Management of Primary Soft Tissue Tumors of the Chest Wall

Anthony Cipriano, MD, William Burfeind Jr, MD*

KEYWORDS

• Surgery • Chest wall tumors • Malignancy

KEY POINTS

- Primary chest wall tumors compose only 1% to 2% of all thoracic neoplasms and approximately 60% are malignant. A majority of patients present with a painless mass.
- Work-up includes thoracic imaging with CT, MRI, and possibly PET. Diagnosis is made with a carefully designed core needle biopsy or incisional biopsy.
- Surgical wide excision with negative margins is the standard treatment of most primary soft tissue malignancies.

INTRODUCTION

The chest wall comprises a variety of tissues, including skin, fat, lymphovascular vessels, fascia, muscle, bone, and cartilage. Overall, it provides protection to vital organs, provides stability for the shoulders and arms during movement, and has evolved into a dynamic structure that assists with respiration. Chest wall masses have broad potential etiologies and present diagnostic and therapeutic challenges. Each of the tissue types composing the chest wall can give rise to benign or malignant primary chest wall masses. Also, due to the chest wall's close proximity to the breast, pleura, mediastinum, and lung, it is at risk from invasion from locally advanced tumors and inflammatory processes. Finally, the chest wall is a large target for metastases from distant tumors and these are the most common malignant chest wall masses.

Primary chest wall tumors compose only 1% to 2% of all thoracic neoplasms.[1] They are best classified according to their tissue of origin (bone or soft tissue) and are further subdivided as either benign or bearing malignant potential. Approximately 60% of primary chest wall tumors are malignant, with increasing malignant risk at the extremes of age.[2] Of these malignant tumors, approximately 55% arise from bone or cartilage and 45% from soft tissue.[3] All chest wall tumors are uncommon and their nomenclature varies throughout the literature. This article reviews the most common primary soft tissue chest wall tumors and focuses on their description, demographics, diagnosis, treatment, and general prognosis.

DIAGNOSIS

A majority of patients with a primary soft tissue chest wall tumor present with a painless, palpable mass. Often, patients have already undergone some form of thoracic imaging for unrelated conditions and a chest wall mass was noticed incidentally. Rapid growth and pain are harbingers of malignant tumor behavior and imply local invasion of adjacent structures, such as periosteal or neurologic structures. There are no reliable clinical

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Department of Surgery, St. Luke's University Health Network, Bethlehem, PA 18015, USA
* Corresponding author. 701 Ostrum Street, Suite 603, Bethlehem, PA 18015.
E-mail address: William.Burfeind@sluhn.org

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features, however, for distinguishing benign from malignant. Initial work-up should include obtaining a detailed history with attention to prior/current malignancies, radiation exposure, recent/current infections, symptoms associated with the mass (ie, pain), time of discovery, rate of growth, and review of prior thoracic imaging. If the mass is palpable, its size and characteristics of soft versus hard and fixed versus mobile should be noted. The initial goal of evaluation and work-up of a chest wall mass should be to determine if it is a primary or secondary lesion because this guides treatment.

Diagnostic imaging cannot reliably distinguish benign from malignant lesions although it can be suggestive of tumor type and differentiating benign from malignant tumors. The advent of cross-sectional imaging techniques, such as CT and MRI, have aided in localizing and determining the extent of chest wall lesions. Chest radiography is often the initial test and does not allow for comprehensive tumor assessment. CT is more sensitive than chest radiography for detecting calcified tumor matrix and cortical destruction—findings suggestive of extracompartmental extension—and delineating the extent of pleural, lung, mediastinal, and nodal involvement.[4] MRI is useful for further characterization of the lesion and offers superior spatial resolution giving precise delineation of tissue planes including bone and vasculature. The imaging features of many chest wall tumors can be suggestive but often are nonspecific.

PET has proved a valuable tool in the evaluation, particularly in regard to staging and response to treatment, of various neoplasms and has shown promise in the imaging of chest wall tumors as well. With regard to sarcomas, PET has been shown more accurate than CT alone in initial staging, evaluation of early treatment response, and both short-term and long-term follow-up.[5] Furthermore, when combined with CT, accuracy in staging, treatment response evaluation, and restaging is increased compared with either PET or CT alone.[5] For restaging sarcomas, PET has been shown to detect high-grade local recurrence, with a sensitivity of 88% and specificity of 92%.[6] It also has a role in distinguishing high-grade soft tissue sarcomas from low-grade sarcomas/benign lesions (95% sensitivity and 75% specificity).[7] PET has found superior to CT for defining the extent of chest wall sarcomas, particularly for tumors greater than 5.5 cm, and may provide some value in planning full-thickness chest wall resection.[8] In 1 study of pediatric patients, ages 18 or less, PET and conventional imaging modalities (CIMs) have been shown equally effective in detection of primary tumors (accuracy 100%).[9] In the same study, PET was superior to CIMs for the detection of lymph node involvement (sensitivity 95% vs 25%, respectively) and bone manifestations (sensitivity 90% vs 57%, respectively). Best results were obtained with combined modalities, with 91% correct therapeutic decisions, a rate significantly better than using CIMs alone (59%). The role of PET/CT in regard to evaluating chest wall tumors is promising, but the evidence is not strong enough to support its use as a sole modality for pretherapeutic staging. It adds useful information, however, when combined with CT and MRI.

Whether a tissue diagnosis is needed prior to definitive therapy depends mainly on the size of the lesion. Small lesions (<2 cm), thought to be benign, should undergo excisional biopsy with clear, greater than 1 cm, margins, which is both diagnostic and therapeutic. If the lesion is larger (>2 cm) or likely malignant, a preoperative tissue diagnosis is preferred. Fine-needle aspirate is acceptable when a metastatic lesion is suspected but often yields insufficient tissue to adequately diagnose and grade soft tissue sarcomas.[1,10,11] If a lesion is benign, it is resected with negative margins and, if malignant, wide excision is performed.[12] It is essential to plan biopsies with future curative resection in mind because reexcision of the biopsy tract is required as part of the definitive resection.

Benign Soft Tissue Tumors

Lipoma

Lipomas are benign mesenchymal tumors of adipose tissue that can occur superficially and deep as well as intramuscularly.[13] They are painless, nontender, well-circumscribed, mobile masses that can occur extra thoracically or intrathoracically.[14] Intrathoracic lipomas can remain completely intrathoracic or can extend through the ribs, giving rise to a dumbbell-shaped lesion.[15,16] The appearance of these tumors on CT and MRI shows a well-circumscribed, homogeneous lesion with the imaging characteristics of fat, which distinguishes it from liposarcomas that almost always contain inhomogeneous nonfatty elements.[17] Grossly, these lesions are well-circumscribed, thinly encapsulated tumors of mature adipose tissue. Histologically, they comprise uniform, mature adipose tissue that lacks atypia or multivacuolated lipoblasts.[13] Malignant transformation is exceedingly rare.[18] Surgical excision is indicated for cosmetic purposes or if malignancy cannot be excluded (**Table 1**).[1]