OSTEOSARCOMA APPEARING AS A PATHOLOGIC FRACTURE

by E. VERHAVEN, H. DE BOECK and P. OPDECAM

Osteosarcoma is the most frequent primary malignant bone tumor. It usually occurs in the second decade of life. Pathologic fracture as the primary manifestation of an osteosarcoma is rather unusual, especially under the age of 10.
The authors present the case of a 9-year-old boy with a diaphyso-metaphyseal femoral fracture, in whom the radiological features of an osteosarcoma were not recognized at the time of the fracture as they were not clearly visible, and at follow-up initially were considered as proliferative callus formation.

Keywords : osteosarcoma ; bone tumor ; pathological fracture.
Mots-clés : osteosarcome ; tumeur osseuse ; fracture pathologique.

SAMENVATTING

E. VERHAVEN, H. DE BOECK en P. OPDECAM.
Pathologische frakturen bij osteosarcoma.

Osteosarcoma worden slechts zelden vastgesteld bij kinderen jonger dan tien jaar. Een pathologische fraktuur als de eerste manifestatie van een osteosarcoma is nog zeldzamer in deze leeftijdsgroep, doch moet steeds uitgesloten worden.
De auteurs beschrijven het geval van een 9 jarige jongen, aanvankelijk behandeld voor een fraktuur van de femurdiafyse, die achteraf een pathologische fraktuur van een osteosarcoma bleek te zijn.

RÉSUMÉ

E. VERHAVEN, H. DE BOECK et P. OPDECAM.
Fracture pathologique et ostéosarcome.

L'ostéosarcome est très rare chez l'enfant de moins de dix ans. Une fracture pathologique — première manifestation d'un ostéosarcome — est encore plus rare dans cette population, mais reste une hypothèse à examiner.
Les auteurs décrivent le cas d'un garçon de neuf ans, traité pour fracture diaphysaire du fémur, qui par la suite s'est avérée être secondaire à un ostéosarcome.

INTRODUCTION

Osteosarcoma, the most common primary malignant bone tumor, is usually diagnosed on clinical, radiological and pathological grounds (1, 2, 3, 6). It can occur in any age group, but more than half of the cases occur in the second decade of life (1, 2, 3, 6).
The incidence of pathologic fracture in osteosarcoma varies from 5 to 10% (6). It usually occurs in a later stage of the disease (1, 2). Pathologic fracture as the primary appearance of an osteosarcoma is very rare, especially under age 10 (1, 2).
The authors describe the case of a 9-year-old boy who was treated for a femoral fracture that afterwards proved to be a pathologic fracture in a high-grade osteosarcoma.

CASE REPORT

A 9-year-old boy was admitted to the hospital after a fall on the left leg while roller-skating in August 1989. He was unable to bear weight. Physical examination revealed a healthy-looking boy with painful mobilization of the left leg.

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Roentgenograms showed a spiral fracture in the middle and lower third of the left femoral diaphysis (fig. 1). Skin traction was applied, which gave good reduction of the fracture. Skin traction was continued for 3 weeks. A radiograph was performed every week to check the reduction. After 3 weeks, a hip spica cast was made under general anesthesia. Another 3 weeks later, the plaster cast was removed because of radiologically abundant proliferative callus at the fracture site (fig. 2 a-b). Clinically, a painless swelling at the distal femur was observed and was attributed to callus formation.

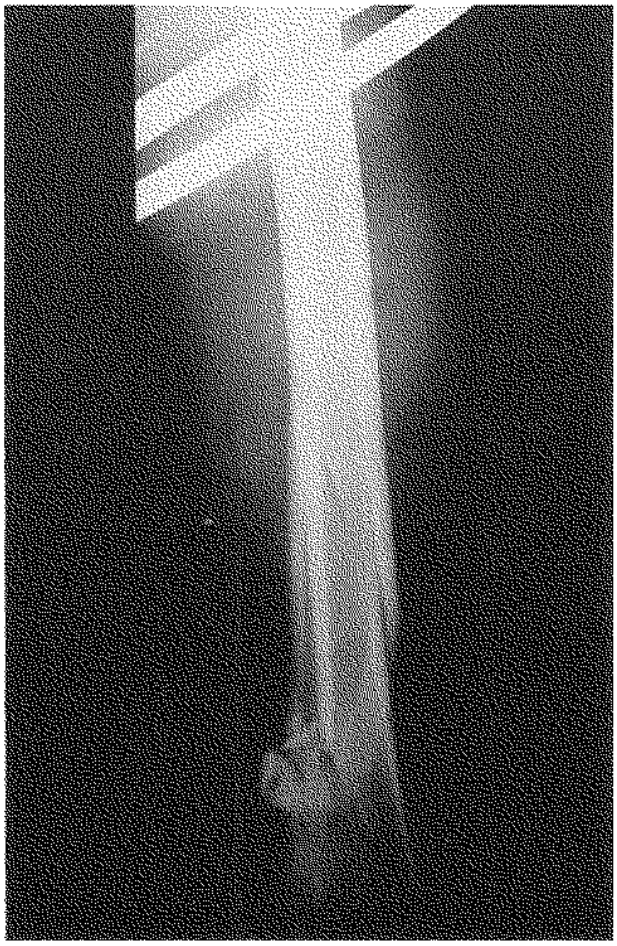

Fig. 1. — AP radiograph showing a spiral fracture of the femoral diaphysis.

Two months later, the boy was admitted again with a large, expanding, warm and painless mass of the left distal femur. Radiographs showed considerable destruction of the left distal femoral metaphysis with soft tissue extension and periosteal reaction. Laboratory studies demonstrated an elevated sedimentation rate and an elevated level of alkaline phosphatase isoenzymes. A bone scan revealed an increased uptake of radioactivity in the entire left femur and more specifically in the distal part. Computerized tomography and MRI showed the presence of a massive bone lesion, with extensive soft tissue extension. MRI also revealed the presence of skip metastases in the femoral neck and head as well as effusions in the hip and knee joints. A radiograph and computerized tomography of the chest did not show any metastases. A needle biopsy was performed and histological examination revealed a high-grade classic osteosarcoma of the osteoblastic type. Chemotherapy, including high-dose methotrexate and citrovorum factor, was administered weekly for 6 weeks. Magnetic resonance imaging of the femur, performed after 6 weeks of chemotherapy, showed regression of the soft tissue extension, but not of the intraosseous involvement. Skin metastases could still be observed in the proximal part of the diaphysis. The knee joint effusion was also still visible. At 6 weeks, a disarticulation of the hip was performed.

Postoperatively, maintenance combination chemotherapy with high-dose methotrexate, bleomycin, cyclophosphamide and adriamycin was started and continued for another 34 weeks (4).

DISCUSSION

Pathologic fractures are a common first symptom for metastatic disease, especially in adults (1). Compared to metastatic disease, primary osteosarcoma is very rare and most commonly affects male adolescents (1, 2, 3, 6). Due to the infrequency of malignant bone tumors, pathologic fractures in the age group under 10 are very rare (1, 2).

Osteosarcomas are usually located in the metaphyseal region of the distal femur or of the proximal tibia (1, 2, 3, 6). They usually appear with a history of pain and swelling in a healthy-looking young patient (1, 2, 3, 6).

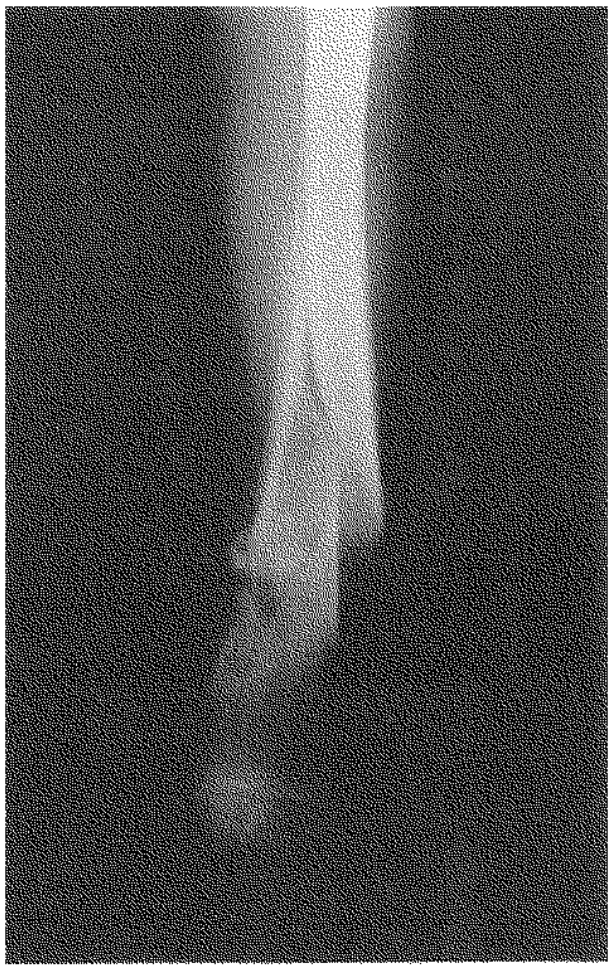

Fig. 2a

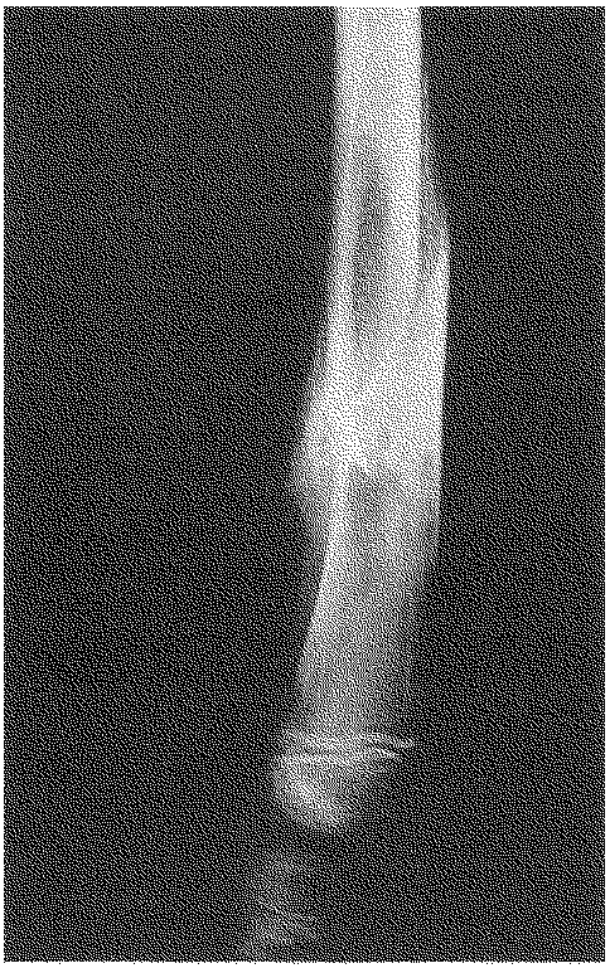

Fig. 2b

Fig. 2 a-b. — AP and lateral radiographs demonstrating proliferative callus formation at the fracture-site 6 weeks later.

Radiologically, destruction of the overlying cortices with invasion of the surrounding soft tissues can be observed. Periosteal reaction gives rise to the Codman's spur or triangle and the perpendicularly radiating "sun-ray" appearance. These are characteristic but not pathognomonic features for malignancy. A differential diagnosis with Ewing's sarcoma, rhabdomyosarcoma and neuroblastoma must be made. Benign processes such as ossifying subperiosteal hematomas and osteomyelitis can also produce similar images (1, 2, 3, 6). Concerning our patient, retrospective analysis of the radiographs made at the time of the accident could perhaps reveal a discrete elevation of the periosteum, but in our opinion the presence of a malignant process was certainly not obvious. The radiographs made after removal of the cast were indicative of the presence of a malignant bone tumor, but were unfortunately misinterpreted as being an abundant callus.

Pathologic fractures are thought to occur in 5 to 10% in an advanced stage of the disease (6). In the case presented, such a fracture appeared at an early stage.

Malignant bone tumors of the distal femur or the proximal tibia are usually diagnosed by pain, often by the presence of a mass and sometimes by a pathologic fracture, in that order (1, 3). Fracture

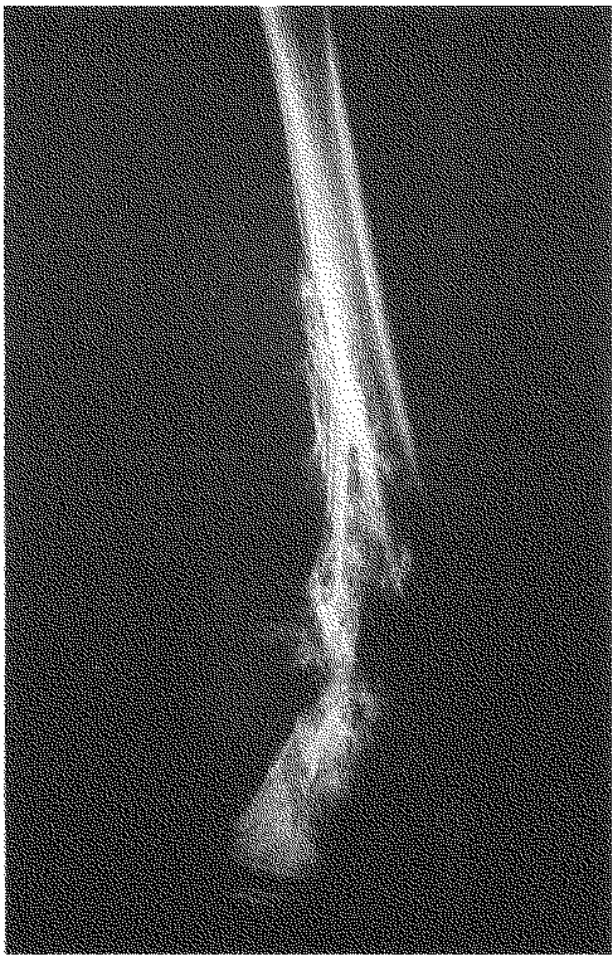

Fig. 3a

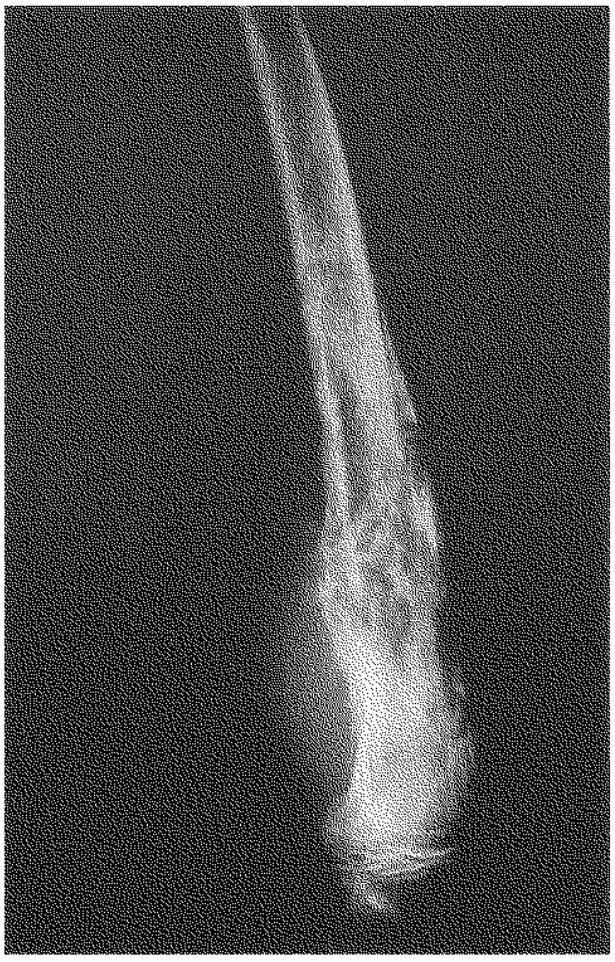

Fig. 3b

Fig. 3 a-b. — At 5 months followup, AP and lateral roentgenograms reveal a large area of bone destruction with periosteal elevation, soft tissue extension and formation of neoplastic bone, very suspicious for the presence of a malignant bone tumor, especially an osteosarcoma.

healing following pathologic fracture in osteosarcoma does not differ substantially from that found after an uncomplicated fracture. During the first months, the reparative process is more dominant than the destructive tumoral process. Therefore, most of these fractures heal well (1). However, the presence of a pathologic fracture influences the natural evolution of a malignant bone tumor. Tumor extension occurs by dissemination via the hematoma or less frequently locally, by breaching the tumor capsule and the reactive zone (1). Early diagnosis is therefore of the utmost importance, as late diagnosis inevitably results in dissemination of the tumor (1). Treatment depends on the grade of soft tissue extension, the presence of skip-metastases and generalized metastases (1, 2, 3, 5, 6). If considerable soft tissue extension or distant skip-metastases are present, radical tumor excision (amputation or disarticulation) is preferable. If not, limb-sparing surgery can be considered (1, 2, 3, 5, 6). Local dissemination can be so extensive that limb-sparing surgery becomes impossible (1, 5).

Considering the large size of the tumor, the proximity of the neurovascular bundle, the proximal extent of the tumor in the medullary canal and the involvement of the knee joint, we decided to perform a hip disarticulation.

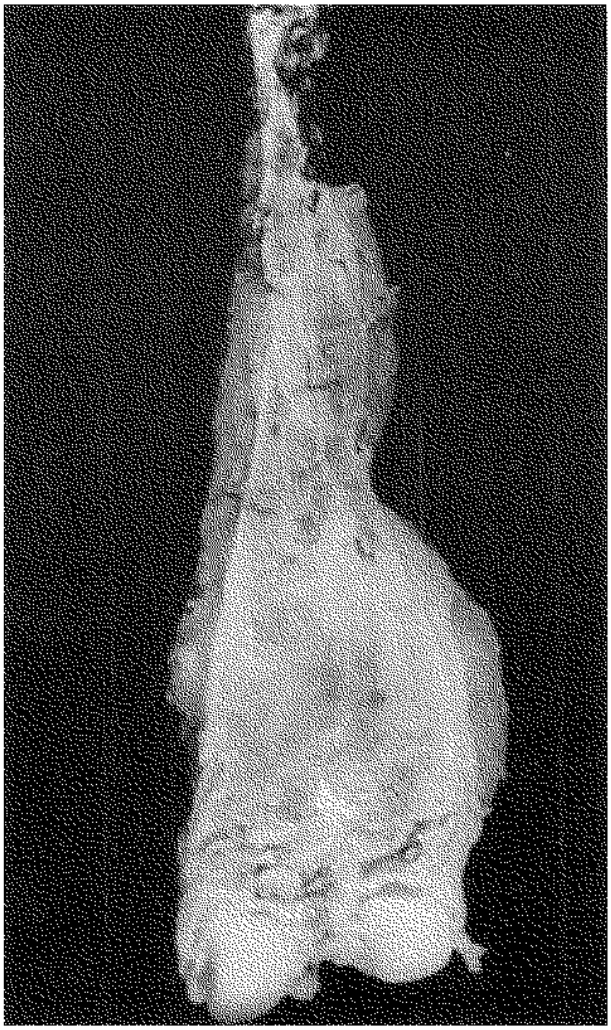

Fig. 4. — Photograph of the resected femur, which clearly shows the extent of the osteosarcoma in the entire femur, indicating its very invasive and malignant character.

CONCLUSION

Pathologic fracture as the primary appearance of an osteosarcoma is very rare in children under 10 years of age.

Radiographs of fractures in children and adolescents must be evaluated for the presence of a pathologic fracture, even though it is unusual in these age groups.

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