Pediatr Radiol (2010) 40:859–868
DOI 10.1007/s00247-010-1604-z

REVIEW

Pathology of the thoracic wall: congenital and acquired

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Received: 24 December 2009 / Accepted: 24 January 2010

Abstract This review aims to cover the main congenital and acquired lesions that arise in the thoracic wall of infants and children. Imaging often plays an essential role in the evaluation of symptomatic and asymptomatic thoracic wall abnormalities. The use of appropriate imaging modalities for each condition will be addressed, as well as the range of benign and malignant conditions that can occur.

Keywords Chest · Congenital · Acquired · Child · Chest wall · Pediatrics

Introduction

The thoracic wall encases and protects the vital structures within the chest cavity. It is comprised of multiple layers, including skin, subcutaneous fat, muscle, bone, cartilage and pleura. Thoracic lesions and pseudolesions of the thoracic wall in children can be classified into normal variants, malformations, infection, tumours and trauma. These lesions can be located in the superficial layer of the thoracic wall and involve skin or subcutaneous tissues, in the intermediate layer affecting muscles and bones, or in the deep layers including the dorsal spine, ribs and intercostals spaces, the sternum, several fascial layers and the parietal pleura. Imaging often plays an essential role in the evaluation of symptomatic and asymptomatic thoracic wall abnormalities. Symptomatic disease usually requires imaging evaluation to determine the localization and to characterize the lesion. Asymptomatic palpable lesions are usually benign conditions, or normal developmental variations, but imaging is often required to confirm [1].

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Congenital and developmental abnormalities

The thoracic wall is usually quite symmetrical, being somewhat narrower in the upper portion than the lower portion. The anteroposterior diameter of the chest in infants is generally wider than in older children. The thoracic index (widest anteroposterior diameter/widest transverse diameter) is about 0.85 in infants and 0.72 in older children [2]. Children with pectus excavatum deformity or an idiopathic flat chest have a decreased thoracic index. In the idiopathic flat chest, the thorax is flat and wide and the thoracic kyphosis is reduced, but the position of the sternum is normal. Children with pectus carinatum deformity or with a "barrel chest" shape, have an increased thoracic index. Congenital and developmental thoracic wall abnormalities typically involve the ribs, costal cartilage and sternum.

When chest radiographs are taken in an infant in a rotated position, the frequently prominent double curvature of the clavicle can simulate a fracture. In older children, the sternal end of the clavicle may show marked cupping that should not be misinterpreted as osteomyelitis or septic arthritis.

Rib anomalies are very common findings with no clinical relevance. They include agenesis or partial aplasia, bifid ribs, supernumerary ribs and bridging between ribs. Cervical ribs may arise from the seventh cervical vertebra and can compress the brachial plexus or the subclavian artery. Intrathoracic rib, a rare incidental anomaly, is sometimes seen on the chest radiograph. It appears as a long bony structure emerging from the vertebral body with

its course running parallel and oblique to the spine (Fig. 1) [3]. Eleven pairs of ribs can be an isolated finding or a part of several syndromes, such as Down syndrome, trisomy 18, and cleidocranial dysplasia [4].

Anterior thoracic wall anatomic variations are frequent in children, the most frequent being asymmetry in the shape or size of the rib cartilage or the position of the sternum [1]. Many asymptomatic palpable chest wall masses in children are developmentally normal anatomic osseous and cartilaginous variants. A prominent convex anterior cartilage, localized thickening, bifid cartilage and chondral nodule are common. The position of the sternum in mild pectus excavatum or carinatum can produce a chest bump, which is frequently referred for imaging study. A palpable mass in the anterior thoracic wall is a cause of concern for parents, who bring their child to the physician for consultation. Cross-sectional imaging can confirm the benign nature of these variants, but is often unnecessary [5].

CT and MR imaging can be performed to study the masses that are often normal variants of bone or cartilage in asymptomatic children. US can be used as an easy alternative method to analyze the anatomic variant or rule out other types of chest wall masses. When evaluating palpable focal chest wall masses in the absence of clinical signs suggesting an aggressive lesion (e.g., pain, interval increase in size or constitutional symptoms), chest radiography and US are usually sufficient to exclude an aggressive condition.

The most common anatomic variations affecting the sternum are tilting of the sternum, pectus excavatum and pectus carinatum. Deviation of the typical horizontal position of the sternum in the transverse axis of the body is referred to as sternal tilting. This variant is usually not visible on chest radiography, but the secondary lateral displacement of the medial end of the clavicle may be a helpful clue. Sternal tilting can be associated with anterior subluxation of the adjacent clavicular head or abnormal convexity of the adjacent rib causing a palpable thoracic wall mass [1].

In pectus excavatum, the most common thoracic wall deformity, the inferior aspect of the sternum is depressed posteriorly, resulting in a mild or severe concavity. It can be a sporadic or inherited isolated lesion, or be associated with Turner syndrome, osteogenesis imperfecta, muscular dystrophy, Marfan and Ehlers-Danlos syndromes, or prolapsed mitral valve, and in severe forms can compromise cardiopulmonary function. On the frontal chest radiograph, the soft tissue depression results in silhouetting of the right heart border, simulating right middle lobe consolidation; the heart is shifted and rotated to the left. The sternum is usually rotated to the right, the anterior rib ends have a steep downward course and the posterior ribs are orientated horizontally. On the lateral view, the chest is narrow and the degree of the sternal depression is well depicted (Fig. 2). CT is an excellent imaging modality to assess the anatomic severity of the pectus excavatum defect, to assess the degree of cardiac shift or compression, to identify associated tracheobronchial compression and to assess the results of surgery [6]. The ratio of the internal transverse diameter of the chest to the anteroposterior diameter of the chest at its narrowest dimension on axial CT, known as the pectus or Haller index, can be used to characterize the degree of chest wall deformity. Children requiring surgical correction for pectus excavatum have a pectus index >3.25, compared to <3.25 in normal controls [7, 8]. In children, there are age-related and sex-related differences in the chest wall configuration [9]. MR imaging can be used to obtain measurements in axial images similar to those of CT and to assess the dynamics of the chest wall and diaphragm [10, 11].

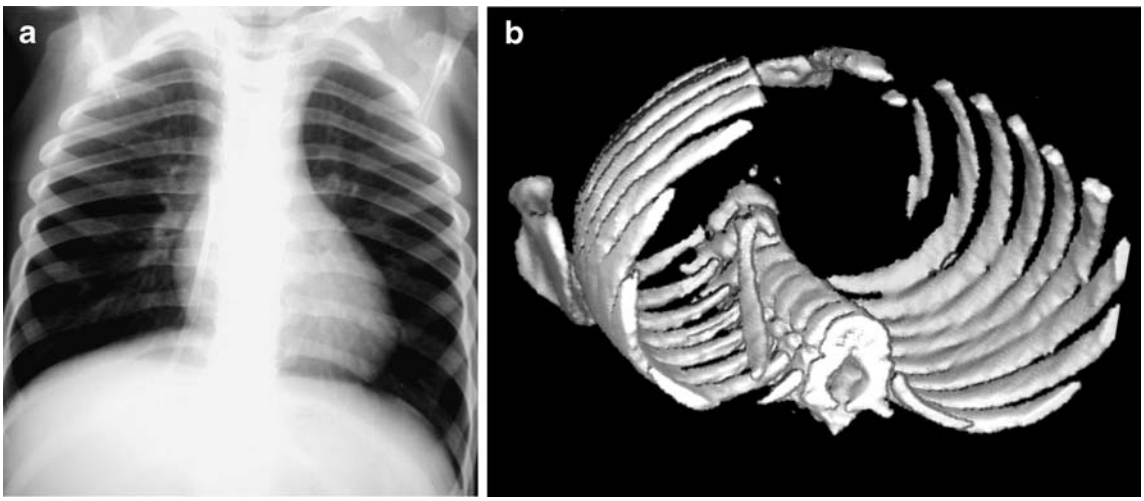

Fig. 1 Intrathoracic rib in a 3-year-old boy. **a** AP chest radiograph. There is an abnormal, dense elongated structure in a right paravertebral position. **b** Shaded-surface display image shows the intrathoracic rib with its anterior vertebral origin and its course running parallel and oblique to the spine

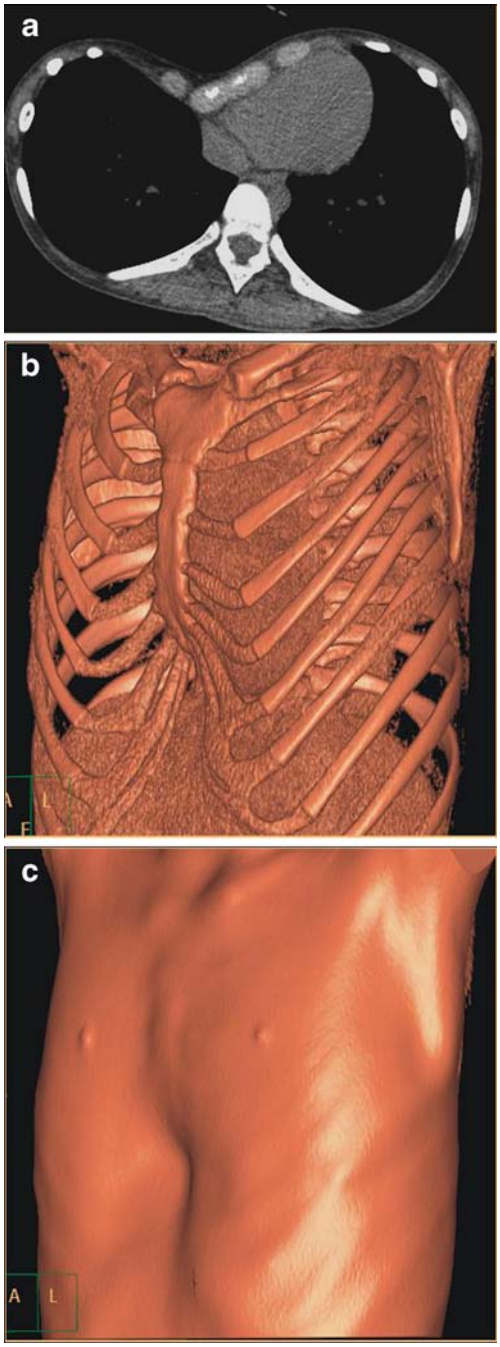

Fig. 2 Pectus excavatum in a 12-year-old boy with anterior chest deformity. (**a**) Axial noncontrast-enhanced CT scan of the chest demonstrates marked depression of the sternum. The Haller index is usually measured in the axial plane. (**b**) Volume-rendered image beautifully depicts the sternal depression and the position of the ribs and cartilage, thereby demonstrating the entire chest malformation. (**c**) Volume-rendered surface image to assess the soft tissues, shows the chest wall morphology and the sternal depression

Pectus carinatum is a less common deformity characterized by convexity of the thoracic wall resulting from anterior protrusion of the superior aspect of the sternum. The deformity seems to be caused by a growth disturbance of the sternum and costal cartilages, with premature sternal fusion. Most of the patients are asymptomatic. Pectus carinatum can be an isolated abnormality, but it can also be associated with Marfan syndrome, Ehler-Danlos syndrome, Morquio syndrome, Noonan syndrome, congenital heart disease and scoliosis among others [12].

Congenital abnormalities of sternal fusion are rare. They include bifid or cleft sternum and complete absence of sternal fusion. The severe end of sternal fusion abnormalities are often associated with ectopia cordis (extrathoracic heart) and pentalogy of Cantrell, a combination of sternal, diaphragmatic, cardiac, and abdominal wall lesions [13].

When there is a defect in bony or soft tissue structures of the thoracic wall, herniation of its content can occur. Pulmonary tissue can pass through a congenital chest wall defect, creating a lung hernia. This is seen as an intermittent bulging in the supraclavicular or intercostal area that appears with crying, coughing, or straining. Hence, during inspiration, chest radiography or CT may fail to show the lung herniation. Lung hernias can also be an acquired condition that occurs following chest tube placement, surgery, trauma, neoplastic disease or infection.

Malformations of the chest wall can be a manifestation of a syndrome or skeletal dysplasia such as short rib-polydactyly syndromes, asphyxiating thoracic dystrophy or Jeune syndrome and achondrogenesis [4]. Respiratory distress at birth is common and is related to small and narrow chests [14]. Thin ribs and small lungs can be found in neuromuscular disorders such as myasthenia gravis, myotonia and spinal muscular atrophy. Thin ribs are also seen in progeria and trisomies 8, 13, and 18. In contrast, thick ribs are seen in thalassemia and mucopolysaccharidosis. Coarctation of the aorta, or superior vena cava, and neurogenic tumours can present with inferior rib notching.

Poland syndrome is a rare congenital malformation of the thoracic wall consisting of unilateral partial or complete absence of pectoralis muscles, hypoplasia of subcutaneous or breast tissues, hypoplasia or absence of ribs and anomalies of the ipsilateral upper limb. Males are affected more often than females, and right side involvement is also more frequent. Hypoplasia of chest wall soft tissues results in a hyperlucency of the affected hemithorax. CT and MR imaging with 2-D and 3-D reformatting can help to determine the extent of the abnormalities and show the available muscles for reconstructive surgery (Fig. 3) [15].

The scapula is also subject to a spectrum of congenital anomalies. The most notable of these affecting the chest wall is Sprengel deformity. In this anomaly, the scapula position is high and rotated. The scapula fails to descend from its cervical origin and becomes fixed to the cervical spine by a fibrous band or an omovertebral bone. Klippel-Feil syndrome has additional anomalies of ribs or vertebrae. CT with 3-D reconstructions helps to delineate the deformity when planning corrective surgery [16].

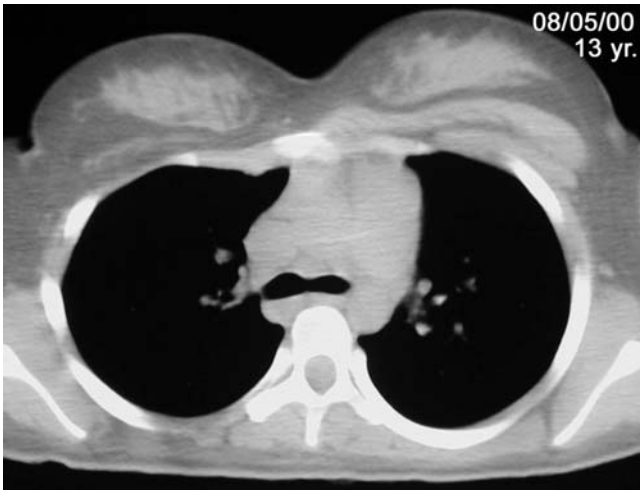

Fig. 3 Poland syndrome in a 13-year-old girl with asymmetrical thorax. Axial CT image shows hypoplasia of the right pectoral muscle. The chest radiograph showed a hyperlucent right hemithorax (not shown)

Cleidocranial dysplasia is characterized by hypoplasia or absence of one or both clavicles with hypermobile shoulders, but other chest wall abnormalities may also be present, such as small scapula, deficient sternal ossification, scoliosis and short ribs [4].

Congenital pseudoarthrosis of the clavicle is a rare and isolated anomaly of the clavicle that presents as a painless mass.

Infection

Chest wall infections, which are relatively rare in children, can occur by haematogenous spread or direct extension. Infection may involve the soft tissues, cartilage and osseous structures and can cause abscess formation. Pyomyositis is the term used when muscles are affected, fasciitis when only subcutaneous fat and fascia are involved, osteomyelitis when there is bone involvement and pyogenic arthritis when there is joint involvement.

Bacterial and fungal infections of the chest wall have been described [17]. The responsible microorganisms include *Staphylococcus aureus* (the most prevalent), *Mycobacterium tuberculosis, Actinomyces, Nocardia, Aspergillus* and *Candida* [18]. Fungal infections are more common in immunocompromised patients. Cat-scratch disease occasionally causes chest wall infections [19]. Patients with these infections present with pain, fever, local erythema and swelling. Recognition of the chest wall infection is sometimes difficult, particularly when it is located in the deep layers.

Chest radiography may show a mass lesion within the chest wall, rib destruction or sclerosis, pulmonary infiltrates, pleural effusion, calcifications and gas in the soft tissues. US, CT, and MR can confirm the presence, location and extent of the infection, show fluid collections, rib destruction and provide a guide for percutaneous drainage. US usually suffices for diagnosing small, superficial, well-delineated lesions and CT or MR are the techniques of choice for imaging complex and deep lesions. On US, an abscess has the typical appearance of an anechoic mass with posterior acoustic enhancement and substantial peripheral flow on colour Doppler, sometimes with swirling echogenic material inside. An abscess on contrast-enhanced CT appears as an isodense or hypodense lesion with a non-enhancing centre and an enhancing rim [20]. Findings similar to those on CT can be seen on T1-weighted MRI sequences and high signal intensity on T2-weighted sequences. Soft tissue inflammation is well delineated by MRI, on fat-suppressed T2-W or short tau inversion recovery (STIR) sequences or T1-W sequences following administration of contrast medium (Fig. 4). MRI is very sensitive for detecting osteomyelitis and is more accurate than bone scan for differentiating between soft tissue

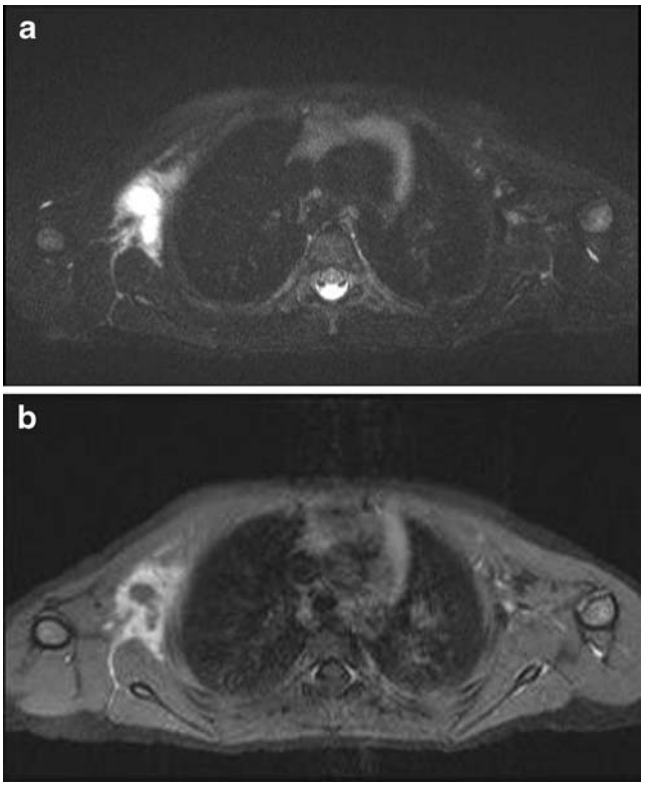

Fig. 4 Chest wall abscess. A 5-year-old boy was referred to our hospital with weakness of the upper right arm, local pain in the shoulder and fever. Radiography and US of the shoulder were normal. An MR study was performed. (**a**) Axial T2-W image with fat saturation (FS) at the level of the upper thorax shows a hyperintense lesion in the right axilla with infiltration of the soft tissues. (**b**) Axial T1-W FS image after gadolinium administration shows peripheral enhancement. The findings are consistent with infectious lymphadenitis with a central abscess. Conservative antibiotic treatment was successful in this boy

inflammation and acute osteomyelitis. MR imaging is more sensitive for marrow oedema without frank bone destruction in early osteomyelitis. Chronic osteomyelitis is shown on chest radiographs or CT studies as areas of destruction and sclerosis. Positron emission tomography (PET) is very sensitive in the detection of clinically silent foci of infection, particularly in immunocompromised patients.

Tuberculous infection can involve the spine but is rare in developed countries. Spinal infection mainly stems from primary pulmonary tuberculosis. The infection is usually limited to the vertebral body, which can be destroyed, and can also affect the contiguous intervertebral disc and the adjacent or distal vertebra. Paraspinal abscess can occur and vertebral collapse can lead to kyphosis or scoliosis and even cord compression. CT and MRI can show the vertebral lesion, epidural extension and delineate the abscess.

Friedrich disease is a disorder of unknown origin. It is an aseptic unilateral or bilateral necrosis that can mimic infection at the sternoclavicular joints [21]. Patients usually present with swelling at the sternoclavicular region. A lesion demonstrating destruction and repair can be seen at the medial end of the clavicle on radiographs and histology shows necrosis without infection. Usually the radiological and clinical features improve very slowly.

SAPHO syndrome, which is associated with chronic recurrent multifocal osteomyelitis, is so named because it is a combination of synovitis, acne, pustulosis, hyperostosis and osteitis. It has a predilection for the anterior chest wall. Sclerosis and periostitis are the main radiological findings [22].

Tumours

The thoracic wall can be affected by benign and malignant tumours. Imaging is performed to determine their location, size and characteristics. Chest radiography is the first imaging technique used and enables an estimation of the location of the lesion, its extension, rib destruction, associated intrathoracic components, pleural effusion and presence of pulmonary metastases. CT and MR imaging demonstrate the mass and osseous changes, define the margins and internal structure of the lesion, lymphatic spread and any associated pleural effusion. These techniques are also useful for staging purposes. CT is better than MR for investigating lung metastases.

Tumours of the thoracic wall are uncommon in infancy and childhood and are usually malignant [23]. Children present with a mass, pain, cough or respiratory distress from a pleural effusion or intrathoracic component. The tumour may be located within the bones or soft tissues of the chest wall. At times it may be difficult to differentiate between benign and malignant lesions, but a sharply marginated osteolytic lesion usually indicates a slow-growing lesion (benign).

Benign tumors

Lymphangiomas and haemangiomas are the most common vascular lesions of soft tissue in the chest wall in neonates, infants and young children. In lymphangiomas, lymphatic vessels predominate. When the dilated lymphatic vessels lead to cyst formation, they are referred to as cystic hygromas. These benign tumours are present at birth and can occur anywhere, but are most commonly located in the neck, chest wall and axilla and can extend into mediastinum. Lymphangiomas of bone are very rare. Bleeding within a mixed lymphangiohaemangioma leads to a sudden increase in size. Possible complications are infection, chylothorax and chylopericardium. Sonographically, the microcystic lesions are predominantly anechoic masses that contain septations. Colour Doppler may demonstrate arterial or venous flow within the septations. Microcystic lesions are more echogenic and can mimic solid soft-tissue lesions. When the lesion is not confined to the subcutaneous layer, MR imaging is the best method to assess its extent. Blood or protein products within the cysts can result in fluid–fluid levels.

Haemangiomas are very common benign soft-tissue tumours composed of abnormally proliferating endothelial cells. They are typically located in the head and neck, chest wall and extremities. Haemangioma of the chest bones is not frequent. These tumours may be present at birth or appear in the first weeks of life. Classically they are divided into capillary, cavernous and mixed types depending on their vascular composition: arterial, arteriovenous, venous and capillary. Haemangiomas can increase in size during a proliferative phase and later involute spontaneously. Imaging may be indicated for deeper or larger lesions. US is often the initial imaging modality used with the lesions typically appearing as well-defined hypoechoic, or less commonly, hyperechoic masses. Large haemangiomas may have a complex echotexture containing dilated vascular channels. Colour Doppler depicts their hypervascular nature demonstrating high-velocity arterial and venous flow. MR imaging is useful for assessing the extent and the relationship with adjacent structures before surgical procedures. Haemangiomas appears as lobulated soft-tissue masses that are isointense to muscle on T1-W images and hyperintense on T2-W images. They often contain tubular signal voids reflecting the high-flow vessels and show diffuse enhancement after gadolinium administration (Fig. 5).

"Vanishing bone disease" or diffuse cystic angiomatosis of bone (Gorham-Stout disease) is a rare condition with spontaneous progressive resorption of bone, and sometimes, pleural effusion (Fig. 6). Rare benign soft-tissue tumours that affect the thoracic wall include lipoblastoma

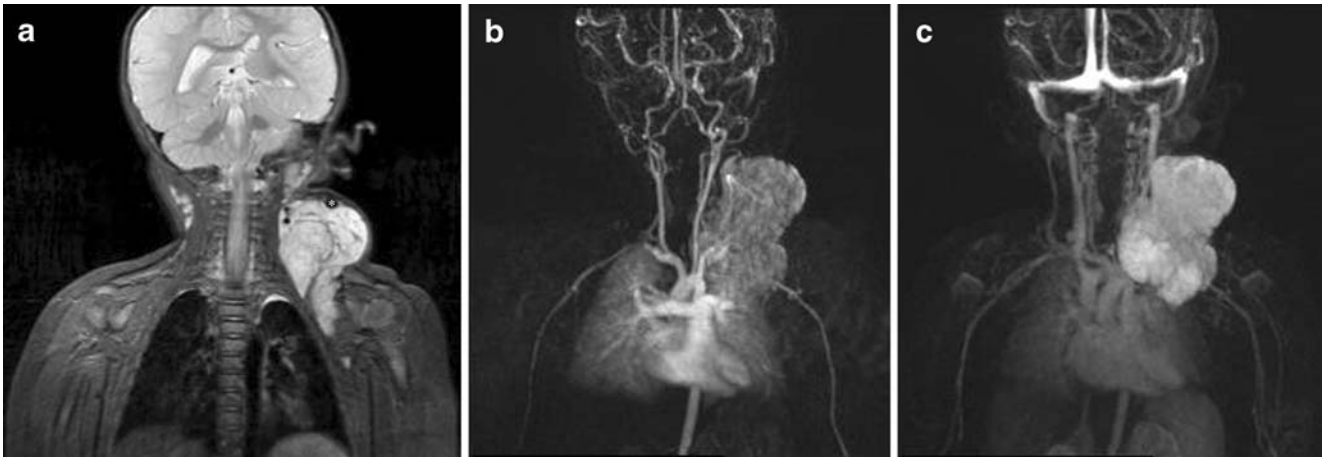

Fig. 5 Congenital haemangioma in the left cervicothoracic region. MRI was performed in a 1-year-old boy with a cervical mass. (**a**) Coronal T2-W FS image shows a hyperintense mass in the left laterocervical region with caudal extension to the thoracic wall; flow voids (*asterisk*) representing arterial vessels are also seen. (**b**) and (**c**) Coronal 3-D maximum intensity projection (MIP) reconstruction of a dynamic MR angiography shows arterial enhancement and a tissue component with delayed enhancement, consistent with congenital haemangioma

and fibrous tumours of childhood, such as fibroma, fibromatosis, fibrous hamartoma of infancy and neurofibromatosis. Neurogenic tumours such as schwannomas and neurofibromas arising from the intercostal nerves or sympathetic ganglia can erode adjacent ribs.

The osseous structures of the chest wall can also be affected by benign lesions. The most common is osteochondroma or exostosis. These consist of excrescences composed of cortical and medullary bone with a hyaline cartilaginous cap. Osteochondromas usually affect the metaphysis of tubular bones, but they can also be found in the ribs, vertebra, clavicle, scapula and sternum, as solitary or multiple lesions. Multiple osteochondromas occur with an autosomal-dominant disorder. When they are located in ribs, pleural effusion or haemothorax may occur. Chest radiography, CT, and MR imaging can depict the lesions and the origin from the parent bone. Malignant transformation is rare [24].

Fibrous dysplasia is a developmental anomaly of bone-forming mesenchyme in which the osteoblasts fail to undergo normal morphologic differentiation. The ribs are commonly affected. Radiologically, this condition is characterized by a focal, expansile, multiloculated lucent lesion or fusiform enlargement with a ground-glass or sclerotic appearance. It can be an incidental finding [25].

Langerhan cell histiocytosis can affect the ribs, clavicles, scapula and vertebral bodies. Vertebral body flattening is classically seen. Lesions in the other locations can have a variable appearance ranging from a poorly defined, expansile, lytic lesion to a sharply marginated, sclerotic lesion. A soft tissue mass adjacent to the bony lesion may be present [26].

Mesenchymal hamartoma of the thoracic wall is a rare, benign lesion that arises from the ribs. Histopathologically it has both a cystic and solid component. The solid component comprises normal maturing mesenchymal tissues including bone, cartilage, fat and fibroblasts. The cystic component reflects haemorrhagic cavities related to secondary aneurysmal bone formation. On chest radiography, mesenchymal hamartoma appears as a partially calcified, extrapleural chest wall mass with involvement of one or more ribs. The rib lesion consists of partial or complete destruction, erosion and enlargement. CT and MR imaging can detect the mineralized matrix and haemorrhagic cystic components [27].

Malignant tumours

Thoracic wall malignancies often involve many layers of the chest because of their aggressive nature. Malignant masses of the chest wall include primary soft-tissue neoplasms, soft-tissue extension of a primary osseous neoplasm or a primary mediastinal neoplasm infiltrating the thoracic wall. In children several malignant tumours can affect the thoracic wall [23, 28, 29].

The most common malignant tumours affecting the chest wall in children (50–65%) are the Ewing sarcoma family of tumours (ESFT), including primitive neuroectodermal tumour (Askin tumour), Ewing sarcoma of bone and extraosseous Ewing sarcoma [30]. It can be difficult to differentiate between these tumours. Hence, electron microscopic identification of neurosecretory granules and immunohistochemical analysis for neuron-specific enolase are required to make the correct diagnosis. Histology of these tumours is to that of small, round, blue-stained cells. They are also associated with chromosome 11 and 22 translocation and MIC2 gene [31, 32].

On initial examination, chest wall tumours can be mistaken for empyema because of the signs and symptoms of inflammation or infection and a pleural mass. Moderate

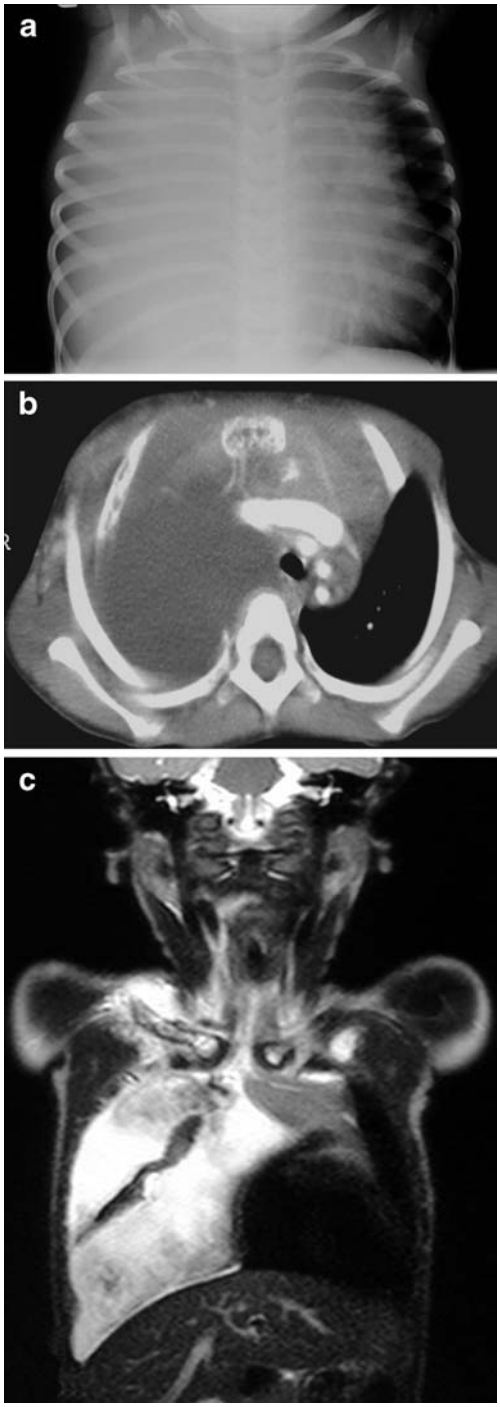

Fig. 6 Gorham disease in a 2-year-old boy with respiratory distress. (**a**) AP chest radiograph shows a dense lesion in the entire right hemithorax with the mediastinum displaced to the left and bone lesions. The right clavicle contains a sclerotic/lytic lesion, and the first two right ribs have an altered bone trabecula pattern. (**b**) Axial CT scan shows the lytic bone lesion in the sternum and the first right rib. A pleural effusion is occupying the right hemithorax and a soft-tissue lesion is seen in the chest wall. (**c**) Coronal T2-W MR image shows an enlarged, hyperintense right clavicle with soft tissue involvement and a large ipsilateral pleural effusion. The results of open surgical biopsy of the clavicle were consistent with bone lymphangiomatosis

to high pyrexia, elevated C-reactive protein and elevated white cell count in combination with an opacity on chest radiography suggest a pleural collection rather than a tumour. Children with a tumour can present with pain, cough, dyspnoea, pleural effusion and a chest wall mass. Plain radiography sometimes shows only pleural effusion, which can hide the underlying lesions, or a soft-tissue mass with erosion or destruction of the adjacent rib (Fig. 7) [33, 34]. US can be of help in making the diagnosis, but sometimes misses the evidence of tumour. CT or MR

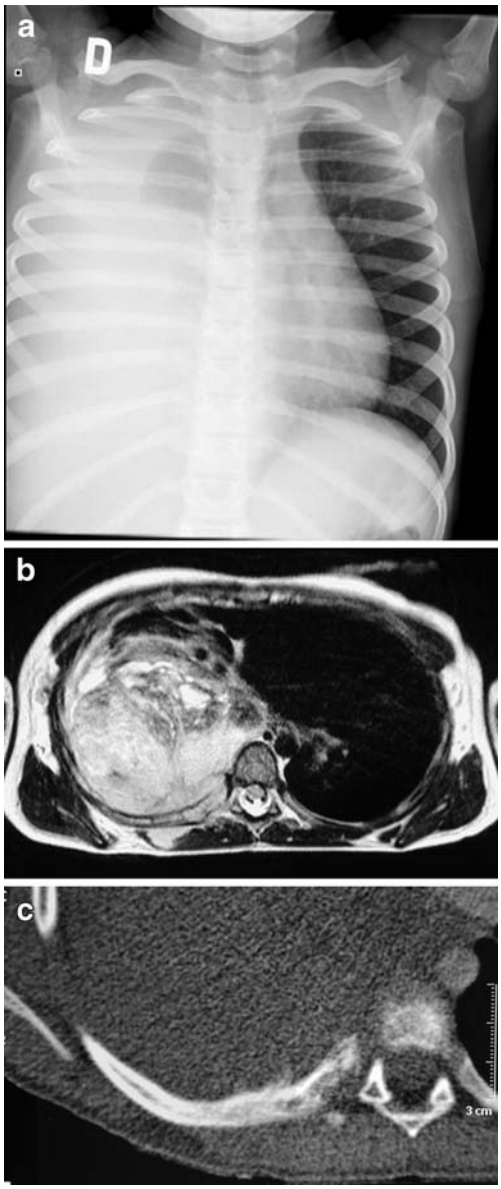

Fig. 7 Askin tumor in an 8-year-old girl who was initially diagnosed with right lung pneumonia and pleural fluid. (**a**) Chest radiograph shows a right lower lobe density and pleural effusion. US of the thorax (not shown) showed a mass in the right thorax. (**b**) Axial T2-W MR imaging at the level of the mass shows an extrapleural soft-tissue mass originating in the posterior arch of the rib of heterogeneous high signal intensity. (**c**) A localized low-dose CT slice also shows the bone involvement

imaging with intravenous contrast is necessary to detect the underlying tumour, its origin and metastases [35]. MR shows a heterogeneous mass with high signal intensity on T2-W images and contrast-enhanced T1-W images. MR may be superior to CT for defining tumour extension and CT may be better at detecting lung metastases. Cross-sectional imaging is useful to assess tumour response to chemotherapy during follow-up [36]. These tumours usually have a poor prognosis and often recur.

Rhabdomyosarcoma is the most common soft-tissue sarcoma in children and the second most common malignancy of the thoracic wall [30]. They can arise from any location, but rhabdomyosarcomas of chest wall origin have a poor prognosis [37]. The embryonal form is the most common histologic subtype of chest wall rhabdomyosarcoma. Chest radiography may show a soft-tissue mass with variable rib involvement and CT and MR can help in assessing tumour extension. CT shows a nonspecific, enhancing heterogeneous mass with adjacent rib destruction and pleural effusion. MR imaging depicts heterogeneous increased signal intensity on T1-and T2-W images and heterogeneous contrast enhancement on T1-W images [38].

Thoracic lymphoma usually presents as mediastinal masses. It can affect the thoracic wall as an isolated soft-tissue mass or by direct mediastinal or parenchymal extension. CT is the most common imaging modality used to evaluate and stage lymphomas. The isolated chest wall mass appears as a homogeneous soft-tissue mass within the chest wall muscles. Lymphomatous extension of a mediastinal mass usually occurs in the parasternal region and will appear as a homogeneous soft-tissue mass obliterating normal fat planes and displacing chest structures. MR imaging is very sensitive in the study of soft-tissue chest wall masses, and shows increased signal intensity in the mass on T2-W images [39, 40].

Other less common malignant soft-tissue masses affecting the thoracic wall in children include neuroblastoma, congenital fibrosarcoma, mesenchymal chondrosarcoma, osteosarcoma and malignant peripheral nerve sheath tumours. Neuroblastoma can invade the thoracic wall by direct extension from the posterior mediastinal mass. MR is the best imaging modality to analyze the extent of the tumour and neural invasion (Fig. 8). The main MR findings are abnormal high signal intensity on T2-W images and heterogeneous enhancement after contrast administration [41]. Congenital fibrosarcoma is rare in children and has a better prognosis than in adults. It typically occurs in the axilla or thigh. MRI shows a mass isodense to muscle on

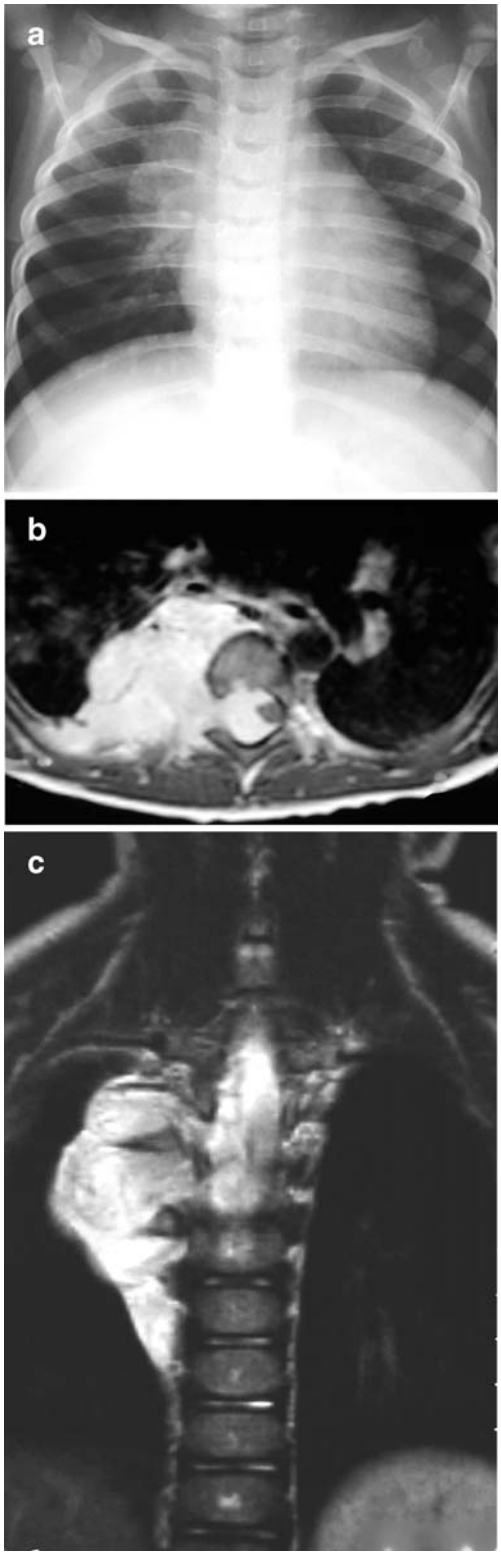

Fig. 8 Neuroblastoma; posterior mediastinal mass involving the chest wall in a 3-year-old boy with posterior chest pain. (**a**) AP chest radiograph shows a dense paravertebral lesion. The fourth and fifth posterior ribs are thinner and the intercostal spaces are wider in this area. The well-defined borders of the elongated mass associated with the bone lesion, confirms the findings of an extrapleural lesion. Axial (**b**) and coronal (**c**) T2-W MR images show a mass located in the posterior mediastinum invading the extradural canal, several foramina and the posterior soft tissue in the right side. MR imaging is the best technique for the study of neurogenic tumours with invasion of adjacent structures

T1-W images and a hyperintense mass with heterogeneous contrast enhancement on T2-W sequences [42]. The ribs and mandible are the most commonly affected bones in mesenchymal chondrosarcoma. CT can detect calcifications in the mass, reflecting the cartilaginous matrix. MR shows a mass with low signal intensity on T1-W images and high signal intensity on T2-W images.

Conclusion

A wide spectrum of disorders can involve the thoracic wall in children. These conditions can be congenital, developmental or acquired. Chest radiography is still the primary screening modality for palpable, symptomatic or asymptomatic chest wall disorders, as well as for symptomatic nonpalpable processes in children. The recent technological advances in cross-sectional imaging (CT and MR) have expanded the capacity for lesion characterization and assessing anatomical location and extension. Radiologists have an important role in analyzing the wide spectrum of pediatric chest wall variations and diseases and can limit the differential considerations, establish a specific diagnosis, and even suggest potential management options.

References

1. Donnelly LF, Frush DP, Foss JN et al (1999) Anterior chest wall: frequency of anatomic variations in children. Radiology 212:837–840
2. Nathanson I (1994) Chest wall abnormalities. In: Loughlin GM, Eigen H (eds) Respiratory disease in children: diagnosis and management. Williams and Wilkins, Baltimore, pp 533–541
3. Kamaruddin K, Wright NB, Pilling DW (1995) Intrathoracic rib. Pediatr Radiol 25:60–61
4. Lachman RS (2006) Taybi and Lachman's radiology of syndromes, metabolic disorders and skeletal dysplasias, 5th edn. Mosby, St Louis
5. Donnelly LF, Taylor CN, Emery KH et al (1997) Asymptomatic, palpable, anterior chest wall lesions in children: is cross-sectional imaging necessary? Radiology 202:829–831
6. Pretorius ES, Haller JA, Fishman EK (1998) Spiral CT with 3D reconstructions in children requiring reoperation for failure of the chest wall growth after pectus excavatum surgery. Preliminary observations. Clin Imaging 22:108–116
7. Haller JA Jr, Kramer SS, Leitman SA (1987) Use of CT scans in selection of patients for pectus excavatum surgery: a preliminary report. J Pediatr Surg 10:904–906
8. Chuang JH, Wan YL (1995) Evaluation of pectus excavatum with repeated CT scans. Pediatr Radiol 25:654–656
9. Daunt SW, Cohen JH, Miller SF (2004) Age-related normal ranges for the Haller index in children. Pediatr Radiol 34:326–330
10. Raichura N, Entwisle J, Leverment J et al (2001) Breath-hold MRI in evaluating patients with pectus excavatum. Br J Radiol 74:701–708
11. Herrmann K, Zech C, Strauss T et al (2006) Cine-MRI of the thorax in patients with pectus excavatum. Radiologe 46:309–316
12. Shamberger RC, Welch KJ, Castaneda AR et al (1988) Anterior chest wall deformities and congenital heart disease. J Thorac Cardiovasc Surg 96:427–432
13. Morales JM, Patel SG, Duff JA et al (2000) Ectopia cordis and other midline defects. Ann Thorac Surg 70:111–114
14. Eich GF (2007) Chest wall abnormalities which cause neonatal respiratory distress. In: Donoghue V (ed) Radiological imaging of the neonatal chest, 2nd edn. Springer, Berlin Heidelberg, pp 347–356
15. Fokin AA, Robicsek F (2002) Poland's syndrome revisited. Ann Thorac Surg 74:2218–2225
16. Cho TJ, Choi IH, Chung CY et al (2000) The Sprengel deformity. Morphometric analysis using 3D-CT and its clinical relevance. J Bone Joint Surg Br 82:711–718
17. Thomas KE, Owens CM, Veys PA et al (2003) The radiological spectrum of invasive aspergillosis in children: a 10-year review. Pediatr Radiol 33:453–460
18. Wong KS, Hung IJ, Wang CR et al (2004) Thoracic wall lesions in children. Pediatric Pulmonol 37:257–264
19. Lew DP, Waldvogel FA (1997) Osteomyelitis. N Engl J Med 336:999–1007
20. Faro SH, Mahboubi S, Ortega W (1993) CT diagnosis of rib anomalies, tumors, and infection in children. Clin Imaging 17:1–7
21. Levy M, Goldberg I, Fischel RE et al (1981) Friedrich's disease. Aseptic necrosis of the sternal end of the clavicle. J Bone Joint Surg Br 63:539–541
22. Letts M, Davidson D, Birdi N et al (1999) The SAPHO syndrome in children: a rare cause of hyperostosis and osteitis. J Pediatr Orthop 19:297–300
23. Shamberger RC, Grier HE (1994) Chest wall tumors in infants and children. Semin Pediatr Surg 3:267–276
24. Lee KC, Davies AM, Cassar-Pullicino VN (2002) Imaging the complications of osteochondroma. Clin Radiol 57:18–28
25. Karmazyn B, Davis MM, Davey MS et al (1998) Imaging evaluation of chest wall tumors in children. Acad Radiol 5:642–654
26. Glass RB, Norton KI, Mitre SA et al (2002) Pediatric ribs: a spectrum of abnormalities. Radiographics 22:87–104
27. Groom KR, Murphey MD, Howard LM et al (2002) Mesenchymal hamartoma of the chest wall: radiologic manifestations with emphasis on cross-sectional imaging and histopathologic comparison. Radiology 222:205–211
28. Dang NC, Siegel SE, Philips JD (1999) Malignant chest wall tumors in chidren and young adults. J Pediatr Surg 34:1773–1778
29. Laor T (2004) MR imaging of soft tissues tumors and tumor-like lesions. Pediatr Radiol 34:24–37
30. Eich GF, Kellenberger CJ, Willi UV (2008) Radiology of the chest wall. In: Lucaya J, Strife J (eds) Pediatric chest imaging, 2nd edn. Springer-Verlag, Berlin Heidelberg, pp 113–336
31. Shamberger RC, Tarbell NJ, Perez-Atayde AR et al (1994) Malignant small round cell tumors (Ewing's-PNET) of the chest wall in children. J Pediatr Surg 29:179–185
32. Ambros IM, Ambros PF, Strehl S et al (1991) MIC 2 is specific marker for Ewings sarcoma and primitive neuroectodermal tumor for n MIC 2 expression and specific chromosome aberration. Cancer 67:1886–1893
33. Saiffuddin A, Robertson RJ, Smith SE (1991) The radiology of Askin tumours. Clin Radiol 43:19–23
34. García-Peña P, Owens CM (2008) Helical multidector chest CT. In: Lucaya J, Strife J (eds) Pediatric chest imaging, 2nd edn. Springer-Verlag, Berlin Heidelberg, pp 47–75
35. Sharif K, Alton H, Clarke J et al (2006) Paediatric thoracic tumours presenting as empyema. Pediatr Surg Int 22:1009–1014
36. Sallustio G, Pirronti T, Lasorella A et al (1998) Diagnostic imaging of primitive neuroectodermal tumour of the chest wall (Askin tumour). Pediatr Radiol 28:697–702

37. Saenz NC, Ghavimi F, Gerald W et al (1997) Chest wall rhabdomyosarcoma. Cancer 80:1513–1517
38. Kim EE, Valenzuela RF, Kumar AJ et al (200) Imaging and clinical spectrum of rhabdomyosarcoma in children. Clin Imaging 24:257–262
39. Carlsen SE, Bergin CJ, Hoppe RT (1993) MR imaging to detect chest wall and pleural involvement in patients with lymphoma: effect on radiation therapy planning. AJR 160:1191–1195
40. Bergin CJ, Healy MV, Zincone GE et al (1990) MR evaluation of chest wall involvement in malignant lymphoma. J Comput Assist Tomogr 14:928–932
41. Watt AJ (2002) Chest wall lesions. Paediatr Respir Rev 3:328–338
42. Coffin CM, Jaszcz W, O'Shea PA et al (1994) So-called congenital-infantile fibrosarcoma: does it exist and what is it? Pediatr Pathol 14:133–150