CASE REPORT

Case Report: Tibial and fibular osteochondroma as an unusual cause of popliteal artery entrapment syndrome [version 1; referees: awaiting peer review]

Jamil Victor de Oliveira Mariúba, Marcone Lima Sobreira, Winston Bonetti Yoshida, Eduardo Savio de Oliveira Mariúba, Hamilton de Almeida Rollo, Regina Moura, Matheus Bertanha, Rodrigo Gibin Jaldin, Rafael Elias Farres Pimenta, Paula Angeleli Bueno de Camargo, Mariana Thais Silva Secondo

Department of Surgery and Orthopedics, Disciplina de Cirurgia Vascular e Endovascular, Faculdade de Medicina de Botucatu, Universidade Estadual Paulista (UNESP), São Paulo, Brazil

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Abstract

Background: Osteochondroma, or osteocartilaginous exostosis, is the most common benign neoplasm of bone, and accounts for 20-50% of all benign tumors. Vascular complications associated with osteochondromas are rare, and include pseudoaneurysm formation, vessel occlusion and vessel displacement. To date, only two cases of popliteal artery entrapment syndrome (PAES) caused by an isolated fibular osteochondroma have been reported.
Case Report*:* This report describes a unique case of PAES. A 33-year-old woman had a history of multiple osteochondroma, including of the proximal tibia and fibula on the left, diagnosed at age two years and monitored clinically by an orthopedist. The patient presented at our facility with a one-year history of a progressive intermittent claudication, left-sided toe pain and pallor in cold weather. After a complete evaluation, we diagnosed an arterial occlusion of the left popliteal artery. We tried several attempts of revascularization, by different forms, without success. The case went to amputation surgery.
Conclusion: We consider this an important case because, although the association of osteochondroma and PAES is rare, physicians should consider it early to avoid acute vascular complications. Moreover, to date, we believe this is the first description of a PAES related with multiple osteochondroma.

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Corresponding author: Jamil Victor de Oliveira Mariúba (jamilvictor@gmail.com)

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Introduction

Popliteal artery entrapment syndrome (PAES) is an infrequent condition, generally caused by embryonic abnormalities and characterized by deviation of the popliteal artery from its normal course and subsequent compression. During muscular exertion, blood flow to the leg muscles is reduced, leading to intermittent claudication, pallor, and coldness[1–3]. PAES may be classified into six types, with medial deviation of the popliteal artery around the medial head of the gastrocnemius being the most common form[3].

Osteochondromas are the most common benign tumors of bone (30%), and generally occur in the metaphyseal area of long bones[4,5]. The vast majority (85%) of osteochondromas present as single lesions and are not hereditary. Conversely, approximately 15% of cases present as multiple osteochondromas, which are characteristic of the autosomal dominant disease hereditary multiple osteochondromas (HMO), or hereditary multiple exostoses (HME); 62% of cases have a positive family history[5,6].

Osteochondromas occur in 3% of the overall population. The solitary form exhibits no gender predominance, whereas the hereditary form has a 1.5:1 male-to-female ratio; it is more common in the Caucasian population, but its overall prevalence is lower than that of solitary osteochondroma (0.9 to 2 cases per 100,000 population)[5,6].

Osteochondromas take the form of cartilage-covered masses that grow exophytically on the surface of bones. They develop in childhood and adolescence, usually as slow-growing, painless lesions. However, depending on the site of the tumor, significant symptoms may develop as a result of complications, including fractures, bone deformity, mechanical joint abnormalities, and neurovascular involvement. Malignant transformation may occur later in life, during adulthood (0.5–5% of cases), but metastases are rare. Proliferation of osteochondromas usually ceases after puberty[5–7].

The treatment of choice is resection, provided the skeleton has matured[5]. Vascular compression, arterial and venous thrombosis, aneurysms, and pseudoaneurysms are common findings, occurring in 91% of cases; pseudoaneurysms are the most prevalent vascular complication[5,8].

Osteochondroma is an exceedingly rare cause of PAES. To date, only two case reports have been published in internationally indexed journals[1,4]. The present report describes a single case of popliteal artery entrapment secondary to multiple osteochondroma, treated by the present authors.

Case report

A 33-year-old woman had a history of multiple osteochondroma, including of the proximal tibia and fibula on the left, diagnosed at age two years and monitored clinically during childhood and adolescence by orthopedic specialists at an outside facility, with family history of osteochondroma (a sister, without complications). As the patient was pain-free and had no fractures or functional changes at the knee joint, resection was not indicated from an orthopedic standpoint.

The patient presented at our facility with a one-year history of left-sided toe pain and pallor in cold weather. Approximately one month before, she had experienced a sudden onset of intermittent claudication at 400 m; her maximum walking distance had decreased progressively to 100 m in the intervening time.

Physical examination revealed pallid discoloration of the hallux, second toe, and third toe of the left foot, with localized pain. The popliteal and distal pulses were absent in the left lower extremity, and a temperature gradient was felt on the affected foot.

A preoperative arterial duplex scan revealed left popliteal artery occlusion. Arteriography also showed occlusion of the superficial femoral artery, consistent with a thrombus.

A thromboembolectomy was performed using a Fogarty catheter inserted through the superficial femoral artery, with removal of fresh thrombi. The catheter could not be advanced past the juxta articular portion of the popliteal artery. Intraoperative angiography demonstrated a patent anterior tibial artery (Figure 1). The popliteal pulse returned at the end of the procedure, but distal pulses remained absent, although flow was present on Doppler analysis.

Postoperatively, the patient underwent CT angiography (Figure 2–Figure 4), which revealed a mass on the head of the fibula and tibial plateau, encircling the juxta articular portion of the popliteal artery. There was no contrast uptake in the arterial lumen distal to the site of the tumor.

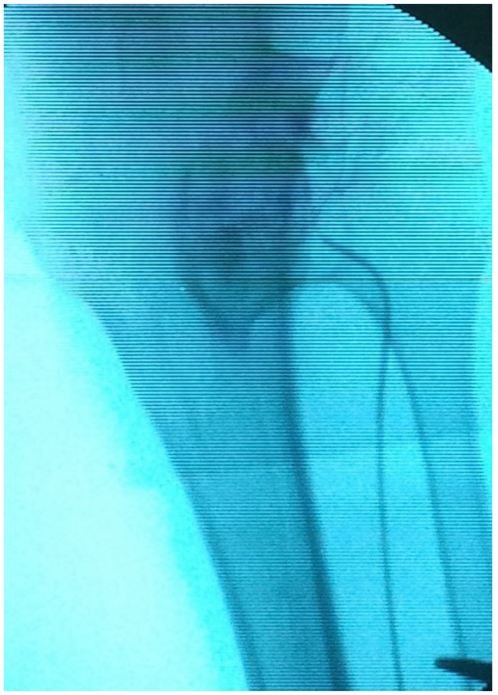

Figure 1. Arteriography showing distal filling of the anterior tibial artery. A bony tumor is visible in the left fibular head.

A second Fogarty thromboembolectomy of the posterior tibial artery was then performed, with creation of a femoral-to-posterior tibial bypass greater saphenous vein graft placed in an inverted position, which was unsuccessful. A femoral-to-anterior tibial graft could not be fashioned due to anatomical limitations (the tumor obstructed the usual course of the bypass graft).

The patient was started on alprostadil (200 micrograms diluted into 200 milliliters of saline every 12 hours) which was discontinued after 24 days due to elevated liver enzymes. Toe pallor progressed to wet gangrene of the forefoot (Figure 5) and the patient developed intractable pain, fever, and confusion. We then performed a knee disarticulation amputation, which healed satisfactorily and produced clinical improvement (sensitivity and motor skills preserved without pain). The patient was discharged to outpatient follow-up (returned at 14 days to evaluate the healing, at 3 months to evaluate physical therapy and each year). She adapted successfully to a prosthesis and leads a virtually normal life.

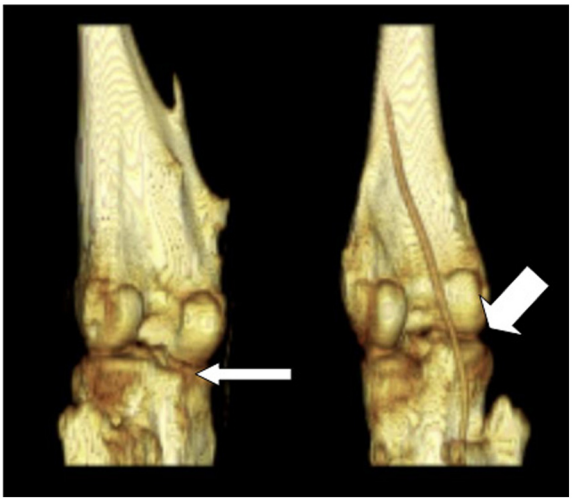

Figure 4. Three-dimensional reconstruction of CT angiogram demonstrating absence of contrast uptake in the left popliteal artery (thin arrow) and contrast enhancement of the right popliteal artery (thick arrow).

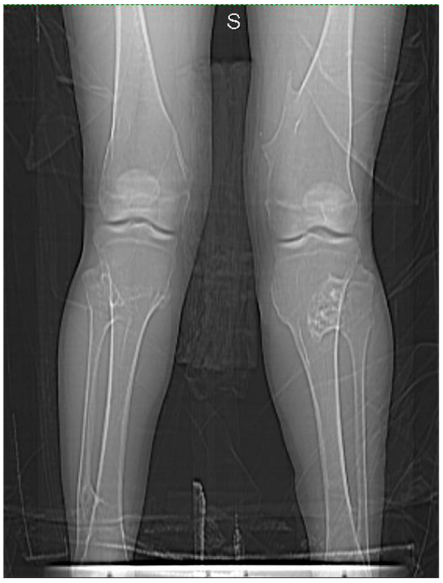

Figure 2. CT scout view showing a mass lesion in the left fibular head.

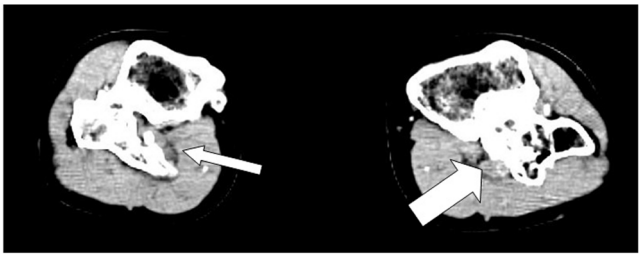

Figure 3. CT angiography views comparing the right (thin arrow) and left (thick arrow) popliteal arteries. The tumor is seen to completely envelop the left artery.

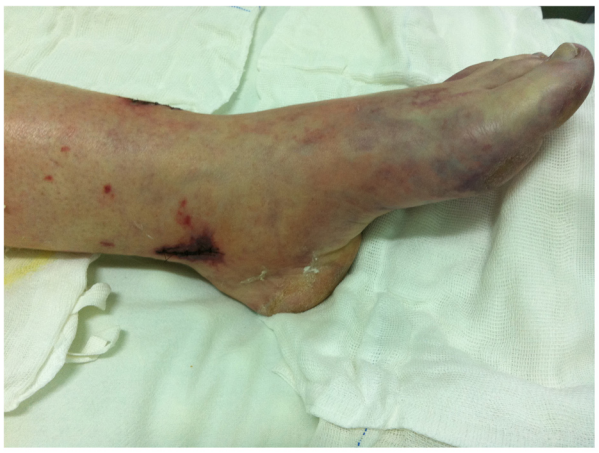

Figure 5. Progression of gangrene in the affected extremity.

Discussion

In younger patients, intermittent claudication, due to peripheral arterial disease, is a rare presentation. Its causes include early-onset atherosclerosis, trauma, tumors, arteritis, and fibrous dysplasias, but the most common etiology is PAES[1–4]. Entrapment may occur due to derangements in embryonic development of the popliteal artery or of the musculotendinous components of the popliteal fossa, leading to deviation of the artery from its normal course or to the development of anomalous structures that compress the artery. Consequently, distal blood flow is reduced during muscle contractions, producing intermittent claudication, pallor, and coldness of the extremity. These signs and symptoms usually remit at rest[1–3].

Resection is usually indicated for asymptomatic, incidentally detected, solitary osteochondromas, so as to prevent future complications. Surgical resection is also recommended for painful or continuously growing lesions, joint involvement, vascular

complications with intermittent claudication, or evidence of malignant transformation[5].

Osteochondromas are generally asymptomatic, and over 130 case reports of vascular complications have been published, with pseudoaneurysm and venous thrombosis being the most common such complications[7]. Eschelman *et al*. reported that among 56 cases of osteochondromas with vascular involvement, only six were tibial and only half of these caused arterial compression, as observed in our patient[9].

Guy *et al*[4]. reported a case of popliteal artery stenosis secondary to osteochondroma, presenting with left lower extremity pain and edema. The pain was initially exertional and resembled intermittent claudication, but eventually became continuous. Distal arterial pulses were palpable in both lower extremities, and a duplex scan of the popliteal arteries showed a normal triphasic waveform bilaterally. On the left side, blood flow velocity decreased during calf muscle contraction, and was followed by reactive hyperemia on relaxation, indicating popliteal artery entrapment. Signs of venous hypertension were also present (edema and varicose veins around the medial malleolus). Plain radiography and magnetic resonance imaging (MRI) revealed a fibular tumor compressing the artery[4].

Holzapfel *et al.*[1] reported another case of popliteal artery entrapment in which Doppler ultrasonography and MRI revealed a partially compressed, but not occluded artery, with preserved distal pulses. Venography showed a completely compressed popliteal vein, mimicking deep venous thrombosis. The patient was treated for four weeks with low molecular weight heparin, to no effect, before being referred for correct diagnosis and management[1].

In 2013, Henry *et al.*[9] reported a case of intermittent claudication in a 23-year-old woman with a solitary tibial osteochondroma, which was treated by division of the soleus muscle and resection of the osteochondroma.

A diagnosis of PAES should be considered in all young patients with intermittent claudication. Pain occurs in the foot and calf muscles, usually after strenuous exercise. Spastic claudication, in which patients are pain-free while running, but paradoxically experience pain while walking, may also occur; some patients report pain when standing on tiptoes[3].

Diagnostic confirmation usually begins with a duplex scan, which enables dynamic visualization of the popliteal artery and demonstrates its patency at rest and stenosis or occlusion in response to functional maneuvers[3]. MRI and CT angiography may aid in diagnosis by identifying the musculoskeletal or tendinous structures implicated in compression[3].

Arteriography plays an important role in diagnosis and surgical treatment planning, and is indicated whenever arterial lesions, such as aneurysmatic degeneration or arterial thrombosis, are suspected. Osteochondroma (osteocartilaginous exostosis) is a cartilage-capped bony projection arising on the external surface of bone containing a marrow cavity, which is continuous with that of the underlying bone[6]. To establish the diagnosis of osteochondroma and to ensure good visualization of the bones, anteroposterior and lateral views of both lower extremities should be obtained, with the foot in passive dorsiflexion and active hyperextension.

In the case reported here, the patient presented with signs and symptoms of acute ischemia. This is an unusual manifestation of osteochondroma with arterial involvement, and this limited our approach to the case, because we were prepared for an embolus and what actually happened was a thrombosis. Even after two attempts at surgical revascularization for limb salvage, a major amputation of the affected extremity was required. Treatment failure in this case was attributed to the extent of local arterial involvement and by chronic distal occlusion.

In patients with osteochondromas in the vicinity of the popliteal artery, this vessel may be encircled by the tumor and chronically compressed, leading to endothelial injury, thrombus formation, and, eventually, critical ischemia. Although the association of osteochondroma and PAES is rare, it should be considered by physicians, particularly in the differential diagnosis of young patients with evidence of vascular involvement. Imaging should be performed early and periodically, so as to prevent diagnostic and therapeutic delay[1].

Consent

Written informed consent for the publication of the patient's clinical details and images was obtained from the patient.

Competing interests

No competing interests were disclosed.

Grant information

The author(s) declared that no grants were involved in supporting this work.

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