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Bone Tumors: the Last in a Series on Bone and Joint Pain in Children

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There are a great number of tumors and tumor-like lesions of bone. The accurate diagnosis of a bone lesion is generally not possible on the basis of its radiographic characteristics. The patient history, physical examination, laboratory studies, and often histologic examination of the lesion are required. The radiographic appearance of a lesion can assist in indicating the aggressiveness of the lesion, coupled with the location and age of the patient, to allow a reasonable differential diagnosis of the lesion (see Table I).

Table I: Bone Tumors and Tumor-Like Lesions: Typical Ages of Patients

AGE (Years)

TUMOR

MALIGNANT

Osteosarcoma

Parosteal Osteosarcoma

Chondrosarcoma

Fibrosarcoma

Fibrous Mistiocytoma

Malignant Giant Cell Tumor

Ewing's Sarcoma

Ameloblastoma

Hemangioendothelioma

Histiocytic Lymphoma

Chordoma

Plasma Cell Myeloma

Skeletal Metastasis

BENIGN

Osteoma

Osteochondroma

Enchondroma

Chondroblastoma

Chondromyxoid Fibroma

Osteoid Osteoma

Osteoblastoma

Nonossifying Fibroma

Desmoplastic Fibroma

Lipoma

Hemangioma

Giant Cell Tumor

Neurilemoma

Simple Bone Cyst

Aneurysmal Bone Cyst

Reference: Resnick and Niwayama. Diagnosis of Bone and Joint Disorders, 2nd edition, Saunders, 1988.]

Just as a brief review, the features that indicate an aggressive lesion are:

- moth eaten or permeative bone destruction;
- cortical erosion, penetration, and expansion;
- periosteal reaction;
- invasion in the adjacent soft tissues.

We all know that if any of these features are present, aggressive follow-up is necessary.

The cause of bone tumors is unknown. They may arise in areas of rapid growth; many originate at the metaphyseal-epiphyseal region and in the medullary bone. Other theories indicate heredity, trauma and radiation as possible factors in the development of bone tumors. As with most neoplasms, there is never just one etiology. Fortunately, most bone tumors are benign or slow growing.

Osteochondromas are the most common benign bone tumor and occur most often in people between the ages of 10 and 20. I think we should be familiar with the common radiographic features of this lesion. It is an osteocartilaginous exostosis which arises from the external surface of a long bone and contains spongiosa and cortex that are continuous with those of the parent bone (see Figure 1). Osteochondromas commonly occur at the metaphysis. These lesions can be monitored radiographically and generally are best left alone. They can, however, affect vascular and tendinous junctions (or even joint range-of-motion) if large enough. A very small percentage will degenerate to a more aggressive lesion.

Figure 1: The most common type of benign bone tumors are the osteochondromas, which commonly occur at the metaphysis.

Two other benign lesions that are frequently found in children are the nonossifying fibroma and the fibrous cortical defect. They occur in about 30-40 percent of normal children; therefore it is important to review their general radiographic appearances also.

The nonossifying fibroma is usually found in children ages 8-20, with there being a male predominance of 2:1. Most lesions are asymptomatic and are found by coincidence when the patient is being x-rayed for some other reason. The radiographic appearance of the nonossifying fibroma is generally so characteristic that biopsy is rarely necessary. The typical appearance is that of a solitary, radiolucent, eccentric and generally ovoid lesion which often thins and may expand the cortex slightly (see Figure 2). The most frequent location is the lower extremity, most commonly the tibia. Since these lesions are asymptomatic, no treatment is necessary. They will often spontaneously regress over a 2-5 year period.

Figure 2: Nonossifying fibromas are most frequently found in the lower extremities, usually on the patient's tibia.

The fibrous cortical defect is most common in the 4-8 age range; again, it is more frequently seen in males, with a ratio of 2:1. The fibrous cortical defect and the nonossifying fibroma are histologically identical. The radiographic features of the fibrous cortical defect are that of an eccentric, lytic geographic lesion which is placed within the metaphysis of long tubular bones. Most lesions appear round and extend parallel to the long axis of the long bone (see Figure 3). The peripheral rim of the lesion is generally sclerotic and there may be more than one lesion. Since most lesions regress totally within two years, no treatment is necessary.

Figure 3: The fibrous cortical defect usually appears within the metaphysis of long tubular bones.

Malignant bone tumors occur as a primary bone tumor, or as a metastasis from another area of the body. Primary bone tumors are rare (less than 1% of all tumors are malignant) and are more common in young men. The incidence of bone tumors in children is approximately five cases per million children each year. The most common malignant bone tumor in children is the osteosarcoma, with Ewing's sarcoma second.

Let's review the radiographic appearances of the osteosarcoma and the Ewing's sarcoma, since there are by far the most common in children. Hopefully, we will never have the opportunity to consider either diagnosis since primary bone tumors are rare.

The radiographic features of the osteosarcoma are relatively well established. The classic presentation is that of a focal lesion in the metaphysis, creating either a mottled "permeative" lesion with a poorly defined zone of transition or a dense "sclerotic" region filling the medullary space (see Figure 4). Amputation offers the best possibility of control when the lesion is accessible and metastasis has not occurred. Prognosis is poor because of the tendency for early metastasis. Often pulmonary metastasis is already present when the primary lesion is discovered.

Figure 4: The most common malignant bone tumor in children is the osteosarcoma.

Ewing's sarcoma is found most commonly in the 10-25 age range. There is a 2:1 male to female ratio. The classic presentation is that of a diaphyseal lesion, permeative in appearance, with a wide zone of transition. A delicate "laminated" or "onion skin" periosteal response is seen in half of the cases (see Figure 5). Early detection is the key to survival. Radiation therapy and conjunctive chemotherapy is the present accepted mode of treatment. Amputation still may be used in cases below the knee.

Figure 5: Half of all Ewing's sarcoma cases present with a "laminated" or "onion skin" periosteal response.

Common symptoms for aggressive bone tumor are bone pain, which is many times worse at night. Occasionally a mass can be felt at the tumor site if the disease is advanced. Generalized leg pain is also a common complaint. Systemic symptoms are generally absent, with the exception of Ewing's sarcoma. In the presence of a Ewing's sarcoma the patient may experience slight to moderate fever; secondary anemia; leukocytosis; and increased erythrocyte sedimentation rate. Benign tumors are generally asymptomatic, with the exception of a palpable mass which is non-tender.

Prognosis varies depending on the type of tumor. The survival rate with primary bone tumor is improving with new treatments and early detection.

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