Waxing and waning of joint motion

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Abstract

Knee pain, stiffness and swelling are common complaints. Trauma and osteoarthritis are the most common diagnoses. However, benign and malignant tumours may also present around the knee. These tumours require characterization prior to definitive treatment. Here we describe a rare cause of knee stiffness with a popliteal mass.

Keywords

Knee pain; knee stiffness; melorheostosis; dripping candle wax sign.

Case report

A 32-year old female presented following a 5-year history of progressive left knee stiffness. Walking up and down stairs had become increasingly difficult because of the stiffness. The patient denied having sustained an injury to the knee and did not suffer from pain or swelling of the knee. There was no significant past or family history of bone or joint disease.

On physical examination there was a fixed-flexion deformity of the left knee with a reduced range of motion, 10–45 degrees of flexion. The left gastrocnemius was wasted, and a diffuse swelling within the popliteal fossa was apparent. The mass in the popliteal fossa was poorly defined and non-tender. There was no demonstrable joint effusion and the knee was stable. Both lower limbs were neurovascularly intact and there were no palpable lymph nodes. Examination of the spine and hips was unremarkable. The differential diagnosis for a swelling in the popliteal fossa includes most commonly Baker cyst, lipoma, neuroma, deep venous thrombosis and more rarely pigmented villonodular synovitis (PVNS).

Plain radiographs of the left knee were obtained and revealed aggregates of calcific density both within the popliteal fossa and extending along the posterior cortices of the distal femur and proximal tibia. This radiographic appearance could be likened to dripping candle wax (Fig. 1). Further radiographs of the pelvis, femur, leg, ankle and foot were obtained and showed the medial cortices of the femur, tibia, navicular, first cuneiform, first metatarsal and phalanges to be similarly involved (Fig. 2). Extraosseous densities, in addition to the popliteal mass, were
identified adjacent to the proximal fibula and medial to the ankle joint. Radiographs of the contralateral lower limb were unremarkable.

Computed tomography (CT) and magnetic resonance imaging (MRI) were used to further characterize the extraosseous mass within the popliteal fossa. Lobulated synovial calcification and proliferation was evident with nodular areas of T2 hypointensity (Fig. 3). These features were thought to be consistent with PVNS. The linear pattern of hyperostosis in the left femur and tibia were again evident on both CT and MRI. These features are pathognomonic of melorheostosis, a rare hyperostotic condition that is distinguished by a linear pattern of distribution along the axes of long bones[1].

Discussion

PVNS and melorheostosis are both exceedingly rare conditions. Melorheostosis has an estimated incidence of 1 case per million[1], while the incidence of PVNS is 1.8 cases per million[2]. The prospect of having arrived at 2 such diagnoses simultaneously in this case demanded diagnostic parsimony. The 14th century logician William of Occum gave us the dictum, Pluralitas non est ponenda sine necessitate—otherwise put, “among competing hypotheses, favour the simplest one”[3], and this case illustrates the ongoing utility of Occum’s razor (or scalpel?) in surgery. In this case, many surgeons would proceed to confirm the diagnosis by biopsy, and this was indeed considered, however the extraosseous calcification in the popliteal fossa and the unilateral hyperostosis of the femur, tibia and foot are best explained as belonging to a single diagnostic entity: melorheostosis.

Leri et al.[1] first described melorheostosis in 1922 as a pattern of hyperostosis with a longitudinal pattern of distribution for the long bones. This is the classic pattern of disease. Further subtypes of disease have been proposed by Freyschmidt et al.[4], including osteoma-like, myositis ossificans-like and osteopathia striata-like variants. The pathogenesis of melorheostosis remains unknown, although most authors agree that it is non-hereditary in nature. The linear
distribution of hyperostosis suggests a developmental origin in the limb buds. Alternatively, Murray et al.[5] have proposed that melorheostosis may be the result of a postnatal segmental sensory nerve neuropathy, similar in nature to herpes zoster infection and shingles. This theory has arisen from the observation that the bone and soft tissues affected by melorheostosis belong to functional units called sclerotomes, analogous to dermatomes or myotomes.

Melorheostosis is not associated with increased mortality and it is primarily contractures of the soft tissues and joints that give rise to clinical symptoms. The lower limbs are most commonly affected. Joint stiffness, pain and deformity are the most common presentations although the dripping candle wax sign may be an incidental finding on radiographs[1]. Premature closing of the epiphyseal plate by hyperostosis may lead to growth disturbance with leg length discrepancies of up to 10 cm documented in the literature[6]. Soft tissue and muscle involvement is common, along with skin changes. The skin over the affected bones may appear thickened, shiny and erythematous. There are 3 case reports of osteosarcoma arising from bone affected by melorheostosis[7–9].

**Teaching points**

Both non-operative and operative approaches to managing melorheostosis have been suggested[11]. Non-operative options include manipulation, serial casting, nerve blocks and oral medication such
as bisphosphonates, non-steroidal anti-inflammatory drugs and nifedipine\textsuperscript{[10]}. Empirical evidence for such treatments is lacking. Operative options aim to restore mobility and include tendon lengthening, limb lengthening, excision of hyperostoses and even amputation. The best outcomes result from bony procedures alone, while joint contractures commonly recur after soft tissue releases. Surgery for the purpose of pain relief is ineffective\textsuperscript{[1,10]}.  

**Fig. 3.** T2-weighted sagittal MRI of the left knee showing a lobulated mass within the popliteal fossa.

**References**