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The Core Curriculum

Osteosarcoma

Objectives

1. Describe the presenting symptoms of patients with osteosarcoma
2. Describe the radiographic features of osteosarcoma
3. Discuss the early management of the patient suspected to have osteosarcoma
4. Discuss the prognosis for osteosarcoma

Discussion

Osteosarcomas by definition produce osteoid. However, although that is their common feature, their behavior and anatomic features can vary widely. Most osteosarcomas seen in the pediatric are primary, which arise as solitary neoplasms. There are an increasing number of secondary osteosarcomas, following a number of pre-existing conditions such as radiation therapy, multiple hereditary exostoses, chronic osteomyelitis, etc. Most, but not all, secondary osteosarcomas affect the adult population. Children with retinoblastoma have a high incidence of secondary osteosarcoma. The incidence of osteosarcoma is about 3 times that of Ewing's sarcoma; it is estimated that 500-1000 new cases occur in the US yearly. The second decade is the most frequent time of diagnosis, with a slight preponderance of males affected. The age of presentation of parosteal osteosarcoma, which has a better prognosis, is older and more often affects females. The bones about the knee are most often affected, involvement of the axial skeleton is much less common than with Ewing's sarcoma.

Pain and swelling are common presenting features. In a recent study from Sweden, only about 20% had night pain. A tumor mass was present about 40% of the time at the first visit. Nonetheless, the diagnosis was delayed about 9 weeks from the time of the first presentation to a physician, strains, tendinitis, or athletic injuries often were suspected. Laboratory values are not particularly helpful. Radiographically, 90% involve the metaphysis. Usually, there is medullary destruction with poorly defined margins, cartical destruction, and reactive periosteal bone. The amount of mineralized osteoid evident is extremely variable. Findings associated with parosteal sarcomas are less dramatic. Osteosarcoma is an aggressive tumor which metastasizes quickly, usually to the lung. Skip lesions in the affected bone can be present up to 25% of the time, complicating treatment. Prognosis is of course better for patients whose lesions have not yet metastasized at the time treatment is begun. Axial or pelvic lesions also carry a poorer prognosis. Tremendous advances in chemotherapy and limb salvage surgery over the past 30 years have pushed survival rates well over 50%, a remarkable increase from the 10-15% survival rate in 1970.

The best results follow early referral of a patient suspected of having osteosarcoma to a center capable of managing the entire course of the disease. Placement of biopsy incisions can be critical to the success of subsequent limb sparing surgery.

The genetic link to retinoblastoma has pointed to the role of tumor-suppressor genes, and that osteosarcoma follows inactivation of these genes. Much work remains to be done before we have a mature understanding of this process.

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