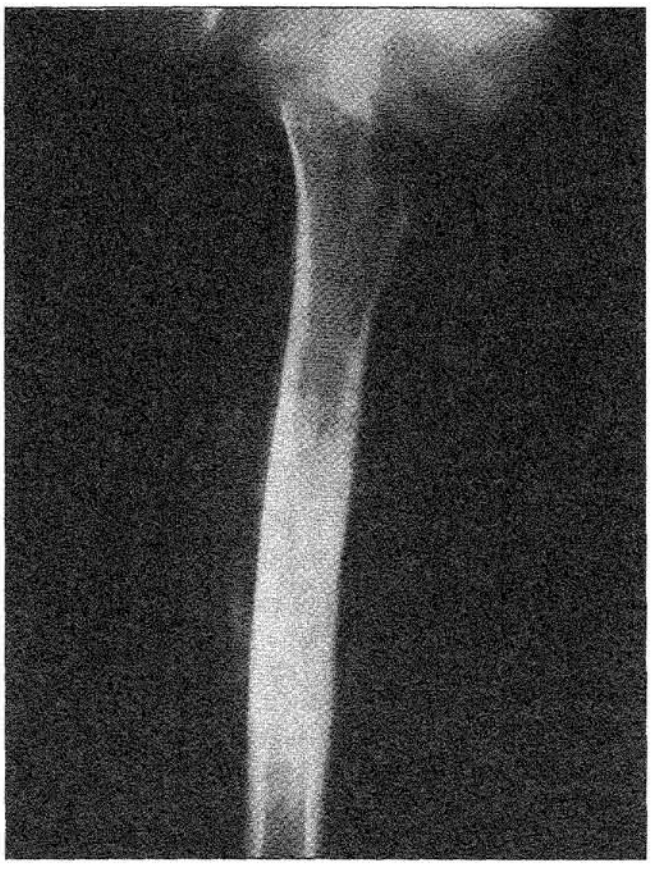
## Clinical History:

Radiograph of the femur (**Figure 1**) of a 13-year-old boy presenting with painful swelling of the mid-thigh. There was no history of trauma nor were there systemic symptoms.

**Figure 1: Lateral radiograph of the femur**



Answer on page 387

## What is the diagnosis?

a) Giant cell tumour

b) Stress fracture

c) Caffey's disease

d) Chronic osteomyelitis

e) Ewing's sarcoma

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# RADIOLOGICAL CONFERENCE

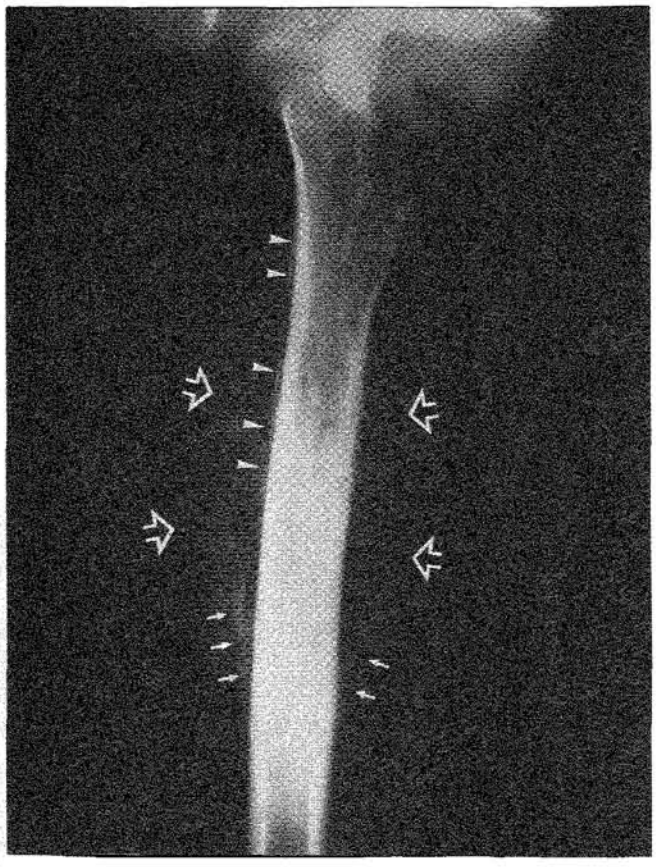
## Answer:

e) Ewing's sarcoma

## Radiological findings

Radiograph (**Figure 2**) shows ill-defined medullary destruction in the femoral diaphysis, with patchy areas of new bone formation. There is extension through the cortex to involve the soft tissues. Prominent periosteal reaction, including Codman's triangles, is present. Features are typical of a malignant bone tumour. In this age group, the two leading differential diagnoses are osteogenic sarcoma and Ewing's sarcoma. The latter diagnosis was confirmed by biopsy in this patient.

**Figure 2: Same radiograph as Figure 1 with the addition of arrows. Mixed lytic-sclerotic permeative destruction is present in the diaphysis of the femur. The lesion margins are ill-defined. Codman's triangles (small arrows) and *lamellated periosteal reaction* (arrowheads) are seen, as is a large soft tissue mass (open arrows)**



## Discussion

### Giant cell tumour

This tumour typically occurs in young adults between the ages of 20 and 40 years. Classically, it is located subarticularly in the epiphysis and metaphysis of a long bone, especially the distal femur. Other frequently affected sites are the proximal tibia, distal radius and the sacrum. Patients often present with pain of variable severity and swelling. Other symptoms include weakness, limitation of joint motion and pathological fracture. Radiographically, giant cell tumour is seen as an expansile lucency with ill-defined endosteal margins. Bony expansion is often eccentric and there may be cortical destruction and soft-tissue extension. Presence of internal septa may produce a multilocular appearance. This diagnosis can be excluded in this patient on the basis of age, site and radiographical appearances.

### Stress fracture

Stress fractures are usually associated with low-grade repetitive trauma, produced by activities such as jogging and marching. Typical sites include the shafts of the tibia and metatarsals. Affected patients usually present with persistent pain of the affected bone. Radiographically, the diagnosis of stress fracture should be considered when horizontal or oblique linear band-like sclerosis is seen in a weight-bearing bone. Occasionally, however, exuberant periosteal reaction covering the entire bone may be present. This aggressive appearance may mimic a bone tumour such as Ewing's sarcoma. Biopsy should be avoided as histologically, new bone formation within pre-existing bone trabeculae may resemble malignancy. Carefully obtaining a history of trauma and repeated radiographs after 1-2 weeks, sometimes supplemented by computed tomography (CT) and magnetic resonance imaging (MRI) for better lesion delineation and assessment of the surrounding soft tissues, are helpful in differentiating unusual stress fractures from bone tumours.

*(Continued on page 389)*

# RADIOLOGICAL CONFERENCE

### Caffey's disease

Also known as infantile cortical hyperostosis, Caffey's disease is typically seen in infants up to the age of 6 months. Multiple bones are involved, especially the mandible, ribs and clavicles. In long bones, the epiphyses and metaphyses are spared. Radiographically, the bones have a dense appearance due to florid periosteal reaction. In this patient, the diagnosis of Caffey's disease can be excluded from the age and lack of lesion multiplicity.

### Chronic osteomyelitis

Acute osteomyelitis may produce lytic lesions and layered periosteal reaction, usually near the ends of the shaft of long bones. It may therefore resemble malignant bone tumours such as Ewing's sarcoma. In chronic osteomyelitis, a geographical area of destruction may be present. The lesion may have sclerotic margins, bone expansion and joint involvement. The finding of a bony sequestrum, or island of dense dead bone, further enhances this diagnosis. However, antibiotic treatment may obscure the febrile and septic clinical course of acute osteomyelitis, and may also alter the normal radiographic progression of osteomyelitis. In summary, osteomyelitis produces a variety of radiographical appearances and should always be considered in the differential diagnosis of a lytic or sclerotic bone lesion.

### Ewing's sarcoma

Ewing's sarcoma is primarily found in children and adolescents, typically between 5 to 15 years of age. The commonest symptoms are pain and swelling. Patients with this tumour may sometimes have fever, increased erythrocyte sedimentation rate, anaemia and leucocytosis. Common sites affected are the femur, pelvis, humerus and shoulder girdle. Classically, it is seen as a permeative, i.e. consisting of multiple small holes, lesion in the diaphyses of a long bone. The tumour may stimulate reactive new bone formation, giving a sclerotic or mixed lytic-sclerotic patchy appearance, as seen in this patient. Ewing's sarcoma often has a lamellated (or onion-skinned) type of periosteal reaction, although 'sunburst' or amorphous types may also occur. Triangular elevation of the periosteum overlying a lesion penetrating the cortex produces the Codman's triangle. All these forms of periosteal reaction are considered to be aggressive.

Ewing's sarcoma may metastasize to other bones, the lungs and lymph nodes. Although radiographs are still the best single imaging modality for diagnosis of this tumour, other forms of imaging such as bone scintiscans, CT and MRI are required for staging prior to commencing treatment. Multi-agent chemotherapy has made a significant difference to the prognosis of Ewing's sarcoma, improving the 5-year survival rate from less than 10% to about 40%. Prognosis is also improved in patients who undergo complete surgical excision. Ideally, patients with this tumour should be treated with a combination of surgery, chemotherapy and in selected cases, irradiation. ■

## References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