

Multiple malignant fibrous histiocytoma of bone

A case report

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A 27-year-old man had tumors in both the femoral and the tibial metaphyses of the left knee. Although histological examination of the biopsy specimens

revealed different findings in the 2 lesions, the final diagnosis was malignant fibrous histiocytoma in both.

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Multiple primary bone tumors other than angiosarcoma or osteosarcoma are very rare. We describe a case of malignant fibrous histiocytoma (MFH) which seemed to have arisen multifocally.

Case report

A 27-year-old man was admitted to our hospital with pain in the left lower leg since 1 year. He had no particular past history or familial history. Plain radiographs revealed osteosclerosis with indistinct margins and destruction of the bone in the distal diaphysis and metaphysis of the femur which showed an increased transverse dimension. The radiographs also revealed radiolucency and destruction of the cortical bone in the proximal metaphysis and diaphysis of the tibia. Laboratory examinations were normal. CT showed an osteosclerotic shadow in the medullary canal of the femur and a soft tissue density mass in the medullary canal and destruction of the lateral cortex of the tibia. Whole body bone scintigraphy showed an abnormally high uptake only in the distal femur and the proximal tibia. On angiography, intense hypervascularity was found in each lesion. On the femoral side, low-to-moderate signal intensity was observed on T1-weighted MR images and a non-homogeneous area of high signal intensity was demonstrated on T2-weighted images. On the tibial side, there was low signal intensity on T1-weighted images and high signal intensity and extraskeletal invasion of the tumor into the muscle on T2-weighted images.

Because of the multiple bone involvement, bone metastasis from some other primary cancer was suspected, but on further examinations (chest radiographs, chest CT, gastrointestinal examination, and abdominal

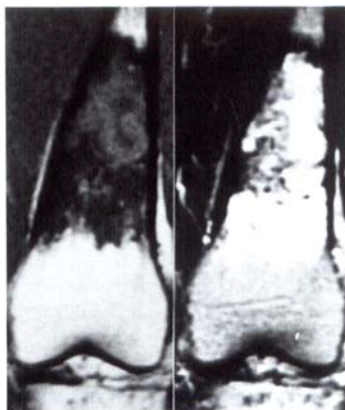
CT) only a renal cyst was detected. No primary focus at any other site could be identified.

Open biopsy from both lesions was performed. Histological examination revealed spindle cell proliferation with bone destruction and storiform pattern in the femoral lesion. Many mitotic figures and necrotic areas were also demonstrated. The tibial lesion was composed of pleomorphic polygonal cells, which produced an extracellular myxoid substance with scattered giant cells. Only one small focus of osteoid formation by tumor cells was demonstrated in the specimen from the tibial lesion. The results of immunohistochemical staining suggested that the tumor cells were not of epithelial origin, and no matrix formation was seen between the cells. Electron microscopic findings suggested that these tumors were not metastatic cancers or other sarcomas. On the basis of these observations, we considered the most likely diagnosis to be MFH of the bone in both lesions.

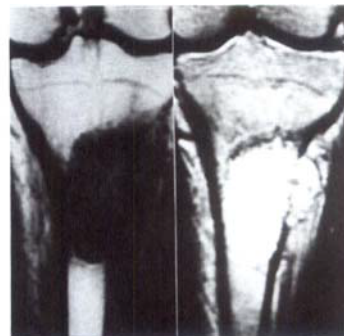
Preoperative chemotherapy, according to Rosen's T-14 protocol, gave a marked effect evaluated by MRI, bone scintigraphy, and angiography. The tumors were resected with a wide margin (17 cm of the distal femur and 14 cm of the proximal tibia). Reconstruction was done with a Kotz prosthesis. Extensive necrosis encompassing over 99 percent of the tumor was histologically demonstrated. Since preoperative chemotherapy was effective, the postoperative treatment was abbreviated due to the patient's request to discontinue treatment. The entire treatment was completed in 19 weeks. Presently, 34 months after surgery, no signs of local recurrence or distant metastasis have been detected and the patient has resumed his job as an office worker.



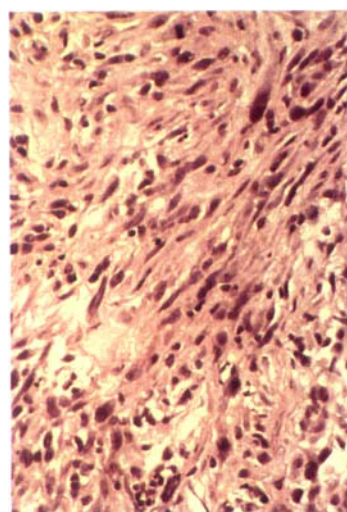
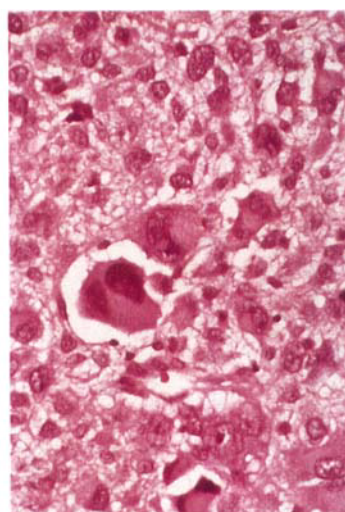
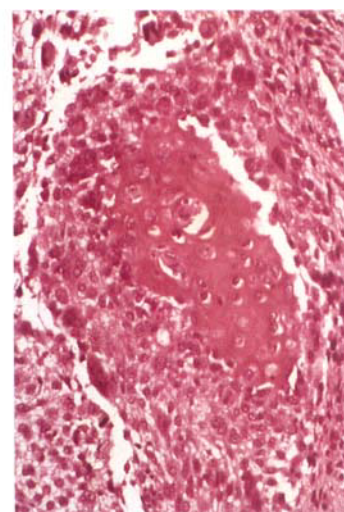
Radiographs.



MR (T1- and T2-weighted images).



MR (T1- and T2-weighted images).

Femoral lesion. Spindle cell proliferation (HE, $\times 200$).Tibial lesion. Pleomorphic polygonal cells with scattered giant cells (HE, $\times 200$).A microscopic small focus of osteoid formation in the tibial lesion (HE, $\times 100$).

Discussion

Reports of multiple MFH date back to Steiner (1944). Chen (1978) and McCarthy et al. (1979) reported 4 patients with multicentric lesions. Dahlin and Unni (1986) observed multiple MFH in only 1 of 52 cases of MFH, and Mirra (1989) found none in 62 cases. Huvos (1991) reviewed 130 cases reported during the past 57 years and noted synchronous multiple MFH in 3 and metachronous multiple MFH in 13.

Wold et al. (1990) suggested that bone metastasis of renal cancer, fibrosarcoma, and fibroblastic osteosarcoma as diseases should be differentiated from MFH

of bone. In our patient, CT findings indicated that the kidney lesion was a simple cyst because of the water density of the lesion without enhancement. Metastasis from other visceral organs was considered unlikely on the basis of findings on various imaging techniques. Fibrosarcoma was also unlikely, because the cells constituting the tumor were short and spindle-shaped, while pleomorphic and diverse cells showing the characteristics of histiocytes were noted. The major portion of the tumor was not matrix-forming, but rather myxoid and pleomorphic, and the lesions were multifocal. The diagnosis of the lesion in the femur was MFH beyond any doubt, while the radiographic findings of

the tibial lesion were osteolytic, without periosteal reaction. Only one microscopic focus of osteoid formation was recognized in the specimen from the tibial lesion. However, the focus of osteoid formation was so small that we diagnosed this disease as MFH, rather than osteosarcoma.

In multiple bone lesions, it is important to determine whether the lesions are multifocal or metastatic. Bone tumors may metastasize via lung metastasis, even when no lung lesions are detected by imaging techniques. In our case, radiologic findings of the lesions were completely different. The difference between the 2 lesions in the histopathological features further reduces the probability of metastasis. We consider the entity in this case to be multifocal MFH of bone.

References

- Chen K T. Multiple fibroxanthosarcoma of bone. *Cancer* 1978; 42 (2): 770–3.
- Dahlin D C, Unni K K. Malignant (fibrous) histiocytoma. In: *Bone Tumors* (Eds. Dahlin D C, Unni K K) Charles C Thomas Publisher, Springfield 1986: 357–65.
- Huvos A G. Malignant (fibrous) histiocytoma. In: *Bone Tumors* (Ed. Huvos A G) W. B. Saunders Co, Philadelphia 1991: 497–521.
- McCarthy E F, Matsuno T, Dorfman H D. Malignant fibrous histiocytoma of bone: a study of 35 cases. *Hum Pathol* 1979; 10 (1): 57–70.
- Mirra J M. Malignant fibrous histiocytoma. In: *Bone Tumors* (Eds. Mirra J M, Picci P, Gold R H) Lea & Febiger, Philadelphia 1989: 766–99.
- Steiner P E. Multiple diffuse fibrosarcoma of bone. *Am J Pathol* 1944; 93: 877–93.
- Wold L E, McLeod R A, Sim F H, Unni K K. Malignant fibrous histiocytoma. In: *Atlas of Orthopaedic Pathology*. (Ed. Bordin G M) W. B. Saunders Co, Philadelphia 1990: 136–41.