A 23-year-old white woman was admitted to the hospital with a four-month history of left shoulder and left pleuritic chest pain. She had received ibuprofen and chiropractic manipulation without relief. She had no history of trauma, cough, dyspnea, hemoptysis, fever, arthralgias or weight loss. She had no significant past medical history. Her physical examination was totally unremarkable except for obvious left-sided splinting with deep inspiration. There was no chest wall mass or tenderness and no abnormality was detected on auscultation of her lungs. Routine laboratory investigations were normal except for an elevated alkaline phosphatase of 115 μ/ml (normal 20-100 μ/ml). The chest roentgenogram and tomograms showed a pleural-based mass on the left associated with an expansive bone lesion in the third rib (Fig 1 and 2). Bone scan revealed increased uptake only at the area of the lesion in the left third rib. A transthoracic needle aspiration was performed which showed a few red blood cells. Because of the persistent chest pain, thoracotomy was performed.

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Figure 1

Figure 2
Diagnosis: Ewing’s sarcoma

During thoracotomy, a 5-cm tumor mass extending from the third rib into the chest wall and adherent to the lung surface was removed with an en bloc resection of the second, third, and fourth ribs and a left upper lobectomy. Histologic examination of the mass showed it to be Ewing’s sarcoma. A bone marrow biopsy showed evidence of metastatic disease. She was treated with local radiation therapy and chemotherapy consisting of cyclophosphamide, adriamycin and vincristine. Eight months after initial diagnosis, the patient became severely neutropenic and thrombocytopenic after a course of chemotherapy and died of Gram-negative sepsis. At autopsy she was found to have Ewing’s sarcoma metastatic to several ribs and vertebral bodies.

The major roentgenographic differential diagnosis of an extrapleural soft tissue mass associated with rib destruction includes bone sarcomas (Ewing’s, osteogenic sarcoma, chondrosarcoma, fibrosarcoma), rib fracture with hematoma or callus, metastases, myeloma, and osteomyelitis of ribs with soft tissue abscess (including tuberculosis, actinomycosis and fungus).1

Ewing’s sarcoma is second in frequency to osteogenic sarcoma among primary bone tumors in children and young adults. It is most common in the second decade and is rare under five or over 30 years of age. There is a 1.5-2:1 male predominance, and the tumor is exceptionally rare in blacks. Histologically, Ewing’s sarcoma is a highly malignant tumor characterized by small, round, anaplastic cells which must be differentiated from non-Hodgkin’s lymphoma, metastatic neuroblastoma, embryonal rhabdomyosarcoma, and metastatic small cell lung carcinoma.2

The clinical presentation is usually with pain and swelling, often associated with fever and weight loss. Over half the patients have a palpable mass at initial presentation. The femur is the most common single bone site of involvement (17 percent) and axial skeleton lesions are found in 50 percent of patients. Ribs are the primary site in only 10 percent of the cases. The tumor affects the diaphysis of long bones, with a prominent soft tissue component. Metastatic disease is found in 50 percent of patients at presentation, with bone, bone marrow, lung and central nervous system representing the most common sites.

Poor prognostic signs include the presence of axial lesions, elevated serum LDH, fever, leukocytosis and multifocal bone involvement or metastatic disease. Local treatment alone, with surgical excision or moderate dose irradiation, gives poor results with less than 10 percent five-year survival. The aggressive nature of the tumor and high frequency of metastatic disease at presentation make systemic therapy essential. The most active chemotherapeutic agents against Ewing’s sarcoma include adriamycin, actinomycin-D, cyclophosphamide and vincristine, which are usually given in combination as CAV or CAVA. In addition, local control is achieved with megavoltage radiation therapy (~6,000 rads) to the primary lesion. Primary surgical therapy is indicated in only a few circumstances, including lesions of ribs, clavicles and feet, often followed by postoperative irradiation.

Aggressive multimodality therapy, continued over 18-24 months, has resulted in 50-75 percent five-year survival in patients with non-metastatic disease.3,4 Distant relapse and uncontrollable metastases are the most frequent causes of treatment failure and the incidence of osteogenic sarcoma is significantly increased in long-term survivors.

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