Chest Wall Tumors

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Introduction

The musculoskeletal structure of the chest wall serves to protect the thoracic and mediastinal viscera, while providing integrity for respiration. The complex relationship of all anatomic components contributes to its function but also makes it susceptible to a wide variety of pathology. Thus, tumors of the chest wall represent a diagnostic and therapeutic challenge. Chest wall masses have a broad differential diagnosis including local extension of intrathoracic lesions, presentations of systemic diseases or inflammatory processes, metastasis, and less frequently, primary tumors of the chest wall, which can be benign or malignant. More than 50 % are malignant and are most commonly a result of direct invasion or metastasis from adjacent thoracic tumors [1]. Overall, primary chest wall tumors make up less than 5 % of thoracic malignancies and vary widely in pathology, as they arise from all anatomic structures comprising the chest wall [2, 3]. This chapter reviews the pathology, options for diagnosis, and standard treatment of the various lesions arising from the chest wall.

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Evaluation

Signs and Symptoms/Clinical Presentation

Chest wall tumors, with their diverse etiology, also have varying clinical presentations. Workup should always begin with a thorough history and physical. They can be symptomatic or asymptomatic, with >20 % being found incidentally on chest radiograph [4]. Pain is the most common symptom for both benign and malignant lesions and it is usually a sign of bony invasion. Extrathoracic tumors are more likely to present as a palpable, enlarging mass, which is a common presentation, along with pain. There is a wide variation of age at presentation. Older patients tend to have larger, more aggressive tumors when compared to their younger counterparts, who are more likely to have smaller benign tumors [4]. Though tumors can present at any age, they are more likely to be malignant at the extremes of age [3]. There are no specific signs or symptoms that distinguish between benign and malignant lesions, which add to the challenge of diagnosis [3]. Symptoms can

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indicate the location of the lesion, for instance, paresthesia and weakness may be present with involvement of neurologic structures.

Diagnosis

To perform an operation with the best possible outcome for the patient, correct diagnosis, preoperative staging, and surgical planning are essential. Given the wide variation in histology of origin, it is frequently necessary to use imaging appearance, location, and clinical information to make a diagnosis rather than imaging alone [5].

Imaging Modalities

Chest Radiograph: Radiographic diagnosis alone can be challenging. As mentioned previously, >20 % of chest wall tumors are found incidentally on chest x-ray [5]. If this is not the case, chest radiograph is often the first imaging modality obtained [6]. The chest radiograph can not only show location and size, it can detect calcification, ossification, or bone destruction, but is otherwise limited in detail. Therefore, additional workup is always indicated, with CT and MRI being the most useful diagnostic modalities (Fig. 8.1).

Computed Tomography (*CT*): If contrast is used, CT scan can provide information about the vascularity of a tumor, location, and composition of a chest wall mass, as well as appraisal of the extent of tumor invasion and involved structures. It is also better at predicting cortical bone involvement. Cartilaginous matrix calcifications are better defined with CT than with MRI which is very useful for preoperative planning [6]. It can also be useful in confirming histologic diagnosis by obtaining a CT-guided biopsy of a lesion. The approach for biopsy should be chosen together with the surgeon, with consideration given for preoperative planning (Fig. 8.2).

Magnetic Resonance Imaging (*MRI*): Accurate tissue characterization can be obtained with MRI due to its superior tissue-resolving features and multiplanar image acquisition, which makes it an important assessment tool [5, 7]. It is the most accurate study for spinal involvement, characterizing soft tissue involvement, and further delineating between vascular, soft tissue, and nerve involvement [3, 6, 8] (Fig. 8.3).

Positron Emission Tomography (*PET*): The clinical significance of standard uptake value (SUV) may be useful but has not been formally established as a diagnostic tool in the evaluation of chest wall lesions [9, 10]. There is some data suggesting that PET may be more accurate for determining the extent of large tumors, especially those >5.5 cm [10, 11]. Though, it is not routinely

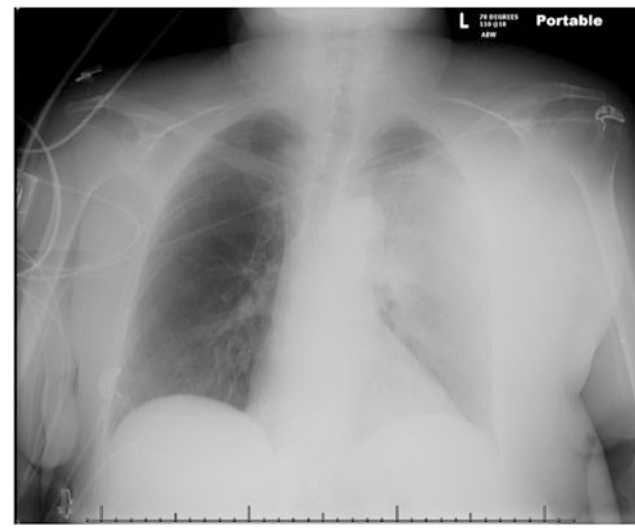

Fig. 8.1 Chest radiograph of large left chest wall tumor found to be poorly differentiated carcinoma with extensive necrosis

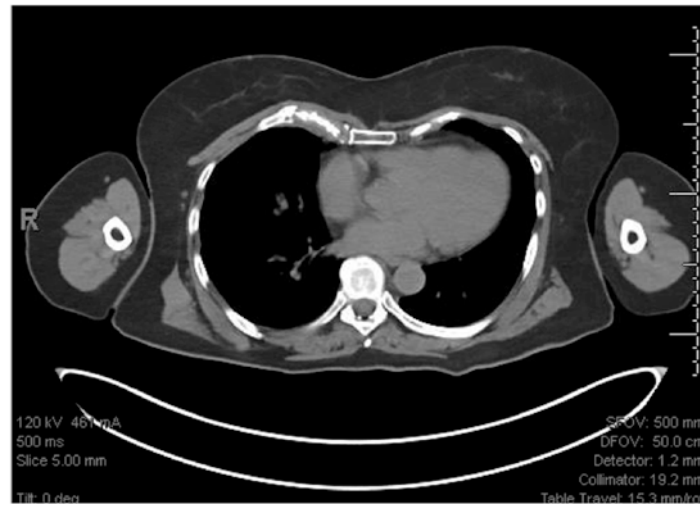

Fig. 8.2 Axial CT scan of chest wall sarcoma demonstrating osseous destruction of right anterior third rib

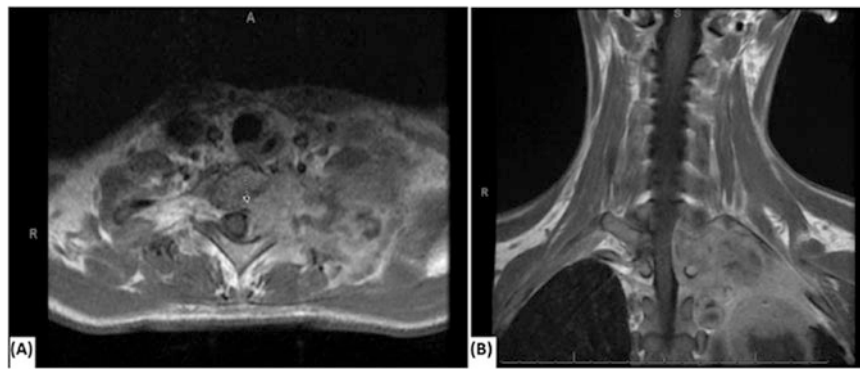

Fig. 8.3 MRI scan of left chest tumor in a patient with Hodgkin's Lymphoma invading the thoracic spine causing cord displacement and rib destruction (**a**) axial view and (**b**) coronal view

discussed as a common imaging modality in the evaluation of chest wall tumors, there seems to be increasing frequency of its mention in the literature. In a small recent study by Petermann et al., PET imaging was found to be superior to CT for defining the extent of chest wall tumors, giving hope that it will be found to be of significant diagnostic value in larger prospective studies, which have yet to be conducted [11]. In the case of malignant disease, it may be useful for defining patients with limited disease versus disseminated disease (Fig. 8.4).

Ultrasound (*US*): There has recently been documented use of ultrasound for more clearly defining tumor margins [6, 10, 12]. Ultrasound may accurately predict the degree of tumor invasion or extension, which could help with preoperative planning to ensure negative surgical margins [10]. Caroli et al. describe the absence of lung sliding on ultrasound as being accurately predictive for lung invasion from a chest wall tumor in 8/8 patients where other imaging modalities were inconclusive [12].

Biopsy

In the majority of cases, radiographic features alone are insufficient to make a complete diagnosis; therefore, histologic evaluation is required. Suitable modalities to obtain tissue diagnosis include fine-needle aspiration, incisional biopsy,

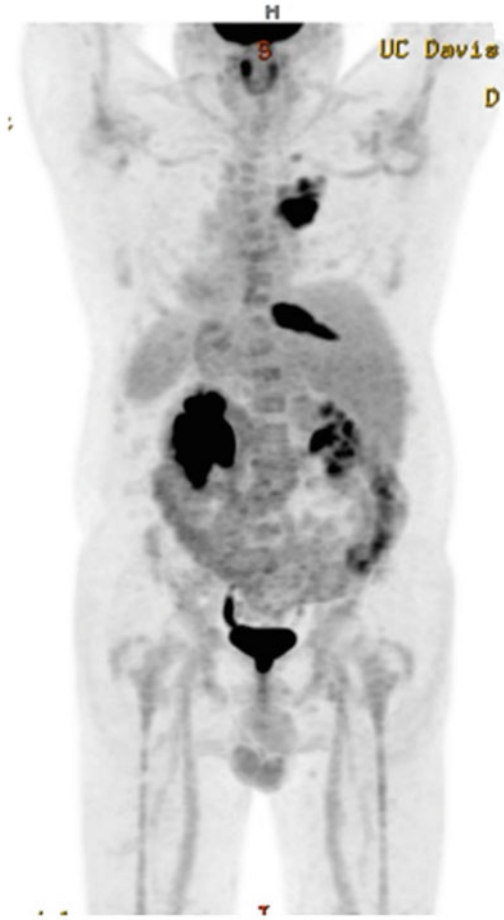

Fig. 8.4 Maximum Intensity Projection image from PET/CT demonstrating isolated metastasis to the right tenth rib from NSCLC

or excisional biopsy [9]. The approach to obtaining a biopsy is guided by the size and location of the lesion, extent of resection, complexity of any associated need for reconstruction, and if there are any associated comorbidities. Typically, lesions less than 2–5 cm undergo excisional biopsy depending on the suspected pathology and the surgical center [2, 3, 10]. An absolute diagnosis prior to resection is usually of value for lesions greater than 5 cm, given the increased likelihood of extensive dissection and complex reconstruction. Therefore, lesions greater than 2–5 cm may undergo needle aspiration or incisional biopsy for confirmation of diagnosis, allowing for better operative planning [2, 3, 10]. Core needle biopsy may determine if it is a benign or malignant process but may not obtain sufficient sampling for further histologic analysis or genetic testing.

It is essential that the entire treatment plan be considered before any biopsy to ensure correct placement of the biopsy location. The lesion is often approached directly in order to avoid contamination of surrounding structures. If a malignant diagnosis is obtained, definitive excision is required, which necessitates total excision of the biopsy tract. It is preferable, for the earlier reasons, that the biopsy be performed at the treating/surgical center rather than at the referring center, or in coordination with the operating surgeon.

Preoperative Evaluation

Once the diagnosis has been made, and the primary excision is determined, it is important to complete a thorough history and physical with careful consideration of any past surgical history or treatment which may affect approaches for resection or reconstruction. Immunosuppression, history of radiation or planned radiation, and previous chest procedures should all be considered. Additionally, full medical assessment should be obtained, including a cardiac and pulmonary function evaluation in cases where there is concern for simultaneous pulmonary resection. As mentioned earlier, complete radiographic analysis is essential to operative planning and may delineate the need for preoperative consultation with neurosurgical or reconstructive specialists. Patients with extensive or complicated lesions are best served with a multidisciplinary approach, which may include cardiothoracic surgery, spine surgery, plastic surgery, radiation oncology, and/or medical oncology [10, 13].

Pathology

Benign

Benign chest wall tumors are less common than malignant lesions and arise from nerve, blood vessel, osseous, cartilaginous, or fatty tissue [5]. It is crucial that malignant diagnoses are defini-

tively ruled out with radiographic and histologic analysis for any of these lesions.

Skeletal Lesions

Benign chest wall tumors of bony origin are less common than malignant bony lesions. However, one should always assume a malignant condition until proven otherwise.

Osteochondroma

Osteocondroma is the most common type of benign bone tumor and is typically found in the femur, humerus, and tibia [14]. In the chest, osteochondromas are most common in the rib or scapula, where they are commonly found at the costochondral junction, and they develop from abnormal growth of normal tissue [5, 10]. Osteochondromas make up 50 % of benign rib tumors [2]. These masses frequently cause pain as they progress with growth of bony exostoses [14]. Peak incidence is in the second decade of life [15]. It is one of the few chest wall tumors where a definitive diagnosis can be made on CT or MRI based on the appearance of the cortex and medullary space blending into the underlying bone [5], and punctate or flocculent calcifications with mineralized hyaline cartilage cap [3]. Surgical treatment is resection and provides complete pathologic evaluation, symptomatic relief, and minimizes the risk of malignant transformation [16]. Cartilage caps greater than 2 cm in adults and greater than 3 cm in children are suspicious for malignant degeneration [3, 10].

Chondromas (Enchondromas)

Chondromas are also typically found at the sternocostal junction arising from cartilaginous tissue [2, 10]. They are relatively common, making up 15–20 % of benign chest wall lesions. Chondromas are usually painless, slow growing, osteolytic lesions, and present between 20 and 30 years of age. The distinction between chondroma and low-grade chondrosarcoma is difficult. Therefore, all chondromas are treated as malignant lesions, and wide excision is recommended [4]. Appropriate wide excision for a malignant lesion is commonly accepted as full thickness resection with 4-cm margins and en bloc removal of one rib above and below the lesion as well as intercostal muscles, pleura, and wide clear margin of adjacent tissue [17, 18].

Fibrous Dysplasia

Fibrous dysplasia typically appears in the lateral or posterior tract of the ribs and is the third most frequent benign chest wall lesion [2]. Normal bone is replaced with fibrous tissue forming a slow-growing mass, which can cause pathologic fractures and result in pain; otherwise, presentation may be as an asymptomatic mass in the posterior aspect of a rib. Fibrous dysplasia appears as a lytic lesion on chest radiograph with a soap bubble or ground-glass appearance that is diagnostic. Only one bone is involved in 70–80 % of the cases [5], most commonly the second rib [10]. In 25 % of cases it affects more than one bone [15]. When this occurs, it may also be associated with cafe-au-lait spots and endocrine abnormalities with the constellation of abnormalities known as McCune–Albright syndrome [3, 15]. It typically occurs in the second and third decade of life with no disparity in gender ratio [3]. Treatment is wide local excision for relief of symptoms and confirmation of diagnosis [4]. Conversely, some sources believe that excision is not necessary if the lesion remains asymptomatic, as imaging is often diagnostic [10].

Eosinophilic Granuloma/Langerhans Cell Histiocytosis

Eosinophilic granuloma, or Langerhans cell histiocytosis, is a less common tumor that can arise in the anterior chest wall [4]. This results from idiopathic proliferation of histiocytes, considered to be of bone marrow origin. These masses tend to present with chest pain, fevers, and an isolated tender mass that has a typical lytic appearance on radiograph and chest CT [19]. Eosinophilic granuloma is a diffuse infiltrative inflammatory process that can affect many organs and can also manifest with an associated leukocytosis. In the chest wall, it can cause destruction of bone cortex and new subperiosteal bone formation that can mimic osteomyelitis or malignancy [4]. Treatment in the literature varies ranging from nonsurgical

approaches such as steroids, chemotherapy, and low dose radiotherapy without surgical excision [3] to wide local excision performed both for diagnosis and symptomatic relief [19, 20]. Most forms are treated nonsurgically, but if the lesion is isolated, then resection and curettage are reported to have good results [10]. This lesion requires a histologic diagnosis, with identification of birbeck granules on electron microscopy [3]. If a procedure is necessary for diagnostic purposes, then it often makes sense to pursue excisional biopsy at the time.

Aneurysmal Bone Cyst

Aneurysmal bone cysts are a rare, benign, locally aggressive entity consisting of an expanding osteolytic lesion with blood-filled cystic spaces. Their etiology is not clear, as they are often associated with abnormal bone or are found in the setting of another underlying bone tumor [10]. They are usually found on the posterior chest wall and 75 % present before the age of 20 [3]. If there is soft tissue extension, then it may be difficult to differentiate from sarcomas. Complete excision is recommended, with cure rates of 70–90 % [10]. Radiation is sometimes used for local control in the setting of aggressive or recurrent tumors [10]. These lesions are not known to metastasize.

Osteoid Osteomas

Osteoid Osteomas are small, tumors of osteoblastic origin that rarely occur as primary chest wall tumors [15]. They often present in the first and second decades with nocturnal pain, which is improved by NSAIDs [3, 10]. The most common location for presentation is the posterior spine and ribs and can also be associated with scoliosis. Radiographically, they have a small radiolucent lesion with thick sclerotic margin of reactive bone and surrounding soft tissue edema. Their characteristic appearance on bone scan is known as the double density sign [15]. Treatment generally involves radiofrequency ablation.

Osteoblastoma

Osteoblastoma is a rare osteoblastic tumor thought to be on the continuum of osteoid osteomas [3, 10]. These tumors also primarily affect the posterior ribs. On radiologic evaluation they appear as well-defined expanding osteolytic lesion, but with a sharp sclerotic rim and lack of a central nidus. They can be locally aggressive with potential for recurrence, and thus wide local excision is the preferred treatment [15].

Giant Cell Tumor

Giant cell tumors are more common and occur most frequently between the second and fourth decades [10], with men having a higher occurrence than women [3]. They are considered locally aggressive with 30–50 % recurrence rates and rare reports of metastasis [3]. Secondary to their locally aggressive nature, wide local excision is recommended [10]. They are osteolytic lesions with cortical thinning and often are associated with a soft tissue mass (Table 8.1).

Soft Tissue Tumors

Soft tissue tumors that affect the chest wall have a similar variability in pathology of that of the bone-based lesions. Cutaneous nevi, lipomas, hemangiomas, lymphangiomas, and neurogenic tumors are some of the benign lesions that can be found in the soft tissue of the chest wall. They are treated with wide local excision to negative margins to avoid local tissue recurrence. Soft tissue tumors have some additional challenges to preoperative diagnosis. Intercostal hernias have been reported to be confused with benign soft tissue lesions, mandating appropriate radiographic confirmation of clinical suspicion based on history and physical exam [21]. Radiographic evaluation may not always definitively address malignancy but will assist in operative planning.

Lipomas

Lipomas usually present as well-circumscribed adipose masses but are often deeper and larger on the chest than on other sites of the body [3]. They are more prevalent in the obese and older patients, with incidence highest at approximately ages 50–70 [3]. As with many chest wall tumor presentations, they can sometimes be challenging to

Table 8.1 Benign bony lesions

Tumor	Origin	Presentation	Imaging characteristics	Treatment
Osteochondroma	Cartilage	Rib, scapula, 50 % of benign rib tumors. Peak incidence in second decade	Cortex and medullary space blends w/ underlying bone, punctate or flocculent calcifications w/ mineralized hyaline cap (>2 cm cap in adults or >3 cm in children = ↑ risk of malignant transformation)	Resection
Chondroma (enchondromas)	Cartilage	Usually painless lesion at 20–30 year of age, most commonly found in anterior ribs	Osteolytic, lobulated appearance with distinct boarders	Wide local excision—distinction from low-grade malignant chondrosarcoma is difficult
Fibrous dysplasia	Fibrous tissue	Lateral or posterior rib. Slow growing. Often w/ pathologic fracture @ 20–30 yo	Lytic lesion. Soap bubble or ground glass appearance = diagnostic	Wide local excision for symptoms and definitive diagnosis vs. no intervention if diagnosis confirmed and remains asymptomatic
Eosinophilic granuloma (langerhans histiocytosis)	Bone marrow	Anterior chest wall, ribs, sternum. Rare. May have pain, fever, leukocytosis	Focal lytic radiolucent lesions with biopsy necessary for diagnosis (birbeck granules on histology)	Most forms treated nonsurgically. If the lesion is isolated, then resection and curettage are reported to have good results
Giant cell tumor	Osteoclast	20–40 yo with ♂ > ♀ 30–50 % risk of recurrence	Osteolytic lesions with cortical thinning, often with soft tissue mass Vascular sinuses w/ giant cells and spindle cells	Wide local excision recommended secondary to locally aggressive nature and rare ability to metastasize
Aneurysmal bone cyst	Unclear	Posterior chest wall, <20 yo, rare, locally aggressive. can coexist with other lesions	Cystic, expanding osteolytic lesions. Free fluid w/ multiseptated hemorrhagic cysts which are not pathognomonic	With complete excision, cure rates 70–90 %. Radiation may be used for aggressive disease or recurrent tumors
Osteoid osteoma	Osteoblast	Nocturnal rib pain that responds to NSAIDs and Tylenol, first–second decade of life. Often posterior, may be associated with scoliosis	Small radiolucent lesion with thick sclerotic margin of reactive bone with soft tissue edema. Characteristic appearance on bone scan = double density sign	Radiofrequency ablation?
Osteoblastoma	Osteoblast	Posterior/posterio-lateral rib mass or pain	Well-defined osteolytic lesion with sharp sclerotic rim and lack of a central nidus	Can be locally aggressive and recurs, wide local excision is recommended

evaluate with radiographic modalities, and are often difficult to differentiate from low-grade liposarcomas [3]. There have been case reports of soft tissue lesions with preoperative imaging suggestive of intrathoracic or chest wall invasion, which were subsequently found to be giant benign lipomas on excision [22, 23]. Chest wall lipomas should be excised for symptomatic relief and complete diagnostic evaluation secondary to the risk of malignant transformation.

Lymphangiomas

Lymphangiomas of the chest wall can be cystic or cavernous in nature and are a result of a developmental malformation. They can be located within the mediastinum or the chest wall itself. Preoperative CT imaging is essential to assess the extent of the lesion, as several reports of giant lesions exist in the literature [24, 25]. Complete surgical excision is required for excellent prognosis and avoidance of long-term lymphatic fistula formation. Nonoperative therapy with radiation or sclerosing agents remains controversial for lymphangiomas of the chest wall, with surgery as the standard of care [2].

Hemangiomas

Hemangiomas arise from blood vessels and can be found within the chest wall or protruding through the chest wall from the thoracic cavities or mediastinum and arise from blood vessels. Often seen as heterogeneous soft tissue masses with fatty, fibrous, and vascular elements on CT scan [3]. Ultrasound may be a useful modality to evaluate flow within the lesion [3]. MRI is used to distinguish benign from malignant lesions based on phleboliths, fat component, and high intensity and fat suppression on T2 imaging technique, but surgical biopsy is required for definitive diagnosis [26]. Hemangiomas of the chest wall can be intramuscular, intercostal, or cavernous. They tend to occur in patients less than 30 years of age and present as painful masses. Treatment is complete surgical excision if symptomatic, but local recurrence rates are as high as 20 % [27].

Neurogenic Tumors

Neurogenic tumors of the chest wall include neurofibromas and neurilemomas that arise from peripheral nerve sheaths and are usually associated with neurofibromatosis. Presentation is often between the ages of 20 and 30 and are seen as slow growing homogeneous masses on CT and MRI [3]. These lesions may undergo cystic degeneration creating a target appearance on MRI. As one may expect, biopsy is extremely painful. Surgical excision is usually only recommended for cosmetic reasons for cutaneous lesions, as there is a low likelihood of malignancy. However, plexiform lesions that are increasing in size or becoming symptomatic should be completely excised without preoperative biopsy [28].

Desmoid Tumors

Desmoid tumors arise from musculo-aponeurotic structures and are considered myofibroblastic or fibroblastic in origin. They can develop anywhere in the body, but are most common in the abdomen and extremities, with only 10–28 % arising in the chest wall [1, 10]. The most frequent location in the chest wall being in the shoulder girdle [18]. Their histology is benign, but because of their aggressive growth rates and tendency to grow into nearby structures or cause compressive symptoms, they can be considered malignant. Desmoid tumors are common in females and males, usually younger than 40 years and can be found in patients with familial adenomatous polyposis, where they are related to a mutation in the APC gene. Desmoids can also occur in sites of previous trauma, scar, or radiation. Presentation can consist not only of a palpable chest wall mass/swelling and associated pain, but dyspnea, cough, shortness of breath, and dysphasia have all been reported [18]. The latter symptoms possibly being the result of tumor mass effect. Resection to tumor-free margins is needed for cure, which is difficult to achieve given high incidence of microscopic positive margins [10, 18]. This clearly reinforces the need for wide margins at the time of original resection. Again, appropriate wide excision for a malignant lesion is commonly accepted as full thickness resection with 4-cm margins and en bloc removal of one rib above and below the lesion as well as intercostal muscles, pleura, and wide clear margin of adjacent tissue [17, 18]. When negative margins are not possible, radiation should be considered,

though the effectiveness of radiation treatment for nonresectable disease, recurrence, or to treat positive margins, remains with uncertain efficacy [10, 29]. Recurrence rates for desmoid tumor are high. Abbas et al. report 5-year probability of developing a local recurrence as 37 %, with an 89 % rate in patients who had positive margins at the time of resection [17].

Elastofibromas

Elastofibromas classically occur in the subscapular region, with peak incidence between 40 and 70 years of age. This lesion has a female predominance and a characteristic layered appearance with mild enhancement on CT scan performed with IV contrast [10]. Recommended treatment is complete excision, with intention of cure (Table 8.2).

Malignant

Skeletal

Bony malignant chest wall tumors account for 55 % of all chest wall masses and have an average 5-year survival of 60 % [4]. Malignant lesions tend to grow faster, manifest more painfully, and present as larger masses than benign lesions [30]. Primary tumors of the sternum are also usually found to be malignant [3].

Chondrosarcomas

Chondrosarcomas are most commonly found on the anterior chest wall and account for 30 % of primary malignant bone tumors [31]. They are the most common malignant bony tumor of the chest wall. Chondrosarcomas are rarely found in patients younger than 20 years of age and are found more commonly in a bimodal distribution in the second–third and fourth–fifth decades of life [4, 10, 30]. These tumors represent a malignant degeneration of benign chondromas, with both tumors having similar clinical presentations of painful, hard, slow growing, fixed masses on the anterior chest wall [32]. Additionally, they can be associated with trauma [4]. Radiographic appearance is typically a well-defined mass with soft tissue attenuation associated with cortical destruction, soft tissue attenuation with foci of dense chondroid matrix calcification [2, 6]. Synchronous or metachronous lung metastases are seen in 10 % of patients at the time of presentation [4]. Pathologic distinction between chondroma and chondrosarcoma is challenging, therefore both tumors are resected with 4-cm margins. Resection is the mainstay of therapy, chemotherapy is largely ineffective, and radiation is reserved for patients who are unresectable or with positive resection margins [10, 32]. Five-year survival is 65–92 %. Tumor-free margin is the largest predictor of local recurrence. Four to ten percent of patients with negative margins will have local recurrence; whereas 73–75 % of those with positive margins will have local recurrence [4, 33].

Osteosarcomas

Osteosarcomas make up 10–15 % of malignant chest wall tumors, commonly occurring in the rib, scapula, and clavicles [4, 30]. These tumors present as painful masses in young or elderly adults. They often present in the second decade of life [10], specifically cited at puberty in one report [3]. Metastatic disease at the time of presentation is common, with the most common sites being lung, lymph nodes, and liver. The mass appears calcified on imaging with both lytic and calcified or sclerotic osteoid areas [6], and may have hemorrhagic or necrotic components [30]. IV contrast will show areas of different enhancements [6]. Treatment is wide local excision in combination with neoadjuvant chemotherapy. The presence of metastases drastically affects 5-year survival, decreasing it from more than 50 % to between 15 and 20 % [2]. Response to chemotherapy, tumor burden, and presence of metastases are predictive of overall survival [4]. Radiation has been used in cases with an inability to achieve an adequate resection; however, osteosarcomas are not very radiosensitive [10].

Ewing Sarcoma

The Ewing sarcoma group of tumors is a spectrum of small round-cell tumors that share the chromosomal translocation t(111;22) and include Ewing sarcoma, and primitive neuroectodermal tumor (PNET), which is also known as Askin tumor when located in the chest wall. These tumors are the third most common malignant chest wall tumors overall but are the most common in the pediatric and young adult populations. Males are

Table 8.2 Benign soft tissue lesions

Tumor	Origin	Presentation	Imaging characteristics	Treatment
Lipomas	Adipose tissue	Well-circumscribed adipose tissue. Found more frequently in older (50–70 yo) and obese patients	Can be difficult to differentiate from low-grade liposarcomas on imaging. Tend to be larger and deeper than at other sites on the body	Excision for symptomatic relief and complete diagnostic analysis
Fibromas (desmoid tumors)	Fibrous tissue, myofibroblastic or fibroblastic	Often present <40 yo. Can occur at sites of trauma, scar, radiation, or in pts with familiar adenomatous polyposis. Pain, mass, swelling, and sx from mass effect have all been reported	Generally have similar enhancement to muscle. (Variable nondescript appearance depending on collagen content and extent of myxomatous degeneration)	Aggressive growth rates, often resulting in compressive symptoms and tendency to grow into nearby structures. High recurrence rates. Must have resection to tumor-free margin for cure. Radiation tx when negative margins not possible
Hemangiomas	Vascular	Painful mass in <30 yo, intramuscular, intercostal, or cavernous	Heterogeneous soft tissue mass with fatty, fibrous, and vascular elements. Ultrasound often used to evaluate flow. MRI can help distinguish between benign and malignant, but surgical biopsy required for definitive diagnosis	Complete surgical excision if symptomatic. High local recurrence rates
Neurogenic tumors (neurofibromas, neurilemomas)	Nerve, peripheral nerve sheath tumors	Usually associated with neurofibromatosis. Slow growing. Often occurring between 20 and 30 yo	Homogeneous mass on imaging. May have cystic degeneration creating a target appearance on MRI. Extremely painful if biopsied	Low likelihood of malignancy. Surgical excision of cutaneous lesions for cosmetic reasons. Plexiform lesions that are symptomatic or growing should undergo complete excision
Lymphangiomas	Developmental malformation of lymph system		Cystic or cavernous lesion	Complete surgical excision is standard of care. Treatment with radiation or sclerosing agents = controversial
Elastofibroma		Mass in subscapular region at 40–70 yo, ♀>♂	CT: characteristic layered appearance with mild enhancement with IV contrast	Local excision is curative

slightly more affected than females with a 1.6:1 ratio [3]. In addition to the typical chest wall tumor presentation of a painful chest mass, these tumors may also present with systemic symptoms such as fever, malaise, and weight loss, as well as pleural and pericardial effusions [3]. On radiographic imaging, these lesions are often seen as a large noncalcified, soft tissue mass with bone destruction, classically associated with an onion peel or sunburst appearance [3]. They may also have components of hemorrhage or necrosis [6]. The Ewing sarcoma family is an aggressive tumor family with high recurrence rates and high likelihood of metastases [34]. Neoadjuvant chemotherapy is typically given followed by wide local excision with good results. Response to chemotherapy is predictive of local recurrence. Myeloablative therapy and stem cell rescue may improve outcomes in patients with primary metastatic presentation, and bilateral whole-lung radiation has also been used to improve event-free survival with lung, bone, or bone marrow metastasis [3]. While radiation has been shown to provide good local control, it has significant oncogenic potential and cardiopulmonary toxicities in such a young population. Metastases reduce 5-year survival to 30 % from 100 % with local disease at presentation [4].

Solitary Plasmacytoma

Solitary plasmacytoma is a rare tumor composed of monoclonal plasma cells, as in multiple myeloma; however, this is a discrete mass without diffuse spread. This tumor affects elderly men who present with pain without a mass. Once surgical biopsy has been performed, surgical therapy stops because this tumor is treated primarily with radiation [35]. Five-year survival is 40–60 % and is most dependent on whether or not multiple myeloma develops instead of control of the primary lesion [4]. Two-thirds progress to develop myeloma within 3 years of diagnosis, worsening prognosis, and the rest may achieve permanent cure (Table 8.3).

Soft Tissue Tumors

Soft tissue tumors commonly present as painless masses, with the anterior chest wall being the most common location for soft tissue malignancies of the thorax [3].

Soft Tissue Sarcomas

Soft tissue sarcomas are the majority of primary malignant chest wall lesions, but they account for only 6 % of soft tissue sarcomas in the body. They are typically found in middle-aged men who present with a painless mass, except rhabdomyosarcomas, which are more common in children [1]. As with many chest wall tumors, the diagnosis cannot be made radiologically and requires tissue confirmation [15]. The mainstay of treatment is wide local excision. For high-grade sarcomas, 4-cm margins and resection of the rib above and below the lesion is recommended, with reexcision for positive margins [15]. If there is any question about the possibility of full resection, then neoadjuvant therapy should be explored. We will review some of the histologic subtypes later.

Malignant Fibrous Histiocytomas (MFH)

Malignant fibrous histiocytomas (MFHs) are found in the chest wall but are very common throughout the body, most commonly in the extremities, abdomen, or retroperitoneum. MFH is commonly found in elderly men, and there is usually a history of previous chest wall radiation [36]. Bimodal distribution has also been reported with peaks between 20 and 30 years of age and then again between 50 and 60 years of age [3]. The masses can grow to be a large size but are usually not painful [28]. MFH has a heterogeneous appearance on CT that can be enhancing or calcified, with ill-defined contours [3]. Treatment is wide local excision, and local recurrence rates are higher than 30 % [36]. Pre- and postoperative neoadjuvant chemotherapy are often utilized [37]. Metastatic lesions are diagnosed in 30–50 % of patients, and 5-year survival is only 38 % [28].

Liposarcomas

Liposarcomas are one of the most common malignant soft tissue tumors in the body, but they are not common in the chest wall [38]. They are rarely found in children and are most common in men ages 40–60 years. It is not unusual for them

Table 8.3 Malignant bony lesion

Tumor	Origin	Presentation	Imaging characteristics	Treatment
Chondrosarcoma	Cartilage	Anterior chest wall, painful firm mass. Most common malignant bony tumor of chest wall. 30–40 yo. Can be associated with trauma. lung metastasis in 10 %	CT-Well-defined mass with cortical destruction, lobulated soft tissue attenuation with foci of dense chondroid matrix calcification of varied shape and densities	Resection with 4-cm margins. 5-year survival 65–92 % with tumor-free margin. 75 % with + margins → recurrence. Radiation tx for recurrent or unresectable disease
Osteosarcoma	Bone	Second most common mal. Chest wall bony tumor. Typically present at puberty, commonly with metastasis	Calcified mass with lytic or sclerotic osteoid bone matrix within the mass. May have hemorrhagic or necrotic components	Wide local excision in combination chemotherapy. Tumor burden, metastasis, and tx response all predict overall survival which ranges from 15 to 50 %
Ewing sarcoma family (includes Askin tumor and primitive neuroectodermal tumor)	Bone	Third most common mal. Chest wall tumor overall. Most common in pediatric and young adults, ♂ > ♀	Large noncalcified, soft tissue mass with bone destruction. May have hemorrhagic or necrotic components. Classically associated with an onion peel or sunburst appearance	Neoadjuvant chemotherapy followed by wide local excision High recurrence rates and high likelihood of metastasis. Response to chemotherapy predicts recurrence Myeloablative therapy and stem cell rescue may improve outcomes in patients with metastatic disease
Solitary plasmacytoma	Bone marrow/ plasma cells	Rare. Elderly men. Pain without associated mass	Discrete mass without diffuse spread. Needs histologic diagnosis	Radiation. 40–60 % 5 years survival. 2/3 → myeloma. Overall survival is dependent on progression to multiple myeloma

to be large and frequently associated with trauma. Treatment is wide local excision, and 5-year survival is 60 % [28]. Local recurrence rates are high, and there is little to no role for chemotherapy and radiation.

Angiosarcoma

Angiosarcoma of the chest wall is a rare vascular tumor that occurs in adults and is associated with chronic lymphedema, irradiation, and chemical exposure [30]. The most common clinical presentation of angiosarcoma of the chest wall is after radiation therapy for breast conservation therapy as part of breast cancer treatment. Patients commonly present 5–10 years after radiation therapy, and 5-year survival after diagnosis is only 16 % [39]. Wide local excision is the only option for therapy, as most patients cannot have more radiation, and chemotherapy is usually ineffective.

Rhabdomyosarcoma

Rhabdomyosarcoma is the second most common malignant chest wall tumor in children, following Ewing Sarcoma. It is uncommon to see in adults. Rhabdomyosarcoma is an aggressive tumor with only ~10 % as being fully resectable [3]. Preoperative workup is essential and should include MRI, CT, abdominal US, and bone scan to rule out metastatic disease. Treatment includes pre- and postoperative chemo and radiation therapy in combination with surgical resection (Table 8.4).

Table 8.4 Malignant soft tissue lesions

Tumor	Origin	Presentation	Imaging characteristics	Treatment/prognosis
Soft tissue sarcomas (liposarcomas, synovial sarcomas, rhabdomyosarcomas, fibrosarcoma, neurofibrosarcoma)	Any soft tissue (adipose, synovium, muscle, fibrous tissue, nerve)	Majority of primary malignant chest wall lesions. Middle-aged men with painless mass (exception: Rhabdomyosarcoma = most common in children)	Heterogeneous appearance with varying levels of enhancement and calcification depending on the histology	Wide local excision with 4-cm margins and rib above and below for high-grade lesions. Reexcision for + margins or recurrent disease
				Most = poor prognosis with survival influenced by histology, tumor grade, tumor burden, and location
Malignant Fibrous Histiocytomas (MFH)	Fibrous tissue	Elderly men, usually with history of prior chest radiation, usually not painful, but may become large	CT: heterogeneous appearance enhancing or calcified with ill-defined contours	Neoadjuvant chemotherapy followed by wide local excision and further chemotherapy. Local recurrence >30 %. 5 years survival 38 %
Liposarcoma	Adipose tissue	Men ages 40–60 yo, may be associated with trauma		Wide local excision. 5 years survival 60 % High local recurrence rates
Neurofibrosarcomas (malignant schwannomas or malignant peripheral nerve sheath tumors)	Nerve	Associated with radiation, neurofibromitosis (29 % will develop this tumor) [3]. Men 40–50 yo, painful mass		Wide local excision and postoperative radiation. 5-years survival = 55 % [28]
Rhabdomyosarcoma	Muscle	Second most common malignant chest wall tumor in children. Uncommon in adults		Pre- and postoperative neoadjuvant chemo and radiotherapy combined with surgical resection. Aggressive tumors with only 10 % fully resectable
Fibrosarcoma	Fibrous tissue		Heterogeneous masses on CT and MRI may have areas of necrosis and hemorrhage	Neoadjuvant chemotherapy followed by resection. Postoperative radiation used if margins +. Likely to have local recurrence and/or metastasize
Angiosarcoma	Vascular	Associated with chronic lymphedema, irradiation, and chemical exposure. Most commonly 5–10 years after radiation tx for breast cancer		Wide local excision. 5-years survival 16 %
				Not candidates for further radiation. Chemotherapy is ineffective

Radiation-Associated Malignant Tumors and Metastatic Disease

Radiation-associated malignant tumors of the chest wall are uncommon but not rare [1]. Cancers of the breast and lung or lymphomas are common indications for radiation to the chest. In a large series from Memorial Sloan-Kettering Cancer Center, in 361 patients, 21 (6%) chest wall tumors arose in patients with a history of radiation to the chest. These patients were all treated with resection and had similar survival as that of patients with tumors arising de novo [40].

Not all chest wall tumors are primary lesions. Metastatic lesions from breast, lung, or unknown primary tumors can be found in the chest wall, and the role for surgical resection is gaining clarity. In the case of unknown primaries, chest wall lesions are treated like a primary tumor with resection for tissue diagnosis and therapy [41]. For breast cancers, formal studies are still lacking, but one large series from Chicago found an increase in survival with control of a chest wall lesion with either surgery or radiation [37]. Chest wall and sternal resection for metastatic cancers is associated with relief of pain from ulceration and bleeding caused by disease recurrence (Fig. 8.5).

Pediatric Tumors

Like tumors in their adult counterparts, pediatric chest wall tumors are varied in their histology, presentation, and age of onset; however, they have their own diagnostic and therapeutic challenges [42]. Approximately 20% of chest tumors in childhood are located in the chest wall, and histologic diagnoses can range from benign to malignant and infectious to noninfectious [43]. There is also wide variability in incidence of disease by country. In underdeveloped countries, tuberculosis of the chest wall is more prevalent than the common North American malignancies to which we are more accustomed. Ewing sarcoma is the most common malignant diagnosis in children vs. chondrosarcomas in adults [44].

Because of the young age of pediatric patients, both survival data and long-term effects of treatment have to be considered in multidisciplinary treatment planning. Chemotherapy and aggressive surgical resection are the mainstays of therapy with a goal to avoid radiation therapy where possible. Preoperative chemotherapy is given to most chemosensitive tumors to allow for improved local control, less extensive surgery, and to treat micrometastatic disease. Scoliosis, restrictive pulmonary function, hypoplasia of soft tissues, and secondary tumors are some of the long-term sequelae that can be seen in children after treatment of a chest wall tumor [44].

Surgical Management

Chest wall resection is the primary treatment modality for chest wall tumors and can be performed with low morbidity and mortality. If tumors are chemosensitive, preoperative chemotherapy should be administered to reduce the tumor burden. Osteosarcoma, rhabdomyosarcoma, Ewing sarcoma, and other small-cell sarcomas should be treated with chemotherapy in a neoadjuvant setting, and then continued postoperatively depending on tumor response. Chondrosarcomas and other adult soft tissue sarcomas are typically excised surgically and irradiated if negative margins cannot be achieved. Resection can prolong survival and provide palliation for symptomatic lesions [45].

Extent of tumor location and invasion may influence preoperative planning and intraoperative approach. Depending on the extent of pulmonary involvement, a double lumen endotracheal tube may be required to allow for concurrent pulmonary resection. In one review of 25 years of data, 34% of chest wall tumors infiltrated into the lung, requiring associated lung resection [12]. Placing a thoracoscope in the chest, away from the lesion, may be useful to determine extent of pulmonary involvement and exact location when the lesion is not palpable. When necessary, a needle can be inserted into the chest under thoracoscopic guidance to aid in operative planning. Appropriate margins are dependent on the histology of the tumor and are a key predictor of recurrence-free survival [46]. For aggressive malignancies that can spread along the periosteum, the entire rib should be resected with costal articulations either posteriorly or anteriorly depending on tumor location. Sections of ribs above and below the tumor

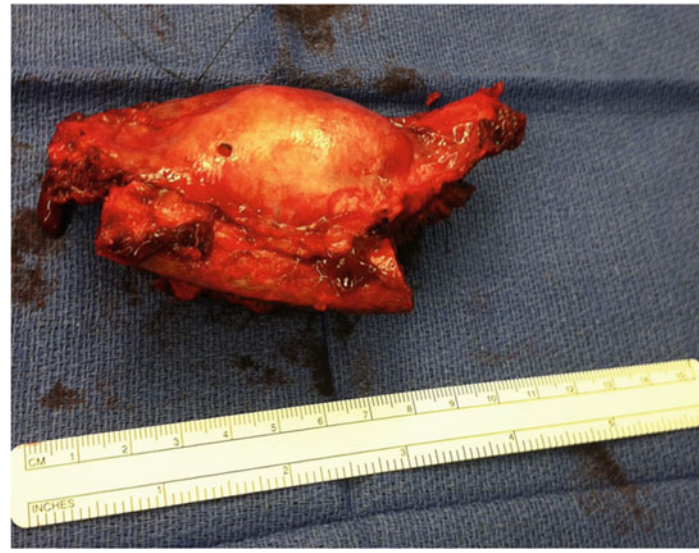

Fig. 8.5 Resected metastatic colon cancer to rib

should be resected as well. For high-grade malignancies, 4-cm margins are adequate, and low-grade malignancies can be treated with 1–2 cm margins. Desmoid tumors are not technically malignant, but their behavior is so aggressive that one should use 4-cm margins for these lesions when possible. Any involved soft tissue, skin, underlying pleura, or lung tissue should be resected with the tumor, provided pulmonary function permits resection.

Adequate oncologic resection should not be compromised for concern over chest wall defect; however, the integrity of the chest wall should be maintained to avoid pulmonary compromise.

Reconstruction

Chest wall reconstruction after large en bloc resection is often accomplished as part of a multidisciplinary team in conjunction with plastic surgery. The choice of material and method of reconstruction largely depends on anatomic location and surgeon preference. Goals of reconstructive therapy include preserving respiratory and body mechanics, visceral protection, and cosmetic outcome.

The main tenants of chest wall reconstruction are as follows [2]:

1. Defects less than 4–5 cm typically do not require reconstruction.
2. Posterior defects covered by the scapula do not require reconstruction.
3. Defects located at the scapular tip must be reconstructed to prevent the scapula from becoming trapped within the defect.
4. Skeletal stabilization is achieved with autologous tissue, mesh, Gore-Tex (W.L. gore and Associates, Inc. Flagstaff, AZ) or methyl methacrylate "sandwich" reconstruction.
5. Soft tissue reconstruction can be performed using myocutaneous or omental flaps.

Autologous tissue flaps can be constructed from a variety of soft tissue donor sites, including: the Latissimus dorsi, Pectoralis Major, Rectus Abdominal muscle flap (VRAM or TRAM), External obliques, Trapezius, or Omentum flap. The specifics and details of these reconstructions are beyond the scope of this chapter. As mentioned previously, it is important to keep in mind location of the defect, locations, and types of prior

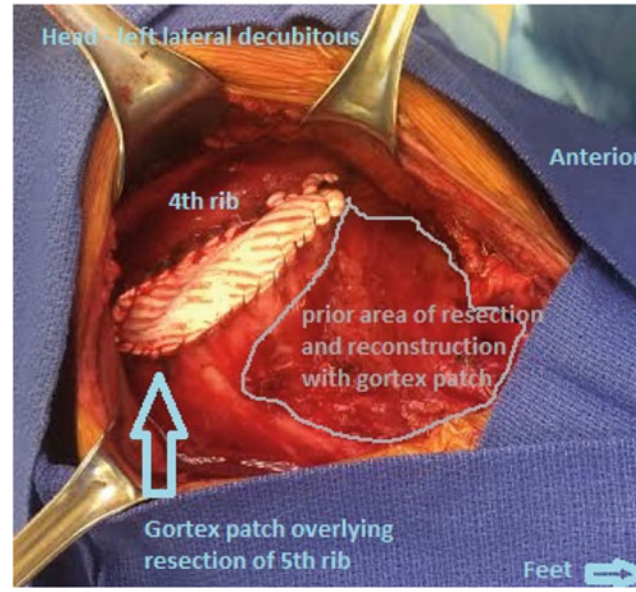

Fig. 8.6 Chest wall reconstruction of the fifth rib space using a gortex patch in a patient with prior chest wall resection and gortex patch reconstruction for desmoid tumor of the chest wall

surgical procedures, prior oncologic history and treatment, or any prior areas of radiation when choosing a suitable donor site for soft tissue reconstruction (Fig. 8.6).

Reconstruction in pediatric patients can be challenging given ongoing growth of the chest wall and intrathoracic components. Large resections and reconstructions often leave chest wall asymmetry despite reconstructive efforts. This asymmetry can become more pronounced as the child grows and may lead to the development of skeletal deformities such as scoliosis [15]. Children should continue to undergo long-term follow-up while growing, with specific attention to chest wall development and associated bony abnormalities including scoliosis screening.

Postoperative Management

In the immediate postoperative period, use of epidural catheters or localized infusion catheters for pain control can minimize postoperative morbidity and mortality. Largely because of increased ability to engage in and adequately perform appropriate pulmonary hygiene. Postoperative pulmonary hygiene is vitally important because large anterior defects can result in weak cough and inability to clear secretions, leading to debilitating pneumonia. Bronchoscopy may be necessary to facilitate adequate pulmonary hygiene in these patients.

Surveillance

Duration and frequency of postoperative surveillance will vary depending on age of the patient, tumor type, presentation, stage, response to treatment, and ability to obtain clean margins at the time of surgical resection.

Summary

Chest wall tumors are a heterogeneous group of lesions that provide an interesting diagnostic and therapeutic challenge for surgeons. Careful preoperative evaluation of the patient, radiographic imaging, and histopathology are required. In general, treatment is wide local excision, with margins for malignant disease being wider (4-cm margins when possible), and adjuvant radiation is typically given in the event of positive margins. Local control is the most important prognostic

factor, with disease-free survival for malignant disease being limited by positive margins. Chemotherapy is rarely effective, but may be helpful as neoadjuvant therapy prior to resection in the event of large tumor burden. For advanced disease or lesions that may result in significant functional loss, multidisciplinary preoperative planning is indicated, involving thoracic surgery, plastic surgery, neurosurgery, radiation medicine, oncology, and physical medicine and rehabilitation. Excellent outcomes for patients with benign, primary malignant, and metastatic lesions of the chest wall can be obtained with complete surgical resection and appropriate reconstruction.

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