A case of osteosarcoma arising in a solitary osteochondroma

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Osteochondroma, the most frequent bone tumor, is composed of trabecular bone covered with a hyaline cartilage cap. A malignant change in a solitary osteochondroma is rare; 1–4 percent undergo malignant transformation as compared with 20–40 percent in patients with multiple osteochondromatosis (Matsuno et al. 1988). It is sometimes difficult to distinguish malignant transformation in an osteochondroma from other malignant tumors occurring on the surface of bone.

Since the dedifferentiated chondrosarcoma (DCS) was described in detail by Dahlin and Beabout (1971), it has been given considerable attention because of its characteristic histologic features and high malignancy. DCS occurs in approximately 10 percent of the patients who have a low-grade chondrosarcoma and histologically show chondrosarcomatous features with additional mesenchymal components (Dahlin and Beabout 1971, McCarthy and Dorfman 1982, Capanna et al. 1988). Most of the cases have been located in the medullary cavity. However, Bertoni et al. (1989) described 7 cases of dedifferentiated peripheral chondrosarcoma on the surface of the bone that previously had had an osteochondroma. Only 16 cases of DCS originating in an osteochondroma have been reported to date (Fennel 1938, Anderson et al. 1969, Frassica et al. 1986, Matsuno et al. 1988, Bertoni et al. 1989).

We report an additional case of dedifferentiated peripheral chondrosarcoma.

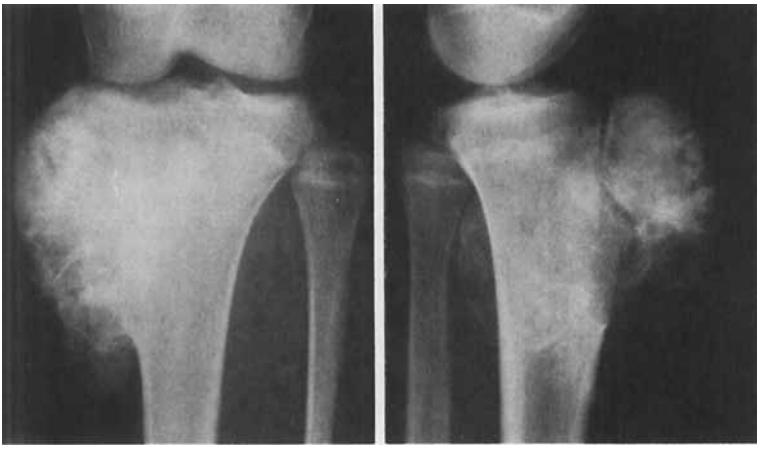
Case report

In February 1988, a 14-year-old Japanese girl was admitted to our hospital because of increasing pain and swelling in her left knee joint of about 2 years' duration. There was no history of trauma. Radiographs showed a juxtacortical tumor, irregularly and heavily ossified with focal lucent areas, at the proximal end of the left tibia (Figure 1). Computerized tomography (CT) demonstrated that this large mass, containing areas of irregular mineralization, arose from the surface of the tibia, and the marrow space was continuous with the lesion (Figure 1). An open biopsy was performed and a histologic diagnosis of chondroblastic osteosarcoma was made. After six courses of chemotherapy with high doses of vincristine and methotrexate, a wide resection of the tibia was performed in July 1988. Postoperatively, the patient received chemotherapy; and she is doing well and has no metastases after 2 years. No osteochondromas were seen in other bones, and no family history of osteochondroma was obtained.

Pathologic findings

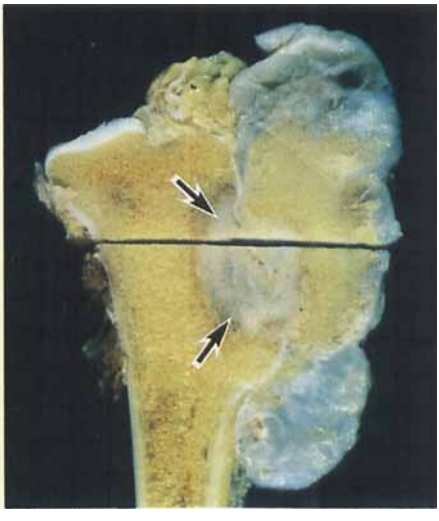
The specimen consisted of a polyploid piece of bone tissue, measuring 12 × 9 × 4 cm, that projected from the surface of the bone. Upon sectioning, the surface of the tumor was found to be covered with a 3-mm up to 3.5-cm-thick cartilage cap. The wide chondroid portion was composed of soft, gelatinous, and translucent grayish white lobules; and in the central portion, a yellowish necrosis was seen. Under the cartilage cap, osseous trabeculae and fat marrow were seen merging with the underlying, normal cancellous bone. Partially, the cartilage cap at the bony stalk showed continuity with a massive, hard, whitish lesion; and bone trabeculae had been destroyed.

Histologically, the hyaline cartilage cap shifted to normal bone trabeculae by an endochondral ossification process, suggesting an osteochondroma. The wide chondroid areas were myxomatous, and cellular, binucleate cells or pleomorphic chondroblasts that had an extensive cytoplasm and a large nucleus were recognized. These depicted a feature of low-grade chondrosarcoma. At the site of the bony stalk adjacent to these chondrosarcomatous areas were anaplastic high-grade foci, with direct formation of irregular tumor osteoid and bone by atypical stromal cells. These malignant osteoblasts of osteosarcoma infiltrated into the medullary region (Figure 1).

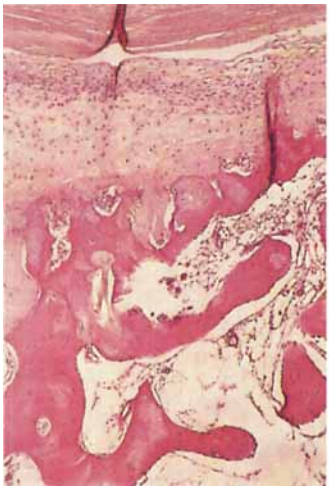


A large, mixed osteosclerotic and lytic lesion of the proximal tibia. The indistinct and irregular margin suggests a malignancy.

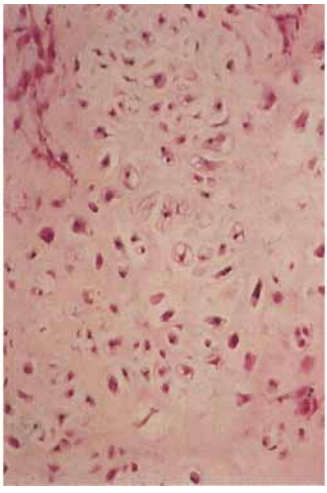
CT scan demonstrates a large tumor arising from the tibia (arrows). The marrow space of the tibia is continuous with that of the tumor mass.



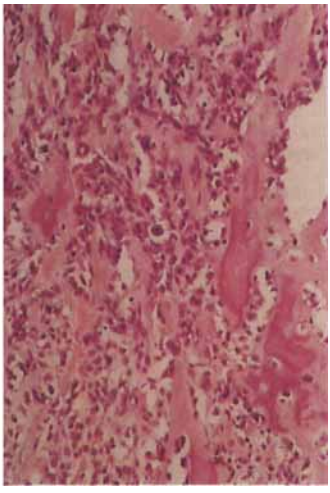
Photograph of the cut section shows various thickness of the cartilage cap and the underlying spongiosa. A whitish, hard lesion can be seen at the bony stalk (arrows).



Cartilage cap of benign osteochondroma. Endochondral ossification is producing regular bony trabeculae with fat marrow. H&E, ×20.



Low-grade chondrosarcomatous area. Irregularly arranged cartilage cells with pleomorphism and atypism are seen. H&E, ×50.



Formation of osteoid by highly anaplastic tumor cells. The appearance is consistent with a diagnosis of osteosarcoma. H&E, ×50.

Figure 1. A 14-year-old girl with an osteosarcoma arising in a tibial osteochondroma.

Discussion

We diagnosed our case as an osteosarcoma arising in an osteochondroma. Malignant transformation in an osteochondroma should be differentiated from other malignant tumors arising on the surface of the bone, especially from a periosteal chondrosarcoma and a periosteal osteosarcoma.

If plain radiographs, a CT scan, or a gross specimen demonstrate an extension of the marrow space of the bone into the lesion, the diagnosis of a preexisting osteochondroma can be easily established. However, the larger tumors may destroy the original osteochondroma in its entirety. A periosteal chondrosarcoma consists of pure cartilage and produces no osteoid matrix (Nojima et al. 1985). A periosteal osteosarcoma has a chondroid matrix with a typically osteoid component and no involvement of the medullary canal (Unni et al. 1976, Bertoni et al. 1982).

Our patient had no history of multiple osteochondromatosis, and the biopsic examination showed a chondroblastic osteosarcoma. However, a cartilage cap covered the surface of the projecting lesion, and there was a continuity of the lesion with the normal marrow space on the radiologic and the macroscopic examination. Microscopically, we recognized features of a low-grade chondrosarcoma, together with an osteosarcoma, but most of the cartilage cap was benign, was maturing into the bone trabeculae, and the spaces between these trabeculae contained fat marrow. From these findings, we confirmed the presence of a preexisting osteochondroma. When an osteochondroma undergoes malignant change, it usually shows features of a chondrosarcoma (Garrison et al. 1982).

However, in recent years, there have been several reports of osteosarcoma, fibrosarcoma, or malignant fibrous histiocytoma that have originated in an osteochondroma (Fennel 1938, Frassica et al. 1986, Matsuno et al. 1988). Bertoni et al. (1989) described 7 cases as dedifferentiated peripheral chondrosarcomas that were composed of additional mesenchymal components recognized together with an osteochondromal lesion. According to that report, dedifferentiated peripheral chondrosarcoma is more frequently seen in younger patients as compared with central DCS cases, and histologically the mesenchymal components were composed of malignant fibrous histiocytoma. A chondrosarcoma and a DCS that arise in the medullary cavity are mostly found in the pelvic bone, femur, and humerus. When compared with these tumors, dedifferentiated peripheral chondrosarcoma occurs at the sites that favor the growth of osteochondroma: in the distal femur and in the proximal tibia.

A central DCS may be fatal (McCarthy and Dorfman 1982, Frassica et al. 1986, Capanna et al. 1988). In dedifferentiated peripheral chondrosarcoma, 5 out of 7 cases reported by Bertoni et al. (1989) had pulmonary metastases and died after a short period of time. The prognosis for patients with periosteal chondrosarcomas, periosteal osteosarcomas, or chondrosarcomas secondary to osteochondroma is better than for patients with dedifferentiated peripheral chondrosarcomas (Unni et al. 1976, Garrison et al. 1982).

When malignancy is suspected in osteochondroma, we believe that a detailed investigation should be conducted, with samplings of an adequate amount of representative tumor.

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