showed a thrombosed aneurysm of the right popliteal artery. Proximally, the popliteal artery coursed posterior to the medial femoral condyle and was separated from

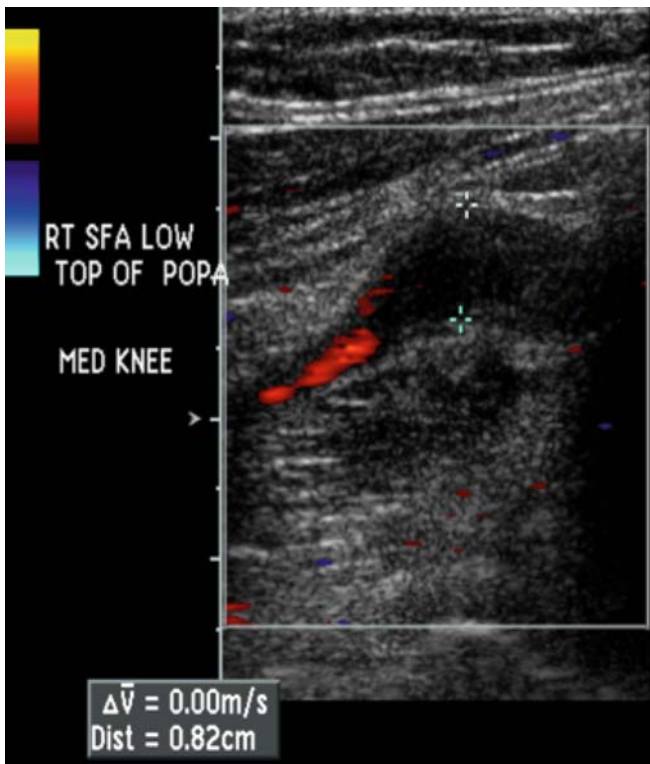


Fig. 1 Longitudinal color Doppler image demonstrating thrombosed aneurysm of the right popliteal artery

the popliteal vein by the medial head of the right gastrocnemius muscle (Fig. 2). The left popliteal artery was patent, without aneurysmal dilatation; however, it had an identical course to its right-sided counterpart (Fig. 3). These findings were consistent with bilateral type I popliteal entrapment. Angiography of both lower limbs was performed and showed an abrupt occlusion of the right popliteal artery proximal to the trifurcation, with one-vessel dominant runoff via the peroneal artery to the right lower extremity. The right peroneal artery reconstituted proximally and was patent to the level of the ankle, where it reconstituted the right posterior tibial artery supplying the plantar arch. The left popliteal artery and its trifurcation were patent. Both popliteal arteries coursed medially and were separated from the popliteal vein by the medial head of the gastrocnemius muscle (Fig. 4). Multiple attempts to access the occluded popliteal artery were unsuccessful. Intravenous heparin was given. The patient's symptoms started to improve, but a plan for future surgical treatment was still discussed with the family.

Discussion

Anderson Stuart was the first to describe the basis of popliteal entrapment in an amputated leg in 1879 [1–3].



Fig. 2 Axial T1WI of the right knee shows a thrombosed aneurysm of the right popliteal artery (black arrow), separated from the popliteal vein by the medial head of the gastrocnemius muscle (white arrow)

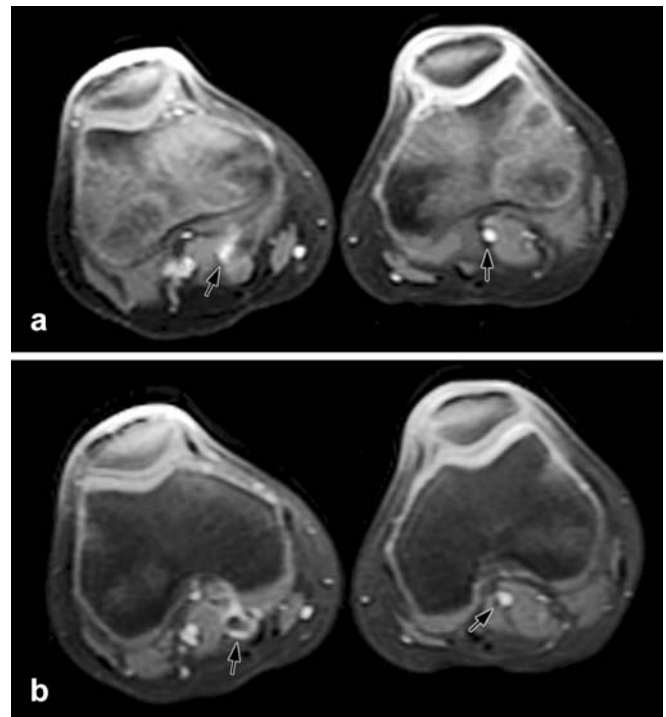
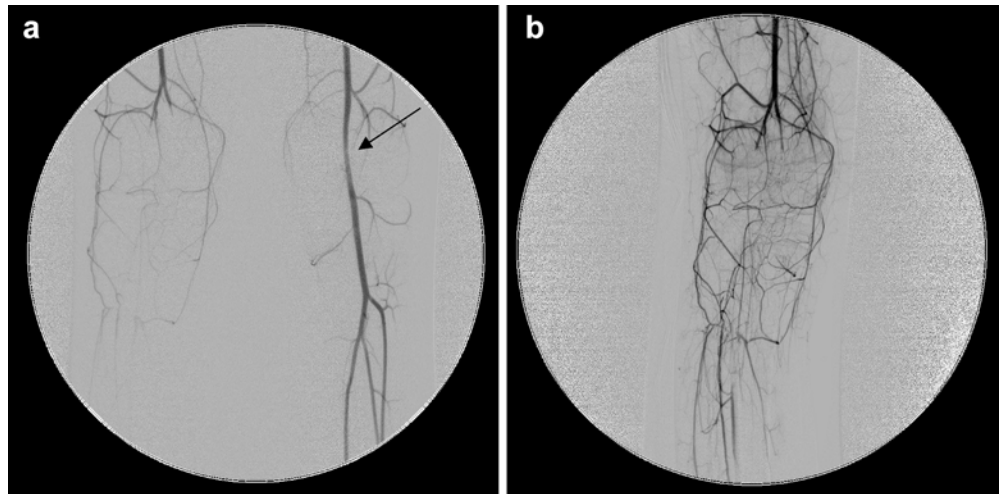


Fig. 3 Axial GRE images of both knees demonstrate bilateral type I popliteal entrapment syndrome. Black arrows pointing to the thrombosed right popliteal artery and a patent but medially deviated left popliteal artery

Fig. 4 Lower extremity angiogram. **a, b** Demonstration of thrombosis in the proximal right popliteal artery (**a**) with excellent collateral supply to the distal right leg (**b**). Note subtle medial deviation of proximal left popliteal artery (*arrow*), consistent with type I popliteal entrapment syndrome



In 1965, Love and Whelan, from Walter Reed General Hospital in the United States, introduced the term “PAES” [3]. It refers to compression of the popliteal artery caused by an abnormal anatomical relationship between the vessel and neighboring musculotendinous structures [4]. It is most often seen in young men with manifestation of calf pain during exercise and intermittent or progressive claudication [1, 3–5]. The arterial compression may cause chronic vascular microtrauma with local premature arteriosclerosis and thrombus formation with distal ischemia. In addition, stenosis and turbulent flow can cause post-stenotic ectasia and aneurysm formation, as in our case [4]. Compression characteristically occurs on passive dorsiflexion and active plantar flexion of the ankle. However, this can also be seen in individuals with normal anatomy [4]. The incidence of PAES in autopsy series may be as high as 3.5%, although clinically significant entrapment is seen in only 0.17% of patients, with a male-to-female ratio of 9:1 [2].

The popliteal artery and vein normally run down in the popliteal fossa between the heads of the gastrocnemius muscle [2]. PAES occurs when there is anomalous course of the artery around a normally placed or anomalously placed muscle [5]. There are various classifications of PAES. The most acceptable, proposed by Whelan and modified by Rich, divides PAES into five subtypes [1]: type I is described as an aberrant medial arterial course around the normal medial head of the gastrocnemius muscle; type II is where the medial head of the gastrocnemius muscle has an aberrant origin arising from the intercondylar region more laterally and inferiorly rather than from the medial femoral condyle, displacing the popliteal artery medially; in type III, the artery is in a normal position, but an aberrant accessory slip from the medial head of the gastrocnemius muscle wraps around the popliteal artery and entraps it; in type

IV the popliteal artery is located deep to the popliteus muscle or beneath fibrous bands in the popliteal fossa; type V is any form of entrapment that involves the popliteal vein [1, 4]. The second type is considered the most common and is found in 57% of patients [1]. PAES is known to be bilateral in one-third of cases, for which surveillance of the contralateral popliteal artery should be carried out [2, 4]. Our patient is considered to have bilateral type I PAES.

In pediatric patients, ischemic symptoms can often be misinterpreted as musculoskeletal pain [2]. Other possibilities would include sterile or septic emboli in right-to-left cardiac shunting, thrombophilia, hypercoagulable states, and small-vessel disease such as vasculitis [2]. The literature reports delays of years, with an average of 12 months, between the onset of symptoms and correct diagnosis [1, 5, 6].

Although PAES is uncommon and difficult to diagnose clinically, it must be considered in the healthy child or young adult presenting with acute limb ischemia or intermittent claudication [1, 3]. Early and correct diagnosis is important because PAES is a progressive condition and early treatment of the symptomatic patient may prevent devastating vascular complications or limb loss. An irreversible lesion of the popliteal artery can manifest like aneurysmal dilatation, thrombosis, or embolism, which can lead to ischemia and threaten the viability of the limb [3]. Careful history and clinical examination are the most important steps in the diagnosis [3, 6]. The radiological diagnosis can be confirmed by non-invasive means such as duplex sonography, CT, MRI, and MR angiography [1, 3]. Digital subtraction arteriography may have a role, but it has the disadvantage of being invasive and not sufficient by itself to evaluate PAES, especially in cases of arterial occlusion [1]. In our case, duplex ultrasonography, MRI, MRA, and DSA were performed.

Popliteal artery entrapment syndrome should be treated surgically regardless of the symptoms [1]. When the diagnosis is made at an early stage and the popliteal artery is intact, the treatment of choice is complete division of the abnormal muscle and transposition of the popliteal artery laterally. If the diagnosis is made after the artery is damaged or occluded, the treatment of choice is a bypass procedure with autologous vein, using

the contralateral long saphenous vein, in addition to division of the anomalous musculotendinous structure [2, 3, 7]. Thromboembolectomy with intra-arterial thrombolysis has been reported as another option prior to surgical transposition [2, 7]. The contralateral popliteal artery should be examined because PAES can be bilateral; that should always lead to surgical correction of the asymptomatic side, as well [2].

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