SYNCHRONOUS MULTICENTRIC OSTEOSARCOMA IN PUERTO RICO: A Pediatric Case Report

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ABSTRACT

Synchronous multicentric osteosarcoma is a very rare condition that might be confused with metastatic osteosarcoma. We report the first pediatric case of synchronous multicentric osteosarcoma reported in the Puerto Rican. The child presented with multiple osteoblastic bone lesions followed by right upper lid ptosis. Orbital CT-Scan showed a lytic lesion involving the sphenoid bone. Management consisted of chemotherapy for forty-two weeks after which she remained without new secondary bone lesions.

Index words: *synchronous, multicentric, osteosarcoma, Puerto Rico, pediatric*

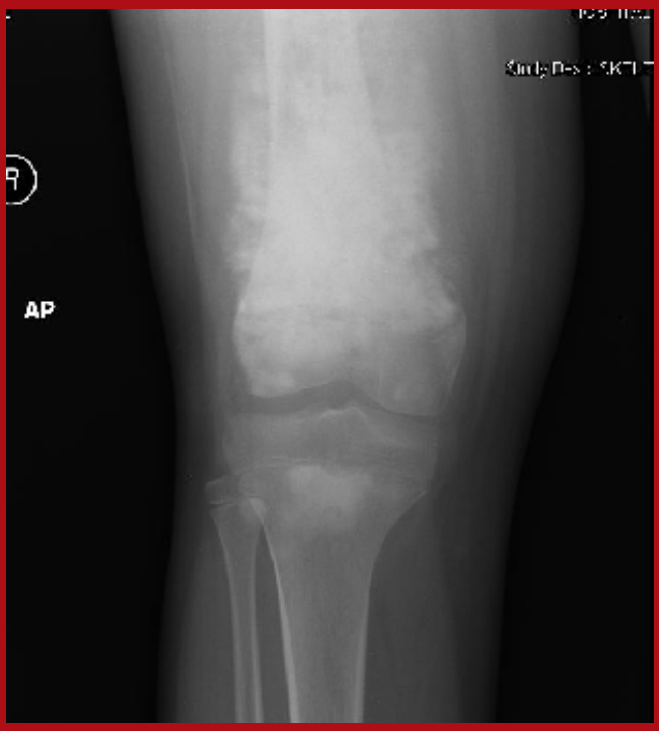
INTRODUCTION

Osteosarcoma is one of the most common bone malignancies of childhood. It usually presents at a solitary site, usually around the knee, and has a tendency to metastasize rapidly, preferably to the lungs. Multicentric osteosarcoma is a variant type of osteosarcoma. It represents around 1.5% of reported cases. Multicentric osteosarcoma is a destructive bone tumor, which presents with multiple lesions throughout the skeleton without evidence of lung disease (1). There exist two categories: Synchronous multicentric osteosarcoma which presents with multiple skeletal lesions that may occur simultaneously, and Metachronous multicentric osteosarcoma, most commonly seen in adults, in which bone lesions appear at different intervals (2,3,4). Synchronous multicentric osteosarcoma is a very rare condition in the pediatric population that may be confused with metastatic osteosarcoma. In this case report we highlight an unusual case of an adolescent female with synchronous multicentric osteosarcoma diagnosed and managed in Puerto Rico.

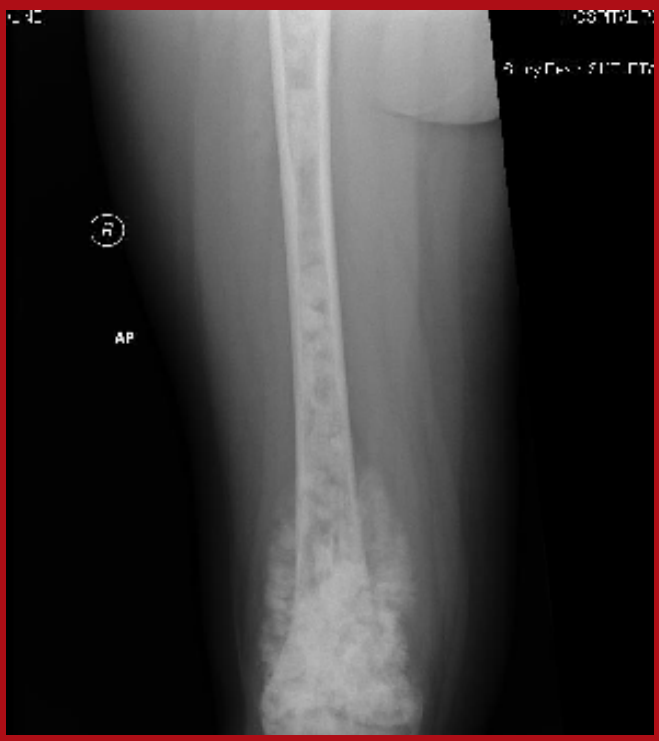
Case History

We report a case of a 12-year-old female previously in good health, who presented with a swollen, painful right knee after a fall from a monkey bar 4 days prior to evaluation. Initial radiographs showed a large lesion in the distal femur. She was transferred to the University Pediatric Hospital of Puerto Rico where a skeletal survey revealed multiple osteoblastic bone lesions. There were no history of constitutional symptoms like fever, weight loss or night sweats. On physical examination, there was moderated swelling and tenderness of her right distal thigh; as well as pain along her right knee arc of motion. The remainder of the exam was normal. Over the following days this patient developed a right upper eyelid ptosis progressing to a cranial nerve III palsy. All routine labs were within normal limits, except for an elevated Alkaline Phosphatase (2781 IU/L), and uric acid (6.2mg/dL).

Plain radiographs showed multiple osteoblastic bone lesions (see Figures 1, 2). Her chest



Figures 1 and 2: Right AP view shows a large heterogeneous osteblastic lesion within the distal femoral metaphysis and epiphysis extending to the diaphysis with a large soft tissue component.



radiograph was normal. A CT-Scan and MRI of her lower extremities revealed multifocal osteosarcoma with a dominant lesion centered at the right distal femoral metaphysis (see Figure 3). Orbital CT-Scan revealed a lytic lesion with a soft tissue component involving the lesser wing of the sphenoid bone (see Figure 4). Thoracic CT-Scan showed no evidence of metastatic disease to the pulmonary parenchyma (see Figure 5). Whole-body bone scintigraphy was compatible with extensive skeletal metastatic disease. It showed increased activity in the calvarium, the right orbital region, right clavicle, humerus, pelvis, both lower extremities, and spine at the level T4, L3 and L4 (see Figure 6). Incisional biopsies taken from the lesions in the right distal and proximal femur revealed a high-grade osteosarcoma. After these findings, the patient was started on a chemotherapy protocol that consisted of Cisplatin, high dose Methotrexate and Adriamycin.

DISCUSSION

This is the first case of a synchronous multicentric osteosarcoma reported in the Puerto Rican population. The accurate origin of the multicentric osteosarcoma cannot be established. Multicentric origin and p53 germ line mutations can contribute to enhanced predisposition to tumor growth (5). The most common localization of multicentric osteosarcoma is the metaphyseal region of longs bones in the upper and lower extremities (6). This patient also presented with irregular bone (sphenoid) involvement without pulmonary disease. After management with chemotherapy for 42 weeks the child remains without evidence of new secondary bone lesions. Surgical intervention will only be considered for palliative treatment. Despite chemotherapy, her prognosis is poor (6).

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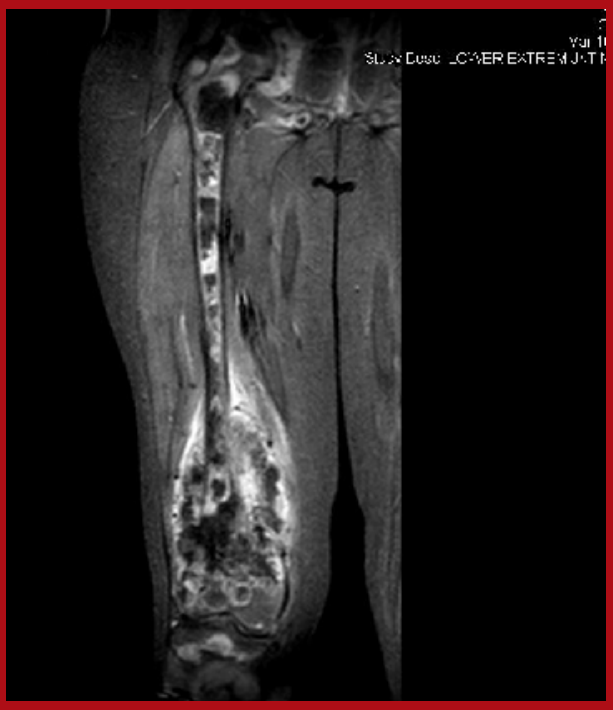

Figure 3: MRI lower extremities reveals a heterogeneous signal intensity mass arising from the distal metaphysis of the femur, with a wide zone of transition and a large soft tissue mass.

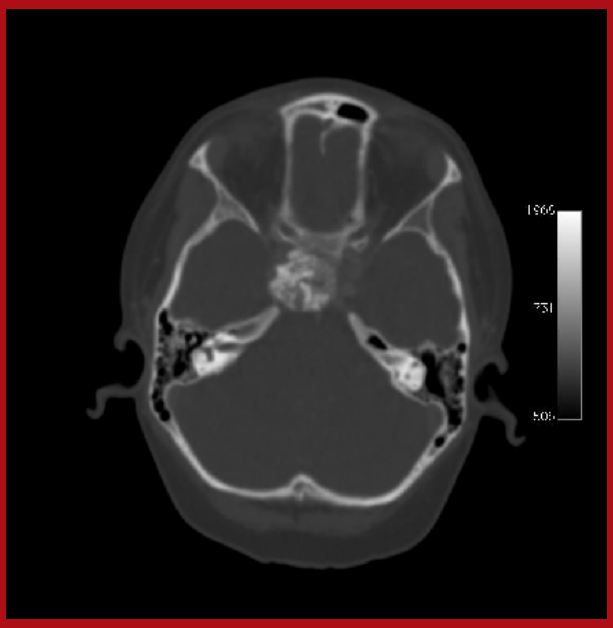

Figure 4: Head CT-Scan reveals a lytic lesion with a soft tissue component involving the sphenoid bone.

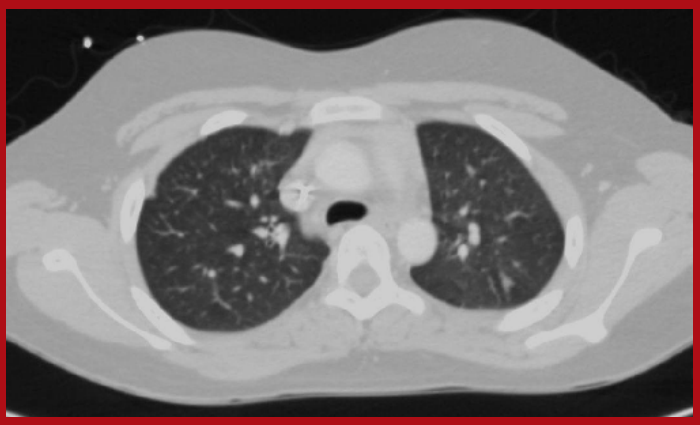

Figure 5: Thorax CT-Scan shows no evidence of metastatic disease to the pulmonary parenchyma.

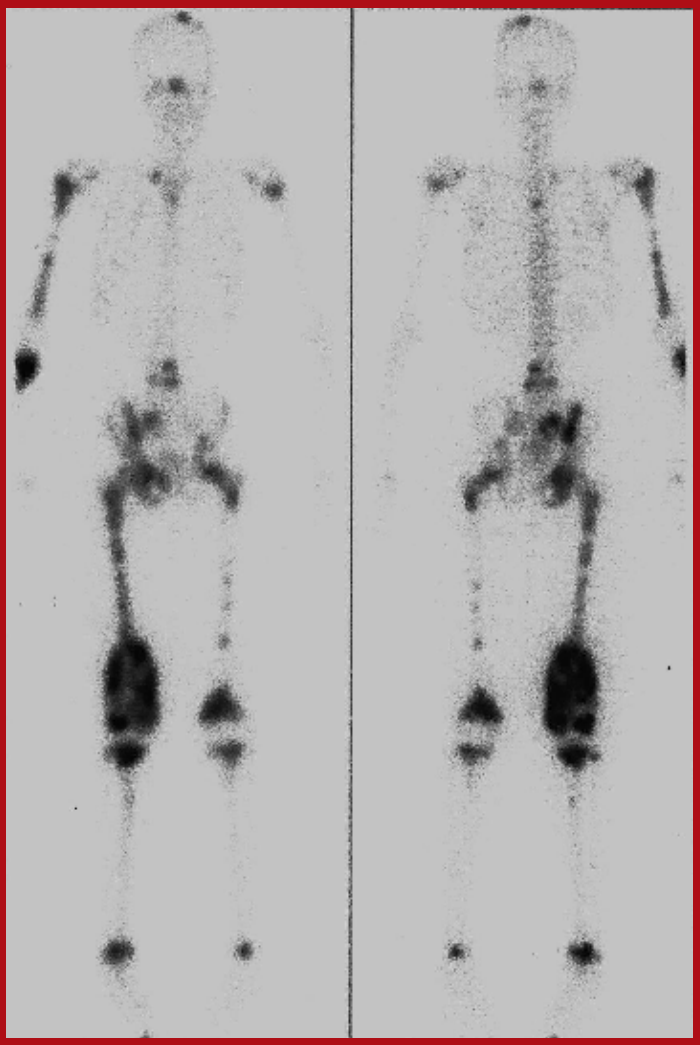

Figure 6: Bone scan findings are compatible with extensive skeletal disease.

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RESUMEN

Osteosarcoma multicéntrico sincrónico es una condición rara que se puede confundir con osteosarcoma metastático. Reportamos el primer caso pediatrico de osteosarcoma multicéntrico sincrónico en Puerto Rico. En su evaluación inicial la paciente presentó con múltiples lesiones del hueso seguido de ptosis del parpado derecho superior. Tomografía computarizada de la órbita demuestró una lesión lítica envolviendo el hueso esfenoide. La paciente fue manejada con quimioterapia por cuarenta y dos semanas luego de la cual permanece sin lesiones secundarias nuevas en el hueso.