Brain and bone scans in primary Ewing's sarcoma of the petrous bone

Case report

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The authors present a case in which primary Ewing's sarcoma of the right petrous pyramid in a 9-year-old girl showed no uptake on a $^{99m}$Tc-pertechnetate nuclide angiogram. Intense uptake was present on a $^{99m}$Tc-polyphosphate bone scan, but a static brain scan was only minimally abnormal. The diagnosis and treatment of Ewing's sarcoma are reviewed.

KEY WORDS □ Ewing's sarcoma □ brain scan □ $^{99m}$Tc-polyphosphate □ bone scan

PRIMARY bone sarcomas are infrequent in childhood, and represent only 2% of all neoplasms in children under 15 years of age. This report describes $^{99m}$Tc-pertechnetate brain and $^{99m}$Tc-polyphosphate bone scans in a child with a primary Ewing's sarcoma of the right petrous bone.

Case Report

A 9-year-old girl was admitted with severe right otalgia and recent onset of a partial peripheral right seventh nerve paralysis. She had a long history of recurrent otitis media, and Teflon tubes had been placed in both ears 3 years previously. These had been replaced 4 months prior to the present admission. She had then experienced gradually increasing pain in the right ear, attributed to recurrent serous otitis; her mother had noticed drooping of the child's right facial muscles 10 days prior to admission. An audiogram at that time suggested recurrent fluid in the right middle ear. A Reuter bobbin was inserted in the right tympanic membrane, and prednisone therapy was begun. There was some initial response, but the pain later increased in intensity and she was admitted for evaluation.

Examination. The physical examination was normal except for a mild conductive hearing loss and lower motor neuron facial weakness, both on the right side. The Reuter bobbin was noted in place, but the tympanic membranes were not inflamed. Abnormal laboratory studies included phosphorus 6.1 mg (normal range of 3.6 to 5.6 mg) lactic dehydrogenase, 215 IU (normal range of 90 to 200 IU), and 15,500 white blood cells with 60% polymorphonuclear cells. A chest film and metastatic bone survey were normal.
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Fig. 1. Tomogram showing the orderly expansion and overgrowth of the right petrous bone (large arrow). There is narrowing of the internal auditory canal (small arrow). The metallic Reuter bobbin in the tympanic membrane is noted (double arrows).

Tomograms of the petrous bones showed asymmetry of the petrous pyramids with apparent overgrowth and expansion of the petrous bone on the right. The internal auditory canal on the right appeared narrow (Fig. 1). The radionuclide cerebral angiogram showed symmetrical normal perfusion. The static scintiphotos showed minimal increased uptake on the right lateral view at the base adjacent to the lateral sinus (Fig. 2). A $^{99m}\text{Tc}$-polyphosphate bone scan revealed intense activity over the right petrous bone, but the skeleton was otherwise normal (Fig. 3). The impression based on the radiographs and scans was monostotic fibrous dysplasia of the right temporal bone. Neurofibromatosis was also considered. A meningioma en plaque could not be excluded. Petrositis was considered unlikely.

Operation. Operative decompression of the right seventh nerve was performed through a middle fossa approach. The petrous bone was diffusely soft, hypervascular, and erythematous. A frozen section showed a chronic inflammatory pattern with atypical cells. It was elected to terminate the procedure at this point to await the permanent sections, which revealed Ewing’s sarcoma (Fig. 4).

Discussion

Ewing’s sarcoma is a tumor of young people: 90% of these patients are under 30 years of age. Males are affected more frequently, with the male to female ratio about 1.6 : 1. The tumor is histologically highly anaplastic, with solidly packed small round cells; glycogen can usually be demonstrated in the tumor cells with special stains.

Most of the primary tumors occur in the long bones (47%), pelvis (19%), or ribs (12%). The skull is rarely involved, probably in less than 4% of cases. The tumor is somewhat more common in the jaws, and the total incidence in the skull and jaws is usually given as 9%. None of the reports we reviewed gave specific anatomical locations of the primary skull tumors. From 14% to 28% of patients have evident metastases at the time of diagnosis. The 5-year survival rate ranges from 8% to 24%. Radiation therapy
with systemic chemotherapy is advocated.\textsuperscript{17,33} A 5-year survival rate of 53\% has been recently reported in a series of patients with localized primary tumors treated with radiotherapy and adjuvant chemotherapy;\textsuperscript{16} the pattern of metastasis in patients receiving such combined treatment may be altered.\textsuperscript{21}

Certain clinical and laboratory factors may be prognostically important.\textsuperscript{26} The radiographic appearance of Ewing's sarcoma is quite variable, but often shows extensive longitudinal patchy bone destruction, abundant lamellar periosteal new bone formation, and an associated soft tissue mass.\textsuperscript{10} The soft tissue mass of Ewing's sarcoma does not ossify, in contrast to the soft tissue mass of osteogenic or chondrogenic sarcoma.\textsuperscript{8} In the present case the apparent orderly bony overgrowth and expansion of the right petrous bone suggested a defect of mesenchymal development, either fibrous dysplasia or neurofibromatosis. Fibrous dysplasia may be monostotic in the skull.\textsuperscript{20,23,24} The bony changes of meningioma \textit{en plaque} may be difficult to differentiate from fibrous dysplasia.\textsuperscript{19} Neurofibromatosis not uncommonly involves the skull\textsuperscript{7,18,24} and may mimic the changes of fibrous dysplasia. Petrositis would be unlikely to cause overgrowth and expansion of the bone.

The scan findings in the present case are rather nonspecific, but point toward a primary bone lesion. The markedly positive bone scan with the minimally abnormal pertechnetate brain scan suggests an osseous process, although this pattern is not rare in cerebral infarcts and some other intracranial lesions.\textsuperscript{1,14} Positive bone and brain scans have been reported in fibrous dysplasia.\textsuperscript{18,31,32,36} The normal radionuclide angiogram and static brain scan findings tend to exclude meningioma.\textsuperscript{5} Bone scanning with \textsuperscript{99m}Tc-labeled agents is effective in delineating the primary lesion and bone metastases in Ewing's sarcoma.\textsuperscript{13} Scanning with \textsuperscript{67}Ga may show a greater extent of involvement of the primary lesion than that demonstrated on the

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**Fig. 3.** Intense $^{99m}$Tc-polyphosphate uptake in the right petrous bone is evident in anterior (left), posterior (center), and right lateral (right).
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bone scan; chest radiographs are most effective in showing pulmonary metastases. Ewing's sarcoma also may show uptake on radionuclide angiography. Although extraskeletal uptake of radiostrontium and 99mTc-labeled bone scanning agents has been reported in several other types of tumor of young people we are not aware of any report of uptake in extraskeletal Ewing's sarcoma. This may reflect the absence of radiographically evident calcification in the soft tissue mass of Ewing's sarcoma.

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References


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