

Unusual presentations of lung lesions in children: difficult to diagnose case series

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We present five cases of unusual and rare presentations of different congenital/acquired lung lesions. These cases were encountered during our daily practice. They were misdiagnosed initially and received wrong treatment. There were challenges encountered during the diagnosis and the management of these patients, which required different modalities, ranging from chest radiography, computed tomography to surgical exploration and biopsy, to reach the final diagnosis. These cases range from one absent lung, bilateral intralobar pulmonary sequestrations, bronchogenic cyst, congenital right diaphragmatic hernia to cystic hydatidosis (hydatid cyst). There should be a high index of suspicion when the patient has an abnormal/atypical presentation, a prolonged course of the disease or abnormal imaging. Healthcare providers should also think of rare chest diseases and refer such patients to a paediatric specialist (pulmonologist) to help in the final diagnosis and

specific management, which may require invasive procedures or specific imaging.

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Introduction

We present in this paper five cases of unusual and rare presentations of different lung lesions (congenital/acquired lung lesions), which represent a wide range of chest diseases. There were challenges encountered during the diagnosis and the management of these patients. These patients were initially misdiagnosed with other common illnesses or even received inappropriate or unnecessary treatment. Different modalities of investigations were required, ranging from chest radiography (CXR) and chest computed tomography (CT) up to surgical exploration and biopsy, to reach the final diagnosis. A summary of the following cases is shown in Table 1.

Case 1

A 2.5-year-old female patient presented with sudden-onset wheezes and respiratory distress (subcostal and intercostal retraction) for the past 4 days. She was referred to our hospital with the suspicion of foreign-body (FB) inhalation, although there was no history of choking. There was no history of similar conditions or chest diseases. Her perinatal history (the period immediately before and after birth) was normal. On examination, the respiratory rate (RR) was 25 breaths/min, there was no fever or clubbing, and growth parameters (weight and height) were within 50th percentiles. Breath sounds were absent over the whole left lung, with an impaired percussion note.

CXR was performed (Fig. 1a). FB inhalation was suspected, and so urgent rigid bronchoscopy was

performed, which revealed a sharp carina and an absent right upper lobe bronchus with a blind end of the left main bronchus. Hence, CT chest was recommended (Fig. 1b). Echocardiography revealed a small restrictive subaortic ventricular septal defect.

Final diagnosis: agenesis of the left lung

Comment

Our case was referred with the suspicion of FB inhalation, due to sudden-onset respiratory distress for few days, which was supported by an abnormal CXR finding of compensatory hyperinflation of the left lung, shift of the heart and the mediastinum and crowded ribs of the left side, which were suggestive of a collapsed left lung. However, there was no history of choking and no signs of respiratory distress; however, FB inhalation was highly suspected.

Paediatric airway FB aspiration is associated with a high rate of airway distress, morbidity and mortality, especially in children younger than 3 years of age [2]. The presenting symptoms of FB aspiration may vary depending on its location, size and chronicity. The child may be comfortable and in no apparent distress or may present in extremis with impending airway failure. Coughing, wheezing, shortness of breath, fever and recurrent

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Table 1 A summary of our cases

Age/sexOnset of symptoms	Symptoms	General exam	Chest exam	Initial diagnosis	Diagnostic test	Final diagnosis/management
2.5 Y/F, Sudden for 4 days	Wheezes and RD	RR 25 breaths/min, no fever, clubbing	Absent BS and impaired note over Lt side	FB inhalation	CXR, CT chest bronchoscopy	Lt lung agenesis/conservative
11 Y/MChronic since the age of 4 months	Recurrent respiratory infections, productive cough, dyspnoea and intermittent fever	RR 28 breaths/min, no pallor, cyanosis, clubbing, or palpable lymph nodes	Barrel chest, retractions, bilateral basal consonating crepetations	BA, supprutive lung disease, immunodeficiency	CXR, CT/angiography biopsy	Intralobar pulmonary sequestration/surgery
^a 4.5-month/FG gradual since 1 month age [1]	Noisy breathing with feeding difficulty	RR 60 breaths/min, HR 140 beats/min; O ₂ saturation 94%, inspiratory stridor, no cyanosis, fever	Retractions, hyper-resonant percussion, inspiratory and expiratory sibilant ronchi	Hyper-reactive airway, BA, FB inhalation	CXR bronchoscopy, CT chest	Bronchogenic cyst/surgery
3.5 Y/MRecurrent for few weeks	Recurrent wheezes, chest infections and abdominal distension	RR 30 breaths/min, HR 90 beats/min, no respiratory, distress, fever or cyanosis	Diminished BS over Rt lung base and BB over Rt middle zone	Lung abscess, hydropneumothorax	CXR, CT chest	Congenital diaphragmatic hernia/surgical correction
4 Y/MIntermittent over 3 weeks	Low-grade fever, RD, cough, progressive abdominal enlargement and Rt abdominal pain	Toxic look, RR 40 breaths/min, no fever, Pallor, jaundice, cyanosis, abdominal enlargement, hepatomegaly	Diminished BS over Rt middle and basal lung, impaired percussion note	Pneumonia/unresolved residual pneumonia, tumor	CXR, CT chest/abdomen, abdominal US pathology	Hydated cyst/antiparasitic albendazole

BA, bronchial asthma; BB, bronchial breathing; BS, breath sound; CT, computerized tomography; CXR, chest radiography; F, female; FB, foreign body; HR, heart rate; Lt, left; M, male; RR, respiratory rate; Rt, right; US, ultrasound; Y, years. ^aPreviously published: Zedan *et al.* [1].

pneumonia may each be the presenting symptoms. Parental recall of a choking or gagging event followed by a cough is highly suspicious. However, this initial event may be short lived and the child may be asymptomatic for one or more weeks, often leading the parents to forget about the inciting episode [3].

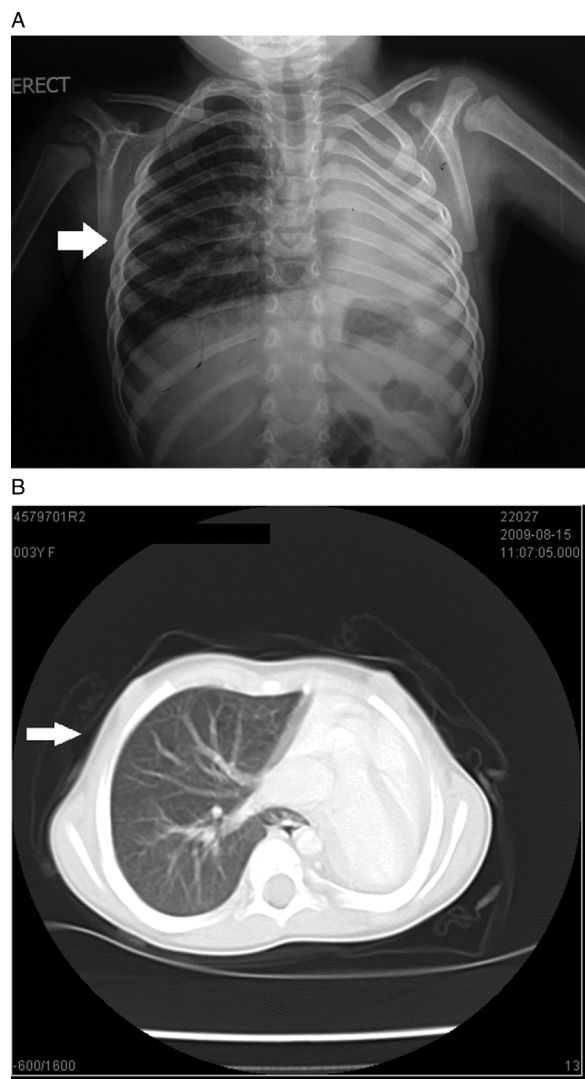
The physical examination can be nonspecific and the radiological evaluation normal, which may result in delayed diagnosis. An elevated index of suspicion is necessary in all cases of pneumonia, atelectasis or wheezing with atypical courses. Early diagnosis is fundamental for optimal management, consisting of the removal of the FB as soon as possible to prevent respiratory sequelae. Localized wheezes or diminished breathing sounds from the affected lung are the main findings [4].

Cervical and thoracic radiographical evaluation is the most important investigation in every patient suspected of having an FB aspiration. Radiographical abnormalities are more frequent when the FB is endobronchial. This location is also more frequent in cases of delayed diagnosis and delayed removal. An expiratory CXR should be requested (in cooperative children) when the standard

inspired film is normal, as this strategy allows the visualization of air trapped by a valve-like effect due to partial obstruction of the bronchial lumen. A mediastinal shift may also be seen. The most frequent radiological findings are obstructive emphysema, atelectasis and consolidation. However, normal findings may occur [4]. CT scans can be used in patients with prolonged signs and symptoms to identify early sequelae or to detect radiolucent materials [5]. Rigid bronchoscopy was performed for the suspected FB, but the amazing finding of an absent right upper lobe bronchus and a blind end of the left main bronchus suggested an abnormal lung pathology or development, which required a CT scan for the chest. CT chest showed a compensatory hyperinflated right lung and an absent left lung, confirming the final diagnosis.

Regarding pulmonary agenesis, the onset of symptoms is remarkably variable. In many cases, the presence of this anomaly usually comes to light during infancy because of recurrent chest infections, cardiopulmonary insufficiency or due to associated congenital anomalies. However, patients with one lung have been reported to survive well into adulthood without much complaints [6].

Figure 1



(a) CXR AP reveals a left opaque hemithorax, a hyperinflated right lung (arrow) and shift of the heart and the mediastinum to the left side. (b) CT chest demonstrates an hyperinflated right lung (arrow) with the heart occupying the left side of the chest. AP, anteroposterior; CT, computed tomography; CXR, chest radiography.

The exact aetiology of this condition is unknown, although genetic factors, viral agents and dietary deficiency of vitamin A during pregnancy have been implicated. Left-sided agenesis is more common and these individuals have a longer life expectancy than those with right-sided agenesis. This is probably due to the excessive mediastinal shift and malrotation of the carina in right-sided agenesis, which hinders proper drainage of the functioning lung and increases the chances of respiratory infections [6]. Nearly 50% of the cases of pulmonary agenesis have associated congenital defects, involving the cardiovascular, the skeletal, the gastrointestinal and the genitourinary systems [7]. Echocardiography for our patient revealed a small ventricular septal defect as an associated congenital anomaly.

The differential diagnosis of lung agenesis also includes lung collapse, a thickened pleura, a destroyed lung and pneumonectomy. The final diagnosis can be established after bronchoscopy and/or bronchography. Surgery is seldom required for agenesis or aplasia, as it can be managed on conservative lines. The prognosis in these cases depends on the functional integrity of the remaining lung and the presence of associated anomalies [6].

Case 2

An 11-year-old male patient presented with recurrent episodes of respiratory tract infections (up to two to three episodes per month) since the age of 4 months. He had productive cough of excess whitish to greyish purulent offensive sputum, associated with dyspnoea and intermittent fever. There was no weight loss or chest pain. The patient was previously diagnosed to have bronchial asthma, but without response to bronchodilators or steroids. There was no family history of chest problems, with an irrelevant neonatal history.

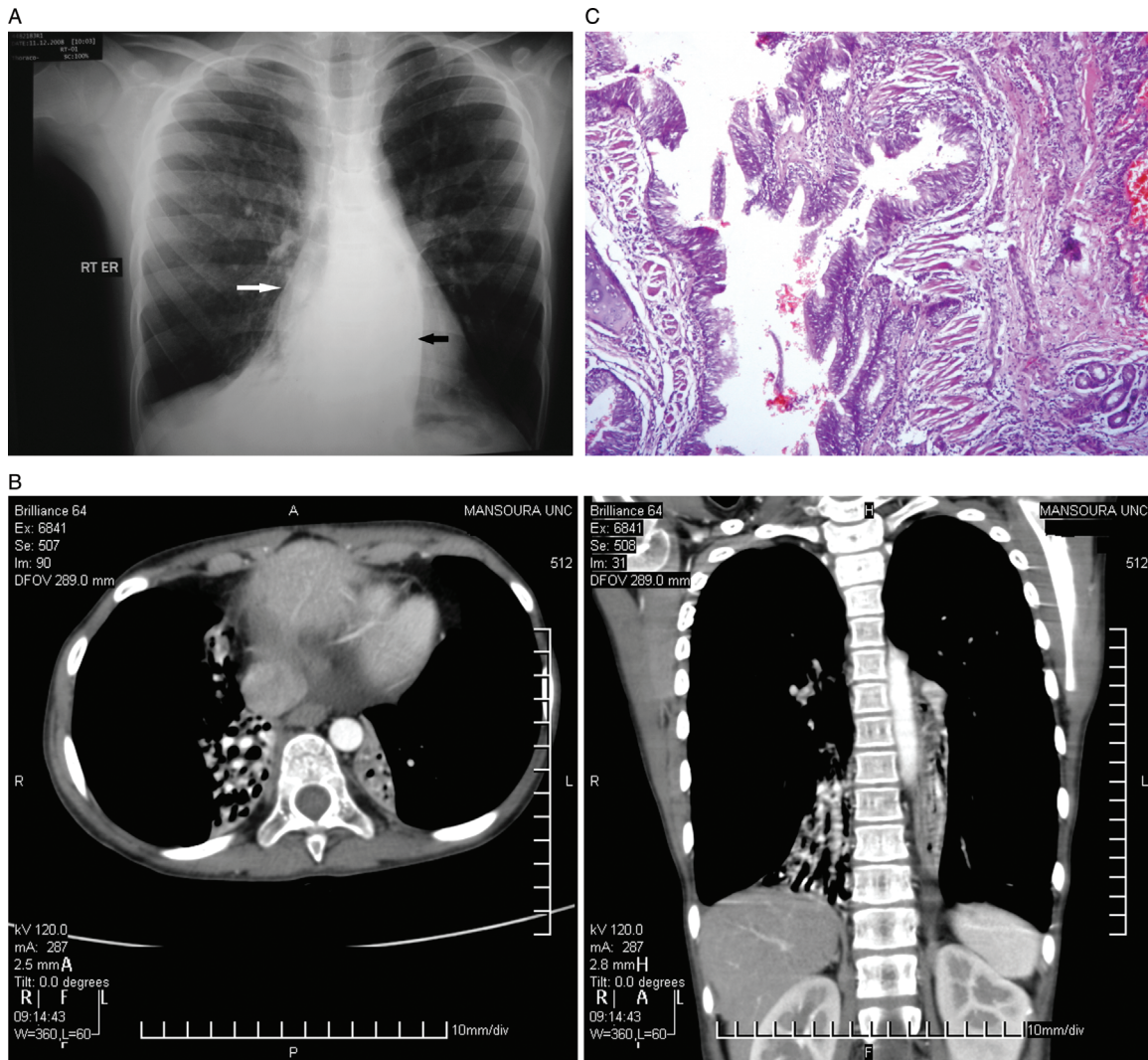
The patient had average growth parameters. On examination, there was no pallor, no cyanosis, no clubbing or palpable lymph nodes. His vital signs were as follows: temperature, 37.0°C; heart rate (HR), 110 beats/min; RR 28 breaths/min, with an average blood pressure for his age and height. Chest examination revealed a barrel-shaped chest, intercostal retractions, with a central trachea. There were bilateral basal medium-sized consonating crepitations.

The white blood cell count was 9000/mm³ (66% polymorph). CXR (Fig. 2a) showed consolidation collapse of the medial segment of the right middle lung lobe. Multislice CT and CT angiography of the chest showed multicystic air-filled lesions occupying the medial segment of the right lower lung lobe and the posterior basal segment of the left lower lobe (Fig. 2b).

Bronchoscopy was performed and the bronchoalveolar lavage revealed suppurative smears with no atypical cells.

Surgical excision of the lesions was performed in two sessions. Pathologic findings of the excised mass revealed acute and chronic inflammatory changes with purulent exudates and variable-sized spaces lined with cuboidal and pseudostratified columnar epithelium, a few smooth muscle bundles and cartilaginous bronchus-like structures (Fig. 2c).

Figure 2



(a) A preoperative CXR film showing segmental consolidation collapse of the medial segment of the right middle lung lobe (white arrow) and retrocardiac collapse in the left side (black arrow). (b) CT chest angiography demonstrates multicystic air-filled lesions occupying the medial segment of the right lower lung lobe and the posterior basal segment of the left lower lobe. These lesions were supplied by descending branches of both the right and the left pulmonary arteries and drained into the right and the left pulmonary veins. The angiographic diagnosis was bilateral intralobar PS. (c) Pathological examination of the excised PS shows acute and chronic inflammatory changes with purulent exudates and variable-sized spaces lined with cuboidal and pseudostratified columnar epithelium (arrow), a few smooth muscle bundles and cartilaginous bronchus-like structures (H&E stain, $\times 200$). CT, computed tomography; CXR, chest radiography; PS, pulmonary sequestration.

Final diagnosis: intralobar pulmonary sequestration

Comment

Pulmonary sequestration (PS) is a congenital malformation consisting of a nonfunctioning bronchopulmonary mass separated from the normal pulmonary tissue. Hence the term PS is applied to a pulmonary lobe or a portion of a lobe that is supplied by an anomalous systemic artery and drains either into the systemic or into the pulmonary veins. The conditions are divided into intralobar PS, in which the sequestration is situated inside the visceral pleura of a normal lobe, and extralobar PS, in which the sequestration is surrounded by its own pleura. Most sequestrations are unilateral; bilateral ones are rare [8].

The child with recurrent chest infections presents the clinician with a difficult diagnostic challenge. Does the child have a simply managed cause for his symptoms, such as recurrent viral respiratory infections or asthma, or is there evidence of a more serious underlying pathology, such as bronchiectasis, PS, immune deficiency, cystic fibrosis and ciliary abnormalities [9].

Repeated episodes of pneumonia are often the presenting feature of structural airway abnormalities (localized bronchial stenosis, bronchomalacia, tracheobronchus and bronchiectasis) or parenchymal lung lesions [PS, cystic adenomatoid malformation and

Table 2 Differential diagnosis of congenital cystic lung disease and parenchymal lung lesions presented as recurrent pneumonia [1]

Disease	Pathogenesis	Classification	Clinical aspect	Radiology
Bronchogenic cyst	Remnants of the primitive foregut containing tissue normally found in the trachea and bronchi	Single, multilocular or multiple location: paratracheal, carinal, paraoesophageal, hilar	Early childhood, but symptoms can develop at any age cough, stridor or wheezing	Cyst appears as an ovoid soft tissue density
Pulmonary sequestration	Mass of nonfunctional embryonic lung tissue characterized by bronchiectasis and varying degrees of cystic changes and is usually supplied by an aberrant systemic artery	Intralobar: lies within a lobe of the lung invested by its visceral pleura Extralobar: has its own investing pleura outside the normal lung parenchyma	Intralobar: asymptomatic productive cough, fever, haemoptysis, recurrent pneumonia and chest pain Extralobar: newborns, frequently associated with other congenital anomalies infection (when there is oesophageal or gastric communication)	Soft tissue mass in the posterior basal segment of the lung
Congenital cystic adenomatoid malformation (CCAM)	Solid, cystic or mixed masses that communicate with the normal tracheobronchial tree	Type 1 (50%): one or more large cysts (2–10 cm) Type 2 (40%): multiple smaller cysts (0.5–2 cm) Type 3 (10%): solid on gross examination, but contain multiple 0.3–0.5 cm cysts on microscopic examination	Newborn: progressive respiratory distress After the neonatal period: recurrent pulmonary infections	Multiple air or air fluid-filled, thin-walled cysts that vary in size
Congenital lobar emphysema (CLE)	Overdistension of (usually) one lobe The most constant pathological finding is cartilage deficiency resulting in bronchomalacia	Left upper lobe: 41% Right middle lobe: 34% Right upper lobe: 21% Bilateral: 20%	Progressive, severe respiratory, distress in infants, with wheezing, cough or recurrent chest infection	Overinflation of the affected lobe

bronchogenic cysts (BC)] [10]. Parenchymal lung lesions should be suspected if one lobe is repeatedly infected or if there is incomplete resolution after treatment. CT and magnetic resonance scanning are helpful in defining the anomaly before surgical excision [11]. Differentiating features of such parenchymal lesions are shown in Table 2.

There are many different causes of recurrent chest infections in children. The clinician has to distinguish between children with self-limiting conditions and those with more severe, progressive, diseases such as parenchymatous lung disease. Although PS is a rare condition, early and accurate diagnosis by CT angiography is essential to ensure that optimal treatment is given and to minimize the risk of progressive or irreversible lung damage.

Case 3

A 4.5-month-old female infant was presented with gradual onset of noisy breathing in association with feeding difficulty. Her symptoms started since 1 month of age with no cough, fever or cyanosis. Her noisy breathing sounds were interpreted as wheezes and treated as asthma, suggested by a positive family history of an older asthmatic brother, with no response to bronchodilators.

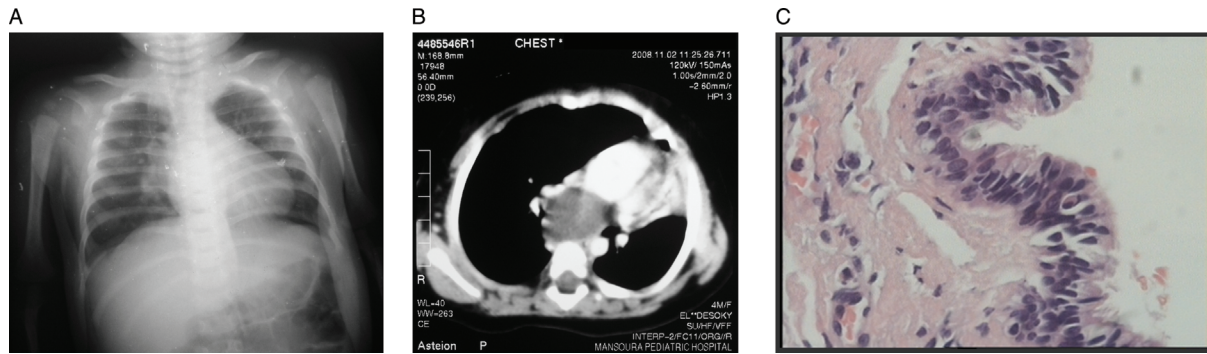
On presentation, she had inspiratory stridor and no cyanosis. She had no fever, the HR was 140 beats/min and RR 60 breaths/min. Her oxygen saturation was 94%. Her growth parameters were normal. Chest examination revealed a normal shape, with suprasternal and subcostal retractions. She had a centrally posed trachea and a hyper-resonant percussion note. Her breath sounds were harsh vesicular with bilateral inspiratory and expiratory sibilant rhonchi.

CXR revealed bilateral hyperinflation (Fig. 3a). Fiberoptic bronchoscopy revealed a wide carina and narrowed right and left main bronchi with external compression on the posterior and the medial walls, caused by a posterior mediastinal mass. CT chest was recommended, which revealed a posterior mediastinal cystic lesion (2–3 cm), suggesting a BC, a duplication cyst or a neuroenteric cyst (Fig. 3b).

The cyst was removed surgically. Pathological examination demonstrated a cystic structure lined with pseudostratified ciliated columnar epithelium (respiratory mucosa). The cyst wall consisted of fibrous tissue, smooth muscle fibres and nerve fibres, with no malignant transformation (Fig. 3c).

After surgery, the infant was stable with no retraction or stridor and a normal chest percussion note. Her

Figure 3



(a) CXR AP shows bilateral overinflation. (b) A CT scan of the chest shows a well-defined, spherical, cystic lesion in the mediastinum. (c) Pathological examination of the wall of the resected cyst shows the respiratory epithelium lining with a pseudostratified columnar epithelium with cilia (H&E stain, $\times 400$). AP, anteroposterior; CT, computed tomography; CXR, chest radiography.

breath sounds turned normal vesicular and equal on both sides.

Final diagnosis: a bronchogenic cyst

Comments

Congenital cystic lung diseases are rare, and vary in presentation and severity. They consist of PS, cystic adenomatoid malformation, congenital lobar emphysema and BC. These four anomalies present a different clinical picture, are often difficult to diagnose and require surgical management [12]. Although there are many similarities in terms of their presenting features, there are particular differences between the diagnostic groups that are important to highlight, especially in relationship to the approaches to imaging and the long-term outcome [13,14] (Table 1).

BCs are remnants of the primitive foregut containing tissue normally found in the trachea and the bronchi [15]. Maier [16] classified BCs by anatomic locations: hailer, paraoesophageal, tracheal, carinal or miscellaneous. Carinal cysts are most likely to produce respiratory problems and are therefore the most frequently reported of BCs [17].

Up to one-third of the BCs remain asymptomatic [13]. Symptomatic cases usually manifest in early childhood, but symptoms can develop at any age. BCs can cause symptoms either due to mass effects or due to direct pressure on an airway leading to air trapping. Secondary infection can also occur. Two-thirds of the patients are diagnosed due to symptoms such as cough, wheezing, stridor, dyspnoea, dysphagia or even chest pain [14].

BCs can cause stridor, which is an alarming and prominent symptom that requires prompt management,

especially in newborns and young children. The differential diagnosis of stridor in infants includes congenital lesions of the upper and the lower airway, inflammatory or neoplastic lesions, vocal cord paralysis, FB and vascular lesions. The presence of mediastinal BCs should also be included in this list [13].

The diagnostic workup of each child presenting with noisy breathing and/ or stridor includes a CXR and fluoroscopy with a barium oesophagogram (or the less-irritant water-soluble gastrographin, especially for infants). If the CXR and the oesophagogram are negative, upper-airway endoscopy is mandated [18].

Case 4

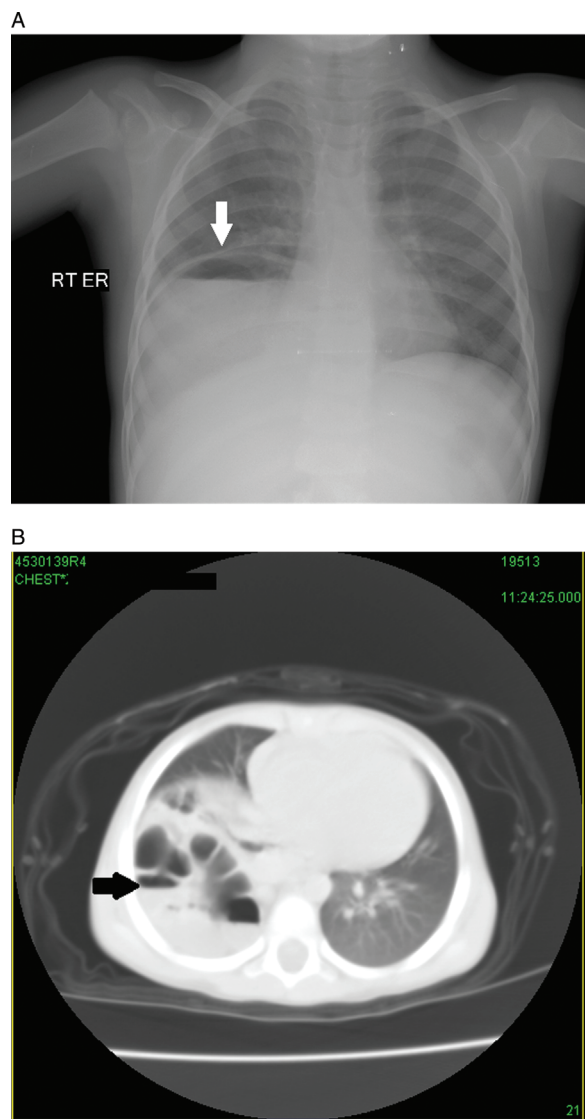
A 3.5-year-old previously healthy male child was presented with the complaint of recurrent wheezes, chest infections and abdominal distension. He received salbutamol nebulizer treatment several times without improvement. At the time of admission, the RR was 30 breaths/min and the HR 90 beats/min. There was no respiratory distress, fever or cyanosis.

Examination revealed no signs of respiratory distress, but there was a diminished breath sound over the right lung base. Bronchial breathing was heard over the right middle lung zone with no dullness on percussion.

Laboratory investigation showed a total leukocytic count of 5900 cells/mm³ (lymphocytes 67%), hemoglobin 12 g/dl and platelets 203 000/ μ l. The C-reactive protein was negative and arterial blood gases were normal.

CXR of the right-sided hydropneumothorax/subphrenic abscess was suggested (Fig. 4a). Hence,

Figure 4



(a) CXR AP shows air and fluid levels with a thin wall (arrow) right-sided hydropneumothorax/subphrenic abscess. (b) CT chest reveals multiple dilated intestinal loops in the right lower lung (arrow), mainly posterior, with a mild leftward mediastinal shift. The size and the shape of the left lung were normal, with normal parenchyma on the left lung. No masses or abnormal cystic air spaces were present. AP, anteroposterior; CT, computed tomography; CXR, chest radiography.

she was started on vancomycin infusion for 1 week without change in the radiological findings; hence, CT chest was recommended. CT chest revealed multiple dilated intestinal loops in the right lower lung, mainly posterior, with a mild leftward mediastinal shift. The size and the shape of the left lung were normal, with normal parenchyma on the left lung. There were no masses or abnormal cystic air spaces (Fig. 4b).

The patient was referred to the paediatric surgery department for surgical correction of diaphragmatic hernia.

Final diagnosis: delayed presentation of right-sided congenital diaphragmatic hernia

Comments

Congenital diaphragmatic hernia (CDH) is generally regarded as a neonatal emergency. There is, however, a small percentage of patients, who present beyond the neonatal period. Late-presenting CDH is associated with a much wider spectrum of clinical presentation, encompassing various combinations of respiratory and gastrointestinal symptoms. The serious respiratory distress occurring in CDH is related to pulmonary hypoplasia, often bilateral, combined with persistent foetal circulation and mechanical respiratory disorders [19]. In 5–10% of the patients, the presentation of CDH is delayed, whereas 7–10% of the patients have no symptoms [20,21]. Most asymptomatic CDH are right-sided, with the diaphragmatic defect covered by the liver [20]. Patients with right CDH often live longer, are asymptomatic and have a normal life [22]. The overall survival of patients with left CDH is about 60% (which makes left CDH a serious neonatal problem associated with high mortality) [23].

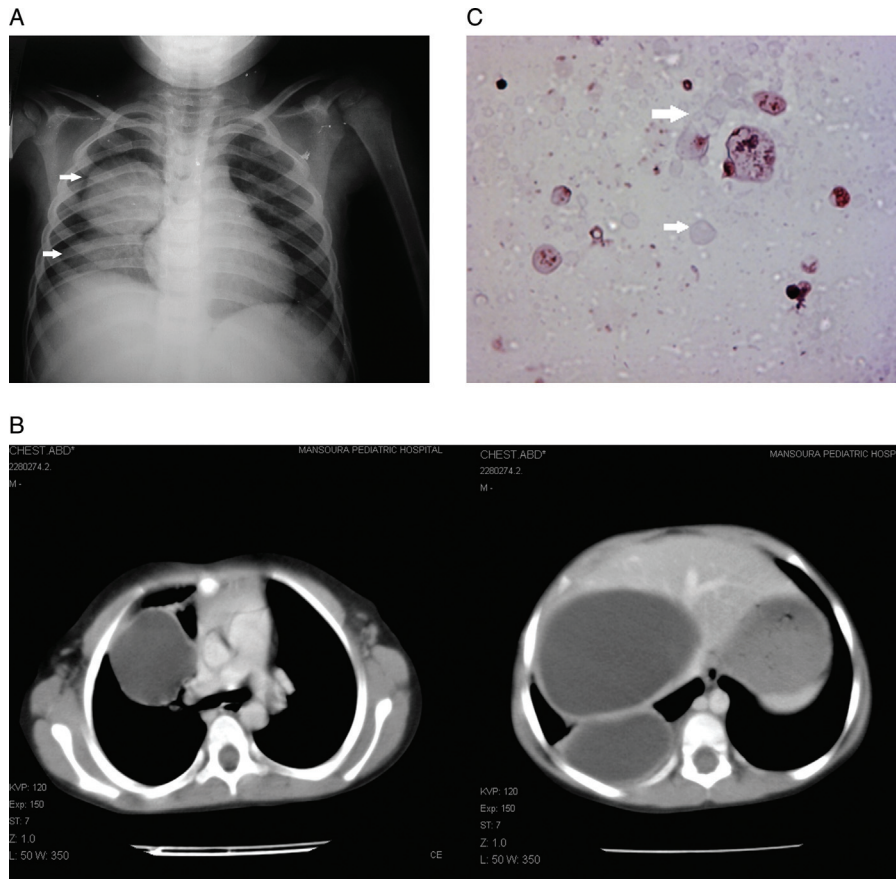
The clinical presentation of delayed CDH is not specific and the diagnosis is often overlooked. There is also a very good respiratory compensatory mechanism [20,24].

Our case was misdiagnosed initially as a lung abscess due to suggestive CXR. Lung abscess is an uncommon condition that can occur at any age. It is believed to be less common in children than in adults. Fever and cough consistently predominate, but are not universal. Unlike the situation with adults, haemoptysis is uncommon as a presenting feature in children with lung abscess. The physical signs elicited in a child with lung abscess most commonly include tachypnoea, a dull percussion note or reduced air entry locally, fever and localized crepitations [25]. The basic diagnostic test for a lung abscess is CXR. However, to distinguish a lung abscess from an empyema, necrotizing pneumonia, sequestration, pneumatocele or an underlying congenital abnormality such as a BC, a contrast-enhanced CT scan is usually considered to be the investigation of choice [26,27].

Case 5

A 4-year-old male child was presented with low-grade fever, respiratory distress and cough for about 3 weeks. There was no history of traveling abroad or of animal contact. He was diagnosed in a general hospital as having pneumonia and received antibiotics and antipyretics for 3 weeks, but with no improvement. His mother noticed

Figure 5



(a) Chest radiograph shows two opacities related to the right lung (arrows). (b) CT chest and abdomen shows multiple cystic lesions in the lung and the liver. (c) Pathological examination of the sampled fluid shows scattered RBCs, lymphocytes with scattered inflammatory cells and many rounded structures with amorphous appearance are seen (arrow); some of them contain rounded eosinophilic granules (picture suggestive of hydatid disease). CT, computed tomography; RBCs, red blood cells.

progressive abdominal enlargement, and the child complained of right hypochondrial pain. An abdominal ultrasound was performed, which revealed a mildly enlarged liver with a large well-defined hypoechoic cystic lesion in the right lobe measuring 8×7 cm with a clear fluid inside and no internal echoes, which cope with a large simple benign cyst.

The patient was referred to our hospital for further evaluation of nonresponding residual pneumonia.

On admission, he had a toxic look with pallor, and the RR was 40 breaths/min (mild tachypnea). No fever was documented. There was no jaundice, cyanosis, purpura/echymosis or oedema in the lower limbs.

An abdominal examination showed diffuse abdominal enlargement, hepatomegaly (the right lobe span was 12 cm and the left span was 5 cm below the costal margin), firm in consistency with rounded borders with no splenomegaly or ascites.

A chest examination revealed diminished breath sound with an impaired note on percussion over the right middle and the basal lung zone.

A CXR was performed (Fig. 5a), which revealed a well-defined homogenous rounded lesion in the right middle lung zone and a nonhomogenous opacity occupying the right lower lung zone. Laboratory results showed a total leucocytic count of 19 000/ μ l (lymphocytes 18%, polymorphs 71%, eosinophils 2%, monocytes 9% and basophils 0%), hemoglobin 12.9 g/dl, platelets 342 000/ μ l, ESR 65/95 and the C-reactive protein was 50 mg/dl, with a normal serum creatinine of 0.6 mg/dl.

A CT chest and abdomen (Fig. 5b) revealed a large well-defined cyst in the right middle lung lobe related to the anterior chest wall measuring about 6 cm in diameter; another similar cyst measuring 6×10 cm was seen in the posterior segment of the right lower lung lobe and a third cyst in the right liver lobe (mainly the anterior sector), measuring 9×6×6.6 cm. All cysts showed an outer

thin nonenhancing wall with content of near water density. CT brain was normal.

A malignant liver cyst or a mass with lung metastasis was suspected, and so an ultrasound-guided cytological aspiration of the liver cyst was performed, which revealed scattered red blood cells, lymphocytes with scattered inflammatory cells and many rounded structures with an amorphous appearance were seen; some of them contain rounded eosinophilic granules (picture suggestive of hydatid disease; Fig. 5c).

He was referred to the Infectious Disease Unit and he received albendazole 15 mg/kg/day for 6 months, which caused marked improvement with regard to his symptoms. Surgical excision was not possible initially because of multiple lesions in the lung and the liver, and so it was planned to be performed after the end of the medical treatment, but the patient was lost to follow-up.

Final diagnosis: cystic hydatidosis (multiple hydatid cysts)

Comments

Human cystic echinococcosis (CE) (cystic hydatidosis) is a chronic zoonotic disease that results from infection with the larval stage of the dog tapeworm, *Echinococcus granulosus*. The disease is highly endemic in most of the countries of the Mediterranean basin, including North Africa and the Middle East [28,29].

In human CE, the liver is the main organ affected, followed by the lung tissue [30].

Patients with hepatic CE frequently exhibit no symptoms because of the slow progression of the cysts [29]; therefore, they may be discovered only accidentally and frequently have complicated forms of the disease. Pulmonary hydatidosis may be revealed during thoracic radiography.

Most symptoms of pulmonary CE are caused by mass effect from the cyst volume, which exerts pressure on the surrounding tissues. The most common symptoms described by the literature are cough (53–62%), chest pain (49–91%), dyspnoea (10–70%) and haemoptysis (12–21%). Other symptoms described less frequently include dyspnoea, malaise, nausea and vomiting and thoracic deformations [31]. The majority of the children and adolescents with lung lesions are asymptomatic despite having lesions of impressive size, assumedly because of a weaker immune

response and the relatively higher elasticity of the lung parenchyma in children and teenagers [32,33].

Radiological studies are the primary step in the detection and the evaluation of pulmonary CE cysts. In CXRs, cysts are well defined as a rounded mass of uniform density that occupies a part of one or of both hemithorax. Combined chest and abdominal CT is a better tool to recognize certain details of the lesions and their surrounding structures, helping to exclude alternative differential diagnoses and can also uncover additional smaller cysts that were not detected by conventional CXR [34,35].

Surgery is the main therapeutic approach. Surgical treatment of CE has two goals: to remove the parasite safely, to prevent intraoperative dissemination and to treat the bronchipericyst pathology and other associated lesions [36,37].

Surgery may involve excision of the cyst or resection of the cyst and the immediate surrounding parenchyma. Despite the lack of consensus, the currently most accepted surgical treatment for lung CE is complete excision using parenchyma-preserving methods, such as cystostomy, intact cyst enucleation or removal after needle aspiration, preserving as much lung parenchyma as possible [38,39].

Resection techniques such as pneumonectomy and segmentectomy should be reserved to cysts involving the whole hemithorax or the whole segment, respectively, and lobectomy should be performed only in large abscessed cysts. To avoid recurrences, the use of presurgical chemotherapy reduces the chances of seeding and recurrence [40].

In children, for simple, accessible cysts, ultrasound or CT-guided percutaneous aspiration, instillation of a scolicidal agent and reaspiration is the preferred therapy. For conventional surgery, the inner cyst wall (laminar and germinal layers) can be peeled easily from the fibrous layer. The cavity should then be topically sterilized and either closed or filled with omentum. Nonpregnant patients with cysts not amenable to aspiration or surgery or with contraindications can be managed with albendazole (15 mg/kg day divided twice daily orally for 1–6 months maximum 800 mg/day). A favourable response occurs in 40–60% of the patients. Alveolar hydatidosis is frequently incurable by any modality, but radical surgery such as partial hepatectomy or lobectomy may cure early limited disease. Liver transplantation is also an option for disease confined to the liver. Medical therapy with

albendazole may slow the progression of alveolar hydatidosis, but if at all feasible, removal of the infected tissue provides the best outcome. Some patients have been maintained on long-term suppressive therapy, but the infection generally recurs if albendazole is stopped [41].

Conclusion

Different congenital and acquired lung lesions can manifest with a wide spectrum of presentations and are an important cause of morbidity and mortality in infants and children. There are many difficulties and challenges encountered when confronted with such cases for the first time. There is always a need for following evidence-based management protocols for children with persistent respiratory symptoms. The diagnosis and the management of these patients usually required different modalities to reach the final diagnosis. However, there should be a high index of suspicion when the patient has an abnormal/atypical presentation, a prolonged course of the disease or abnormal imaging findings. Different available modalities ranging from CXR, CT, MRI to surgical exploration and biopsy should be used to reach the final diagnosis.

Healthcare providers (Paediatricians/General Practitioners) should also think of rare chest diseases. They should also refer such patients to a paediatric specialist (pulmonologist) to help in the final diagnosis and specific management, which may require invasive procedures or specific imaging.

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Conflicts of interest

There are no conflicts of interest.

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