Clinical Quiz

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Polyostotic fibrous dysplasia

Clinical Presentation

A 25-year-old female patient presented to the orthopedic outpatient department with pain and gradually progressive deformity involving both the lower extremities for the last 6 years. She also complained of limping. There was no history of trauma or any family history of such deformities. Examination revealed an angular deformity involving the left femur. Radiograph of the left hip with proximal femur and radiograph of both the legs antero-posterior view were performed (**Figures 1 & 2**).

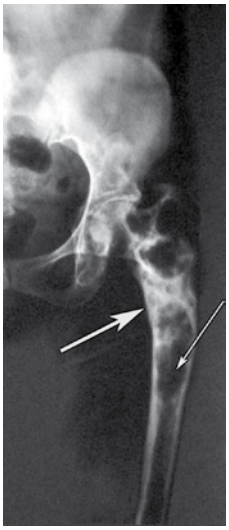

Figure 1 - Radiograph of the left hip.

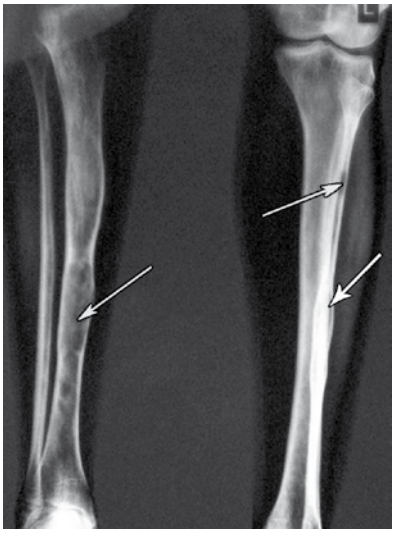

Figure 2 - Radiograph of both legs in the antero-posterior view.

Questions

1. What are the features seen on the radiographs?

2. What is the diagnosis?

3. What is the management?

Clinical Quiz

Answers

1. Radiograph of the left hip with proximal femur shows multiple radiolucent lesions involving the metaphysis and diaphysis of the left femur with ground glass appearance (thin arrow). Also, note the thinning of the cortex and endosteal scalloping. The bone is enlarged and deformed leading to a shepherd's crook deformity of the proximal femur (thick arrow). The radiograph of both legs in the antero-posterior view shows multiple radiolucent lesions involving the right tibia and left fibula (arrows) with ground glass appearance.

2. This 25-year-old female patient has polyostotic fibrous dysplasia. The diagnosis is usually made on the basis of clinic-radiological examination. Young female presenting with deformities of the long bones with classical radiological findings of multiple radiolucent lesions, ground glass appearance, cortical thinning and endosteal scalloping are suggestive of polyostotic fibrous dysplasia. Enlargement of the bone with progressive deformity leading to shepherd's crook deformity of the proximal femur is diagnostic of fibrous dysplasia.

3. Management is mainly conservative and depends upon the symptoms and size of lesion. For symptomatic or large lesions of fibrous dysplasia, initiation of long-term bisphosphonate therapy is found to be helpful in reducing symptoms and increasing cortical thickness.[1,2] Surgery is not required for an asymptomatic lesion unless there is a risk of pathological fracture. For patients with symptomatic dysplasia in the femur, surgical treatment alternatives include curettage and bone grafting.[3] If surgical treatment is required for fibrous dysplasia in long bones, intramedullary nails are recommended.[4]

Discussion

Fibrous dysplasia is a benign, gradually mounting non-familial, congenital disorder in which the normal cancellous bone is replaced by immature woven bone and fibrous tissue. Approximately 20-30% of fibrous dysplasia is polyostotic and 80% are monoostotic. The sites of involvement are the femur (91%), tibia (81%), pelvis (78%), ribs, skull and facial bones (50%), upper extremities, lumbar spine, clavicle, and cervical spine, in decreasing order of frequency.[5] Two thirds of patients are symptomatic before they reach 10 years of age. Often, the initial symptom is pain in the involved limb, usually associated with a limp, a spontaneous fracture, or both.[5] Radiographically, the lesions of fibrous dysplasia have a lucent or ground glass appearance and cause thinning of the cortex and endosteal scalloping. The bone may be enlarged or deformed leading to shepherd's crook deformity of the proximal femur as was seen in our patient (**Figure 1**). Non-surgical management of fibrous dysplasia using bisphosphonates is currently under investigation. Intravenous pamidronate has been shown to improve bone pain, decreases bone turnover, and improve the radiographic appearance.[6] Malignant transformation occurs in less than 0.5% of cases.[7]

References

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