Tumors of the Lung and Chest Wall

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Primary pulmonary tumors of the lung are infrequent in infants and children; the majority of pulmonary neoplasms in children are due to metastatic disease. The approximate ratio of primary pulmonary tumors to metastatic neoplasms and non-neoplastic lesions of the lung is 1:5:60 [98]. Although primary pulmonary tumors are rare in children, the majority are malignant. In a review of 383 primary pulmonary neoplasms in children by Hancock et al. [33], 76 % were malignant and 24 % were benign. This incidence is similar to that previously reported by Hartman and Shochat and Weldon and Shamberger [35]. Table 26.1 demonstrates the spectrum of primary pulmonary neoplasms seen in children. This chapter presents the more common benign and malignant primary pulmonary tumors and discusses the management of pulmonary metastatic disease in the pediatric population.

Lung Tumors: Benign

Inflammatory Myofibroblastic Tumor (Plasma Cell Granuloma)

Inflammatory myofibroblastic tumor (IMT) has also been called inflammatory pseudotumor, histiocytoma, and fibrohistiocytoma [46]. This lesion, which is seen frequently in adults, occurs rarely in children younger than 10 years (approximately 8 % of all cases of IMT). However, IMT is the most common benign tumor in children and accounts for slightly more than 50 % of all benign lesions and approximately 20 % of all primary lung tumors [35]. These tumors usually present as peripheral pulmonary masses, but occasionally present as polypoid endobronchial tumors [2, 6]. The pathogenesis of IMT is not well understood, but an antecedent pulmonary infection has been reported in approximately 30 % of cases. The mean age at presentation in children is 7 years, and 35 % of the children are between 1 and 15 years of age (Fig. 26.1) [2, 6, 96]. Many children are asymptomatic at the time of presentation, but fever, cough, pain, hemoptysis, pneumonitis, and dysphagia may be present. The natural history is that of a slow-growing mass starting as a focus of organized pneumonia

Table 26.1 Primary pulmonary neoplasms in children

Type of tumor	No. of patients (%)[a]
Benign (n = 92)	
Plasma cell granuloma	48 (52.2)
Hamartoma	22 (23.9)
Neurogenic tumor	9 (9.8)
Leiomyoma	6 (6.5)
Mucous gland adenoma	3 (3.3)
Myoblastoma	3 (3.3)
Benign teratoma	1 (1.1)
Malignant (n = 291)	
Bronchial "adenoma"	118 (40.5)
Bronchioalveolar carcinoma	49 (16.8)
Pulmonary blastoma	45 (15.5)
Fibrosarcoma	28 (9.6)
Rhabdomyosarcoma	17 (5.8)
Leiomyosarcoma	11 (3.8)
Sarcoma	6 (2.1)
Hemangiopericytoma	4 (1.4)
Plasmacytoma	4 (1.4)
Lymphoma	3 (1.0)
Teratoma	3 (1.0)
Mesenchymoma	2 (1.7)
Myxosarcoma	1 (0.3)

Modified from Hancock et al. [33]
[a]Percent of benign or malignant tumors

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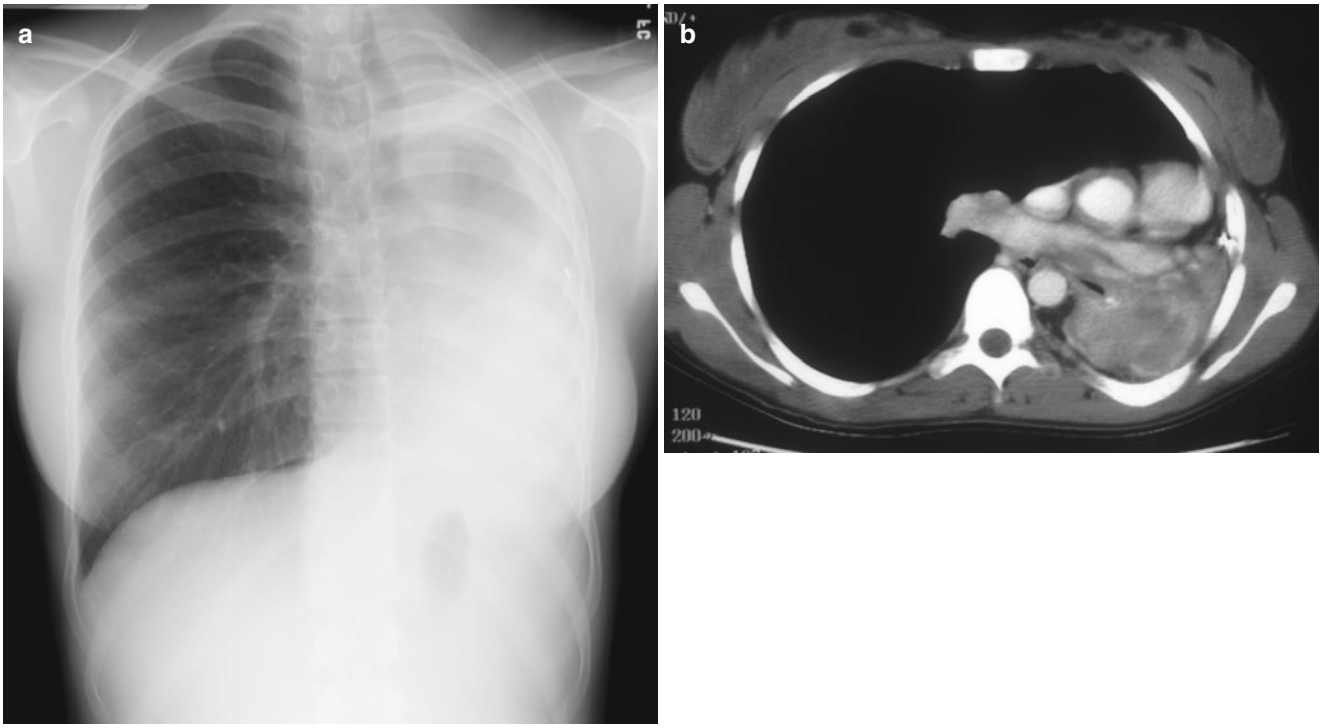

Fig. 26.1 (**a**) Chest radiograph of an 18 year old female who presented with a 6-month history of increasing cough and shortness of breath and recent hemoptysis. Study revealed complete opacification of the left chest. (**b**) Subsequent computed tomography (CT scan) demonstrated occlusion of the mainstem bronchus and a calcified mass in the lung. Pathology on the left lung demonstrated inflammatory myofibroblastic tumor treated with a pneumonectomy

with a tendency for local invasion. However, rare cases of rapid growth have been reported [100]. Extension of the tumor beyond the confines of the lung is common. At least four deaths have been reported due to tracheal obstruction or involvement of the mediastinum by massive lesions.

Treatment consists of a conservative pulmonary resection with removal of all gross disease if possible. Primary hilar adenopathy may be present, and local invasion with disregard for tissue planes mimics malignancy. A frequent problem is establishing the benign nature of the lesion prior to resection. Malignant fibrous histiocytoma of the lung, an extremely rare tumor in children, can mimic IMT radiographically and must be considered in the differential diagnosis [66]. Recurrences following resection occur if the lesion is incompletely resected. Nonsteroidal anti-inflammatory drugs have been used to treat large inoperable lesions, with encouraging results [88].

Hamartoma

Pulmonary hamartoma is the second most frequent benign lesion seen in children. This lesion usually presents as a parenchymal lesion which can be quite large. Approximately one quarter are calcified, and "popcorn-like" calcification is pathognomonic [23]. Two endobronchial lesions have been reported. Four tumors occurring in the neonatal period were quite large and were associated with significant respiratory distress; all were fatal. An interesting association seen in Carney triad is the combination of pulmonary hamartoma, extra-adrenal paraganglioma, and gastrointestinal smooth muscle tumors (GIST); the majority of these patients are young women. The tumors seen in this triad have an unpredictable but often indolent biologic behavior [101]. Conservative pulmonary resection is the treatment of choice; however, lobectomy or even pneumonectomy may be required, especially for large lesions and endobronchial lesions when sleeve resection is not possible or distal parenchyma has been destroyed by chronic obstruction.

Lung Tumors: Malignant

Bronchial Adenoma

Bronchial adenoma is the most frequently encountered primary malignant pulmonary tumor. These are a heterogeneous group of primarily endobronchial lesions. Although adenoma implies a benign process, all varieties of bronchial adenomas occasionally display malignant behavior. There are three histologic types: carcinoid tumor (most common), mucoepidermoid carcinoma, and adenoid cystic carcinoma.

Carcinoid tumors account for 80–85 % of all bronchial adenomas in children [74]. The presenting symptoms are due to bronchial obstruction: cough, recurrent pneumonitis, and hemoptysis (Fig. 26.2a, b). Symptoms are often present for months due to the rarity of this lesion and diagnostic challenges; occasionally, children with wheezing have been treated for asthma, delaying diagnosis as long as 4–5 years. Metastatic lesions are reported in approximately 6 % of cases, and recurrences occur in 2 %. Bronchial lymphadenopathy is often related to chronic inflammation and not to metastatic disease. There is a single report of a child with a carcinoid tumor and metastatic disease who developed the classic carcinoid syndrome [52]. Bronchial adenomas of all histologic types are associated with an excellent prognosis in children, even in the presence of local invasion [86].

Endobronchial biopsy may be hazardous because of the risk of hemorrhage, and endoscopic resection is not recommended due to the risk of tumor remnants in the wall of the bronchus. Bronchography or computed tomography (CT) may be helpful to determine the degree of bronchiectasis distal to the obstruction, because the degree of pulmonary destruction may influence surgical therapy (Fig. 26.2c, d) [3]. However, Tagge et al. [90] described a technique for pulmonary salvage despite significant distal atelectasis. Conservative pulmonary resection with removal of the involved lymphatics is the treatment of choice. Sleeve segmental bronchial resection is possible in children and is the treatment of choice when feasible [27, 45, 75]. Adenoid cystic carcinomas (cylindroma) have a tendency to spread submucosally, and late local recurrence or dissemination has been reported. In addition to *en bloc* resection with hilar lymphadenectomy, an intraoperative examination of the bronchial margins should be carried out if the margins are close to avoid leaving residual tumor. Mucoepidermoid carcinoma of the bronchus has also been described in children as young as 4 years [81], and they are defined as low- or high-grade lesions.

Bronchogenic Carcinoma

Although bronchogenic carcinoma is rare in children, it was the second most common malignant lesion reported by Hancock et al. [33] Interestingly, squamous cell carcinoma was rare, with the majority of tumors being either undifferentiated carcinoma or adenocarcinoma. The term bronchioalveolar carcinoma has been used in most cases [65]. These tumors are associated with both cystic adenomatoid malformations and intrapulmonary bronchogenic cysts. Survivors are rare as mortality exceeds 90 %. The majority of children present with extensive local involvement and/or disseminated disease, and the average survival is only 7 months (Fig. 26.3). Complete resection, followed by adjuvant chemotherapy is recommended for the rare localized lesion.

Pulmonary Blastoma

Pulmonary blastoma is a rare malignant tumor that arises from mesenchymal blastema. This tumor is an aggressive lesion, with metastatic disease at presentation in approximately 20 % of cases [20, 33]. They may arise from the lung, pleura, and mediastinum [69]. These tumors are classified into three types: Type I (purely cystic), Type II (cystic and solid), and Type III (completely solid) [26]. Type I tumors are difficult to distinguish radiographically from cystic adenomatoid malformation (Fig. 26.4a, b) [40]. Occasionally, they may arise in an extralobar sequestration or lung cyst. The majority of cases occur in the right hemithorax. Frequent sites of metastases are the liver, brain, and spinal cord. Local recurrence is frequent, and the mortality rate is approximately 40 % [17, 33, 44]. The majority of children present before 4 years of age, and symptoms include persistent cough, chest pain, episodes of pneumonia that are refractory to antibiotics, and hemoptysis. Diagnosis is established by CT of the chest, bronchoscopy, and biopsy. Because most are located peripherally, resection is usually possible by segmental or lobar resection. The use of multimodal neoadjuvant chemotherapy and radiation following surgical resection has shown promising results in a few patients with extensive disease and dissemination [44, 69]. Dramatic response of the primary may facilitate resection. Chemotherapeutic agents that have been used include actinomycin D, vincristine, cyclophosphamide alternating with courses of doxorubicin, and cisplatin. Histologic evaluation of the tumor shows an exclusive mesenchymal composition, including primitive tubules, immature blastema, and spindle cell stroma. Some demonstrate elements of embryonal rhabdomyosarcoma arising within a multicystic lesion.

An important consideration is the association of primary lung tumors with congenital cystic pulmonary malformations. These lesions may be asymptomatic and be discovered incidentally. In some instances, the natural history of the lung cyst is unknown, and a few may regress [51]. Although some authors recommend simple observation, most pediatric surgeons argue against prolonged observation of cystic lesions because of an increased risk of infection, pneumothorax, sudden cyst enlargement with potential respiratory compromise, and associated malignancy [1, 16, 51, 60, 68, 80, 89]. As mentioned above there is evidence suggesting a relationship between type IV cystic adenomatoid malformation and type I pulmonary blastoma. While complete lobectomy with negative margins is adequate treatment for these patients, close observation is recommended [39, 58, 73]. If patients with asymptomatic cystic malformations are observed without resection, they should be followed closely and evaluated frequently.

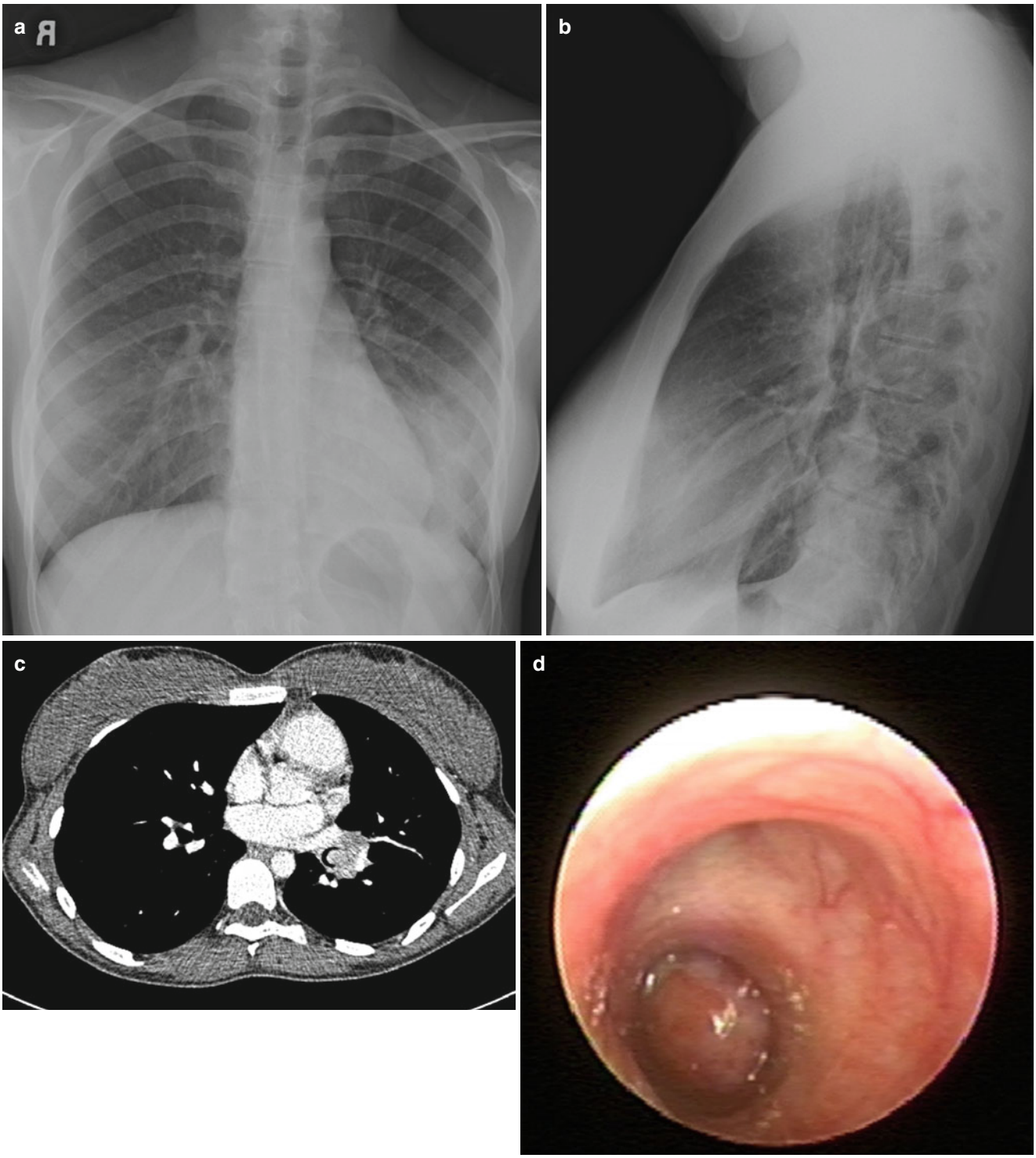

Fig. 26.2 (**a**) This 16-year old female who presented with a pneumonia and pleural effusion which was treated with catheter drainage and antibiotics. Follow-up chest radiographs (**a**, **b**), however, continued to show collapse of the left lower lobe for over a year which led to a CT scan (**c**). This demonstrated an endobronchial lesion extending into the left bronchus intermedius and occluding it with complete collapse of the left lower lobe. Bronchoscopy (**d**) localized the lesion to just distal to the origin of the upper lobe and lingular bronchi

Rhabdomyosarcoma

Rhabdomyosarcoma of the lung is rare and accounts for only 0.5 % of all childhood rhabdomyosarcomas [94]. Many of the lesions are endo-bronchial in origin. This is an important issue because 4 % of benign tumors and 8.6 % of malignant tumors enumerated in Table 26.1 were associated with previously documented cystic malformations [33]. Tumors that developed in these malformations included 11 sarcomas, 9 pulmonary blastomas, 3 bronchogenic carcinomas, and 2 mesenchymomas.

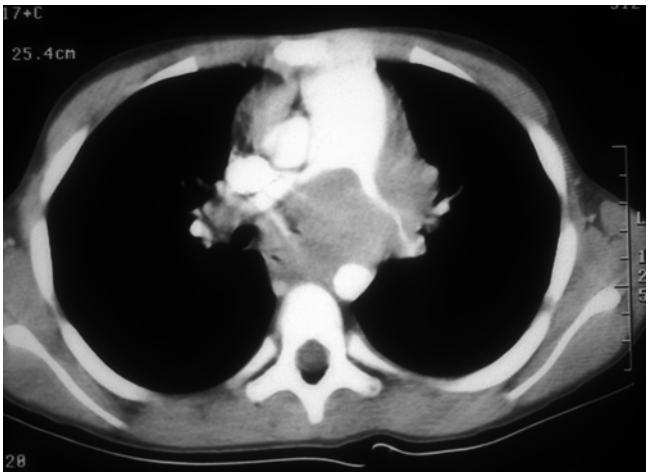

Fig. 26.3 This CT scan of an 8 year old boy who presented with a cough shows a large lesion arising in the hilum of the lung and extending into the mediastinum surrounding the left pulmonary artery. Needle biopsy demonstrated a bronchogenic carcinoma which had a rapidly progressive course despite intensive chemotherapy

Treatment of Metastatic Disease

The histology of the primary tumor and its response to combined-modality therapy govern the management of the pulmonary metastases [48]. Pulmonary metastases should not be resected until the primary tumor is eradicated and other sites of metastatic disease are excluded. Pulmonary metastasectomy is considered most frequently for osteosarcoma [47].

Osteosarcoma

Children with osteosarcoma should be considered for resection of pulmonary metastases once the primary lesion is controlled. The overall disease-free survival is approximately 40 % in children who develop metachronous pulmonary metastases. Multiple factors, such as number of pulmonary nodules and time of recurrence, define the prognosis of children with osteosarcoma and pulmonary metastases [36, 93]. Roth et al. [76] showed that patients with fewer than four pulmonary nodules had an improved survival over those with more than four lesions. According to Goorin et al. [28], a complete resection of all pulmonary lesions is an important determinant of outcome, and penetration through the parietal pleura is associated with an

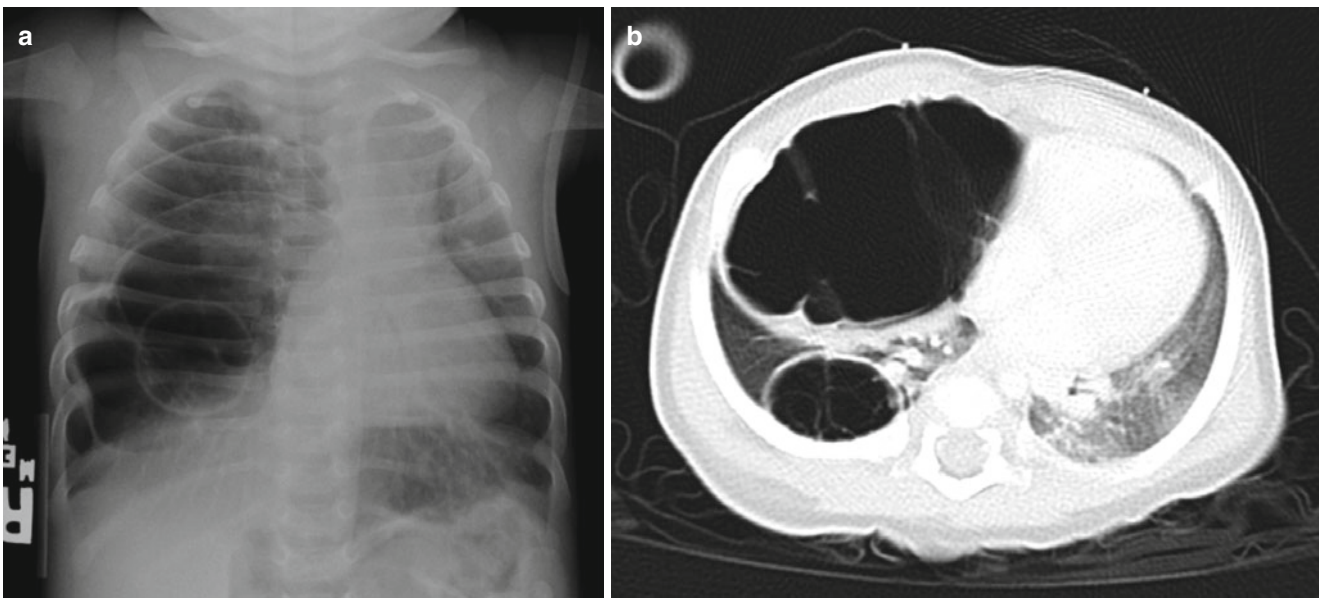

Fig. 26.4 (**a**) Chest radiograph of an 11 year old female who presented with shortness of breath and a cough demonstrated a right sided pulmonary mass. (**b**) Her CT scan revealed a multilobular mass and needle biopsy established the diagnosis of pulmonary blastoma. An extrapleural pneumonectomy and post operative radiotherapy were utilized for local control and she remains free of disease 16 years later

adverse outcome. Although somewhat controversial, the outlook seems to be somewhat improved, even in patients presenting with pulmonary metastases, if all metastatic lesions are resected [59]. Harris et al. [34] reported a 68 % rate of survival in 17 patients with fewer than eight pulmonary nodules at presentation following chemotherapy, resection of the primary tumor, and pulmonary metastasectomy. An aggressive attempt at surgical resection of pulmonary metastases is indicated in osteosarcoma, possibly irrespective of the number of lesions or the interval between presentation and identification of metastases. A number of recent studies have shown a survival advantage in patients with repeated metastasectomy including patients with as many as five recurrences [10, 13, 17].

Soft Tissue Sarcoma

The usefulness of resecting pulmonary metastases in patients with soft tissue sarcoma depends on the histologic subtype. Rarely is pulmonary resection of metastatic lesions required in rhabdomyosarcoma, and resection of pulmonary metastasis in Ewing's sarcoma is not efficacious [38, 48]. Several European protocols are being designed to better define the role of pulmonary resection in Ewing's sarcoma. The remaining sarcomas should be considered for resection if complete excision is possible and the patient's primary tumor .has been completely resected. The time to development of pulmonary metastases, number of lesions, and tumor doubling time are all significant prognostic factors in soft tissue sarcomas. Historically, 10–20 % of these patients can be salvaged by resection of pulmonary metastases [55].

Wilms' Tumor

Rarely is pulmonary resection of metastatic disease required in children with Wilms' tumor. In a review of the National Wilms' Tumor Study by Green et al. [29], no advantage of pulmonary resection was found compared with chemotherapy and radiation therapy alone. In an attempt to avoid pulmonary radiation, deKraker et al. [21] suggested a protocol using primary pulmonary resection after chemotherapy for pulmonary metastases. Only 5 of 36 patients ultimately required resection of pulmonary metastases following chemotherapy, as most patients had a complete response with chemotherapy alone. One encouraging finding was that only 4 of 36 children required whole lung irradiation. Because the results of chemotherapy and whole-lung irradiation are excellent for children with Wilms' tumor and pulmonary metastases, pulmonary resection of metastases should be reserved for selected cases.

Tumors of the Chest Wall

Epidemiology

Tumors of the chest wall are rare in the pediatric population with an incidence of less than 2 % of all tumors in children [24, 50]; up to two-thirds of these lesions are malignant [99]. The majority arise from the bony structures of the chest wall (55 %), as opposed to soft tissue (45 %) [15]. Collectively, a 60 % 5-year overall survival rate for all tumors has been reported, with a recurrence rate of 50 % (local and distant). The 5-year survival rate after relapse is only 17 % [49].

Presentation

Masses of the chest wall typically present with respiratory symptoms or pain, with pain being the most frequent in malignant lesions. Many have extensive protrusion into the pleural cavity with limited if any external findings. In infants and young children the benign lesions are often found incidentally by caregivers, while older children and young adults may present with larger masses that have been present and growing for some time until they produce respiratory symptoms. Incidental discovery on routine chest imaging has been reported to be as high as 20 % [77]. They can be found anywhere on the thorax, and the tissue of origin is generally mesenchymal in nature, regardless of whether the tumors are malignant or benign. Hence, sarcomas are the most common malignant tumors, while carcinomas are almost nonexistent. The symptoms of respiratory compromise or dysfunction – tachypnea, hypoxia, cough, dyspnea on exertion – and these symptoms may have been present for quite a while before seeking medical advice. Symptoms stem from parenchymal compression from the mass intruding into the pleural space and onto the lung or from malignant effusions, both of which interfere with normal respiratory mechanics. Pulmonary function tests may be indicated prior to proceeding with any intervention based on respiratory symptoms.

Diagnostic Adjuncts

Imaging studies should include first, erect, posterior-anterior and lateral chest radiographs to evaluate the location, size, presence of calcifications, osseous involvement, and the presence of pulmonary parenchymal disease. Next, an ultrasound exam is recommended to determine the echo features (solid versus cystic, degree of homogeneity and vascularity of the mass). Axial imaging (computed tomography [CT] or magnetic resonance imaging [MRI]) is performed next. The advantages of CT reside in its ability to clearly define the lung parenchyma and pleural space in relation to the osseous, vas-

cular and soft tissue components of the thorax (and hence mass), and the fact that it is a rapid technique requiring minimal to no sedation, even in the youngest of patients. The negative aspects of CT are the radiation exposure with subsequent risk of a secondary malignancy [12]. The benefits of MRI versus CT include better definition of the soft tissue components, as well as enhanced evaluation of the osseous and neural structures to determine the extent of central or peripheral nerve involvement and/or the presence of skip lesions or metastases. Unfortunately, this technique is time consuming and generally requires sedation or even general anesthesia to obtain optimal studies. Motion artifact from the heart and lungs can also interfere with this technique limiting its utility, but this obstacle is being overcome with the use of cardiac-gated, respiratory-triggered protocols [91, 92]. Diagnosis cannot be established by radiographic studies [91, 92]. Additional imaging studies may be required to assess the presence of metastases in malignant lesions (brain and abdominal CT, bone scan, positron emission tomogram [PET] scan). Recent reports have suggested that the combination of PET and CT scans yields more accurate data in assessing the primary tumor, local and regional lymph node basins, evidence of recurrence and for response to ongoing therapies [72, 79]. Once initial studies have been performed, retrieval of tissue for histopathological evaluation and diagnosis is warranted.

Diagnosis

Biopsy options include small or large specimen approaches. If a mass is small (less than 3 cm) or thought to be benign, then an upfront excisional biopsy may be warranted. However, the incision should be oriented so that a future re-excision can be performed, if needed, without compromising oncologic principles. A rim of normal tissue should be excised circumferentially around the mass. If the mass is large (greater than 4–5 cm), fixed to surrounding structures or involving many structures in the thorax, or if it is considered malignant by imaging, then either an incisional biopsy or core-needle biopsy is warranted. Placing the incision in-line with any future resection is of paramount importance, regardless of the technique utilized, and either approach will yield enough tissue for histopathologic and cytogenetic analyses [41]. Once a diagnosis is confirmed, then disease-specific treatment algorithms may be initiated.

Therapeutic Principles

Though treatment regimens are tumor-specific, there are certain general principles that apply. For malignant lesions, multimodality therapy is the accepted paradigm for the majority of lesions, while simple extirpation is the rule with benign entities. With surgery, the most important concept to emphasize is the need for complete resection with negative pathologic margins to decrease the risk of recurrence and need for subsequent therapy. Surgical extirpation also mandates wound reconstruction. Large defects (greater than 5 cm except for posterior and superior lesions where the defect will be buttressed by the scapula) require the use of prosthetic (rigid [silicone, Teflon (DuPont), methyl methacrylate] or flexible [prolene mesh (Ethicon), PTFE mesh, marlex mesh (Chevron Phillips Chemical), Gore-tex (WL Gore & Associates)]) materials and/or autologous tissues (pedicle or free flaps [latissimus dorsi, rectus abdominis or pectoralis major]) to reconstruct the chest wall to assure normal chest wall mechanics and prevent respiratory embarrassment.

Tumor Types

Chest wall tumors are separated into benign and malignant cohorts (Table 26.2), as well as primary and secondary lesions. Specific tumors and their treatment will be outlined in the subsequent sections.

Chest Wall Tumors: Benign

Aneurismal Bone Cyst (ABCs)

ABCs can be found anywhere on the chest wall, and they generally arise in the ribs. They have characteristic patterns of appearance on both chest radiographs and MRI [91], and they can grow to be quite large producing local destruction to the adjacent tissues (Fig. 26.5a, b). Surgical extirpation with complete excision is the treatment of choice, and recurrence is rare. Histologically, the lesions are blood filled cysts composed of fibrous tissue and giant cells.

Chondroma

Chondromas are slow-growing, painless masses that usually arise in the costal cartilages. On imaging studies, they are lytic lesions with sclerotic margins, and unfortunately, they are difficult to distinguish radiographically from their malignant brethren, chondrosarcomas. Hence, complete resection with a wide margin of normal tissue is advocated [82].

Desmoid

Desmoid tumors are fibrous neoplasms that can be found anywhere in the body. They are thought to be benign, but they have also been reported to undergo malignant

Table 26.2 Pediatric chest wall tumors

Benign
Aneurysmal bone cyst
Chondroma
Desmoid
Fibroma
Fibrous dysplasia
Lipoblastoma
Lipoma
Mesenchymal Hamartoma
Osteochondroma
Osteoma
Vascular malformations
Malignant
Chondrosarcoma
Ewing's Sarcoma Family
Fibrosarcoma
Langerhans cell histiocytosis
Leiomyosarcoma
Leukemia
Liposarcoma
Lymphoma
Neuroblastoma
Rhabdomyosarcoma
Osteosarcoma

degeneration [82]. Desmoid tumors infiltrate adjacent and surrounding tissues, and they are known to travel down fascial planes and to encase neurovascular structures in the mediastinum or the thoracic inlet. MRI is the radiographic procedure of choice to best define the extent of involvement and the structures involved. Treatment is wide local resection with negative margins, but recurrence rates from 10 % (negative margins) to 75 % (positive margins) have been described by some authors [8, 22, 31]. If a complete resection is not feasible and vital structures would be sacrificed during operative extirpation, then multimodality therapy consisting of radiation (50–60 Gy) and cytotoxic (vinblastine and methotrexate) and cytostatic (tamoxifen and diclofenac) chemotherapy is recommended, though the optimal regimen is not well established [7, 53, 54, 85].

Fibrous Dysplasia

Fibrous dysplasia is a benign condition where normal bone is replaced by fibrous tissue. These lesions are generally not large, and patients present with pain, often from a pathologic fracture. On plain radiographs, these lesions are described as lytic in nature with a characteristic "soap bubble" appearance [42]. Treatment is based on symptoms and concerns for

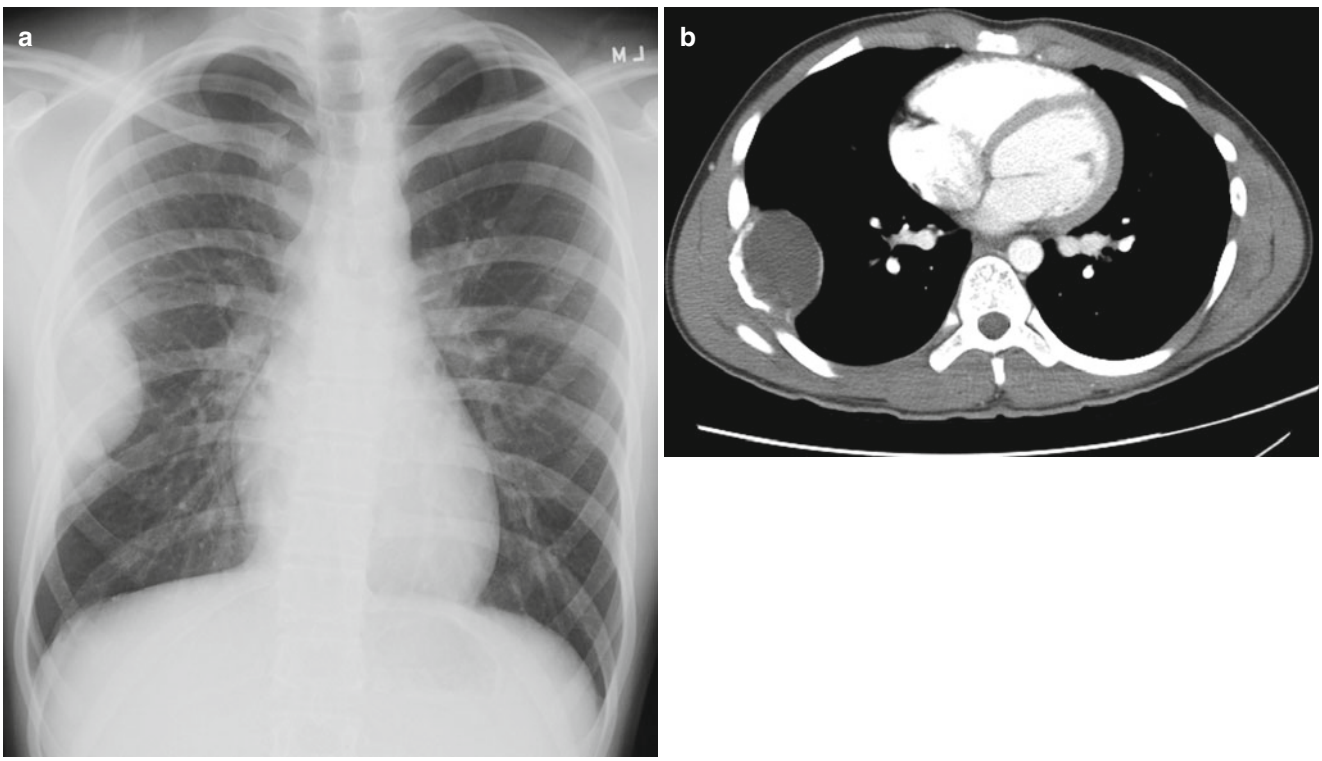

Fig. 26.5 (**a**) Chest radiograph obtained on a 15-year old boy complaining of chest pain after trauma during a hockey match. It revealed a mass lesion based around the 7th rib. (**b**) Follow-up CT scan demonstrated a cavitary lesion of the rib. Because of initial concern regarding malignancy, a percutaneous needle biopsy was obtained but diagnosis was inconclusive. Subsequent open procedure and curettage confirmed a diagnosis of aneurysmal bone cyst

possible fracture secondary to the inherent structural weakness the lesion produces in the bone. Simple excision is the recommended procedure.

Mesenchymal Hamartoma

Mesenchymal hamartomas (MH) are masses found in infants or young children that can also be discovered antenatally. The lesions are generally well circumscribed and though emanating from the chest wall (one or several ribs), they abut or compress, as opposed to invade, thoracic structures (Fig. 26.6a, b). Hence, symptoms at presentation are often from respiratory embarrassment. Parents may also note a palpable mass. These lesions are well defined by radiographic features on cross-sectional imaging, including mineralization and hemorrhagic cystic structures [32]. Histopathologically, these lesions consist of chondroid tissue with blood-filled, endothelial-lined spaces interspersed with osteoclastic giant cells. Treatment strategies have traditionally consisted of complete resection with subsequent chest wall reconstruction, but considering the large size of these lesions and the small volume of the chest cavity in the infants in which they occur, concern over the future complications of scoliosis and respiratory compromise from this approach has been considerable. In light of the fact that they are not know to undergo malignant degeneration [5], expectant observation [71, 83] or other less morbid approaches (radiofrequency ablation [9]) have been described. With expectant observation, the relative size of these lesions decreases as the children grow, resulting in less physiologic impact.

Osteochondroma

Osteochondromas are composed of bony and cartilaginous elements and are more commonly found in males (3:1 ratio) [82]. The lesion can present with pain from a pathologic fracture or compression of nearby nerves, or it can be asymptomatic if it grows into the thoracic cavity. The lesion is well characterized on plain radiographs, and arises from the cortex of the rib at the metaphysis and has a "cartilage cap" [42]. Malignant degeneration has been documented [91], and resection is warranted in all postpubertal patients, with symptoms, or if the mass is growing.

Chest Wall Tumors: Malignant

The majority of malignant tumors in infants and children are sarcomas, and the most common of these tumors will be addressed individually in the following sections.

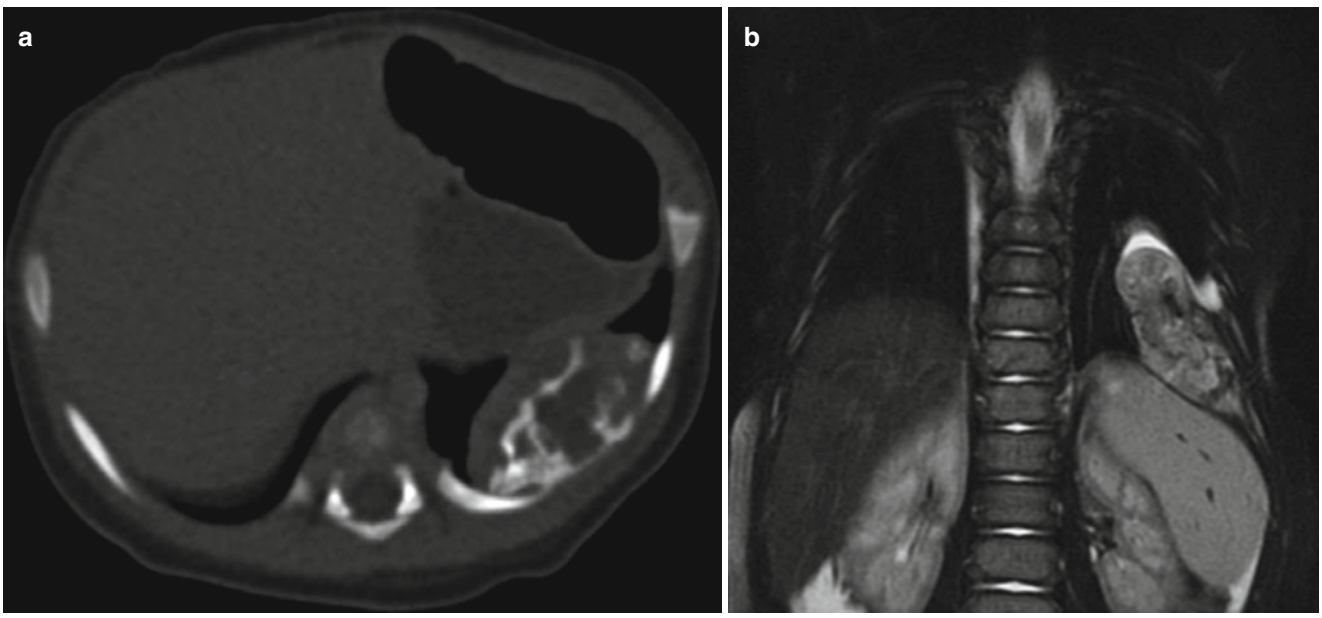

Fig. 26.6 Antenatal ultrasound revealed a calcified chest mass in this infant. Chest radiograph at birth showed partially calcified lesion in the chest and CT scan (**a**) was obtained to better define the lesion. It demonstrated a lesion arising from the ventral aspect of the 10th rib which was heterogeneous with areas of internal spiculated or plate-like ossification/calcification throughout the mass which was felt to be consistent with a mesenchymal hamartoma. MRI obtained at 6 months of age (**b**) demonstrated a heterogeneous mass with amorphous areas of signal void consistent with previously demonstrated calcification. Uptake of contrast was also heterogeneous. This has been followed expectantly and when seen 2 years later the lesion was decreasing in size as the child continued to grow.

Chondrosarcoma (CS)

Chondrosarcomas are derived from cartilaginous elements (costal cartilages) that are the most common primary malignant bone tumor of the chest wall in adults [87], and they are more common in males [82]. CS have been associated with a prior history of trauma [63], as well as being known to form from malignant degeneration of the benign counterpart discussed previously [87]. Some 10 % of patients will present with metastatic disease [15], especially in the lungs and brain. Primary therapeutic intervention is complete surgical resection with a margin of normal tissue of at least 4 cm [82] secondary to the high risk of local recurrence (up to 75 % with positive margins) even with negative margins at the initial operation (10 %) [25]. These tumors are not chemotherapy responsive, and the role of radiation is only for those lesions that are unresectable or with known positive margins. Five-year survival has been reported to be from 60 to 90 % [25, 43], and favorable prognostic factors are the absence of metastases at presentation and a complete resection [15, 25].

Ewing's Sarcoma Family/Primitive Neuroectodermal Tumors (EWS/PNET)

EWS/PNET are the most common malignant chest wall lesion in the pediatric population [83]. They are aggressive tumors requiring multimodality therapy; survival is poor despite intensive therapy, particularly in those who present with metastatic disease. Patients present with respiratory symptoms or pain; metastases to the lung, bone or bone marrow are common (25 %) [15]. EWS/PNET are defined histologically as sheets of small, round cells with scant cytoplasm and are characterized by a balanced gene translocation (EWS/FLI1) (t11:22[q24:q12]) [62]. Imaging studies demonstrate characteristic bony destruction described as lytic or sclerotic lesions [92]. Treatment involves an initial biopsy followed by neoadjuvant chemotherapy (4 cycles) with vincristine, actinomycin, cyclophosphamide, and Adriamycin (Adria-VAC) alternating with etoposide and ifosfamide. This regimen has demonstrated a great deal of success in shrinking the tumor to improve survival and facilitate complete resection [30, 84] (Fig. 26.7a, b). In fact, with the use of neoadjuvant chemotherapy, complete surgical resection with negative margins was possible in 71 % of patients versus 37 % who underwent primary surgical intervention [84]. The extent of surgery should include all involved structures and a soft tissue or osseous margin. Postoperative adjuvant therapy utilizes the same preoperative chemotherapy regimens, but not radiotherapy if complete resection is achieved. This should be the goal, despite the known radiosensitivity of this tumor [97], due to the late effects of radiotherapy (scoliosis, pneumonitis, cardiotoxicity, secondary malignancy, growth retardation, and breast hypoplasia or aplasia) [84]. The use of radiotherapy for residual disease after surgical extirpation, unresectable tumors and for patients who presented with a malignant pleural effusion where is an accepted therapeutic intervention. A recent European consensus conference advocated

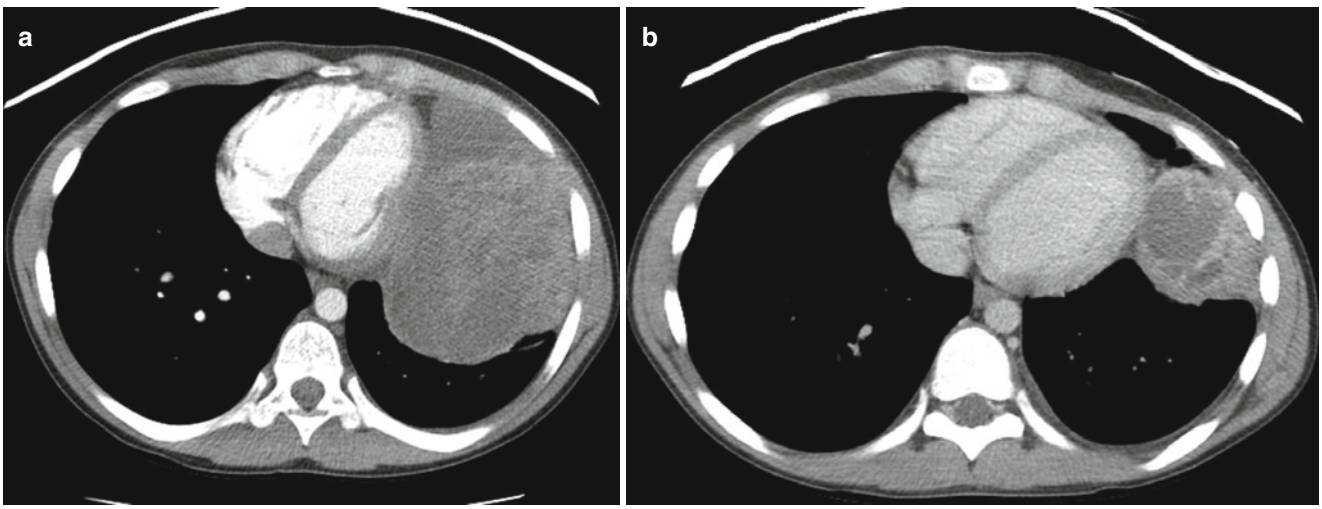

Fig. 26.7 This 10 year old boy presented with left-sided chest pain several days after falling from a tree. Chest radiograph was read as a pulmonary contusion. He had two other episodes of chest pain subsequently and radiographs were felt to demonstrate pneumonias. Six months later a chest radiograph and (**a**) CT scan demonstrated a chest wall mass and open biopsy revealed a Ewing's sarcoma. He received six cycles of 5-drug chemotherapy and the CT scan at that time (**b**) showed remarkable regression of the tumor. Pathology of the resection specimen including portions of three ribs showed no residual tumor

surgery over irradiation in all cases [95]. Five-year overall survival utilizing the above protocol was around 70 % for patients with nonmetastatic disease [14], and the 8-year survival was roughly 30 % for patients with metastatic disease [64]. In patients presenting with metastatic disease, the European Intergroup Cooperative Ewing's Sarcoma Studies Group demonstrated improved survival with the use of myeloablative chemotherapy followed by stem cell rescue at the conclusion of conventional treatment protocols [70].

Fibrosarcoma (FS)

FS (also known as infantile or congenital fibrosarcoma) are malignant tumors found throughout the body in infants that present as large masses that often involve, invade and surround adjacent structures. FS have been found in the chest wall, and several reports have documented the success of multimodality therapy in combating these tumors [57, 67]. FS can be distinguished from other myofibrous and sarcomatous lesions by the presence of a unique gene rearrangement between the TEL gene (12q13) and TRKC gene (15q25) [57]. FS are chemotherapy sensitive, and reports demonstrating the effectiveness of neoadjuvant chemotherapy with vincristine, actinomycin, cyclophosphamide, and Adriamycin followed by surgical extirpation are well accepted (Fig. 26.8a–c) [57, 67]. A recent report [67] from Europe demonstrated that 5-year overall and event-free survival rates were 89 % and 81 %, respectively. The authors report that in their series complete surgical extirpation was rarely feasible, and that conservative surgical approaches should be adopted. Furthermore, 71 % of patients responded to alkylating agent- and anthracycline agent-free regimens, hence, this regimen should be started first to limit long-term toxicity.

Osteosarcoma (OS)

OS of the chest wall can be primary or secondary tumors (prior sites of irradiation or from pre-existing osseous lesions [Paget's disease]) [82]. Lesions are primarily of the

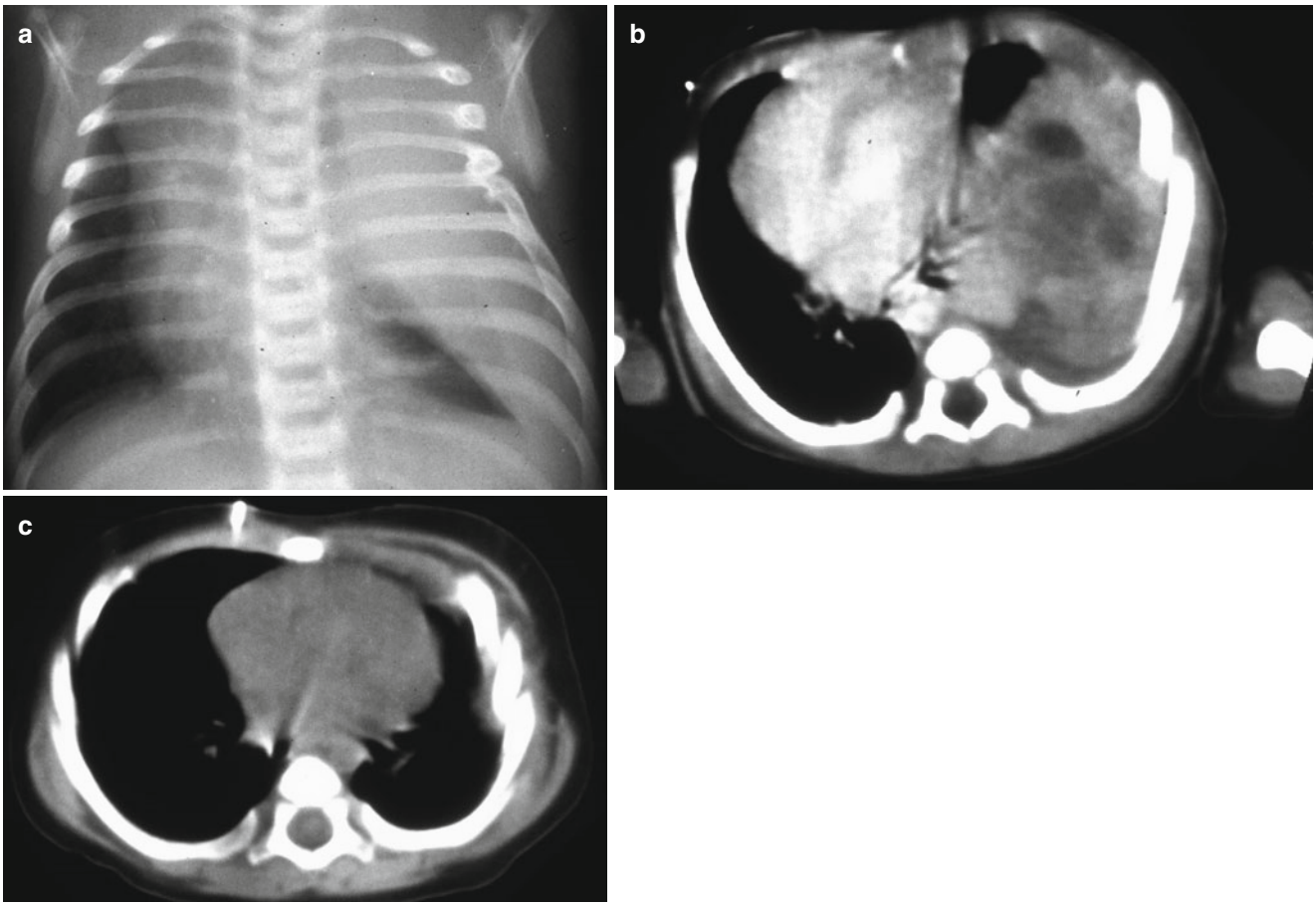

Fig. 26.8 (**a**) Chest radiograph of a newborn with respiratory distress demonstrates a large left pleural "filling defect". (**b**) CT scan demonstrated a solid lesion with some areas of presumed necrosis. Needle biopsy revealed an infantile fibrosarcoma. (**c**) Follow-up CT scan after 5 courses of Adria-VAC (300 mg/m^2 Adriamycin and 2 courses of VAC) produced remarkable regression of the tumor facilitating resection

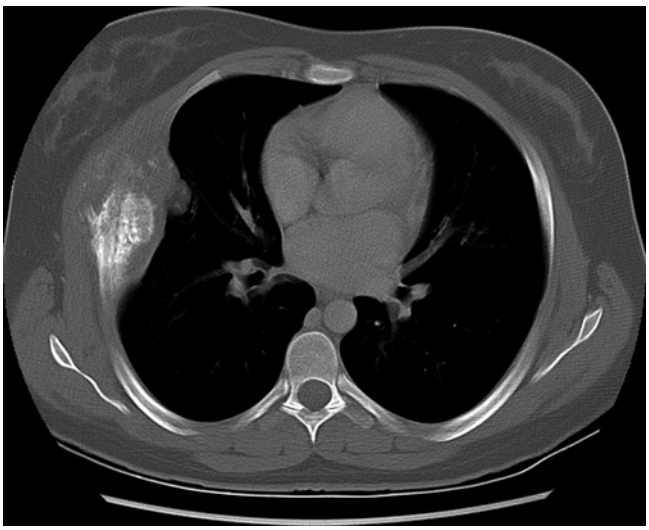

Fig. 26.9 A 15 year old female presented with a 2 month history of stabbing right sided chest pain. Chest radiograph revealed a mass within the thoracic cavity and a subsequent CT scan shown here demonstrated a large mass arising from the right 5th rib with osteoid matrix and irregular well-defined margins. Initial incisional biopsy was not diagnostic, so a resection involving segments of three ribs was performed and it was shown to be an osteosarcoma

ribs (Fig. 26.9), and on imaging, they can be confused with chondrosarcoma [56]. Chest radiographs will demonstrate a "sunburst pattern," and axial imaging concentrating on regional (bony skip lesions) and distant (lung, liver, brain) metastases must be obtained [82]. Pre-therapy biopsy is required to establish the diagnosis, and neoadjuvant therapy precedes extirpative procedures. Overall survival rates are poor (15–20 %) [15], but in the presence of nonmetastatic disease 5-year survival rates can exceed 50 % [15]. Prognosis is related to the presence of metastases, the degree of tumor burden and the response to chemotherapy [11].

Rhabdomyosarcoma (RMS)

RMS of the chest wall is a rare tumor and accounts for no more than 7 % of all RMS in Intergroup Rhabdomyosarcoma Studies (IRS) [4, 19, 61]. The chest wall site is deemed an unfavorable site, and therefore, this is an adverse prognostic factor [4, 61]. Other adverse prognostic factors have been reported to be histopathologic findings (alveolar versus embryonal), tumor burden and size, incomplete resection, and presence of metastatic disease (including lymph node metastases) [4, 18]. Despite advances in the treatment of RMS over the last 40 years, unfavorable sites carry an overall survival of only 55 % (versus 90 % for favorable sites) [4], and those with truncal RMS have been reported to have a failure-free survival rate of no greater than 67 % [78]. These tumors require multimodality therapy, and neoadjuvant chemotherapy followed by surgical extirpation. Radiation is reserved for lesions with positive margins following surgery or unresectable tumors. A report from Saenz and colleagues documented the utility of radiation (median dose of 44 Gy) to salvage some patients with residual disease [78]. However, the necessity for complete surgical resection has been called into question by a recent report from the Children's Oncology Group (COG) [37], in which the outcome of patients enrolled in IRS I-IV with chest wall RMS were analyzed. The report documents that regardless of clinical group (I-III) and other tumor-specific factors (histological subtype, tumor size), the only critical factor to influence failure-free and overall survival was the presence of metastatic disease. In the face of metastases, patients with chest wall RMS had an overall and failure-free survival of 7 % and 7 % versus 49 % and 61 %, respectively, in the cohort without metastases ($p<0.001$). Therefore, the authors suggest where gross total surgical resection will produce significant morbidity or physical debilitation, less aggressive operative approaches should be entertained.

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