



POSNA

The Core Curriculum

Ewing's sarcoma

Objectives

1. Describe the presenting symptoms of Ewing's sarcoma
2. Discuss the prognosis for patients with Ewing's sarcoma
3. Describe radiographic findings secondary to Ewing's sarcoma
4. Discuss initial management of patients with Ewing's sarcoma

Discussion

Ewing's sarcoma is the second most common malignancy of bone in children and adolescents. There is a definite predilection toward white children; it is very rare in children of Asian or African descent. Most cases present between ages 5-25. Ewing's sarcoma can occur in virtually any bone, flat or long. Almost one half of cases involve the femur, tibia, or fibula. Tumors in the hands or feet have a better prognosis. Pain and swelling are the presenting symptoms. Night pain was not a predominant symptomatic feature in a recent series from Sweden. A history of trauma associated with the pain was frequent, and misdiagnosis was common. The average delay from initial presentation to the physician to diagnosis was 19 weeks. Differential diagnoses include osteomyelitis, tendinitis, and strain. Laboratory findings are nonspecific, an elevated ESR and white count are usual. Radiographically, endosteal based cortical erosion is a major finding, but the findings can be variable. When the lesion has penetrated the cortex, there can be surface erosion of the cortex as well. A soft tissue mass contiguous to the affected part of the bone is almost always present.

If a child is suspected of having Ewing's sarcoma, biopsy should be deferred to a center having the capability of definitively managing the problem. Although Ewing's tumor was largely managed nonoperatively, recent advances in treatment have made surgical management feasible in more cases. 5-year survival rates have increased from 5% to 50% over the last 30 years. Prognosis is dependent on whether metastases are present at the time treatment is begun; survival rate in these patients is about 20%. Chemotherapy is the mainstay of treatment, with surgery and radiation therapy used for local control. Radiation therapy has been associated with a number of significant complications in the past secondary to high doses, as has surgery. At the present time, the relative roles of surgery and radiation therapy for local control is being studied. Surgery on the pelvis has recently been demonstrated to improve outcome; which is important as involvement of the axial skeleton portends a worse prognosis than the appendicular.

A chromosomal translocation (11:22) {q24;q12} is consistently found associated with Ewing's sarcoma, as well as other neuroectodermal tumors, including extraskeletal Ewing's, peripheral neuroepithelioma, and Askin's tumor. All these tumors are sometimes referred to as the Ewing's family of tumors (EFT). A promising recent advance in therapy for these tumors is CD99, which appears to have an effect on the unregulated growth of tumor cells, and can be used in conjunction with conventional chemotherapy.

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