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Spinal cord Ewing's sarcoma metastasis: presentation of one case

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Cervical spinal Ewing's sarcomas are rare and cause problems in diagnosis. We present an unusual case of a primary extraosseous Ewing's sarcoma arising from the spinal cord. An 18-year-old woman with fever, headache and back pain lasting one month was admitted to the hospital. Whole body bone scintigraphy was performed with 1110 MBq technetium-99m methylenediphosphonate. Scintigraphy clearly showed abnormal technetium-99m methylenediphosphonate accumulation in the level of the 5th and 6th cervical vertebrae. Magnetic resonance imaging could also confirm this examination finding. After the scintigraphic study, the patient underwent surgery. Pathological diagnosis of the operation specimen was Ewing's sarcoma.

Key words: extraosseous Ewing sarcoma, spinal cord, technetium-99m methylenediphosphonate bone scintigraphy

INTRODUCTION

EWING'S TUMOR was considered the most lethal of all bone neoplasms. Modern therapeutic approaches with radiation and chemotherapy have improved the prognosis considerably. Ewing's tumor is rare, making up about 6% of all malignant bone tumors. There is a distinct predilection for males. Most patients are young; however, it is unusual to see Ewing's sarcoma in patients younger than 5 years, an age group in which metastatic neuroblastoma is more common. Any portion of the skeleton may be involved, but the long bones are the most typical sites. The lesion usually involves the shaft of the bone. Patients with Ewing's sarcoma usually have localized pain or swelling at presentation. However, some patients have systemic manifestations, such as fever, weight loss, and an increased erythrocyte sedimentation rate, suggesting the possibility of osteomyelitis.

On roentgenograms, Ewing's tumors tend to be extensive, sometimes involving an entire bone. The lesion is usually a permeative, destructive process, with multiple tiny areas of destruction involving the bone. Typically, Ewing's tumor produces a pronounced reactive new bone formation of the periosteum, giving rise to an onion-skin appearance.

The p30/32^{MIC-2} gene product, CD99, is a cell-surface glycoprotein expressed in Ewing's sarcomas and primitive neuroectodermal tumors. Strong membrane staining for CD99 is consistently seen in Ewing's sarcoma with monoclonal antibodies 12E7, HBA-71, and O13. CD99 expression can be seen in other malignant neoplasms, including sarcomas, carcinomas, and hematopoietic lesions, such as lymphoblastic lymphomas and acute lymphocytic leukemias.[1–4] Cytogenetic and molecular genetic studies can also be useful adjunctive tools in diagnosting Ewing's sarcoma. The t(11; 22) (q24; q12) chromosomal translocation can be identified in most Ewing's sarcomas and primitive neuroectodermal tumors.[5–7]

The differential diagnosis includes metastatic neuroblastoma and malignant lymphoma. Other small cell malignant lesions, such as small cell osteosarcoma and mesenchymal chondrosarcoma, are ruled out by the presence of matrix within the neoplasm. Metastatic neuroblastoma usually is not a problem, because most often it is already known that the patient has a neuroblastoma. The diagnosis can be very difficult. If the lesion shows clear-cut rosette formation and a neurofibrillary background, the diagnosis of neuroblastoma is obvious. However, some neuroblastomas do not show such differentiation. Malignant lymphomas usually affect older patients, and the infiltrate is polymorphic. In a small percentage of

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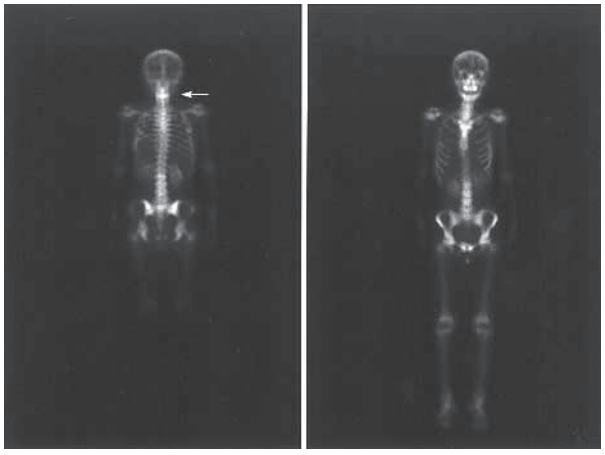

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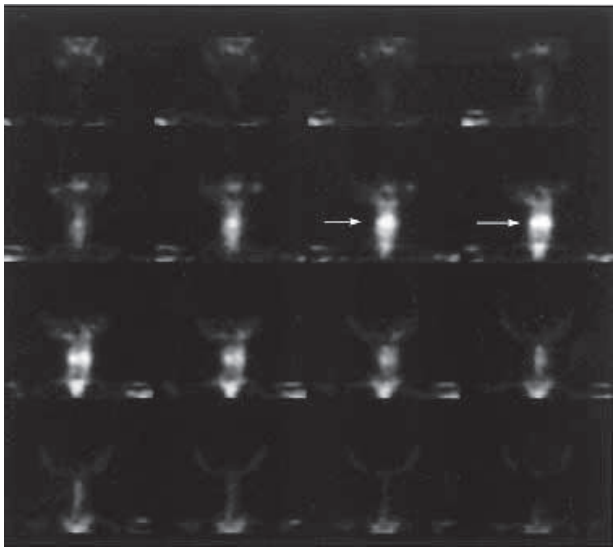

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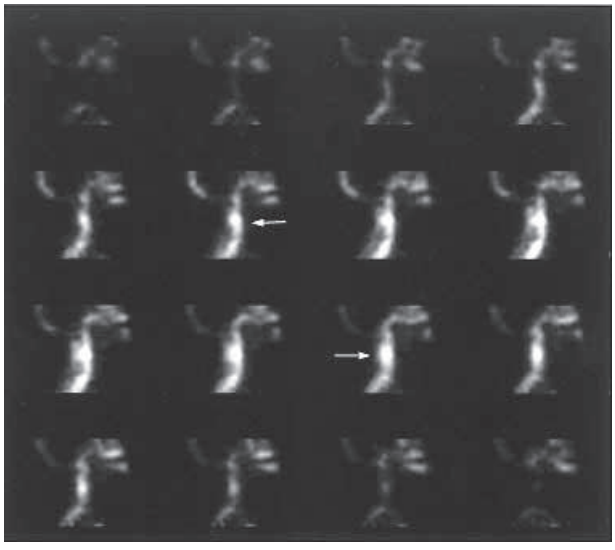

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Fig. 1 Whole body bone image (A) and tomographic images (coronal (B), sagittal (C)) of an 18-year-old woman with fever, headache and back pain demonstrate abnormal technetium-99m methylenediphosphonate accumulation at the level of the 5th and 6th cervical vertebrae (*arrows*).

cases, it can be impossible histologically to differentiate lymphoma from Ewing's sarcoma.

Nuclear medicine is important in the diagnosis, staging, and long-term surveillance of a number of pediatric cancers. Skeletal scintigraphy is used to evaluate Ewing sarcoma. The therapeutic response of primary bone and brain tumors can be assessed using Tl-201 and Tc-99m MIBI scintigraphy. Pediatric oncologic applications of positron emission tomography are being investigated extensively. Cervicothoracic lesions of Ewing sarcoma are not uncommon in children. All cervicothoracic lesions except superficial lesions extend from the neck to the thorax through the thoracic inlet. Evaluation of this area involves multiple imaging modalities. Magnetic resonans imaging is the method of choice for assessing the full extent of cervicothoracic lesions and their relationships to neurovascular structures.[21–25]

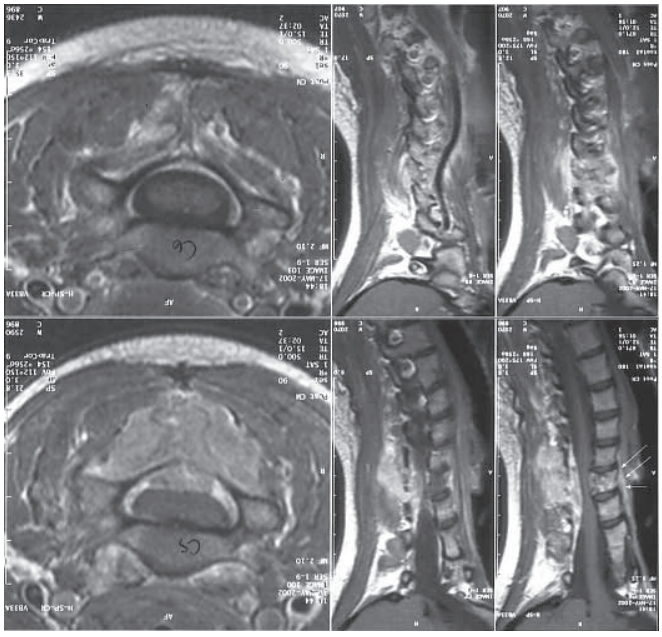
CASE REPORT

An 18-year-old woman with fever, headache and backpain lasting one month was admitted to the hospital. The patient underwent whole body bone scintigraphy. The patient was injected intravenously with 1110 MBq technetium-99m methylenediphosphonate and imaged 4 hours later. The site of injection was distant from any suspected osseous pathology. The patient was routinely scanned using a whole Starcam 3200 SPECT (General Electric Co.) camera with LEAP (low energy all purpose) collimator and 10 cm/min imaging speed and tomographic images were also obtained. We obtained 1000 k counts for whole body imaging with 256 × 256 matrix.

Scintigraphy clearly showed abnormal technetium-99m methylenediphosphonate accumulation in the 5th and 6th cervical vertebrae (Fig. l). Magnetic resonance imaging could also confirm this examination finding (Fig. 2). After the scintigraphic study, the patient underwent surgery. Pathological diagnosis of the operation specimen was Ewing's sarcoma.

DISCUSSION

Ewing sarcoma is a malignant tumor of bone that comprises approximately 10% to 15% of all primary bone tumors. The tumor cells are believed to be of neuroectodermal origin. Patients with this disease have fever, anemia, leukocytosis, and increased erythrocyte sedimentation rate at admission. Pain is the most common symptom.



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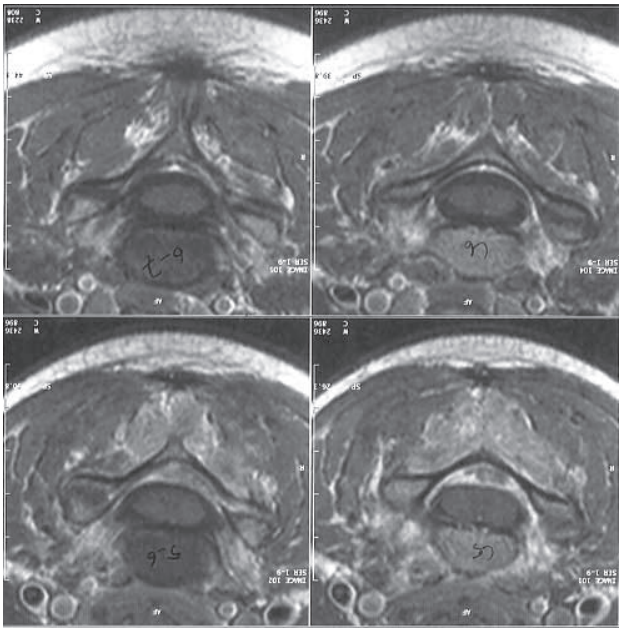

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Fig. 2 Magnetic resonans imaging shows lesion at the level of the 5th and 6th cervical vertebrae.

About 70% of patients experience pain, which may be dull to severe and becomes persistent for about 1 month prior to diagnosis.[8–12]

More than 80% of Ewing sarcomas occur during the first two decades of life. The tumor is uncommon after age 30. The youngest patient reported was 5 months old. Krieun has shown that the distribution of Ewing sarcoma varies with age and mirrors the distribution of marrow.[13] The most common site of the primary tumor was the pelvis (25%) followed in order of frequency by ribs, femurs, spine, tibias, fibulas, scapulas, and other bones.[13,14]

Evaluation of the primary tumor is based on plain radiographs followed by skeletal scintigraphy and magnetic resonance imaging. On radiographs, the lesion is typically lytic, usually with poorly defined margins. On magnetic resonance imaging, Ewing sarcoma frequently is associated with a prominent soft tissue mass that contains areas of necrosis or hemorrhage. Evaluation of soft tissue extent is particularly crucial, where most of the tumor may be extraosseous.

Gallium-67 citrate scintigraphy, which is typically abnormal in Ewing sarcoma, has been reported to be more accurate than imaging with technetium-99m methylenediphosphonate for determining both the response to therapy and relapse.[18] Thallium-201 scintigraphy typically shows intense accumulation in this disease, which appears to be limited to the tumor itself and not the surrounding bone reaction. Some studies have shown a correlation between tumor regression following chemotherapy and reduced uptake of thallium-201.[19,20]

This case clearly showed that technetium-99m methylenediphosphonate whole body bone scan and tomography are good tools for Ewing sarcoma detection. The scintigraphic appearance in Ewing sarcoma is one of intense uptake of bone tracer in the lesion.[15–17]

Patients with small extremity tumors and no clinically detectable metastases at diagnosis have a favorable outcome, with cure rates in excess of 70%. Patients with large, centrally located tumors and those with metastatic disease at diagnosis have a poorer prognosis. Although small, single-institution studies report survival rates up to 50% in these two higher risk groups, it is unlikely that more than 33% are truly cured with available treatment, given the potential for late relapses.[21]

CONCLUSION

Among the proposed imaging methods, magnetic resonance imaging is the best choice for the evaluation of patients with extraosseous Ewing sarcoma, but technetium-99m methylenediphosphonate whole body bone scan and tomography have good diagnostic value for Ewing sarcoma. Because of the importance of metastatic disease in the choice of primary and follow-up therapy for this tumor, skeletal scintigraphy should be done at presentation and at frequent intervals thereafter.

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