Unusual location of bone sarcoma in children

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Authors: S. JERBI, A. Khalfali, G. Abid, O. Bradai, N. chouchane, H. HAMZA; Mahdia/TN
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Learning objectives

Bone sarcomas comprise a group of several different cancerous tumors of the bone. The most common bone sarcomas diagnosed in children are osteosarcoma and Ewing's sarcoma.

Our purpose, through this paper review, is to expose the unusual locations of bone sarcoma in children and clarify the role of imaging in the positive diagnosis and staging.

Background

Unusual locations of bone sarcoma are exceptional and radiographic appearance has no semiologic particularity. So they must be suspected especially in children when radiological exam reveal an aggressive osteolytic lesion. MRI remains the most useful examination for better tumor characterization and the evaluation of locoregional extension. The final diagnosis of bone sarcoma requires histopathological verification.

Findings and procedure details

Sarcomas are the most common bone tumors in pediatric setting.

The sarcomas of long bones are the most common and are dominated in children by osteosarcoma and Ewing's sarcoma.

1/ Osteosarcoma:

Osteosarcoma is the most common malignant bone tumor in children. Its usual seat is the metaphysis of long bones.

Its frequency is estimated at about 2%.

All types of osteosarcoma can be met at the clavicle and scapula:

- Chondroblastic. born in the body of the scapula.
- Osteoblastic.
- Telangiectasic: usually born at the complex acromioclavicular glenoid.
Patients usually present with bone pain, occasionally accompanied by a soft-tissue mass or swelling.

**Location:**

They typically occur at the metadiaphysis of tubular bones in the appendicular skeleton.

-Common sites include:
  - femur: ~ 40% (especially distal femur)
  - tibia: ~ 16% (especially proximal tibia)
  - humerus: ~ 15%

-Other less common sites include: fibula, innominate bone, clavicle, mandible (gnathic osteosarcoma), scapula, maxilla, vertebrae

**Imaging findings:**

They are variable and frequently overlap with those of multiple benign and malignant entities, creating substantial diagnostic challenges.

Radiography is almost always the initial imaging modality. Once the diagnosis is suspected, magnetic resonance imaging (MRI) is essential to determine the distribution of the tumor within the bone and the extent of any associated soft tissue mass. Computed tomography (CT) scanning is less sensitive than MRI in local evaluation of the tumor, but it is used in the staging of metastases.

*Conventional radiography:

Radiographic appearances are variable.

- Most lesions show a mixture of lytic and sclerotic areas. Lesions appear aggressive; they appear either moth eaten (type II of Lodwick), with ill-defined edges, or, occasionally, they appear permeatives (type III of Lodwick), with multiple small cortical holes. After chemotherapy, surrounding bone may form a better-defined shell around the tumor, in which case it appears more geographic.

- Soft tissue extension of osteosarcoma is common; seen as a soft tissue mass.

-Periosteal reactions are commonly seen once the tumor extends through the cortex. A spectrum of changes occur; these include Codman triangles and multilaminated, spiculated, and sunburst reactions, all of which indicate an aggressive process.

*CT:
It may be helpful locally when the radiographic appearances are confusing, particularly in areas of complex anatomy. It may be particularly helpful in visualizing flat bones (such as scapula), in which periosteal changes may be more difficult to appreciate. Cross-sectional images provide a clearer indication of bone destruction, as well as the extent of any soft tissue mass, than do radiographs.

CT scanning may depict small amounts of mineralized osseous matrix not seen on radiographs.

*MRI:

It is the modality of choice in evaluating the local extent of disease because of its excellent bone marrow and soft tissue contrast and multiplanar capabilities.

MRI is the most important imaging technique for the accurate local staging of osteosarcoma

It assesses soft tissue involvement which appears in:

- Low signal intensity on T1WI with heterogeneous gadolinium enhancement
- High signal intensity on T2WI

The intraosseous and extraosseous extent of the tumor must be assessed.

Important features of intraosseous disease are the longitudinal distance of bone containing tumor, the involvement of adjacent epiphyses, and the presence or absence of skip metastases.

The most accurate sequence for determining the longitudinal extent of disease is the T1-weighted spin-echo sequence. The maximum longitudinal extent of the tumor should be measured, and its maximal distance from the articular surface of the nearest joint should be recorded. The longitudinal extent is usually maximal within medullary bone, but occasionally, intracortical extension is more extensive.

The use of short-tau inversion recovery (STIR) may significantly lead to an overestimation of disease, because edema and marrow hyperplasia may show high signal intensity similar to that of tumor.

The presence of hemorrhagic areas and levels Fluid evokes telangiectasic osteosarcoma.

**Prognosis:**

The most significant of all factors used to determine prognosis for the child with osteosarcoma is the extent of the disease at diagnosis and whether it has metastasized
or not. For children or teens with localized disease, the following factors are considered: resectability of the tumor, determined by location and tumor size, and response of the tumor to chemotherapy.

**2/ Ewing's sarcoma:**

Ewing's sarcoma represents 6% of primary malignant bone tumors and is the third in frequency among sarcomas, after osteosarcoma and chondrosarcoma. The male is the most affected.

It occurs mainly between 5 and 25 years with a frequency peak between 10 and 20 years. Approximately 60% of cases are located in long bones, approximately 2/3 on the metaphysis and third on the diaphysis.

The presence of growth cartilage protects generally the epiphysis.

The most affected are the long bones which are, in descending order: the femur, tibia, humerus, fibula and bones of the forearm.

Most common symptoms are progressive localized pain and swelling

Additional symptoms may include

- Fever
- Weight loss
- Anemia
- Leukocytosis
- Elevated erythrocyte sedimentation rate and increased serum lactic dehydrogenase (LDH).

**Differential diagnosis**

On the radiological images differential diagnosis are acute osteomyelitis, eosinophilic granuloma, neuroblastoma metastases and other bone sarcomas. These conditions may have clinical and radiological pictures extremely similar. Histological examination revealed a monomorphic proliferation of small round cells compacted, whose cytoplasm contains many grains of PAS positive glycogen, which is important to rule out a malignant lymphoma or neuroblastoma metastases.

**Imaging findings:**

Radiological symptoms of Ewing tumor are similar as osteosarcoma. They depend on the location, the importance of periosteal reaction and the presence of a bone condensation.
Most lesions are visible on conventional radiographs. However, their degree of spread is better evaluated with MRI.

**Conventional radiographs:**

Common manifestations on conventional radiography, including:

- Poorly marginated, lytic, destructive lesion: Permeative (type III of Lodwick) or moth-eaten (type II of Lodwick) appearance.
- Rarely, they can be sclerotic
- Cortical rupture is common with soft tissue invasion.
- Periosteal reaction is common: Lamellated due to successive layers of periosteal development or spiculated appearance when new bone is laid down perpendicular to cortex along Sharpey's fibers.
- Codman's triangle - formed between elevated periosteum with central destruction of cortex
- Osteosclerosis may be present secondary to reactive bone formation

Other, less common, manifestations

- Thickened cortex
- Expansion of bone
- Pathologic fractures

On the flat bones such as iliac bone and the scapula, in addition to the lamellar periosteal bone, it can occur a reactive condensation that masks osteolysis.

On the flat bones of the trunk and extremities bones, tumor may manifest as an ivory bone and can simulate bone condensing osteosarcoma or condensing metastasis; However the tumor does not produce calcified tissue and the condensing appearance is a normal trabecular thickening in response to tumor infiltration

**CT:**

To evaluate bone destruction and extra-osseous involvement

**MRI:**

It is the method of choice for tumor staging. It assesses soft tissue involvement:

- Low signal intensity on T1WI with heterogeneous gadolinium enhancement
- High signal intensity on T2WI

**Prognosis:**

- 60-75% five-year survival

- Predictors of poorer prognosis:
  - Large tumors
  - Non-resectable lesions, such as those in the pelvis
  - Older age
  - Elevated leukocyte count and sedimentation rate at presentation

**Images for this section:**

**Fig. 1:** Right shoulder Radiography: Moth eaten and permeative osteolytic lesion of the right clavicle
Fig. 3
**Fig. 5:** Fig.2, 3, 4, 5: CT scan. Osteolytic lesions of the 2/3 internal right clavicle with rupture of cortical and multi lamellar periosteal reaction
Fig. 7: MRI: heterogeneous tumoral process of the right clavicle low signal intensity on T1WI with heterogeneous gadolinium enhancement and high signal intensity on T2WI.
Fig. 8: Fig.8: conventional radiography: Opacity occupying the entire left lung field with deviation of mediastinum to the right associated to an osteolytic process of the fourth posterior arch of left side.
Fig. 11: Fig.9, 10, 11: CT scan. Axial slices in bone and soft tissue windows showing important osteolysis of the right acromion with soft tissue invasion and heterogeneous enhancement after injection of contrast product.
Fig. 12: Conventional radiography: Opacity occupying the entire left lung field with deviation of mediastinum to the right associated to an osteolytic process of the fourth posterior arch of left side
Fig. 14: Tumoral process occupying the left lung field with heterogeneous enhancement after injection of contrast product facing to moth eaten osteolytic lesion of the fourth posterior left arch with multi lamellar periosteal reaction. Note an ipsilateral pleural effusion.
Conclusion

Unusual locations of bone sarcoma are exceptional and radiographic appearance has no semiologic particularity. So they must be suspected especially in children when radiological exam reveal an aggressive osteolytic lesion. MRI remains the most useful examination for better tumor characterization and the evaluation of locoregional extension. The final diagnosis of bone sarcoma requires histopathological verification.

Personal information

References

