Tumors of the Chest Wall

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37.1 Introduction and Epidemiology

Chest wall tumors (CWTs) in children comprise a wide range of benign and malignant neoplasms. These tumors are rare in infants, children and young adults, accounting for 1.8% of solid tumors [1]. However, high proportions of CWTs are malignant, and represent a diagnostic and therapeutic challenge to pediatric thoracic surgeons. The chest wall can be affected by: primary malignant neoplasms arising from different structures; secondary tumors which can invade the chest wall from the adjacent structures (e.g., breast, pleura, mediastinum, lung); metastatic tumors [2].

37.2 Etiology and Pathogenesis

CWTs in children are rare tumors and little is known about their etiopathogenesis. CWTs are classified as "benign" or "malignant" based on behavior, tumor type, and tissue of origin. CWTs are primarily of mesenchymal origin and comprise a broad spectrum of lesions arising from the skeletal or soft tissues of the chest wall. Among malignant tumors, soft-tissue tumors of the chest wall are more common than skeletal bony tumors. More than half of malignant CWTs are soft-tissue sarcomas whose histological diagnosis, total resection and local control often represent a challenge [3]. The most commonly encountered primary CWTs in children are small, round-cell tumors, and include Ewing's sarcoma (which is also known as primitive neuroectodermal tumor, PNET, and Askin's tumor) [4]. The category of these tumors is so named due to the lack of distinction among these lesions with regard to their neuroectodermal differentiation. After the pioneering work of Shamberger et al, these tumors are also classified as a single entity: malignant small round cell tumors [5]. In this group are also included the less common chondrosarcoma, malignant fibrous histiocytoma, osteosarcoma, synovial sarcoma and fibrosarcoma.

Benign tumors are less common in most reported series, though they may be under-reported. Benign tumors such as eosinophilic granuloma, aneurysmal bone cyst, hamartoma, osteoma, osteochondroma, chondroma as well as metastatic tumors such as osteogenic sarcoma, neuroblastoma and Wilms' tumor, are rarer than primary malignant tumors. Complete resection is often the only method of oncological management in many of these tumors.

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Furthermore, reconstruction of the wide chest-wall defects in a child is a surgical challenge.

37.3 Clinical Features

Because of their low incidence, the time between symptom onset and the diagnosis of a CWT is often long [6]. The first symptom is often a palpable, enlarging mass (Fig. 37.1). Diagnosis of a CWT due to an incidental discovery on imaging as part of screening or for examination of an unrelated condition is less common. Pain is a common symptom of masses originating from the bone (benign and malign) due to their growth and periosteal damage, whereas soft-tissue neoplasms are often painless.

Neurological symptoms such as paresthesias and weakness, can be present if the growth of the mass involves neurological structures such as the spinal cord or brachial plexus. If eosinophilic granuloma and Ewing's sarcoma are also present, the systemic symptoms of fever, malaise, fatigue, and weight loss can also be observed.

There are no specific signs or symptoms that can be used to discriminate between benign and malignant neoplasms. Rapid growth, involvement of surrounding structures, fixation to underlying tissues and cortical destruction are indirect and non-specific signs of malignancy. Although clinicians often associate pain with malignant CWTs, pain is not a reliable predictor of malignancy, so the diagnosis must be arrived at very carefully [6].

37.4 Diagnosis

Accurate history-taking and a complete physical examination represent the first steps of the work-up. They provide important information on the location, size and features of the neoplasm. The rapidity of the growth of the mass, pain, local inflammation/infection signs, and neurological or systemic symptoms and/or signs must be evaluated carefully. Based on this information, imaging studies should be planned to define the: effect of the lesion on bone; response of bone to the tumor; characteristics and composition of the matrix and cortex of the tumor; evidence of a soft-tissue mass [7]. Frequently, the first imaging examinations are plain radiographs of the chest: they can reveal bony erosion of the lesion, lytic lesions, mediastinal lymphadenopathy or invasion, and the presence of large pulmonary metastases. Ultrasound echography can help in distinguish solid from cystic tissue hamar-

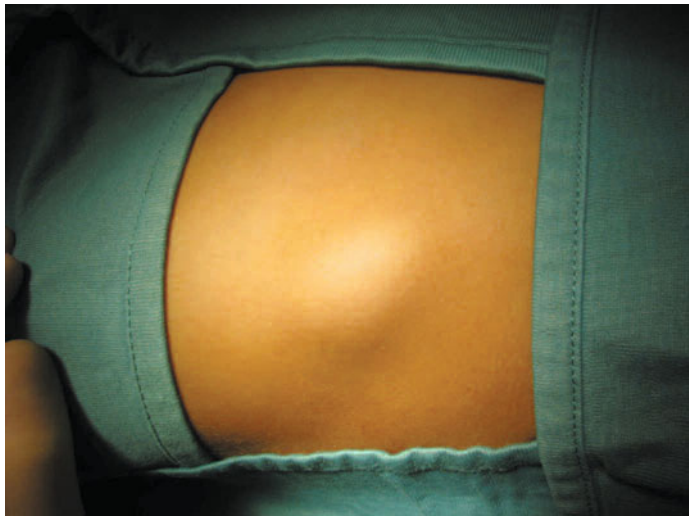

Fig. 37.1 Swelling of left side of the rib due to a benign lesion on the chest wall

tomas, hemangiomatous and lymphagiomatous lesions in superficial tumors. Computed tomography (CT) and magnetic resonance imaging (MRI) are considered essential for defining the mass and surgical planning. Chest CT can be used to assess the extent of bone, soft tissue, pleural and mediastinal involvement, as well as pulmonary metastases. It is also more sensitive than plain radiographs for defining calcification of the tumor matrix or bony cortical destruction [8].

MRI is more accurate than CT for defining soft tissues, vascular and nerve involvement, and spinal cord or epidural extension [9]. Typical malignant signs are destruction of bone and a "sunburst" pattern, whereas benign bony masses are generally smaller with distinct geographic margins. Malignant neoplasms are frequently deep to the fascia and look dark on T1-weighted MRI and bright on T2-weighted MRI. Benign soft tissue tumors are frequently small and superficial, and some benign tumors have classic features on MRI. Bone scintigraphy can be done to exclude bony metastases. Though imaging characteristics can be suggestive, a biopsy is often necessary for a definitive diagnosis of many soft-tissue tumors.

Many factors, such as the site, size, and type of lesion should be considered in the choice of the type of biopsy. Fine-needle aspiration (FNA) biopsy is a relatively inexpensive method with a low prevalence of complications and high diagnostic value for distinguishing between malignant and non-malignant neoplasms. In metastatic and primary bone tumors, FNA biopsy is quite easy to carry out and often accurate tissue diagnosis can be achieved. Unlike open biopsy, FNA does not require surgical incisions, so this procedure should be considered in the triage of bony lesions due to its low risk or morbidity [10]. False-positive results have major therapeutic implications; this is usually due to inadequate sampling or misclassification with regard to the exact subtype of malignant tumor. Nevertheless, the advantages must be balanced against the limitations of this procedure. Excision biopsy is advisable for all rib tumors and small lesions that can be removed without the need for the rebuild of bony structures. Incision or Tru-Cut® needle biopsy is recommended for soft-tissue and unresectable masses. An appropriate biopsy incision is important because the tumor may be resected later. Biopsy specimens should be taken from the peripheral and inner core of the mass to be analyzed for histological, cytogenetic and biological studies.

37.5 Therapeutic Management

37.5.1 Benign Soft-Tissue CWTs

Mesenchymal benign tumors include a heterogeneous group of soft-tissue lesions. In particular, the myofibroblast is the main cell in many fibromatoses tumors [11]. In this group of lesions, the most common neoplasm of early infancy is infantile myofibromatosis. In one study, 15% of patients with infantile myofibromatosis had multiple lesions. [12, 13]. The tumors are mainly present at birth or develop during the first week of life. They can be solitary or multicentric. Most of these tumors undergo spontaneous regression and are managed conservatively [14]. Notably, children with multicentric infantile myofibromatosis with visceral involvement are associated with a higher prevalence of morbidity and mortality.

The second most common lesion in this group is desmoid fibromatosis tumor. These tumors are slow-growing, bland fibrous neoplasms originating from the musculoaponeurotic structures throughout the body. The term desmoid, coined by Müller in 1838, is derived from the Greek word *desmos,* which means "tendon-like". Desmoid fibromatosis tumors often appear as infiltrative, usually well-differentiated, firm overgrowths of fibrous tissue. They arise from the muscle and fascia and extend along the tissue planes with aggressive behavior. The phrase "aggressive fibromatosis" describes their marked cellularity. This disease course and the tendency for recurrence makes the treatment of these relatively rare fibrous tumors challenging. These tumors typi-

cally present in the third decade, but 20–30% of cases have been reported in children, mostly in girls (probably due to the influence of oestrogen) [15]. Although considered benign, these neoplasms are often aggressive with multiple recurrences after incomplete resection. Hence, resection with negative margins is required to prevent local recurrence because they have local recurrence rates of ≤70% [6, 16]. Furthermore, they have been seen in association with Gardner's syndrome (mutation of the adenomatous polyposis coli gene) and in the scars of previous thoracotomies [9]. About 33% of patients with Gardner's syndrome develop desmoid fibromatosis tumors, whereas only 2% of children with desmoid fibromatosis tumor have a diagnosis of Gardner's syndrome [17]. Aggressive fibromatoses describes a more extensive infiltrative disease of fibrous scar tissue, and it has been treated with wide excision or, if unresectable, chemotherapy and radiation, without reducing the rate of recurrence [18].

Lipoblastomatosis is an uncommon benign tumor of brown fat that may be observed in childhood or infancy. It is diffuse lobulated, spreads along the tissue planes and typically recurs [19]. Lipoblastomatosis cannot be distinguished clinically from lymphangioma because they have a similar consistency. The differentiation between water-filled and solid soft-tissue tumors can be obtained by ultrasonography. Study of these masses should be completed with MRI to define the extension of the tumor and its relationship with surrounding tissues. After complete excision of the mass, the tissue should be sent fresh for histological and cytogenetic evaluation that allows differentiation between lipoblastomatosis and liposarcoma [20].

Giant-cell fibroblastoma is a rare type of soft tissue tumor characterized by painless nodules (usually 2–6 cm in diameter) in the dermis (the inner layer of the two main layers of tissue that comprise the skin) and subcutaneous (beneath the skin) tissue. It is a solitary, blue–gray, non-tender mass which is mostly located on the back, anterior chest wall, thigh, or groin. These tumors may come back after surgery but do not spread to other parts of the body. It is of important to distinguish giant-cell fibroblastomas from sarcomas to avoid inappropriately aggressive treatment [21].

In addition, a great number of hamartomatous lesions (e.g., lymphangioma, hemangioma, lipoblastoma, fibroblatic tumors) which involve the chest wall have been described in early infancy and childhood. These lesions are not described here because their presentation and management is not different from to those present elsewhere in the body.

37.5.2 Benign Bony CWTs

The chest wall can be affected by a wide group of benign bony and primary lesions that should be distinguished from non-neoplastic conditions such as cysts, infections, and fibromatoses because they require different management.

The most frequently observed benign neoplasm of the skeleton is osteochondroma. This tumor contains bone and cartilage and usually occurs near the end of a long bone. It accounts for almost half of all rib tumors [22]. This tumor takes the form of a cartilage-capped bony spur or outgrowth on the surface of the bone and frequently grows during the "pubertal spurt" until skeletal maturity. The most frequent clinical features are pain and asymmetry of the chest wall ("bony protuberance"). It is sometimes referred to as "osteocartilaginous exostosis". If an exophytic bone lesion contains a cartilaginous cap >1 cm in height, or if there is associated pain, there is thought to be a higher risk for the lesion being a chondrosarcoma. After puberty, complete resection should be done in patients with painful lesions or tumors that have increased in size [23–25].

Chondromas are benign tumors composed of mature hyaline cartilage. They generally have limited growth potential and are not locally aggressive [26, 27]. These tumors are called "enchondromas" if they occur in the

medullary canal of the bone [28] and "periosteal" or "juxtacortical" chondromas if they occur on the surface of the bone. Chondromas can also arise from the synovial sheaths of tendons or in the soft tissues adjacent to the tendons in the hand and feet of adults. In such cases, they are referred to as "soft-tissue" or "synovial" chondromas. The clinical and imaging features of chondroma and chondrosarcoma are similar and are seen as expansile lesions causing thinning of the bony cortex. Given appropriate treatment, patients with benign chondromas generally have a good prognosis, and most remain asymptomatic. Enchondromas are rare in the axial skeleton. If, however, they are found in the ribs, sternum, pelvis, or scapula, they should be treated with wide local resection and histopathological evaluation to rule out the possibility of chondrosarcoma, which is common at these sites [29].

Osteoid osteoma is a benign osteogenic tumor, typically noted in childhood [30], that rarely involves the chest-wall bones. Its hallmark is a local pain, which worsens during the night, and promptly improves after the consumption of aspirin or other non-steroidal anti-inflammatory drugs. Osteoid osteoma is more frequent in males and it is generally <1.5 cm in greatest dimension. Symptomatic lesions can be resected with excellent results. Osteoblastoma is a rare, benign, bone-forming tumor of the chest wall which is described as a "sternal tumor".

Fibrous dysplasia is a cystic expansive benign neoplasm characterized by a developmental anomaly of the medullary cavity of the rib. Fibrous replacement of the medullary canal is the hallmark of this disease. It is reported to be a solitary, slow-growing non-tender mass in the posterior–lateral aspect of the rib. It is associated with Albright's syndrome in which multiple bony cysts, skin pigmentations, and precocious sexual maturity are observed. The diagnosis is often incidental after radiography. The radiographic aspect is a cystic lesion with thinning of the cortex and a central "ground glass" appearance without calcification [31]. Local excision is curative and should be done for painful enlarging lesions that cause a diagnostic dilemma; conservative management is indicated for asymptomatic lesions [32].

Eosinophilic granuloma is an expansive lesion of the rib which involves the reticuloendothelial system. It is characterized by eosinophilic and histiocytic infiltrates. This tumor is commonly seen between 5 years and 15 years of age. Three types have been reported: eosionophilic granuloma, Letterer–Siwe disease, and Hand–Schuller–Christian disease. Eosinophilic granuloma is limited only to bone involvement, without the systemic symptoms that characterize Letterer–Siwe disease and Hand–Schuller–Christian disease [33]. The systemic diseases have a chronic course and require specific management, including corticosteroids and chemotherapy. The radiographic features of this lesion are: expansile bony lesion in the posterior–lateral aspect of the rib cage; irregular destruction of the cortex of the bone seen as "scalloping"; and new periosteal bone formation. The differential diagnosis should exclude Ewing's sarcoma or osteomyelitis, which requires different management. The excision biopsy of a lesion is indicated for the diagnosis and is curative in a solitary eosinophilic granuloma.

Mesenchymal harmartoma is a typical tumor found in infant which is present at birth in 40% of cases. It is a non-neoplastic proliferation of normal skeletal elements, predominantly cartilage [34]. Malignant degeneration has not been reported [34]. These lesions are mostly intrathoracic but extra-pleural, and can involve a single or multiple ribs (usually in their posterior or lateral portion). It has a typical appearance on chest radiography. However, the intrathoracic component is best evaluated with cross-sectional imaging [35], and the easiest imaging to carry out in infants is CT. The most common presentation is respiratory distress due to the mass effect [36]. Sometimes, it can be seen by prenatal ultrasound as a large thoracic mass or pleural effusion in the fetus [37, 38]. Conservative resection should be done only to relieve symptoms, whereas extensive resection

of multiple ribs should be avoided due to the risk of severe scoliosis. Spontaneous regression has also been reported [36–39].

37.5.3 Malignant CWTs

37.5.3.1 Ewing's sarcoma/PNET

Ewing's sarcoma/PNET is the most common CWT in all pediatric series. It was described for the first time by Askin in 1979 [40]. These lesions are known as malignant small-cell sarcomatous tumors or as PNET due the origin from embryonal neural crest cells. Histological differentiation is needed with other undifferentiated, small-round-cell tumors such as undifferentiated neuroblastoma, embryonal rhabdomyosarcoma, and lymphoma [41].

Ewing's sarcoma/PNET is an extremely aggressive tumor with frequent metastatic spread and local recurrence. It should be considered to be a systemic disease at presentation [42]. The peak incidence of Ewing's sarcoma is 13–16 years, with a male:female ratio of 2:1. Localization to the chest wall accounts for up to 6.5% of cases. The first presenting symptom is increasing pain that can be associated with cough, pain fever, malaise, anemia and increased erythrocyte sedimentation rate.

Plain radiographs highlight the non-pathognomonic signs of lytic destruction and regeneration. This tumor must be differentiated from other lesions such as osteomyelitis and osteogenic sarcoma by clinical and imaging findings. CT is the best method to evaluate the bony extent of the lesion (Fig. 37.2) and small pulmonary metastases. The imaging features are a round, ovoid, multinodular or lobulated neoplasm which can be circumscribed but is rarely encapsulated. Imaging also reveals areas of hemorrhage and necrosis in large tumors. Involvement of the soft tissue of the chest wall can be defined more precisely by MRI [43].

Three histopathological patterns have been defined: compact sheets of cells; a nesting arrangement of cells with an intervening fibrovascular stroma; and serpiginous bands of cells with necrosis. In all of these small cells, almost uniform nuclei with scant cytoplasm are found. The periodic acid–Schiff (PAS) negative cell profiles are arranged around an acidophilic focus of hyaline or fibrous nature rather than a neurofibrillary composition. Cy-

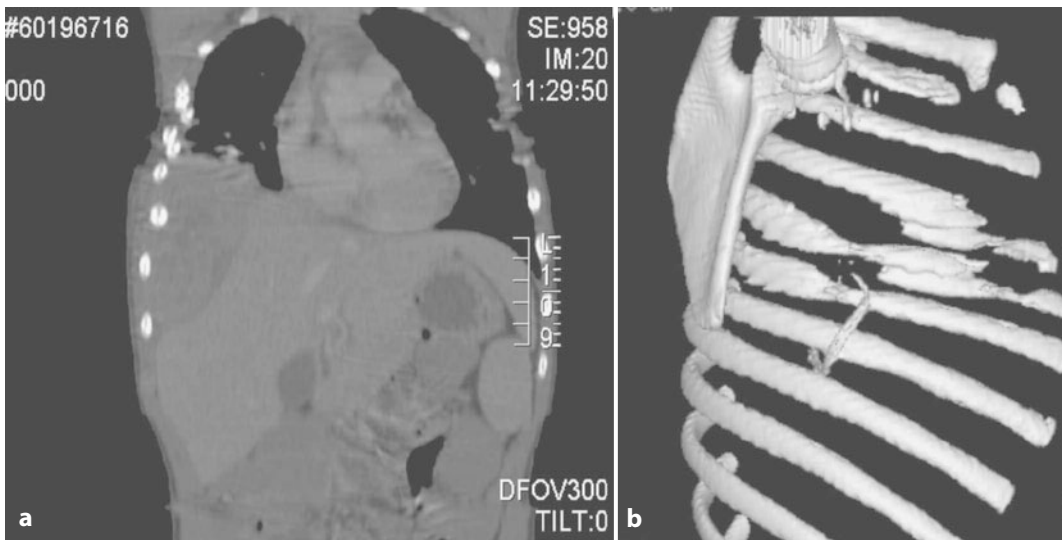

Fig 37.2 Coronal CT showing a large Ewing's sarcoma in the anterior–lateral region of the chest wall (**a**). 3D reconstruction permits the observation of the bone destruction in the ribs (**b**)

toplasmic glycogen is detected at PAS staining to distinguish Ewing sarcoma's from PNET. A characteristic balanced translocation between chromosome 11 and 12 [t(11:22)(q24;q12)] has been revealed in both tumors. The translocation points of these two tumors have now been evaluated: they are analogous [44].

These tumors should be treated using a multimodal approach (surgery, radiotherapy, chemotherapy) because they tumors have a high tendency to metastasize locally and also through hematogenous mechanisms mainly to the lungs and bones [45].

Ribs are expendable, so the whole rib from the vertebra to the costochondral junction can be removed. In lesions located laterally or interiorly on the chest wall, it is possible to leave the posterior portion of the rib. This might help in the reduction of long-term scoliosis. To achieve control of local and systemic micrometastasis, local resection of the entire involved rib with partial resections of the ribs on either side of the tumor, followed by radiotherapy on the site of the lesion, can be done. In general, the radiotherapy dose is 50–65 Gy [46]. Also, chemotherapy is employed to prevent recurrence and metastasis of the tumor [47]. Extra-osseous metastasis has also reported to be associated with an increased risk of distant metastasis and poor survival rates. Many authors have used radiotherapy after resection to improve outcome [48].

37.5.3.2 Rhabdomyosarcoma

Rhabdomyosarcoma is a typical tumor found in infancy. It accounts for 15% of all solid tumors in children, with two age peaks at 2 years to 5 years and around puberty. The prevalence in the chest accounts for 7.4% of all cases of rhabdomyosarcoma [49].

It appears as a rapidly growing mass which originates from the striated muscles of the trunk. At the beginning it is not painful, and sometimes it looks inflamed due to necrosis or hemorrhage. Hence, the diagnosis can be challenging and it should not be mistaken as an abscess.

Biopsy has the aim of confirming the diagnosis and characterizing the histological subtype of rhabdomyosarcoma. The most common type is the alveolar (which looks like pulmonary alveoli), followed by the embryonal subtype. There is also a mixed form where embryonal and alveolar components are present. The pleomorphic subtype contains large, elongated cells with many nuclei or giant nuclei, and it is rare in children.

Rhabdomyosarcoma of the chest wall is treated in a multimodal way that does not differ from the treatment in other localizations, and involves chemotherapy, surgery and radiotherapy.

Surgery has a pivotal role because complete remission cannot be achieved with chemotherapy alone in most cases, even if most tumors respond to vincristin, adriamycin and cyclophosphamide. Preoperative chemotherapy helps to reduce the size in a bulky tumor so as to achieve complete resection with a clear margin; this is fundamental because it has a significant impact on outcome. Debulking and mutilating surgery should be avoided. Radiotherapy has a role in residual, recurrent and metastatic disease.

The stage of the tumor at presentation is correlated with survival. In particular, chest-wall rhabdomyosarcoma has a poor prognosis [49-51]. Risk factors that affect the prognosis include an alveolar sarcoma subtype, advanced stage at presentation, difficulties in local resection and early relapse compared with other sites [52]. Overall five-year disease-free survival in children is 65% as reported by the results of the Third Intergroup Rhabdomyosarcoma Study. Local disease control on the chest wall is fraught with many difficulties, especially if relapse occurs. Radiotherapy is often undertaken to obtain local control of relapsing disease, but results in pulmonary fibrosis. A combination of radiotherapy and surgery may cause secondary restrictive defects that impair the development of the thoracic cavity and may cause scoliosis [53].

37.5.4 Other Tumors

Congenital infantile fibrosarcoma is present at birth, and most cases occur in the first 3 months of life. This tumor has a dense cellularity and mitotic activity but is less anaplastic than conventional fibrosarcoma and has good chemosensitivity. Hence, it must be differentiated from mesenchymal hamartoma and fibroblastomatosis. Preoperative chemotherapy makes resection possible with minimal morbidity. Resection is indicated in most instances of the residual tumor. Local recurrences have been reported with rare instances of metastasis [54].

Osteogenic sarcoma is a highly malignant bony tumour that is extremely rare in children. Osteosarcoma is a primary malignant tumor of the skeleton and is characterized by the direct formation of immature bone or osteoid tissue by tumor cells. Osteosarcoma arises predominantly in the long bones and rarely in soft tissues. Most patients present with a rapidly enlarging mass which is often reported as being painful. The age at presentation ranges from 10 years to 25 years. Plain radiographs, CT, MRI, angiography and dynamic bone scintigraphy are used for the: diagnosis; evaluation of the extent of tumour involvement; decision of the type of surgical procedure; type of reconstruction (if required). The management plan for these tumors is similar to commonly reported sites within the long bones of the limbs (i.e., incisional biopsy to confirm the diagnosis followed by chemotherapy). In the past, all patients with osteosarcoma were treated by amputation, but the cure rate was <10% and almost all patients died within 1 year from diagnosis. Today, for localized osteosarcoma at onset (80% of cases) treated in specialized Bone Tumor Centers with preoperative and postoperative chemotherapy in association with surgery, the percentage of patients cured is 60–70%. Surgery is conservative (limb salvage) in >90% of patients. The prognosis is more severe (cure rate, »30%) for tumors located in the axial skeleton and in patients with metastasis at onset.

Chondrosarcoma is a rare tumor in childhood, with a peak of incidence in men of 30–40 years [22]. It originates from the costochondral arch or the sternum on the anterior chest wall. It grows slowly, usually presenting with pain in a previously asymptomatic lesion with frequent local recurrence and a high risk of late metastasis. Well-differentiated chondrosarcoma is indistinguishable from chondroma. Given that misdiagnosis as chondroma is not uncommon in differentiated chondrosarcoma and results in inadequate excision, an accurate histological diagnosis is important. Plain radiographs show the tumor destroying the cortex (causing a mottled-type calcification within the tumor) whereas the tumor edges cannot be defined. CT gives better definition of the extent of the tumor, which is relevant for preoperative management to plan the resection biopsy (rather than incisional or needle biopsies). Chondrosarcoma does not respond to chemotherapy or radiotherapy, and an inadequate excision can be the cause of local recurrence. Therefore, the outcome of these patients is related to an initial wide excision, which results in cure as well as preventing local recurrences and distant metastasis [23]. Sometimes, complete excision with free margins cannot be done due to the size and location of the mass [52].

37.6 Outcome and Follow-up

The aim of chest-wall surgery is complete elimination of the local tumor with restoration of adequate protection of the thoracic organs without impairment of physiological functions. This will enable adequate growth in the lung and chest wall and an acceptable cosmetic result [55].

Given that most CWTs are treated primarily with resection, a correct surgical indication should be based on evaluation of the histology, location, degree of local invasion, and metastases of the tumor. A standard thoracotomy incision allows palpation of the lesion and enables a biopsy to be taken or resection to be

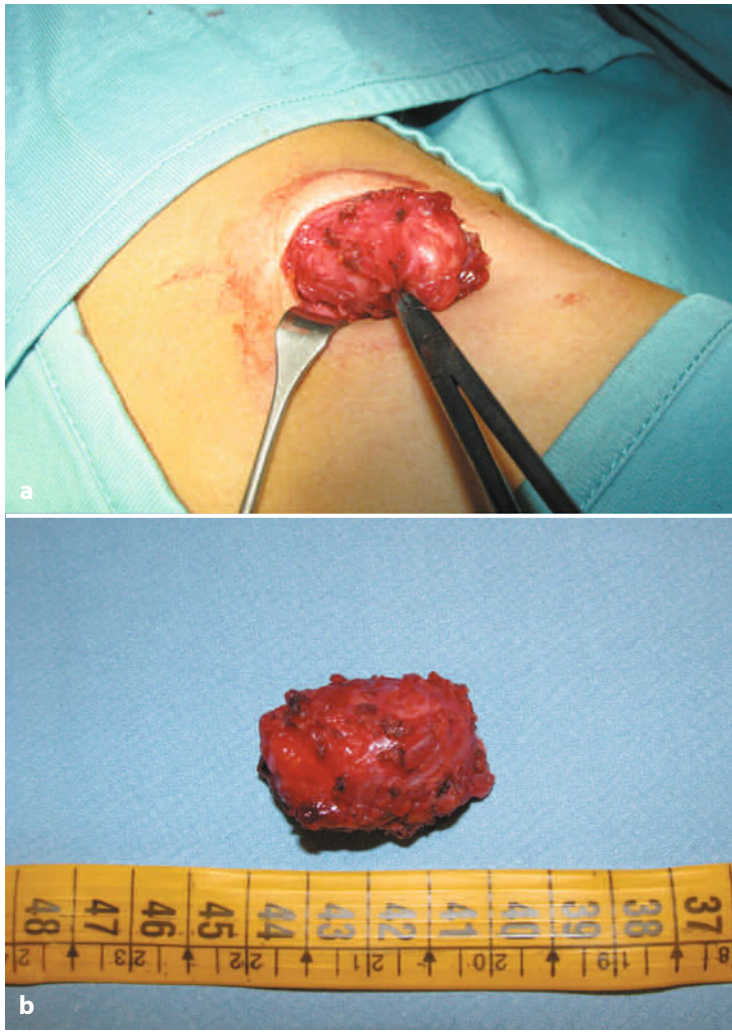

Fig. 37.3 Intraoperative image (**a**) of a standard thoracotomy in a child with rhabdomyosarcoma (**b**) of the chest wall

carried out (Fig. 37.3). Video-assisted thoracoscopic surgery may facilitate localization of the mass if it is detectable from the pleural surface. If the lesion is small and cannot be evaluated visually or by palpation intraoperative localization and resection can be done. This is achieved by placement of a coil wire by interventional radiology or injection of methylene blue into the surrounding tissues under imaging guidance.

Aiming to prevent local recurrence, resection of all tumors must ensure negative margins (even if the margin size depends on the specific tumor type). Benign tumors can be excised with negative margins, whereas a wide excision with a minimum 4-cm margin is required for malignant tumors. Thoracic and reconstructive surgeons are involved in the choice of reconstruction, which depends on the experience and preferences of the center.

Scoliosis is a well-known complication of chest-wall surgery. The severity of the curve is directly related to the number of ribs involved in the resection. In addition, the risk factors for severe scoliosis are resection of posterior segments or lower ribs and irradiation of the spinal column in patients affected by malignant tumors [56, 57]. The convexity of the

curve is towards the resected side. Scoliosis can be progressive until the child is fully grown, so it must be evaluated carefully in the long-term follow-up. This may require further surgical correction of scoliosis with insertion of a Harrington rod or the use of rib-expansion devices.

Severe scoliosis is also a risk factor for secondary impairment of pulmonary function [51] Thus, periodic evaluation of forced vital capacity, forced expiratory volume in one second, and functional residual capacities should be included in the follow-up of patients who undergo extensive chest-wall surgery.

A risk of 20% of secondary malignancy (including secondary sarcomas, acute myeloblastic and lymphoblastic leukemias) [58] has been reported. These tumors are most common within the first 5 years after the diagnosis in patients treated for Ewing's sarcoma/PNET [59–61]. Irradiation with a cumulative dose >60 Gy [62] and a chemotherapy regimen including alkylating agents are common risk factors [63, 64]. A typical complication is wound infection, in which the risk factors are tumor ulceration and use of omentum in soft-tissue reconstruction [65].

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