Ewing's Sarcoma

The Ewing's sarcoma is the second most common primary malignant bone tumor seen in children and is the fourth most common malignant tumor overall. This tumor is a very primitive mesenchymal sarcoma that has a mysterious etiology. With the advent of the electron microscope and immunohistochemical studies, most experts today feel that this tumor probably represents a poorly differentiated member of a larger family of neural tumors, distinct from the neuroblastoma. In 90 per cent of cases, cytogeneticists will find a reciprocal translocation in chromosomes 11 and 22. That is also found in patients with the diagnosis of primitive neuroectodermal tumor [PNET] and Askin's tumor. Other round cell tumors that have a similar histological appearance include the embryonal rhadomyosarcoma, the mesenchymal chondrosarcoma, and the metastatic neuroblastoma. It is very important to separate out the large group of histiocytic lymphomas seen in an older age group that have a similar appearance with H&E staining. With special staining techniques, most lymphomas will be positive for common leukocyte antigen. Likewise with metastatic embryonal rhadomyosarcoma, specific immunohistochemical studies will reveal muscle markers such as actin, desmin, and myoglobin not found in the Ewing's sarcoma.

Ninety per cent of patients with Ewing's sarcoma are from 5 to 25 years of age, with males being affected slightly more than females. The two most common locations for the Ewing's sarcoma are the femur and pelvis, followed by the tibia, humerus and scapula, but it can be found in any location in the body that includes myelogenous tissue, including the spine, rib, foot and hand. Radiographically, the Ewing's tumor is found typically in a diaphyseal-metaphyseal location in the medullary canal with very diffuse, permeative lytic destruction of the surrounding cortical structures that looks like hematogenous osteomyelitis. Likewise, the clinical appearance of Ewing's sarcoma can mimic infectious disease with an elevated temperature, white count and sedimentation rate. Because of the high incidence of necrosis with Ewing's sarcoma, it is common to find liquefied necrotic debris in the tumor site that gives the clinical appearance of osteomyelitis. With early breakthrough into the subperiosteal tissues, the radiographic finding of a reactive periostitis is quite common, creating a multilaminated "onionskin" appearance on a routine X-ray. Another characteristic radiographic finding is the "hair on end" appearance that is created by reactive new bone formation along the perpendicular periosteal blood vessels running between the periosteum and the subadjacent cortex.

Prior to 1970, the prognosis for survival in Ewing's sarcoma was extremely poor with only about 10 per cent of patients surviving their disease. Currently, with the use of adjuvant systemic chemotherapy in non-metastatic Ewing's sarcoma, the survival rate now runs approximately 70 per cent. However, in about 20 per cent of patients with Ewing's sarcoma who develop metastatic disease to other bones or to the lung, the survival rate drops to about 30 per cent. Whenever possible, the orthopaedic oncologist will attempt a wide local resection of the primary tumor site, a technique similar to that used for osteosarcoma. If wide surgical margins are obtained, the chance for survival is probably better than if radiation therapy and chemotherapy had been used without surgery. However, in cases where the surgical margins are positive at the time of the surgical resection, postoperative radiation therapy is indicated. The chance for local recurrence with chemotherapy and radiation therapy alone is about 20 per cent or higher. A devastating complication of radiation therapy is pathological fracture that usually results in intramedullary fixation that may fail and ultimately result in amputation. Secondary sarcomas occur with the use of radiation therapy for Ewing's sarcoma in about 10-15 per cent of cases. The primitive neuroectodermal tumor accounts for approximately 10 per cent of all Ewing's-like tumors and carries about the same prognosis for survival. The clinical management for this entity is essentially the same as for Ewing's sarcoma.