Popliteal pseudoaneurysm secondary to multiple hereditary exostoses

A 20-year-old man with multiple hereditary exostoses presented with a pulsatile swelling in the posterior-medial distal thigh. He gave a history that 5 weeks previously, after squatting down then standing, he experienced a sharp pain behind his right knee. Magnetic resonance (MR) imaging demonstrated a large haematoma associated with the popliteal artery. Ultrasound imaging visualized a 9-cm popliteal artery pseudoaneurysm with flow in the sac. Computed tomography (CT) prior to emergency open surgery further characterized the arterial injury (Fig. 1). A small hole in the posterior-lateral popliteal artery was simply closed with a single suture and the pseudoaneurysm and exostosis excised (Figs 2,3). At follow-up, no arterial compromise was evident. Elective excision of the contralateral exostosis is planned.

Multiple hereditary exostoses is a rare genetic autosomal-dominant benign bone disorder with a male predominance affecting somewhere between 1:50 000 and 1:100 000 in Western populations.1 Mutations have been identified in the EXT family of genes with about two-thirds of patients displaying a mutation in the tumour-suppressor EXT1 gene on chromosome 8q23-q24 and the remaining third the EXT2 gene on chromosome 11p11-p12.2 The condition leads to cartilage-capped tumours (osteochondroma) growing outward from metaphyses of long bones, growth plates or from the surface of flat bones throughout the body. Patients typically have multiple osteochondromas often arising from long bones around the knee, and in the majority of cases they are usually asymptomatic. Malignant degeneration occurs rarely in solitary osteochondromas but where multiple osteochondromas are present the lifetime risk is estimated between 1% and 5%.2 Histopathological assessment of malignant change can be challenging but the presence of a cartilaginous cap >1.5 cm is suspicious of secondary peripheral chondrosarcoma.3 Indeed, for low-grade chondrosarcomas, the radiological depth of the cartilaginous cap is a more reliable marker of malignant change than cellular features on histology. Thus, standardized radiological reporting has been devised to aid diagnosis...
with a cap greater than 2 cm 100% sensitive and 98% (MR)/95% (CT) specific for malignancy. Where possible, CT or MR imaging should be performed prior to surgical intervention.

Vascular compromise as a result of hereditary multiple exostoses is rare with around 100 cases in the literature, about 40 involving popliteal pseudoaneurysms. Other than pseudoaneurysm formation, claudication, acute limb ischaemia, distal embolization, local compression symptoms (peroneal nerve palsy) and venous thrombosis have all been reported. A high degree of suspicion must be maintained with only 39% of cases presenting with pulsatile mass on examination. Ossification of the cartilaginous cap is thought to yield a sharp spike capable of penetrating the adjacent artery, and a history of vigorous or repetitive exercise or direct trauma often precedes presentation.

The management of popliteal pseudoaneurysms in general has evolved with increasing use of minimally invasive and endovascular techniques. Thrombin injection, coil embolization and endovascular-covered stent placement have all been used to manage pseudoaneurysms in the acute setting. Endovascular stent grafting, unless combined with ultrasound-guided aspiration of haematoma, fails to deal with any mass effect from the pseudoaneurysm but more importantly does not address the underlying bony lesion, risking recurrence. In addition, patients with multiple hereditary exostoses are young and active, representing a very different cohort to patients with degenerative popliteal aneurysms in whom endovascular management has been trialled. Even in the elderly, the durability of endovascular stent graft repair has been an issue, with a recent review noting 42/251 stent graft failures at mean post-operative time of 10.8 months leading to a secondary intervention rate of 18.7%. In this review, a fracture rate of 5.6% was noted, but in a younger, more active population with a bony lesion near the vessel a higher fracture rate may be expected.

For this reason, open surgery remains the mainstay of treatment offering a durable repair and the opportunity to excise the causative exostosis. Vasseur et al., in reviewing 103 cases of arterial compromise (83% of which involved the popliteal artery), found simple arterial repair was sufficient in 46% of cases with a further 15% managed by resection of the damaged region and end-to-end anastomosis. The remaining cases were managed with surgical graft replacement (vein where possible) and in a small number of cases angioplasty or arterial ligation. Excision of the exostosis can be easily performed via standard open surgical exposure and achieved with nibblers or bone cutters. The bone should be sent for histological examination.

**References**


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