Ollier's disease (multiple enchondromatosis)

Objectives
1. Describe the radiographic features of solitary enchondroma
2. Describe the clinical and radiographic features of Ollier's disease
3. Describe orthopaedic problems secondary to Ollier's disease
4. Discuss risk of chondrosarcoma in relation to Ollier's disease

Discussion point
1. What is Maffucci's syndrome?

Discussion

Solitary enchondroma is relatively common. The most common perception of its genesis is that a segment of growth plate does not ossify and persists as an ovoid or linear radiolucent extension of cartilage cells into the metaphysis and sometimes the diaphysis. In children, they can cause pathologic fracture, but are more often incidental findings on radiographs taken for some other reason. Metaphyseal bone affected by large enchondromas does not funnelize normally and may appear expanded due to the absence of remodeling. Forty percent of solitary enchondromas involve the hands or feet, usually the phalanges.

Ollier's disease is an uncommon condition of multiple enchondromatosis. Most patients have bilateral involvement, but more severe on one side. Unilateral involvement also occurs. Genu varum is very common, other angular deformity can result depending on the location of the lesions. Leg length discrepancy is common; but bone affected with an enchondromatous lesion can be lengthened, sometimes dramatically. The incidence of malignant differentiation of multiple enchondromatosis is high, 30% in one report from the Mayo clinic. The rare Maffucci's syndrome consists of multiple enchondromatosis accompanied by soft tissue hemangiomas, and, in an historical quirk, was described even before Ollier's disease. The potential for malignant degeneration in Maffucci's syndrome is also very high. Cytogenetic study of a low grade chondrosarcoma in a patient with Ollier's disease revealed a deletion at the same site as was found in patients with chondrosarcoma. Undoubtedly, there will be reports of further investigation into the cytogenetics of these neoplasms in the near future.

References


