
Osteosarcoma Arising in Paget's Disease of the Calvarium

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Paget's disease of the bone occasionally transforms into sarcomatous tumors such as osteosarcoma. While Paget's disease demonstrates markedly increased uptake of bone scanning radiopharmaceutical, osteosarcomas complicating this condition often show decreased uptake. An unusual scan appearance of the osteosarcoma is presented in which the tumor showed intense uptake of the tracer on the bone scan. The extent of the sarcoma as delineated by the bone scan corresponded closely to the size of the lesion as seen on computerized tomography and magnetic resonance imaging and was confirmed at surgery.

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Paget's disease (Osteitis deformans) is a common disease entity in the western world. Prevalence is estimated to be ~5% of the population over 40 years of age (1). A case of osteosarcoma arising in pre-existing Paget's disease that has the unusual scintigraphic finding of a very marked increase in uptake of technetium-99m (methylene diphosphonate [^{99m}Tc]MDP) into the tumor is presented. The size of the lesion as noted on computed tomography (CT) and magnetic resonance imaging (MRI) corresponded closely with the lesion size as noted by scintigraphy. The determination of tumor margins was important to reduce the extent of surgical resection.

CASE REPORT

An 82-yr-old male with a 15-yr history of Paget's disease of the calvarium presented with a complaint of recent headache and a right temporal mass. A previous bone scan dated August 1983 (Figs. 1A and 1B) demonstrates increased accumulation of the radiotracer, typical of that seen in patients with Paget's disease. Also, increased radiotracer uptake is seen beyond the expected confines of the bone, an unusual finding and never ascribed before to uncomplicated Paget's disease. A recent bone scan dated September 1985 (Figs. 2A and 2B) was similar, but a new focal lesion in the right parietal region with more intense uptake had appeared in the pagetic bone. Past and recent skull x-rays demonstrated the typical "cottonwool" appearance of mixed osteolytic and osteoblastic pattern of Paget's disease. Multiple imaging modalities were used to

determine the extent of the lesion for resection. CT of the head (Figs. 3A and 3B) revealed the tumor with intracranial extension but with intact dura. MRI of the head (Figs. 4A and 4B) with multiple spin echo sequence showed calvarial destruction and hypertrophic vessels at the site of the tumor. These findings were compatible with sarcomatous degeneration of Paget's disease but they can also be seen in other tumors complicating Paget's disease such as chondrosarcoma, fibrosarcoma or malignant giant cell tumor. Carotid angiography showed tumor neovascularity. A pre-operative biopsy established this lesion as an osteosarcoma. The combination of imaging procedures accurately determined the exact extent of the tumor. This was important because the usual surgical resection margins extended into the base of the skull and precluded a surgical cure. The usual resection includes conservative margins of normal tissue, which were safely reduced by the exact definition of the tumor margins.

DISCUSSION

Paget's disease is infrequently complicated by the development of osteosarcoma. Other tumors known to arise in pre-existing Paget's disease include; chondrosarcoma, fibrosarcoma, and rarely malignant giant cell tumors or malignant fibrohistiocytomas (2). Osteosarcomas arising in Paget's disease account for between 20 and 30% of all osteosarcomas arising in patients over 40 years of age (3). The largest published series (4) of 4415 patients with Paget's disease found the incidence of sarcomatous degeneration to be 0.9%. The range reported in the literature varies from 0.15 to 5% (5,6).

One of the larger series published (2) described 17 Paget's patients in which 13 had decreased localization of [^{99m}Tc]MDP in the region of the sarcomatous degeneration while in the remaining four patients this de-

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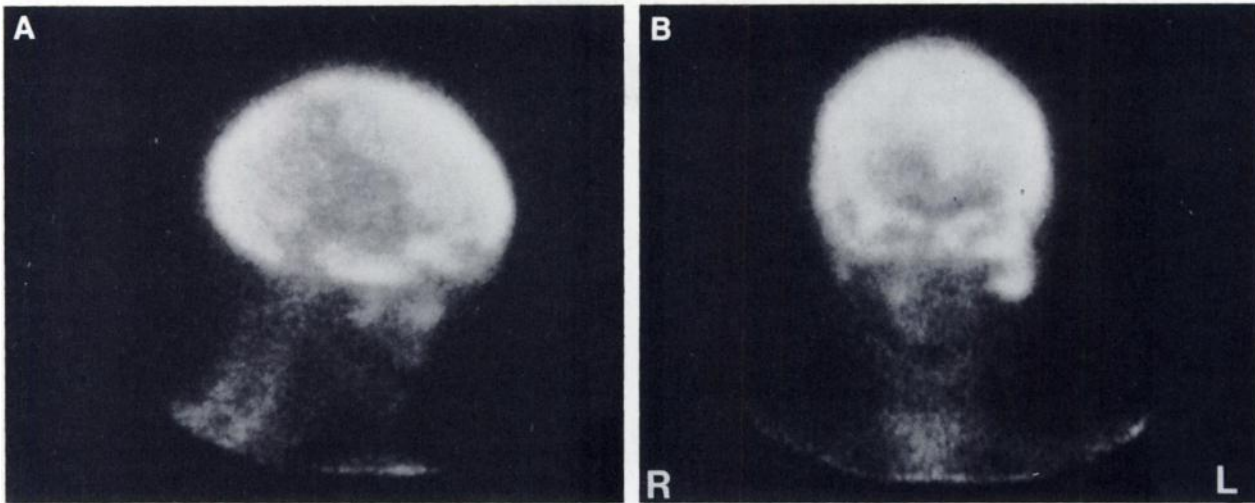


FIGURE 1

A,B: Right lateral and anterior views, respectively, of bone scan of August 1983. Images of the skull show changes of Paget's disease.

crease was less obvious. An isolated case report confirms this decrease (7). Shirazi and Fordham (8) describe the bone scan findings as that of extraskelatal extension of tumor activity beyond the margins of the bone. The presented case report, in addition also demonstrates increased accumulation of the radiotracer within it.

Shirazi et al. (8) describe the limited usefulness of bone scans in differentiating Paget's disease from an osteosarcoma because both produce intense uptake. However, many case reports show decreased uptake in the sarcoma. The case presented is unusual because intense localization of the tracer in the sarcoma is greater than that of the adjacent pagetic bone. Also, the bone scan in this patient was concluded to be an important part of the work up to define the tumor margins prior to surgery, although CT and MRI were helpful also.

SUMMARY

A case of osteosarcoma arising in pre-existing Paget's disease of the calvarium is presented in which the bone scan was helpful in defining tumor margins prior to resection. The scintigraphic findings in this case of increased radiotracer accumulation in the tumor in excess of the adjacent pagetoid bone is unusual.

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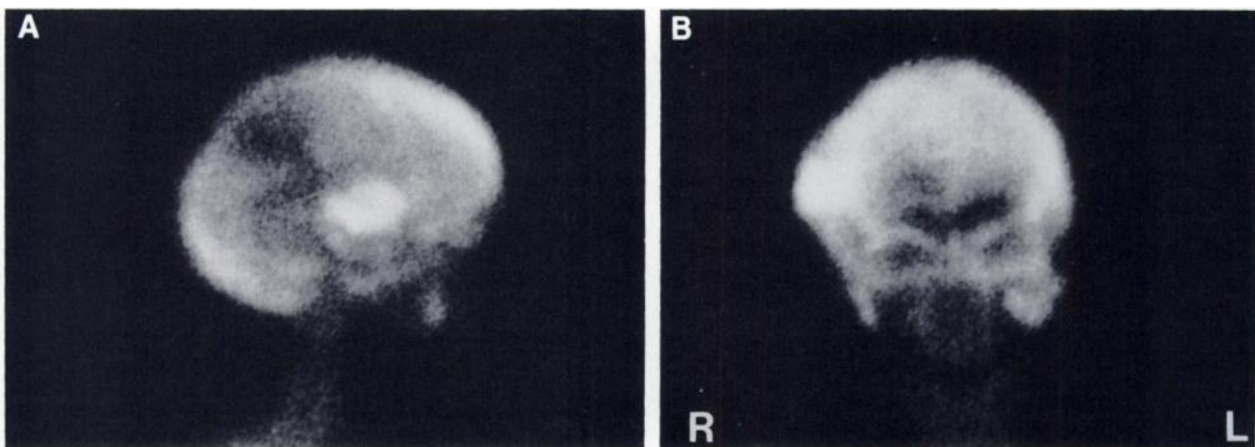


FIGURE 2

A,B: Right lateral and anterior views, respectively, of the bone scan of September 1985 show a focal area of intense uptake in the right temporal region, in addition to the changes of Paget's disease.

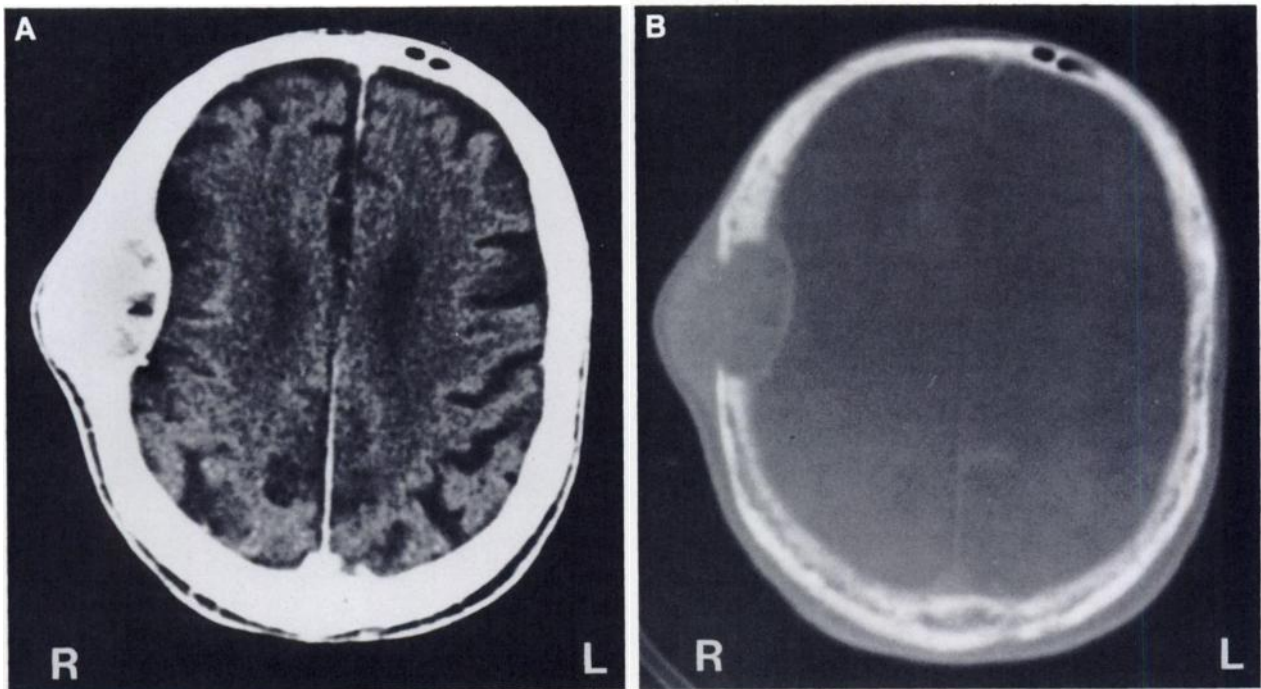


FIGURE 3

A,B: CT of the head shows a well-defined right temporal mass, with rest of the calvarium showing changes of Paget's disease. The dura appears intact.

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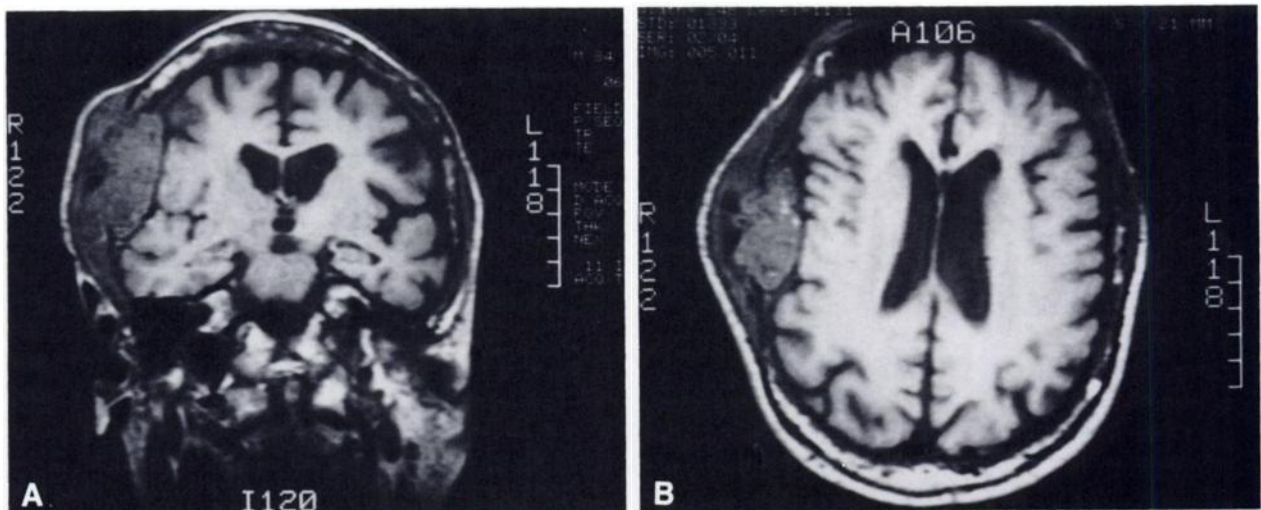


FIGURE 4

A,B: Coronal and Transaxial MR images of the head, respectively (using a GE 1.5 Tesla MR imaging system) show T2 weighted images with a TR of 800 msec and TE of 25 msec and abnormal signals from the calvarium consistent with osteosarcoma complicating Paget's disease.