Case Report

Melorheostosis With Heterotopic Ossification

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Melorheostosis is an uncommon, non-hereditary form of hyperostosis characterized by linear hyperostosis along the major axes of long bones.\(^1\)\(^2\) It is thought to be an embryonal mesodermal disorder that affects both the osseous and soft tissues. The segmental distribution of melorheostosis corresponds most closely to that of the sclerotome, which is defined as an area of the skeleton innervated from a single spinal segment. Heterotopic cartilage and bone are occasionally present in the para-articular soft tissues of affected joints,\(^3\)\(^4\) but are rare in the soft tissues surrounding intact joints without any bone abnormality. In the latter situation, it may be difficult to diagnose the heterotopic ossification as a part of melorheostosis.

This article presents a case of melorheostosis with heterotopic ossification at the popliteal region, where the distal femur and proximal tibia showed no melorheostotic changes, and the common peroneal nerve penetrated the heterotopic bone without any neurologic symptoms. A possible correlation between the heterotopic ossification and the segmental nerve supply to bone and soft tissue also is discussed.

**CASE REPORT**

A 38-year-old woman presented with progressive restriction of flexion in her right knee of 3 years' duration. Physical examination revealed a subcutaneous, bony, hard mass (4×6 cm) in the right popliteal region. Range of motion in the right knee was from 0° extension to 110° flexion. No abnormalities were detected with routine neurological examination.

Plain radiographs revealed irregular calcifications close to the capsule of the right knee without any bone involvement (Figure 1). Computed tomography (CT) revealed a small radiolucent hole within the heavily calcified mass (Figure 2). In addition, hyperostotic lesions (compatible with melorheostosis) in the fibula, calcaneus, cuboid, the lateral three metatarsal bones, and the lateral three digits of the foot were detected (Figure 3). The remainder of the skeleton was normal. Technetium 99m-methylene diphosphonate bone scan showed highly increased radionuclide uptake in the areas of radiographic hyperostosis and moderate increased radionuclide uptake in the heterotopic calcifications.

En bloc excision of the calcified mass in the popliteal region and biopsy of the cuboid were performed. The upper part of the bony hard mass was located intermuscularly between the biceps femoris and the semimembranosus and...
Concerning the etiology of melorheostosis, such as a vascular disturbance, an inflammatory process, or a congenital origin. In 1979, Murray and McCrede drew attention to the peculiar segmental distribution of this disease and detected a good correlation between hyperostotic lesions and the sclerotomes, representing the zones of the skeleton supplied by individual spinal sensory nerves.

In the present case, the sclerotic lesions of the bones corresponded with S1-S2 segments of the sclerotome map. Interestingly, the soft-tissue ossification in the popliteal region also was located in S1-S2 segments of the myotome. In addition, the common peroneal nerve, which penetrated the heterotopic bone in the present case, originates from the nerve roots of L4, L5, S1, and S2. These findings strongly support Murray and McCrede’s hypothesis that ossification of soft tissue may be related to involvement in a corresponding myotome. Since the heterotopic ossification proved to be histologically equivalent to the bony lesions of melorheostosis, this suggests the hyperostotic changes of both types of lesions may be evoked due to some pathologic conditions of the same spinal nerve segment.

Another conspicuous finding from this case was the entrapment of the common peroneal nerve by the heterotopic bone without any neurologic abnormalities. Generally, peripheral nerve compression or palsy by heterotopic bone is rare. In our case, the peroneal nerve was not invaded by the heterotopic bone, suggesting bone formation may be inhibited by contact with the nerve. Although the inhibitory mechanism of bone invasion into nerve remains unknown, our observations indicate that certain nerve structures or nerve secretions may inhibit bone growth into nerves.

Finally, conditions that may produce a calcified or ossified para-articular mass include myositis ossificans, synovial osteochondromatosis, tumoral calcinosis, and soft-tissue sarcomas such as extraskeletal osteosarcoma or calcified synovial sarcoma. When a heterotopic ossification is seen adjacent not only to sclerotic bones but also to intact bones, melorheostosis should be included in the differential diagnosis.

REFERENCES