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Skeletal Radiology

Case report 641 *

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Radiological studies

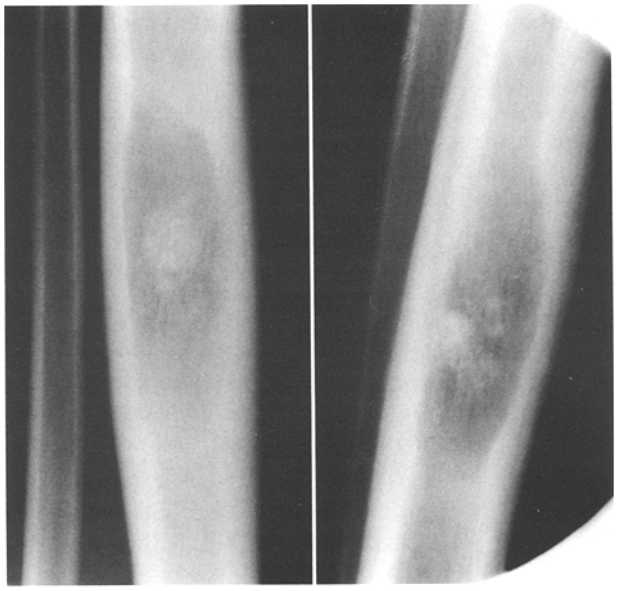

Fig. 1. AP (*left*) and lateral radiographs (*right*) show a radiolucent lesion in the midshaft of the tibia with central areas of spotty calcification. The cortex is partly enlarged with scalloping on its inner surface

Clinical information

A 12-year-old boy complained of pain in the left leg after a slight trauma 4 months previously. Radiographs revealed a slightly expansile, radiolucent area in the tibial diaphysis with foci of central spotty calcification. The cortex was in part thickened, showing scalloping at its endosteal surface and laminar periosteal bone formation (Fig. 1). After a previous needle biopsy, an open biopsy was performed.

The radiographic appearance suggested a primary, most probably a malignant tumor of bone (e.g. a cartilage tumor) and even including in the differential diagnosis an unusual Ewing tumor and osteosarcoma. Conventional central chondrosarcoma seemed to be unlikely because of the age of the patient, at which time malignant cartilage tumors are rare [1, 2, 5, 6, 20] with the exception of mesenchymal chondrosarcoma [12, 14] and clear cell chondrosarcoma [19]. A benign cartilage tumor was excluded due to the endosteal erosion of the thickened and expanded cortex together with periosteal laminated bone formation, suggesting permeative growth of the lesion.

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Diagnosis:
Chondroblastoma-like osteosarcoma

The open biopsy material included fragments of cortical bone and of a soft, friable, highly vascular tissue with hemorrhagic, partly cystic areas.

Microscopically broad areas of closely packed, rounded, oval or polyhedral cells with well-defined cytoplasmic borders were noted. The cytoplasm was eosinophilic, and the nuclei were round or reniform, frequently indented and containing one or two small nucleoli. In some areas, several larger cells with bigger and evidently hyperchromatic nuclei were found, but mitotic figures were usually scarce or even more numerous in some areas (Fig. 2). Multinucleated, osteoclastic-type giant cells were dispersed among the tumor cells and were more numerous in areas of cartilaginous differentiation or around hemorrhagic foci. These were frequently found together with wide cystic blood spaces, lined by the tumor cells and/or multinucleated giant cells, simulating an aneurysmal bone cyst (Figs. 2, 3). The general appearance of the lesion was that of a chondroblastoma with aneurysmal-bone-cyst-like areas, a feature for which we suggested the term "cystic chondroblastoma". Some chondroid areas showed heavy calcification followed at times by enchondral ossification, but the characteristic "chicken-wire" pattern was lacking. The tumor revealed an evident tendency to penetrate the widened Haversian spaces of the cortex (Fig. 2A), which was partially destroyed and showed at its surface evident and active periosteal new bone formation.

The histological features inclined us to confirm the diagnosis of cystic chondroblastoma made by the referring pathologist, Dr. Donato de Próspero of São Paulo, Brazil, who sent us this case in consultation because of its exceptional diaphyseal location. We agreed with his diagnosis, in spite of the diaphyseal location.

Discussion

It is well-known that chondroblastoma principally affects the epiphysis or apophysis, especially of the long bones, although it may not infrequently extend into the metaphysis [4, 3, 10, 11, 17]. However, chondroblastomas originating in the metaphysis are very rare, accounting for no more than approximately 15 cases reported in the literature, which is less than 5% [13, 16, 18]. A distinctly diaphyseal location of this lesion, with the exception of one case mentioned by Salzer et al. [15], has not yet been reported. Because of the presence of areas of certain nuclear atypism and hyperchromatism and the conspicuous invasive growth pattern, we emphasized the aggressive features of the lesion and advised a segmental en bloc resection. A few days after this treatment a local recurrence and invasion of soft tissues was found at the medial aspect.

A submitted photograph of the resected specimen showed (Fig. 4A) clearly the cystic hemorrhagic aspect of the lesion with wide areas of necrosis and destruction of the cortex at its medial site, partly due to the previous biopsy. It was also apparent that the operative sections passed through the tumor margins.

The histological sections of the recurrent tumor showed, besides the cystic vascular pattern, extensive areas of necrosis and chondroblastoma-like structures, foci with conspicuous pleomorphism, bizarre and hyperchromatic nuclei, atypical mitoses, and evident osteoid formation by atypical osteoblasts (Fig. 4B). These features favored a diagnosis of chondroblastic osteosarcoma, highly vascularized or telangiectatic. The short period between the first submitted biopsy, diagnosed as cystic chondroblastoma, possibly aggressive, and the recurrence, showing the pattern of a highly malignant osteosarcoma, clearly excludes a malignant transformation of a pre-existent benign or aggressive chondroblastoma. On the other hand, the diaphyseal location of the tumor made this possibility highly unlikely. It is most probable that due to a sample error evident signs of malignancy and/or tumor bone formation were missed, and the lesion was a chondroblastic osteosarcoma from its inception. That diagnosis is also more in agreement with the radiological features and the clinical evolution. Dr. Unni[1], who had seen some of the slides, concurred with this diagnosis.

Reviewing the section of the first biopsy, we found in one of the three slides an isolated area which appeared to be tumor bone formation and not reactive bone secondary to the previous needle biopsy.

In *summary*, it seems evident that this tumor was from the beginning an osteosarcoma, predominantly chondroblastic and highly vascularized, with wide areas resembling cystic chondroblastoma. Chemotherapy, consisting of doxorubicin, methotrexate, and cisplatin, was started immediately. A month later multiple pulmonary metastases developed.

The questions arise of whether or not a malignant variant of chondroblastoma exists and whether this case should be diagnosed as such. We and other authors have observed that a few cases of chondroblastoma may pursue a more aggressive course and recur locally with invasion of joint spaces, adjacent bones, and/or soft tissue [7, 9]. Many of these complications may be attributed either to incomplete initial curettage or to accidental dissemination of tumor during the operation.

However, from our experience and that of others [9, 11], it can be stated that the reported cases of malignant evolution may be divided into the following categories:

1. Chondroblastoma with sarcomatous transformation, with or without prior radiation therapy treatment ("secondary" malignant tumor).
2. Chondroblastoma exhibiting benign-appearing lung metastasis. All cases reported with the exception of that of Kyriakos et al. [9] and which pursued a malignant course, developed subsequent to some form of operative treatment of the primary tumor.
3. Chondroblastoma-like chondrosarcomas or chondrosarcomas resembling chondroblastoma.

To these possibilities we must add the chondroblastic variant of osteosarcoma, in which chondroblastoma-like features mask the correct diagnosis.

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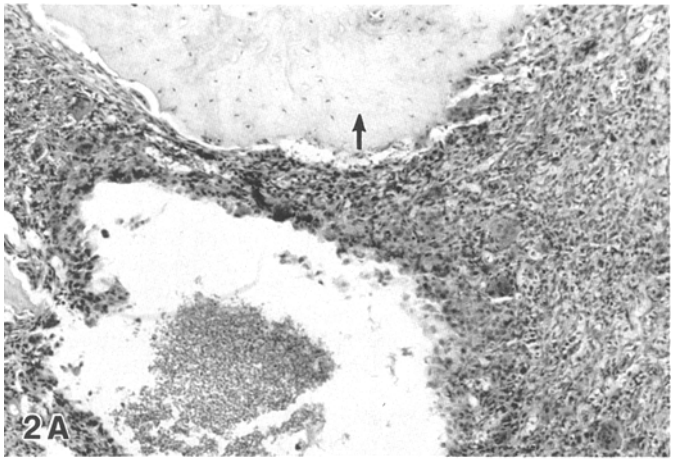

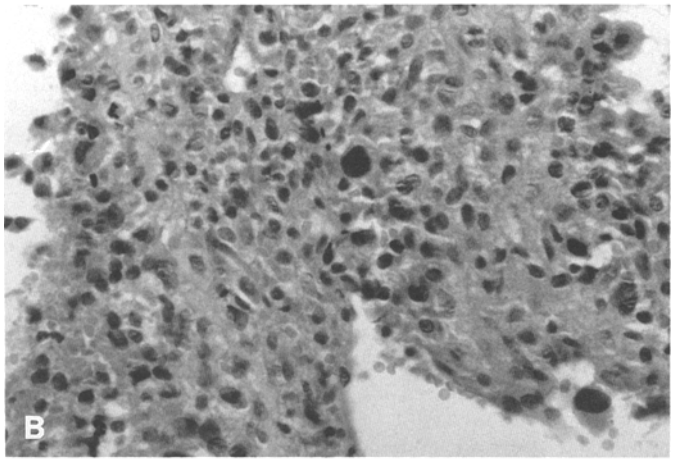

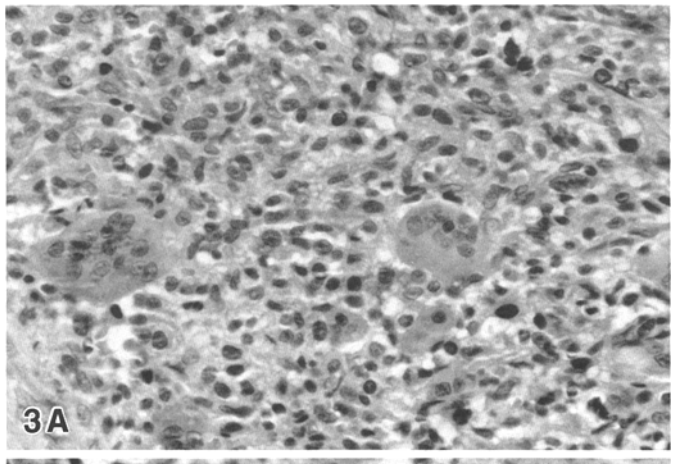

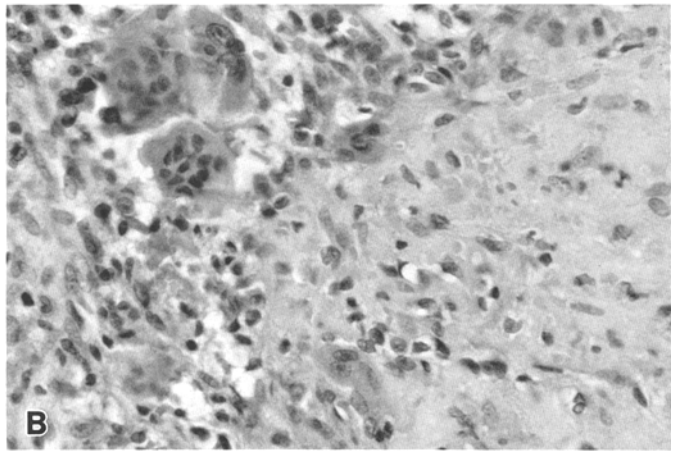

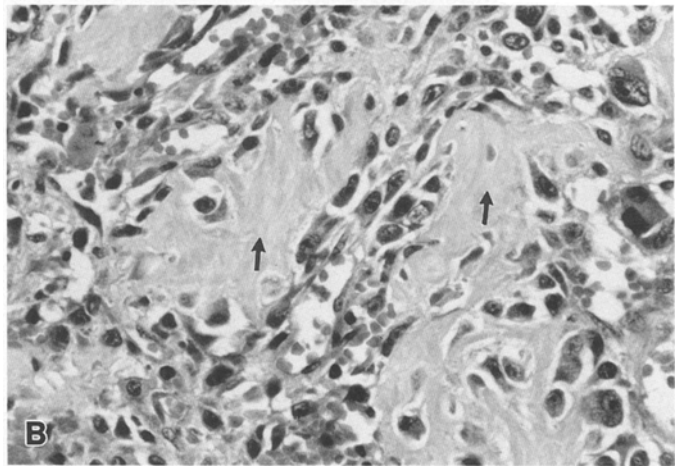

Fig. 2. Photomicrographs of the specimen obtained at open biopsy show at lower (**A**) and higher (**B**) magnification compact cellular areas alternating with large cystic spaces, filled in part with red blood cells. The tumor penetrates the cortex (**A**, *arrow*) and presents in **B** several polyhedral cells with large hyperchromatic nuclei, but atypical mitoses are absent. (**A** H&E, ×100; **B** H&E, ×400)

Fig. 3A, B. Photomicrographs of a solid area show at higher magnification a compact mass of rounded and polygonal stromal cells with distinct cell borders mixed with some scattered osteoclast-type giant cells and an area of chondroid formation in **B.** The stromal cells do not present features of atypism or evident mitotic activity. (**A, B** H&E, ×400)

Fig. 4. A Photograph of the resected specimen shows the hemorrhagic cystic pattern of the tumor presenting extensive areas of necrosis and invasion of soft tissue, at the site of the previous biopsy.
B Photomicrograph of an area of the recurrent tumor shows irregular osteoid trabeculae formation (*arrows*) by atypical osteoblastic tumor cells. (H&E, ×400)

The existence of a "primary" malignant chondroblastoma is questionable. The question must be answered affirmatively if the term malignant chondroblastoma means, as quoted by Kyriakos et al. [9], "a tumor with benign-appearing histological features capable of aggressive behavior with metastasis and death of the patient [8]". Unfortunately, there are as yet no histological parameters that permit determination of which metastases are destined to cease their growth or to progress and kill the host.

Whether the difference between this concept of malignant chondroblastoma and chondroblastoma-like chondrosarcoma or osteosarcoma is purely semantic or whether there are in fact two different entities must await the report of further cases with good documentation and follow-up information.

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