A comprehensive review of osseous Ewing sarcoma: clinical data, skeletal location and imaging features.

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Learning objectives

Osseous Ewing sarcoma represents the second most common primary tumor of bone in children and adolescents. Although the clinical presentation is usually nonspecific and there is a wide skeletal distribution the imaging features of osseous Ewing sarcoma often suggest the diagnosis.

We propose to attend this objectives:

- To evaluate clinical data and primary lesion location of osseous Ewing sarcoma.

- To review the imaging features and pathological correlation of osseous Ewing sarcoma.
Background

Osseous Ewing sarcoma is the most common tumor of the Ewing family of tumors, which also includes extraskeletal Ewing sarcoma, primitive neuroectodermal tumor and Askin tumor [1]. These tumors share a karyotype abnormality with translocation involving chromosomes 11 and 22 [t(11;22)(q24;12)] and histologically they demonstrate crowded sheets of small round blue cells divided by a small amount of fibrous stroma [1,2].

Osseous Ewing sarcoma represents the second most common primary tumor of bone in children and adolescents, exceeded in prevalence only by osteosarcoma. Overall, it is the fourth most frequent primary malignant tumor of bone after multiple myeloma, osteosarcoma and chondrosarcoma [1-3]. Ewing sarcoma is more frequent in the first 3 decades of life, with 95% of cases reported between the ages of 4 and 25 years and has a slight male predilection (M:F 1,5:1) [1,4].

The clinical presentation is usually nonspecific, with local pain and a mass or swelling, and symptoms are often present for more than 6 months before diagnosis [4]. Fever and increased erythrocyte sedimentation rate simulating an infectious origin (osteomyelitis-like) are also common clinical features at presentation [1]. Pathologic fractures are uncommon as an initial clinical presentation and additional clinical manifestations are related to the site of involvement (e.g.: pleuritic chest pain in rib lesions; neurologic deficits and paraplegia in spine lesions) [1,4].

There is a wide skeletal distribution for Ewing sarcoma and both long and flat bones are affected because no bone is immune to tumor development [6]. However the patient’s tumor is most likely to arise in the pelvis or the long bones of the lower extremity (Fig.1) [6]. Long tubular bone involvement is more common proximally than distally, and the majority of long-bone lesions are actually metadiaphyseal [1]. Epiphyseal extension may occur, although lesions centered in the epiphysis are rare [1,4].

Imaging features of osseous Ewing sarcoma often suggest the diagnosis [1]. The evaluation of the primary lesion should start with a plain radiography if a bony mass is palpable. Computed Tomography (CT) scanning is helpful in delineating any bony involvement and looking for pulmonary metastasis. Magnetic resonance imaging (MRI) is accurate in the assessment of the intramedullary tumor and determining the exact extend of disease and the relation to adjacent blood vessels and nerves [2-4]. The definitive diagnosis is made by biopsy providing sufficient material for conventional histology, immunohistochemistry and molecular biology [4]. Imaging, particularly MR imaging, is also vital to evaluate response to neoadjuvant therapy, direct surgical resection, and detect local recurrence or metastatic disease [5].
Fig. 1: Distribution of Ewing Sarcoma. Most frequent locations are the large long bones and the pelvis.

Findings and procedure details

We retrospectively reviewed the cases with histologically and/or genetically proven diagnosis of osseous Ewing sarcoma treated (or referred) at our institution from January 2003 to December 2012. We analyzed all patient files for clinical data, i.e. age, gender, symptoms and location of primary lesion. Secondly, we evaluated the available radiographs, CT scans and MRI sequences for primary tumor characterization, and we did a review of the main imaging features.

Our population of 29 cases with proven diagnosis of osseous Ewing sarcoma demonstrated a male to female ratio 1,9:1 in agreement with the male predominance of the disease reported in the literature. The patient's age ranged from 1 to 52 years, again consistent with previous reports. In the majority of our cases the lesions occurred in the pelvis or lower extremity (68,9%). There was a fairly even distribution of lesions among the pelvis (34,5%) and the humerus (17%).

The clinical presentation was mainly nonspecific, with pain (69%), and a mass or swelling (41%). Other symptoms were related to the site of involvement and included claudication or decreased muscle strength in hipbone lesions, paresthesias in spine lesions and cough in ribs lesions. The time for diagnosis was on average 5 months (Table1).

Radiographic features in Ewing sarcoma of bone are usually very aggressive, reflecting the high-grade nature of this malignant lesion. Bone destruction with a moth-eaten to permeative pattern is common and almost all tumors present with poor margination (wide zone of transition) [1,4,6]. Cortical destruction with an associated soft-tissue mass is also common, although Ewing sarcoma may also transverse the Haversian system and cause a large soft-tissue mass outside the bone despite the absence of cortical destruction (Fig.2 and 3) [1]. Periosteal reaction is frequent and usually aggressive in appearance, either lamellated (onionskin) (Fig.4) or spiculated (sunburst or hair-on-end) (Fig.2) [1,4]. In some patients, Codman triangles may be present at the margins of the lesion (Fig.2 and 3). These result from the elevation of the periosteum and central destruction of the periosteal reaction by the tumor [6,7]. Sclerotic components are less common but may occur in approximately 25% of cases, usually in the intraosseous component of lesions and in flat-bone lesions [1].

Less common radiographic features of Ewing sarcoma of bone include cortical thickening, pathologic fracture and expansible bone remodeling (Fig.6). Periosteal Ewing sarcoma without involvement of the medullary canal is rare but can also cause extrinsic erosion of bone and have radiographic appearance similar to that of periosteal osteosarcoma [1].
Spinal lesions superior to the sacrum are uncommon and mostly affect the lumbar or thoracic elements. These lesions are usually centered in the posterior elements with extension into the vertebral body, while only one third is primarily within vertebrae (commonly with extension into the posterior elements). Spinal lesions typically reveal a large associated soft-tissue mass with spinal canal invasion (Fig.5) [9-11].

Ewing sarcoma arising from the bones of the head and neck region is extremely uncommon. When it occurs in the jaw, mandible is more frequently affected then the maxilla (Fig.6) [12].

The appearance of Ewing sarcoma at CT is similar to that at radiography, with aggressive bone destruction and a large associated soft-tissue mass. Large areas of focal cortical destruction and continuity between the intraosseous and soft-tissue components are common. The soft-tissue component is commonly homogeneous and similar in attenuation to that of muscle (Fig.2 and 4). Contrast enhancement is common at CT and is usually either diffuse or peripheral nodular [1,4].

MR imaging is the optimal radiologic modality in evaluation of bone and soft-tissue tumors, including Ewing sarcoma [1-2,4]. MRI provides accurate information about tumor size and is superior in delineating the extent of the neoplasms and their relation to the surrounding structures [4]. MR imaging of Ewing sarcoma of bone usually reveals marrow replacement and cortical destruction with an associated soft-tissue mass commonly circumferential but asymmetric about the osseous involvement (Fig.2 and 3) [1]. The signal intensity is usually homogeneous and intermediate on T1-weighted images, with heterogeneous gadolinium enhancement (Fig.2). On T2-weighted images, Ewing sarcoma is typically homogeneous and low to intermediate in signal intensity. This signal intensity and homogeneity are likely related to the high degree of cellularity in Ewing sarcoma [1,13-14]. Although large areas of cortical destruction are common at MR imaging, subtle cortical involvement with linear intermediate signal intensity channels that extend through the low signal intensity cortex, allowing continuity between the intraosseous and extraosseous components is also frequent, and may be the only evidence of cortical involvement and destruction in 40% of cases at MR imaging (Fig. 2 and 3). This pattern of intracortical involvement is often subtle at CT and appears as lower attenuation linear channels extending through the high-attenuation cortex (Fig. 2) [1].
Table 1: Clinical data of cases with histologically and/or genetically proven diagnosis of osseous Ewing sarcoma treated (or referred) at Portuguese Institute of Oncology from January 2003 to December 2012.

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Fig. 2: Ewing sarcoma of the proximal left humeral metadiaphysis in a 16-year old boy with pain and enlarging mass in the left proximal upper extremity. (a.) Frontal radiograph shows a permeative lytic lesion with poor margination, cortical destruction and an associated soft-tissue masse. An aggressive spiculated periosteal reaction and Codman triangle is present. (b.) Axial and (c.) coronal CT images reveal the intramedullary centered lesion with marrow replacement, the hair on end periosteal reaction. Low-attenuation channels allow continuity between the medullary and soft-tissue components. (d.) Coronal MR images TSE T1-weighted before and (e.) after intravenous contrast administration demonstrates the lesion centered in the marrow canal with asymmetrical circumferential soft-tissue extension. The lesion is heterogeneously intermediate in signal intensity on both T1-weighted and (f.) T2-weighted images with diffuse enhancement on the postcontrast image.

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Fig. 3: Ewing sarcoma of the fibula in a 9-year old girl with swelling in the left distal lower extremity. (a.) Frontal radiograph shows a permeative lytic lesion with hair-on-end periosteal reaction and Codman triangle. (b.) Axial T1-weighted and (c.) axial T2-weighted MR images demonstrate the lesion centered in the marrow canal with focal cortical destruction and an associated soft tissue masse. The lesion is heterogeneously intermediate in signal intensity on both the T1-weighted and T2-weighted images.

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Fig. 4: Ewing sarcoma of the pelvis in a 16-year old boy with left hip pain for 7-months. (a.) Frontal radiograph shows an ill-defined osteolytic lesion involving the left iliac bone with aggressive periosteal reaction. (b.) Axial and (c.) coronal CT images show the cortical destruction and onionskin periostitis of the left iliac crest.

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Fig. 5: Ewing sarcoma of the lumbar spine in a 22-year old man with 2-month history of a progressive left predominant back pain. (a.) Axial and (b.) coronal CT images reveal an osteolytic lesion with focal cortical destruction that involves the left hemivertebra, pedicle, hemilamina and transverse process of L3. (c) Bone scintigraphy (posterior view) show intense uptake at the third left hemivertebra. Axial T1-weighted MR image (c.) before and (d.) after endovenous contrast administration demonstrate an enhancing mass occupying the left hemivertebra, pedicle, hemilamina and transverse process. The mass extends to the vertebral foramen, deforming the dural sac.

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Fig. 6: Ewing sarcoma of the right mandible in a 1-year old girl with a progressive right facial mass. (a.) Sagital and (b.) coronal CT images reveal an aggressive osteolytic lesion with expansible bone remodeling, cortical destruction and periosteal reaction, involving the body and ascending ramus of the right mandible. (c.) Axial T1-weighted and (d.) post-contrast coronal fat-suppressed T1-weighted demonstrate a heterogeneously mass intermediate in signal intensity with diffuse enhancement on the postcontrast image.

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Conclusion

Osseous Ewing sarcoma is a highly malignant bone tumor that predominantly occurs in young patients with a nonspecific presentation. The most common sites of primary Ewing sarcoma are the pelvic bones and the long bones of extremities. Imaging features often suggest the diagnosis, most frequently with a moth-eaten to permeative bone destruction in the metadiaphysis or diaphysis of a long bone with an associated soft-tissue mass. Radiography and CT scan are good imaging modalities to describe periosteal reaction, lesion's matrix and cortical changes, but MRI is the imaging modality of choice for local staging of primary bone sarcoma.
References


