

MDP Bone Scan: A Key Step in Assessment and Staging of a Rare Presentation of Ewing Sarcoma in a Young Adult Man

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Abstract

A Spanish speaking young man who presented with a painful supra-orbital head mass causing progressive diplopia and epistaxis. Initial clinical and radiological work-up suggested an esthesioneuroblastoma (i.e., olfactory neuroblastoma) with its associated good prognosis. However, further history (obtained through an interpreter) caused the primary physician to order a nuclear bone scan. The results from this scan dramatically changed the clinical approach, tumor work-up, diagnosis and treatment of this patient who was found to have an atypical metastatic Ewing sarcoma.

INTRODUCTION

Ewing sarcoma (ES) typically presents as a tumor originating in the diaphyseal marrow of the long and flat bones which spreads throughout the bone cortex to form a non-osteoid soft tissue mass. Most frequently ES affects Caucasian boys under the age of 15, with a prevalence of approximately 2-3 cases per 1 million population. It is a small blue cell tumor of the primitive neuroectodermal type. From 15 to 30% of cases present with metastatic disease although aggressive spread of this neoplasm to the head or skull is very rare [1]. This case report demonstrates the importance of nuclear medicine bone imaging in the evaluation of a young adult with an anterior skull base tumor, initially diagnosed as an olfactory neuroblastoma. The Tc-99m MDP bone scan strongly suggested a primary lower extremity ES with metastatic spread to the anterior skull base, calvarium and appendicular skeleton and confirmed histologically.

CASE REPORT

A 19 year old Spanish speaking man presented to our ER with a one month history of a palpable, hard, right supraorbital facial mass, causing progressive worsening diplopia and "old blood" brown color epistaxis. The patient reported hitting his head and attributed the swelling to trauma. He denied fevers, loss of appetite, weakness or numbness of his extremities. A complicated, expansile extra-axial lesion, seen in the pre-surgical CT scan [Fig 1.],

appeared to originate in the sinuses which involved both the inner and outer table of the calvarium.

Figure 1

Figure 1: Presurgical CT of the head with contrast shows an expansile, extra-axial, anterior skull base lesion affecting the sinuses, orbit and calvarium with imaging characteristics similar to ENB.



The magnetic resonance (MR) imaging report from an outside hospital described a complicated, 6 cm diameter, retro-orbital solid and cystic lesion centered in the right

paranasal sinuses with calvarium invasion. Because of the patient's age, history, epistaxis, tumor location and radiographic pattern the initial imaging findings were interpreted as most consistent with esthesioneuroblastoma (ENB), i.e. olfactory neuroblastoma [2,3]. Initial intraoperative frozen section analysis of the lesion appeared concordant with the radiological interpretation. Surgical resection of the anterior skull base lesion was performed and histological evaluation yielded a non-specific diagnosis of a small round cell tumor.

Subsequent patient history and complaints of chronic left lower leg pain and swelling, (attributed by the patient to prior trauma to the leg approximately 2 years ago), led to referral to nuclear medicine for skeletal assessment by MDP bone scan and plain X-ray of the left lower extremity.

Whole body images obtained 3 hours post injection of 26.8 mCi (992 MBq) of Tc-99m MDP showed diffuse increased activity in the mid and distal aspect of the left fibula [Fig 2, A] concordant with the lesion also imaged with plain radiography [Fig 3] and MRI [Fig 4]. The nuclear bone study also showed increased activity in the right proximal femur [Fig 2, B] and focal increased activity in both the anterior and posterior aspects of the rib cages [Fig 2, C]. Activity in the mid-frontal bone was consistent with the recent midline craniotomy, but residual neoplasm could not be ruled out. Other bilateral temporal-parietal "hot" areas [Fig 2, D] were confirmed as lytic foci in the calvarium in the post operative CT scan [Fig 5].

Figure 2

Figure 2: Anterior and posterior whole body 99m-Tc MDP bone scan shows increased uptake in the left fibular (A), the contralateral femur (B), the ribs bilaterally (C) and the calvarium (D).

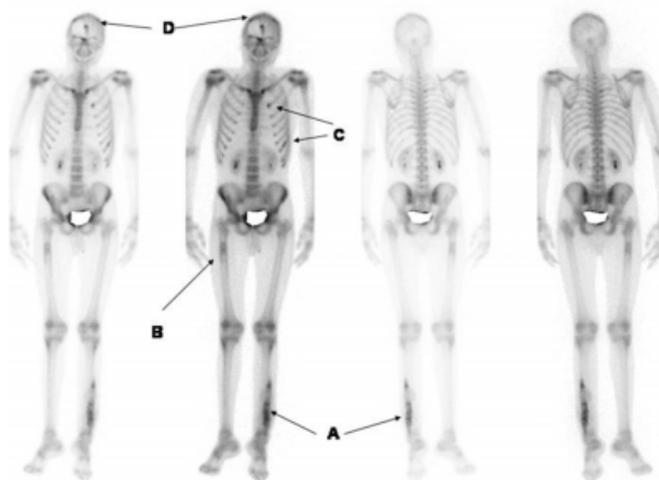


Figure 3

Figure 3: X ray of left lower extremity shows fibular lesion which had significant increased activity on the MDP bone scan.



The nuclear medicine bone scan findings offered the alternative diagnosis of a primary left lower extremity tumor

with metastatic spread to the calvarium, ribs and contralateral extremity, suspicious for ES. The suggestion of ES was also raised in the subsequent report of the MRI study of the left lower extremity [Fig 4]. Surgical excision of the left fibular lesion and histological analysis found a small blue cell tumor, with widespread membranous immunoreactivity for CD-99 and type 1 EWS/FLI-1 fusion transcript concordant with a Ewing sarcoma. Further analysis of the extraaxial lesion also gave positive CD99 stain and type 1 EWS/FLI-1 fusion transcription. It is presumed that the fibular lesion was the primary tumor, since metastatic spread of ES from the skull base to the appendicular skeleton is very rare.

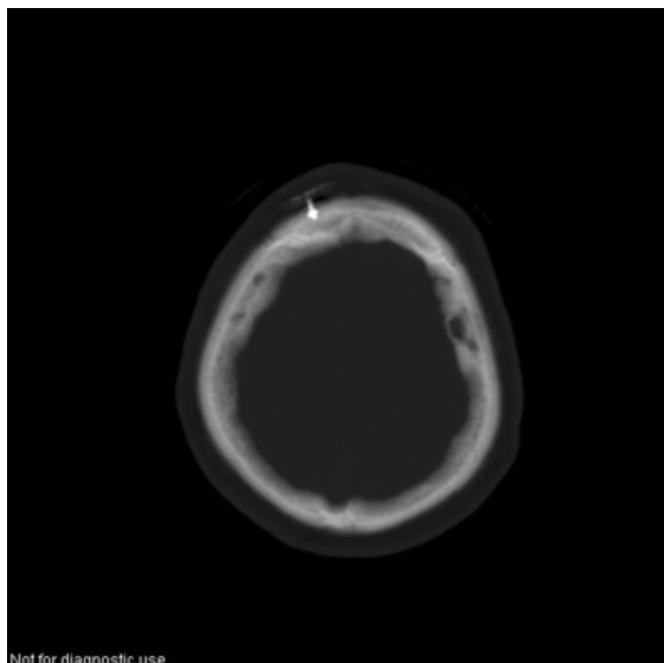
Figure 4

Figure 4: MRI of the left lower extremity, T1 post contrast, shows fibular lesion with complex soft tissue component.



Figure 5

Figure 5: CT of the head, bone windows, confirm focal areas of increased uptake on the MDP bone scan.



DISCUSSION

The differential diagnosis for anterior skull base tumors is extensive, with a spectrum from benign angiofibroma to malignant and metastatic lesions including melanoma, squamous cell carcinoma and esthesioneuroblastoma (ENB) [4]. ENB is a rare polymorphic tumor typically arising from the olfactory mucosa. This tumor most commonly affects young men, two-thirds presenting between 10-40 years of age [5,6]. Microscopic diagnosis is challenging and problematic as ENB has variable histology which may be mistaken for other malignancies [5] while other malignancies may be mistaken for ENB [7]. ENB is no longer classified as a primitive neuroectodermal tumor of the Ewing's type because it does not have FLI-1 expression which differentiates it from ES [8].

The clinical importance of distinguishing ES from ENB is the divergence in therapy and survival. ENB may have indolent growth with a patient surviving with a known tumor for longer than 20 years. Typical 5 year survival is 50-80% with surgical resection and adjuvant radiation therapy as clinically appropriate [6]. Yet ENB can also express itself as a highly aggressive neoplasm capable of rapid widespread metastases which can limit patient survival to a few months [9]. Ewing sarcoma is usually treated with chemotherapy and local resection with less than 30% five-year survival in cases

with disseminated disease [10].

The difficulty in radiographic and pathologic assessment of anterior skull base lesions is redemonstrated in this instance. Both initial imaging and histological findings suggested ENB. Correct diagnosis was made only after subsequent patient complaints and discovery of the primary tumor by MDP bone scan, confirmed by MRI, surgical excision and pathology.

Metastatic spread of ES to the calvarium is so rare there is no precise estimation of the incidence but case reports are known [11]. Despite the rarity of this finding, this report demonstrates the importance of nuclear medicine bone scan for evaluation for metastatic disease in the clinical assessment of anterior skull base lesions. Furthermore, metastases may arise not only from a distant site, as in this case, but also from primary anterior skull base lesions in approximately 5-10% of all presenting tumors [12]. We believe the judicious use of nuclear medicine studies is warranted in the diagnostic evaluation of complex anterior skull base lesions for evaluation of metastatic disease [13] since the prognosis, therapy and outcome for these lesions can be so disparate for the patient.

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