Chest Wall Sarcomas and Induction Therapy

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KEYWORDS

• Chest wall • Sarcoma • Induction therapy • Tumor

Chest wall sarcomas are uncommon in clinical practice. However, only a small number of surgeons have a working knowledge of the cause, evaluation, treatment, and prognosis of patients with primary or secondary chest wall sarcomas. Often, this unfamiliarity leads to inappropriate diagnostic studies, delays in treatment, and frustration for the patient and surgeon alike. In most instances, early referral to a specialized center with experience in these unusual chest wall tumors should be considered. In addition, although there are extensive formal guidelines for treatment, including the use of multimodality treatment of extremity and retroperitoneal sarcomas, because of the rarity of chest wall sarcomas, no such formal guidance is available.

This article reviews the clinical presentation, diagnostic procedures required for evaluation, and the overall treatment strategy for patients presenting with chest wall sarcomas. The limited situations in which induction therapy before resection is appropriate are discussed. This article provides an approach to streamlining the evaluation and treatment of patients with chest wall sarcoma.

PATHOLOGY

Sarcomas are malignancies arising in connective and supportive tissues. The chest wall contains several distinct tissues at risk for sarcoma development, including fat, muscle, bone, cartilage, and blood vessels. The chest wall is also in close proximity to several organs that may seem to be a palpable chest wall sarcoma by direct extension. These organs include extension from the breast, the lung, and the mediastinum. In addition, because of the large surface area of the chest wall, it can be the site of metastasis from a distant sarcoma; however, pulmonary parenchyma metastasis remains the most common site. The primary framework for classification of chest wall sarcoma is shown in **Box 1**.

CLINICAL EVALUATION

The initial evaluation of these patients begins with a careful history, noting the symptoms associated with the mass and the history of its growth. Prior radiographs, if available, are reviewed to determine the rapidity of the growth. On physical examination, it is important to rule out other sites of distant disease as well as to delineate the proximity to vital structures and to determine the patient's fitness to undergo aggressive therapy. The most common study on presentation is a chest radiograph and a computed tomography (CT) scan of the chest. For those lesions in proximity to vital structures such as the brachial plexus, magnetic resonance imaging is useful to delineate tissue planes and surgical relationships.[1] No radiographic test can delineate a benign and malignant chest wall mass, and a definite diagnosis of chest wall sarcoma cannot be made on any imaging study.

An important management decision is whether or not a preoperative tissue diagnosis is required. This decision is particularly important in those, albeit limited, situations in which induction treatment should be considered.

For small chest wall lesions less than 3 cm, an excisional biopsy with wide margins is often performed for diagnosis and will likely be acceptable for treatment purposes. When a lesion is 3 cm or larger, resection carries a higher morbidity. Additional considerations include the potential need for extensive reconstruction and resultant functionality. In some cases, there may be a role for induction therapy. In this situation, it is advisable to obtain a preoperative tissue diagnosis. Possible

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Box 1
Framework for classification of primary and secondary chest wall sarcomas

Primary sarcoma of the chest wall

Soft tissue sarcoma

Bone and cartilage sarcoma

Secondary masses of the chest wall

Tumor invasion from contiguous breast, lung, or mediastinum

Metastasis from distant sarcoma sites to the chest wall

approaches include percutaneous fine-needle aspiration, core needle biopsy, or incision biopsy.

The usefulness of fine-needle aspiration for tissue diagnosis of any chest wall mass, including chest wall sarcomas, remains debated. On the positive side, a fine-needle aspiration is an outpatient procedure that can be performed on the patient's initial visit with little or no morbidity. Although this approach has been reported as an effective technique in these situations in our routine clinical practice, we often find the cytology of fine-needle aspirate is reported as nondiagnostic.[2,3] An alternative is a core needle biopsy, which provides tissue for histologic evaluation, as does the traditional incisional biopsy, often yielding the definitive diagnosis. In keeping with standard principles of sarcoma surgery, it is important to perform core and incisional procedures in a manor that allows excision of the biopsy track at the time of definitive resection. A tissue diagnosis is required in all cases considering induction therapy.

PRIMARY SOFT TISSUE SARCOMAS

There are diverse types of primary sarcoma of the chest wall as listed in **Box 2**.

PRIMARY SARCOMAS OF THE CHEST WALL: SOFT TISSUE

Primary malignant soft tissue sarcomas of the chest wall are generally reported in small series that also include lesions arising in bone and cartilage. Gordon and colleagues[4] reported the largest surgical series of patients with soft tissue sarcomas of the chest wall in 1991. The study included 149 patients who had undergone resection at the Memorial Sloan-Kettering Cancer Center in New York. The 5-year overall survival rate was 66%. The study also included 32 patients who had desmoid tumors, which are not

Box 2
Primary malignant chest wall masses according to tissue of origin

Chest wall sarcomas: soft tissue

Liposarcoma

Rhabdomyosarcoma

Leiomyosarcoma

Malignant fibrous histiocytoma

Angiosarcoma

Chest wall sarcomas: cartilage and bone

Chondrosarcoma

Osteosarcoma

Ewing sarcoma

Synovial cell sarcoma

histologically classified as sarcomas or as malignant. A large retrospective study containing 55 surgically treated patients with soft tissue sarcomas of the chest wall was reported from a single institution in Brazil in 2005.[5] In this series, fibrosarcoma accounted for nearly 53% of the cases. With wide surgical resection they reported disease-free survival rates of 75% at 5 years and 64% at 10 years. Histologic grade of the tumor and the type of surgical resection were found to be independent prognostic factors for disease-free survival. This finding is in keeping with other studies that suggest that age, gender, symptoms, and size do not significantly affect survival.[6]

Liposarcomas commonly present as retroperitoneal masses or extremity masses; primary liposarcomas of the chest wall are exceedingly rare. Most chest wall liposarcomas are low grade and wide surgical resection with a 4 cm R0 resection is considered the optimal treatment; attempts at debulking are considered palliative and offer little advantage to the patient.[7] The role of preoperative adjuvant radiation on the initial presentation of a chest wall liposarcoma is not defined. Radiation therapy is often reserved for patients with recurrent disease who are then offered radical reoperative surgery requiring complex reconstructions.[8] Likewise, at present, there does not seem to be any role for induction chemotherapy. However, there is an interesting report of complete regression of an anterior chest wall liposarcoma with the administration of interferon-α and tumor necrosis factor-α. However, there has not been any follow-up report or duplication of the results.[9]

Rhabdomyosarcoma tumors arise from a primitive muscle rhabdomyoblast. These tumors are found in pediatric populations. The major sites of