UNUSUAL EVOLUTION OF A BENIGN-LOOKING CORTICAL DEFECT OF THE PROXIMAL HUMERUS. A CASE OF INTRACORTICAL OSTEOSARCOMA ?

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A case of an osteosarcoma following a pathological fracture through a small benign-looking cortical lesion of the proximal humerus is described. The unusual evolution from a small cortical lesion to a malignant lesion raises the question whether this is a case of an intracortical osteosarcoma that has some distinct features from those of previously reported cases.

Keywords : intracortical osteosarcoma ; cortical lesion.
Mots-clés : ostéosarcome intracortical ; lésion corticale.

INTRODUCTION

The intracortical osteosarcoma is the rarest subtype of osteosarcoma. The lesion was first described by Jaffe in 1960 (3). Since then, eight additional cases have been reported (1, 2, 4, 5, 6, 7, 8, 9). The typical intracortical origin separates this tumor from conventional medullary osteosarcoma as well as from periosteal and parosteal osteosarcoma.

CASE REPORT

An 8-year-old girl was seen in the emergency room with a history of a fall on her left shoulder. Radiographs of the humerus revealed an undisplaced transverse fracture through a small radiolucent zone at the proximal metaphyseal-diaphyseal junction (Fig. 1). This was considered as a benign lesion requiring no further diagnostic evaluation. The girl was treated with a plaster cast for 4 weeks. A radiograph made after removal of the cast showed callus formation around the fracture. The fracture was stable on clinical examination and physiotherapy was started. A radiograph made 8 months after the accident revealed a bone lesion which was clearly larger than the initial lesion (Fig. 2). It was interpreted as a benign bone cyst in a healing phase after a fracture. The girl had no complaints and had normal use of her arm and shoulder. Four months later she presented with a 3-week history of increasing pain in her left shoulder. On physical examination a painful swelling of the proximal humerus was noted. Radiographs revealed a destructive lesion of the proximal metaphysis (Fig. 3). The girl denied having had pain before the trauma, and the past medical history was unremarkable. MRI revealed a destructive process in the lateral cortex with intramedullary extension. The lesion extended proximally but did not cross the proximal physis. A biopsy was performed. This histologic examination showed conventional osteosarcoma. The patient was treated with chemotherapy for 4 months followed by resection of the proximal two-thirds of the humerus. Reconstruction was performed using an endoprosthesis. Histopathologic examination of the resected specimen confirmed the diagnosis of osteosarcoma. At 34 months after the diagnosis there was no evidence of local recurrence or distant metastasis.

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Fig. 1. — The initial radiograph shows a small cortical lesion.

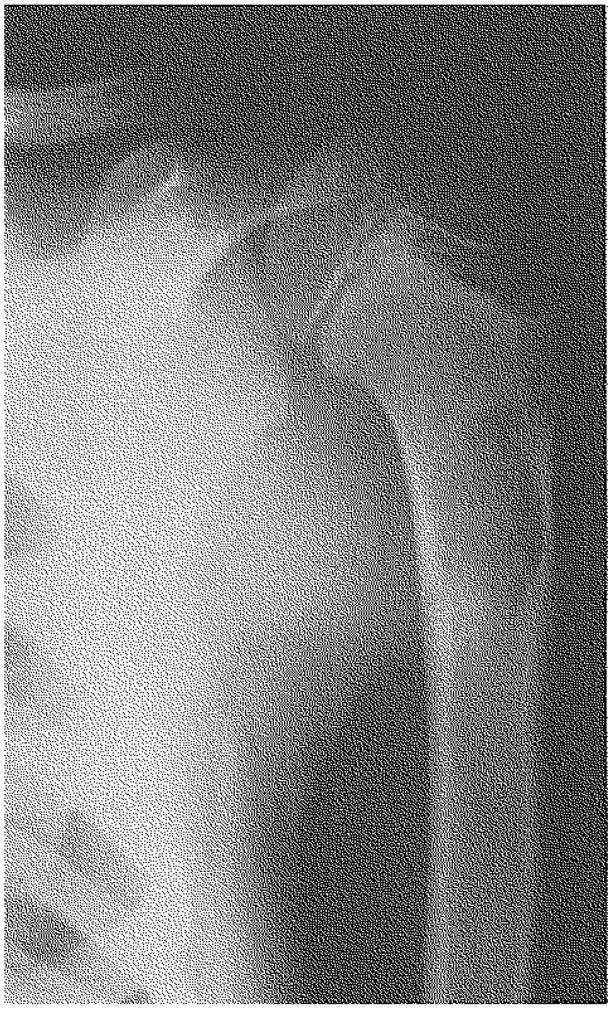

Fig. 2. — Radiograph showing the aspect of the lesion in an advanced stage. Note healing of the fracture.

DISCUSSION

Benign bone cysts are the most common cause of single pathologic fractures in the upper humerus in childhood. The typical radiographic appearance includes a transverse fracture with little or no displacement. Usually the fracture itself heals normally and in about 20% of the cases, the healing of the fracture causes the bone lesion to heal as well. The radiographic appearance of the fracture and of the bone lesion, which showed regular margins without an endosteal or periosteal reaction as in the case presented here, had the characteristics of a benign lesion and was so diagnosed. Therefore no additional investigations were performed. The initial radiographs were reevaluated after the diagnosis of an osteosarcoma was made. The small lytic lesion was located in the cortex (Fig. 1) and showed the radiologic features of intracortical osteosarcoma : a rounded lytic, intracortical lesion less than 4 cm in maximal dimension, as described by Mirra *et al.* (7). Because there is no histologic proof of the true nature of the lesion at initial presentation, the diagnosis of intracortical osteosarcoma can be questioned. This report however clearly shows the

Fig. 3. — The lesion now has all characteristics of a classical osteosarcoma.

progression from a small lytic lesion (Fig. 1) through a more extensive lesion (Fig. 2) to the classical radiographic appearance of an osteosarcoma (Fig. 3). Intracortical osteosarcoma is a rare entity. Jaffe was the first to distinguish intracortical osteosarcoma from conventional centrally located osteosarcoma (3). This author presented two cases of an intracortical osteosarcoma. Kyriakos in 1980 reported a case of an intracortical osteosarcoma in a 24-year-old man (4). The 20-year lapse before presentation of this third case and the paucity of cases reported so far attests to the rarity of this tumor. Picci *et al.* in 1983 reported a case of an intracortical osteosarcoma and suggested that this might be an early manifestation of classical osteosarcoma (8). In our case the evolution of a benign-looking lesion to a destructive lesion typical of an osteosarcoma seems to confirm this hypothesis. All reported cases had similar radiologic characteristics at initial presentation. Our case however has some distinct features from those reported previously. It is the youngest patient reported to date. It is the first case occurring in the humerus. (The previously described cases were located in the tibia or in the femur.) All the previously described cases of intracortical osteosarcoma were located in the diaphysis, whereas in our case the lesion was located at the metaphyseal-diaphyseal junction. Nevertheless, follow-up clearly shows the progression from a small lytic lesion to the classical radiographic appearance of an osteosarcoma. The paucity of reported cases precludes definite statements regarding prognosis and natural evolution of an intracortical osteosarcoma. It seems that some of these tumors progress slowly or not at all, while others evolve into a classical osteosarcoma. The case of our patient and all previous reports illustrate the difficulty in making the correct diagnosis at initial presentation. The initial diagnoses have all been benign. A thorough evaluation and especially a high index of suspicion for malignant lesions is necessary to make the diagnosis.

REFERENCES

1. Anderson R. B., McAlister J. A. Jr., Wrenn R. N. Case report 585. Skeletal Radiol., 1989, 18, 627-630.
2. Blasius S., Link T. M., Hillmann A., Rödl R., Edel G., Winkelmann W. Intracortical low grade osteosarcoma. A unique case and review of the literature on intracortical osteosarcoma. Gen. Diagn. Pathol., 1995/1996, 141, 273-278.
3. Jaffe H. L. Intracortical osteogenic sarcoma. Bull. Hosp. Joint Dis., 1960, 21, 189-197.
4. Kyriakos M. Intracortical osteosarcoma. Cancer, 1980, 46, 2525-2533.
5. Kyriakos M., Gilula L. A., Becich M. J., Schoenecker P. L. Intracortical small cell osteosarcoma. Clin. Orthop., 1992, 279, 269-280.
6. López-Barea F., Rodriguez-Peralto J. L., González-López J., Sánchez-Herrera S., Sánchez-Del-Charco M. Intracortical osteosarcoma. A case report. Clin. Orthop., 1991, 268, 218-222.

7. Mirra J. M., Dodd L., Johnston W., Frost D. B., Barton D. Case report 700. Skeletal Radiol., 1991, 20, 613-616.
8. Picci P., Gherlinzoni F., Guerra A. Intracortical osteosarcoma : Rare entity or early manifestation of classical osteosarcoma ? Skeletal Radiol., 1983, 9, 255-258.
9. Vigorita V. J., Ghelman B., Jones J. K., Marcovic R. C. Intracortical osteosarcoma. Am J. Surg. Pathol., 1984, 8, 65-71.

SAMENVATTING

H. DE BOECK, F. HANDELBERG, J. OTTEN. Ongewone evolutie van een goedaardig uitziend corticaal letsel van de proximale humerus. Een geval van intracorticaal osteosarcoma ?

De auteurs beschrijven een geval waarbij een klein corticaal defect van de proximale humerus op enkele maanden tijd evolueerde naar een osteosarcoom. De ongewone evolutie van dit radiologisch goedaardig uitziende letsel laat vermoeden dat het om een geval van een intracorticaal osteosarcoom gaat.

RÉSUMÉ

H. DE BOECK, F. HANDELBERG, J. OTTEN. Evolution inhabituelle d'une lésion corticale d'allure bénigne au niveau de l'humérus proximal. Un cas d'ostéosarcome intracortical ?

Les auteurs décrivent le cas d'une petite lésion corticale de l'humérus proximal qui a évolué en quelques mois vers un ostéosarcome. Cette évolution peu classique d'une lésion décrite comme radiologiquement bénigne, peut faire supposer qu'il s'agit vraisemblablement d'un ostéosarcome intracortical.