
Case 4

Ewing Sarcoma

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History

A 13-year-old female with right knee pain radiating down the lateral side of the right lower leg (Fig. 4.1).

Diagnosis

Ewing sarcoma of the bone

Findings

- Large mass originating from the right proximal femur that is hyperintense on proton density and isointense on T1 imaging.
- Mass replaces the bone marrow of the right proximal fibula while permeating and destroying the bone cortex seen as areas of higher signal intensity within the dark cortex (arrowheads).
- Extensive infiltration of the mass into the surrounding soft tissue (curved arrows).
- Intense hypermetabolic activity on the fusion images compatible with malignancy.
- Periosteal reaction appearing as a hypointense double line at the bone cortex (thin arrow) on T2 images.
- Edematous changes are seen associated with the mass as T2 hyperintensities without FDG activity.

Discussion

Ewing sarcoma is the second most common malignant primary bone tumor of childhood, with the most common being osteosarcoma. It accounts for 3% of all pediatric cancers. Ewing sarcoma typically occurs between the ages of 4 and 25 years with a peak prevalence between the ages of 10 and 15 years. It has a slight predilection for male patients and marked predominance for Caucasians. It tends to affect the femur followed by the ilium, tibia, humerus, fibula, ribs, and sacrum. The most common presenting symptom is pain localized to the site of the tumor. Ewing sarcoma is a small round blue cell tumor with a karyotype abnormality of a translocation involving chromosomes 11 and 22. It shares this translocation and microscopic features with other Ewing sarcoma family tumors such as primitive neuroectodermal tumor and Askin tumor. Treatment involves systemic chemotherapy with either surgery, radiation, or both. Ewing sarcoma can be aggressive with up to 30% of cases demonstrating metastatic disease at presentation, most commonly to the bone, liver, or lung.

On imaging, Ewing sarcoma typically presents with a moth-eaten destructive permeative pattern and lucent bone lesions on conventional radiographs. It is often associated with a large soft tissue component. MRI reveals marrow replacement, cortical destruction, and a circum-

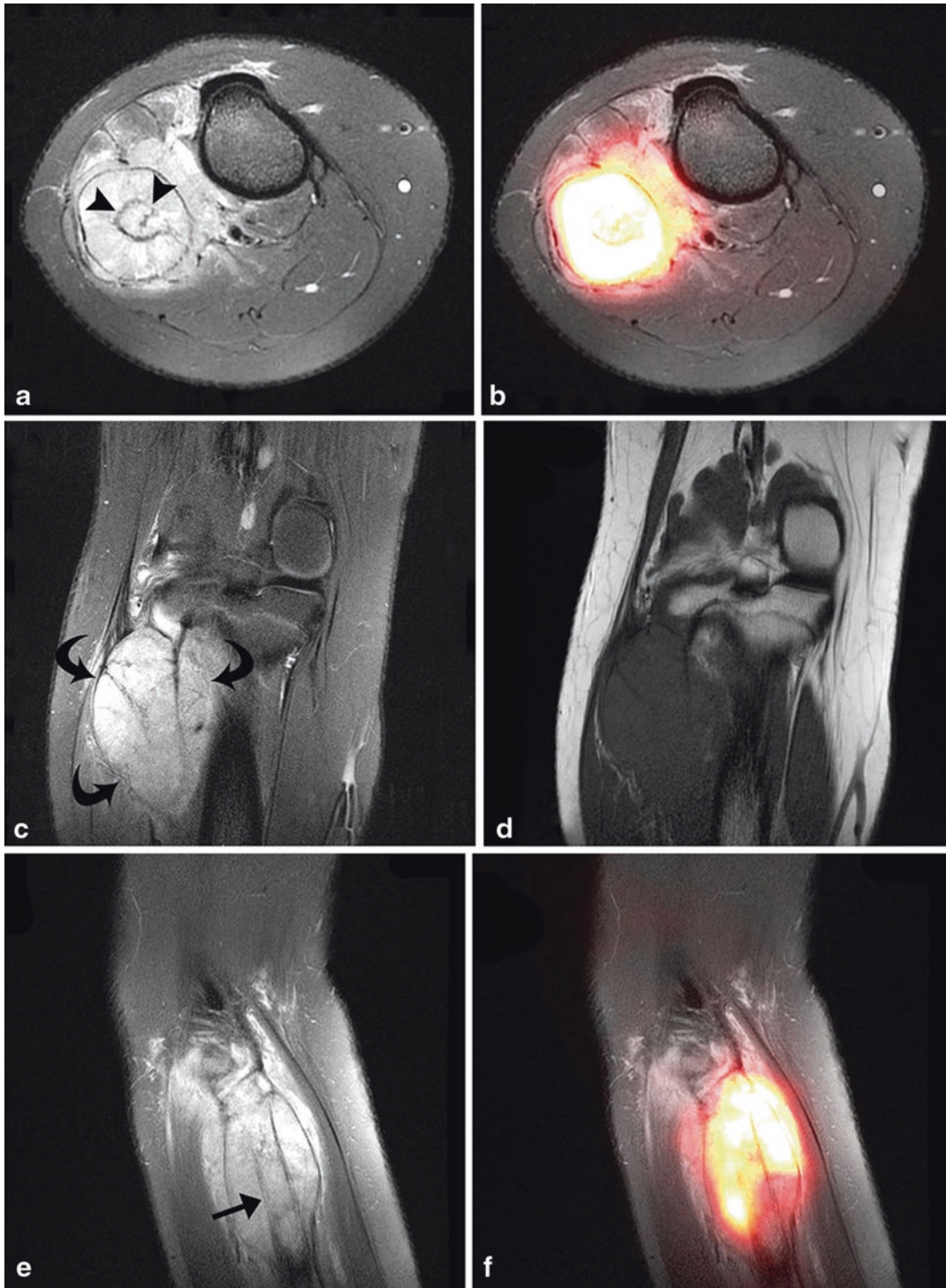


Fig. 4.1 Proton density with fat suppression axial (a), FDG PET/MR proton density with fat suppression axial fusion (b), Proton density with fat suppression coronal (c),

T1 TSE coronal (d), T2 with fat suppression sagittal (e), and PET/MR T2 with fat suppression sagittal fusion (f)

ferential soft tissue mass. It is usually homogeneous and low to intermediate signal intensity on T1-weighted images. On T2-weighted images, it is typically low to intermediate signal intensity due to its high degree of cellularity. High signal on T2-weighted and proton density (shared T1 and T2 weighting) images predominates in larger lesions and may represent hemorrhage or necrosis. In addition, linear low T2-weighted signal striations can be seen as the tumor extends through the Haversian canals and neurovascular channels of the cortex. The tumor enhances avidly post-contrast administration.

FDG PET provides unique information in regard to the metabolic activity of Ewing sarcomas. The functional information obtained by PET can estimate histologic tumor grade which is helpful to stage, restage, and assess treatment response in patients. FDG PET can help predict patient prognosis before and after neoadjuvant therapy. FDG PET has been shown to be superior to other modalities in detecting lymph node

or osseous metastases as well as identifying tumor recurrence. Because the biochemical properties of the tumor change earlier than its morphology, functional information obtained by PET imaging can determine response to treatment. FDG PET combined with MR is useful in surgical planning as it can help delineate tumor margins. PET/MR imaging allows for superior evaluation of Ewing sarcomas as it provides important physiological information and accurate anatomical localization.

Suggested Reading

- Bestic JM, Peterson JJ, Bancroft LW. Use of FDG PET in staging, restaging, and assessment of therapy response in Ewing sarcoma. *Radiographics*. 2009;29(5):1487–500.
- Murphey MD, Senchak LT, Mambalam PK, Logie CI, Klassen-Fischer MK, Kransdorf MJ. From the radiologic pathology archives: Ewing sarcoma family of tumors: radiologic-pathologic correlation. *Radiographics*. 2013;33(3):803–31.