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Case Reports

Primary mixed malignant tumor of bone in an 18-year-old male: Report of a case with radiologic-pathologic correlation



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ABSTRACT

We report a case of primary malignant mixed tumor (MMT) of bone in an 18-year-old boy with X-ray, CT, MR, scintigraphic, FDG PET, and pathologic correlation. Primary MMT of bone is a highly aggressive tumor and presents both a diagnostic and clinical treatment challenge. This tumor is extremely rare and to the best of our knowledge, this is the first report of the diagnostic imaging findings for primary MMT arising from bone in a patient of this age group.

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1. Introduction

Myoepithelial tumors consist of a family of morphologically similar lesions with an evolving nomenclature based on anatomic origin: pleomorphic adenoma of salivary gland, chondroid syringoma of skin and myoepithelioma or mixed tumor of somatic soft tissue and bone. Malignant forms, though rare, have been described in most sites and generally carry a poor prognosis [1–3]. Previous descriptions of this entity have described radiologic findings in adult patients [4,5]; however, X-ray, CT, MR, and FDG PET imaging findings in a pediatric-age patient have not been previously described. Furthermore, the lesion in this case exhibits a linear dense lace-like matrix on CT which has not been previously described with this lesion.

2. Case report

A previously healthy 18-year-old male developed progressive pain and swelling about the right knee over a 2-month period. He had no family history of malignancy. Five months prior to his diagnosis, he experienced minor trauma while playing and heard his knee “pop” when it hit the trampoline. The ensuing pain and swelling caused him to limp for several days, but the symptoms completely resolved within a week. Otherwise, his past medical history was negative. Three months after his knee injury he

developed new pain and swelling of the knee; this progressed rapidly over the next 2 months by which time the knee mass measured approximately 15 cm in diameter. The patient reported a 15-pound weight loss in the months leading up to the diagnosis. He did not have any other systemic symptoms.

Initial radiographs demonstrated an aggressive lesion in the medial condyle of the distal right femur (Figs. 1A and B). CT obtained to characterize matrix showed an aggressive lesion with a focus of dense linear matrix (Fig. 2) with intervening lucent areas. The lesion was felt to be indeterminate at the time, with overall features suspicious for an aggressive lesion such as a high-grade osteosarcoma. Much less likely considerations included secondary malignant degeneration of a cartilaginous lesion such as an enchondroma into chondrosarcoma, given the question of possible cartilaginous component on CT. Whole body Tc-99 MDP bone scan demonstrated abnormal uptake in the right femoral lesion, but no other abnormal areas (Fig. 3). A subsequent MRI demonstrated a large soft tissue mass associated with the osseous lesion and abnormal marrow signal intensity extending to the proximal femoral neck (Figs. 4A–C). Staging PET scan demonstrated metastasis to the lungs and right inguinal and external iliac lymph nodes (Fig. 5).

The incisional biopsy specimen from the right leg consisted of multiple fragments of pink tan soft tissue with focal calcifications. Hematoxylin and eosin stained microscopic sections demonstrated epithelioid cells in cords and solid sheets amid a myxoid to cartilaginous stroma (Fig. 6A and B). Focal areas showed bone arising from endochondral ossification of the cartilage. The tumor cells expressed cytokeratins and S-100. Cytogenetic analysis was remarkable for a clonal $t(X;19)(q13;p13.3)$ translocation. Bone marrow aspirate also showed metastatic disease.

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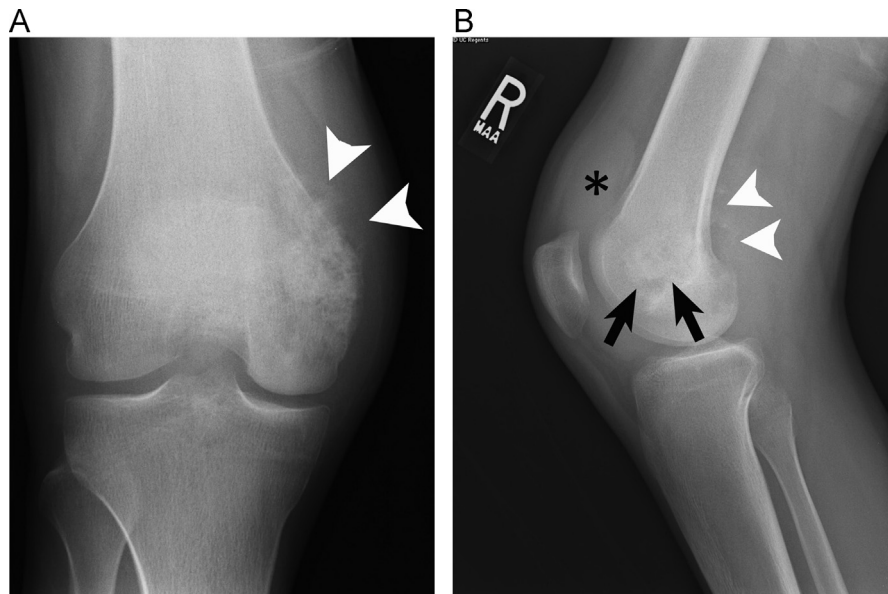


Fig. 1. A: Frontal radiograph of the right knee obtained at diagnosis demonstrating an aggressive lesion involving the medial condyle of the right femur with sunburst periosteal reaction (white arrowheads). B: Lateral radiograph obtained at diagnosis demonstrating a large joint effusion (black asterisk), “sunburst” periosteal reaction (white arrowheads), and a dense sclerotic metaphyseal lesion (black arrows). Overall findings were suggestive of an aggressive lesion.

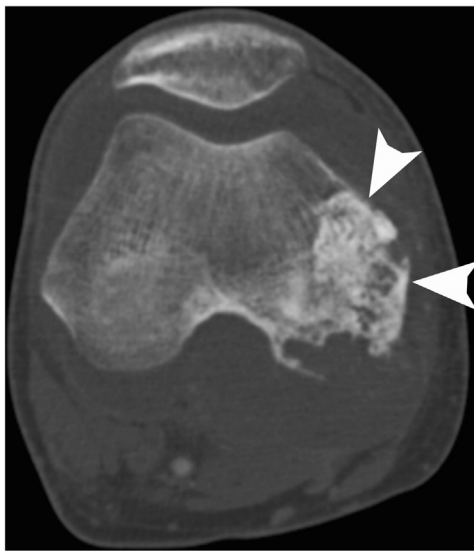


Fig. 2. Axial image from a contrast enhanced CT of the right knee showing the aggressive lesion in the posterolateral aspect of the right medial femoral condyle. Linear areas of increased density are noted (white arrowheads).

This patient was treated with vincristine, adriamycin, and cytoxan, alternating with ifosfamide and etoposide, following the Children’s Oncology Group Ewings sarcoma protocol AEWS-0031. The patient initially showed decrease in size of the knee mass, radiographic resolution of most of the pulmonary nodules, and resolution of the bone marrow disease by morphologic assessment of a bone marrow aspirate 2 months later. However, he developed a new right mid-thigh lesion the following month, proven to be malignant mixed tumor by fine needle aspiration. He then developed numerous firm pink keloid-like nodules on the right knee and leg, the largest being 2 cm in height. Subsequent follow up radiographic and PET imaging (Fig. 6) showed progression of disease. He received additional treatment with an aurora kinase inhibitor and subsequently with the combination of irinotecan, temozolomide, and temsirolimus, but he died of progressive disease 7 months from diagnosis.

3. Discussion

Although the nomenclature is evolving, the term “mixed tumor” refers to the presence of both epithelial and mesenchymal, often chondroid, elements [6]. These tumors most commonly occur in the salivary glands, where they are known as “pleomorphic adenomas” and exhibit benign characteristics. Less commonly, mixed tumors occur as primary skin neoplasms and are termed “chondroid syringomas”, and may exhibit more aggressive clinical behavior [3,6,7]. The origin of these tumors in somatic soft tissue and bone is controversial, but recent evidence suggests rearrangement of the *EWSR1* gene at 22q12 is common in this category [2]. The present case demonstrated typical histologic and immunophenotypic characteristics of mixed tumor, and a novel $t(X;19)(q13;p13.3)$ translocation. It remains to be seen whether $t(X;19)$ produces a fusion gene with analogous pathogenetic mechanisms to the more common *EWSR1* fusions.

Malignant forms of mixed tumor can occur at any site but primary MMT of bone is extremely rare [4,5,1]. Of the reported cases of primary MMT of bone, the average age at presentation ranged between 25 and 44 years [5]. The limited number of reports of this entity in older adult patients occurring in other sites suggests that this lesion may have a period of relatively slow, indolent growth followed by more rapid progression of local invasion and distal metastasis [7]. Treatment recommendations for this lesion are not specific to this tumor, but suggest it be treated as a high-grade sarcoma; there are no reports, however, of a successful treatment regimen.

From an imaging standpoint, this case proved diagnostically challenging. The wide zone of transition between the lesion and normal bone and sunburst pattern periosteal reaction suggested an aggressive lesion. Both radiographs and CT demonstrated confluent dense matrix. The MRI findings of corresponding T1 and T2 hypointensity, suggested osteoid matrix, as cartilaginous matrices are typically T2 hyperintense given increased fluid content. The lesion was initially considered most suspicious for a high-grade osteosarcoma given the aggressive radiographic appearance and metaphyseal location (common for osteosarcoma). However, the overall distribution of metastatic disease, particularly in the bone marrow, was not typical for osteosarcoma.

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