

Classic Osteogenic Sarcoma

Osteosarcoma is the most common primary malignant tumor of bone, making up 20 per cent of all primary malignancies, with approximately 500-1000 new cases diagnosed each year in the United States. The classic or most common form of osteosarcoma is seen typically in children and young adults, with a definite male preference. It occurs in the metaphyseal areas of fast growing bones with the most common location being the distal femur, secondly the proximal tibia, and thirdly the proximal humerus. Fifty per cent of the lesions will be found around the knee joint. This tumor is rare in small bones such as the hand or the foot, or in vertebral segments. Patients usually present with spontaneous symptoms of pain in the area, followed several months later with a tumor mass that is usually diagnosed by biopsy within six months after the onset of symptoms. The radiographic appearance is typically a permeative lytic lesion seen in the metaphyseal area of a long bone with cortical breakthrough and periosteal elevation creating a Codman's reactive triangle, followed later by a sunburst pattern of chaotic bone formation in the soft tissue outside the periosteal sleeve. In a small percentage of cases, a so-called skip lesion will appear as a separate nodule of tumor activity totally separate from the primary which, when found, suggests a very poor prognosis for survival. Fifty per cent of osteosarcomas are of the osteoblastic type, but in a smaller percentage of cases, there will be a prominence of cartilage or fibrous tissue that does not seem to influence the prognosis for survival.

The staging process for this disease includes a magnetic resonance imaging (MRI) study of the primary tumor to identify soft tissue invasion by the tumor and define the medullary extent of the tumor which helps the operating surgeon determine the level of amputation or limb salvage resection. A bone isotope scan is performed to rule out the possibility of other bony foci in the skeletal system and a CT scan of the chest is obtained to rule out the possibility of metastatic disease to the lung. The final staging process includes a biopsy of the primary site performed in such a way as to not contaminate vital structures that might interfere with the potential for a limb salvage resection later.

Prior to 1970, the prognosis for survival with this disease was only 20 per cent even though early amputation was performed at a high level. Pulmonary metastasis was the reason for a fatal outcome in these early cases, however, with the advent of multi-drug chemotherapy the prognosis for survival has increased to approximately 60 per cent. The drugs most commonly used for systemic control of the disease include high dose methotrexate, Adriamycin, cisplatin, and ifosfamide. These drugs are administered through a central venous line on a cyclic basis every three to four weeks for

approximately two months prior to surgical removal of the tumor. Chemotherapy is continued for approximately four months after surgical treatment.

At the present time, 90 per cent of patients with osteosarcoma are treated by limb salvage resection. The most common type of reconstruction consists of a total joint replacement such as a rotating hinge at the knee. A smaller group of patients are treated with allograft reconstruction or combinations of the above. Excisional arthrodesis was a popular technique many years ago but now patients prefer a reconstruction that involves normal joint motion. The prognosis for survival is influenced by the degree of tumor necrosis produced by the preoperative chemotherapy protocol, so that if at the time of surgical resection if there is more than 90 per cent necrosis of the tumor, the patient has a much better prognosis for survival (approximately 85 per cent at five years). Pulmonary metastasis is still the major concern following treatment for osteosarcoma and, if this does occur, aggressive surgical resection of the lesions through the chest wall is frequently performed. There is a 30 per cent survival rate at five years following this protocol. As with other forms of cancer, recent molecular genetic studies have revealed a high incidence of abnormality in the P53 suppressor genes found in this tumor.