

IMAGES IN CLINICAL RADIOLOGY



Melorheostosis of the foot

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A 48-year old male patient of Arabic origin presented to the orthopedic consultation with a history of slow progressive pain, joint stiffness and swelling of the left foot. Local tenderness was present and a painful, hard mass was clearly palpable. Conventional radiography (Fig. A) revealed undulating and sclerotic enlarged areas (candle wax-appearance) in the lateral cuneiform bone, the third metatarsal bone and the proximal, intermediate and distal phalanx bone of the third row. Cortical hyperostosis was also apparent in the middle phalanx and metatarsal bone of the second row. Areas of extraosseous bone formation could be seen, most clearly adjacent to the metatarsal head and the middle phalanx of the third row. A diagnosis of melorheostosis was made. A CT-scan, which is not strictly required to make the diagnosis, confirmed the radiographic findings (Fig. B). The patient was treated conservatively.

Comment

Melorheostosis was first described by Leri and Joanny in 1922. It is a rare, non-hereditary disease. Characteristically, the cortical lesions are progressive and may result in narrowing of the medullary canal and stenosis of an adjacent lumen, foramen, or of the spinal canal. Motor or sensory nerves may be compressed and become symptomatic. The cortical hyperostosis may extend into nearby joints and cause loss of motion. Extensive soft tissue masses may develop, most of which are adjacent to the involved bone, but some may be unconnected to the bone. The soft tissue masses become more ossified over time. Patients with melorheostosis may have associated lesions such as vascular malformations, neurofibromatosis, hemangioma, arterial aneurysms, linear scleroderma, tuberous sclerosis, hemangiomas, and focal subcutaneous fibrosis. The cause of melorheostosis is unknown, but one theory proposed is that the lesions arise from an abnormality of the sensory nerve of the affected sclerostome. A single bone may be involved (monostotic) or several bones may be involved (polyostotic). The affected bones are usually related to the same sclerostome. Isolated cases of malignancy have been reported in association with melorheostosis. Melorheostosis is usually apparent in early childhood and even in the first few days of life. In children the condition affects mainly the bones of the extremities and pelvis, and may result in limb length inequality, deformity, or joint contractures. About 50% of persons affected will develop the symptoms by 20 years of age. The condition occurs in both sexes. Melorheostosis has an estimated incidence of 0.9 cases per million persons. Biopsy shows a variable degree of marrow fibrosis, along with markedly irregular bone constituted of mixed areas of lamellar and woven bone. A mixture of osteocartilaginous, fibrovascular, and adipose tissue is seen in the soft tissue masses. The clinical course is slowly progressive. Severe symptoms may require treatment by sympathectomy or even amputation.

Reference

1. Kalbermatten N.T., Vock P, Rüfenacht D., Anderson S.E.: Progressive melorheostosis in the peripheral and axial skeleton with associated vascular malformations: imaging findings over three decades. *Skeletal Radiol*, 2001, 30: 48-52.

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