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Case 1: Postinfectious Bronchiolitis Obliterans

Javier Lucaya and Joaquim Piqueras

A 5-year-old boy who, 1 year prior to admission, had suffered adenovirus-induced pneumonia (Fig. 2.1). Since then, he has experienced frequent episodes of cough. Physical examination on admission revealed decreased breath sounds and some crackles in the left hemithorax. Inspiratory and expiratory AP chest X-rays and lung CT were obtained.



Fig. 2.1

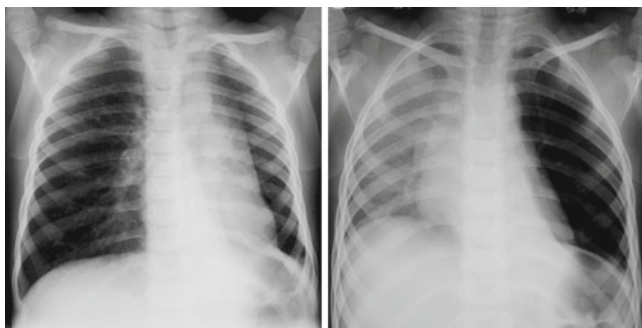


Fig. 2.2

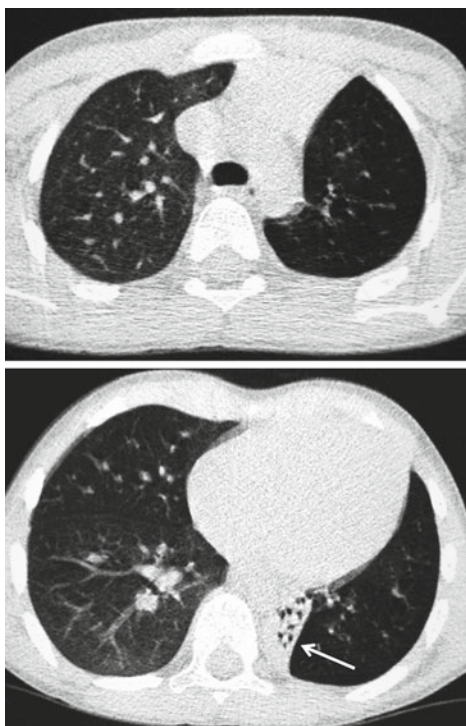


Fig. 2.3

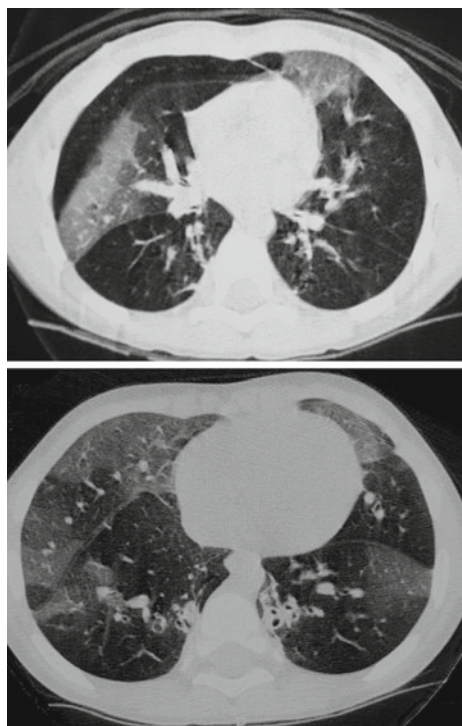


Fig. 2.4

Bronchiolitis obliterans (BO) is an uncommon and severe form of chronic obstructive lung disease in children and adults that results from an insult to the lower respiratory tract. Pathologically, BO is characterized by bronchiectasis of the large airway and obliteration of the small airways. Known etiologies include infection, particularly adenovirus and mycoplasma pneumonia, aspiration, toxic inhalation, lung and bone marrow transplantation, and connective tissue disorders. Presenting signs and symptoms include chronic cough, wheezing, exercise intolerance, tachypnea, and frequent respiratory illnesses. Crackles are a very common finding on auscultation.

A classic manifestation of postinfectious BO is the Swyer-James-MacLeod (SJM) syndrome. First thought to be of congenital origin, SJM is now considered a postinfectious form of BO.

In children with postinfectious BO, chest X-rays may show five different patterns:

- (a) Unilateral hyperlucency of increased volume.
- (b) Complete collapse of the affected lobe.
- (c) Unilateral hyperlucency of small- or normal-sized lung.
- (d) Bilateral hyperlucent lungs.
- (e) Mixed pattern of persistent collapse and hyperlucency and peribronchial thickening (Fig. 2.2). When assessing a chest X-ray showing an asymmetrical degree of pulmonary aeration, the lung showing more vascularity is usually the normal one.

Chest CT is the imaging technique of choice. Characteristic diagnostic features include pulmonary hyperlucency and vascular attenuation, bronchiectasis, and/or a mosaic attenuation pattern. Lobar collapse, usually associated with bronchiectasis (*arrow*), occurs in 20% of cases (Fig. 2.3). Bilateral involvement (Fig. 2.4) of another patient will be present in 50% of cases. The involved areas on the high-resolution computed tomography correspond to those showing infiltrates on chest X-rays at the time of the initial pneumonia.

In children, the finding of a mosaic pattern of lung attenuation is practically synonymous with peripheral airway obstruction and is most commonly seen in asthma and BO. In our experience, the finding of associated bronchiectasis strongly favors the diagnosis of BO.

Pulmonary function tests (PFT) continue to be an important diagnostic tool in patients with BO. Unlike what occurs in asthma, patients with BO usually fail to respond to bronchodilator therapy. The results of PFTs in patients with BO characteristically show irreversible obstructive lung disease. The diagnosis of BO is usually established by radiologic and PFT findings but may be missed by lung biopsy due to sampling error.

Comments

Imaging Findings

Case 2: Thoracic Rib Invasion from Actinomycosis

Paul Marten and Rodrigo Domínguez

A 38-month-old girl with findings on computed tomography (CT) (Fig. 2.5) of multiple rib periostitis as an otherwise subtle chest wall spread



Fig. 2.5

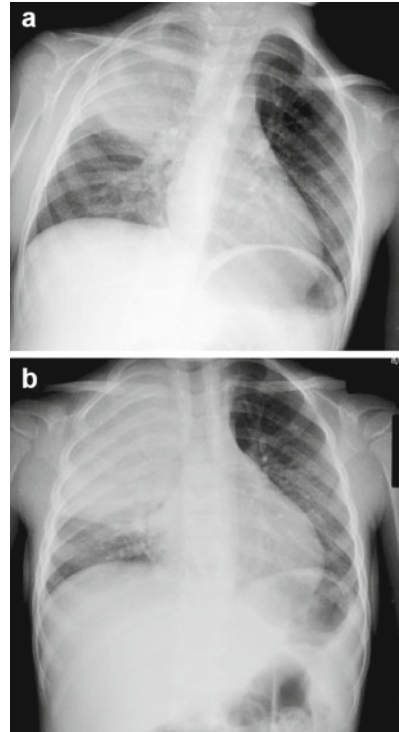


Fig. 2.7

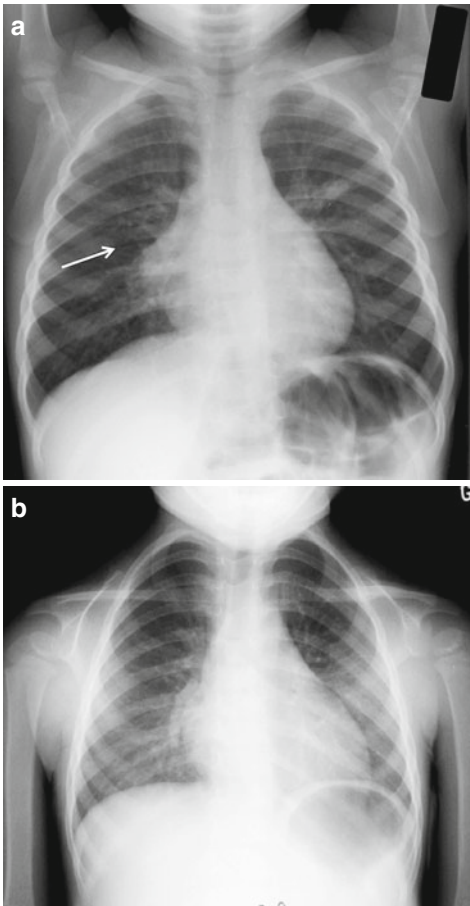


Fig. 2.6

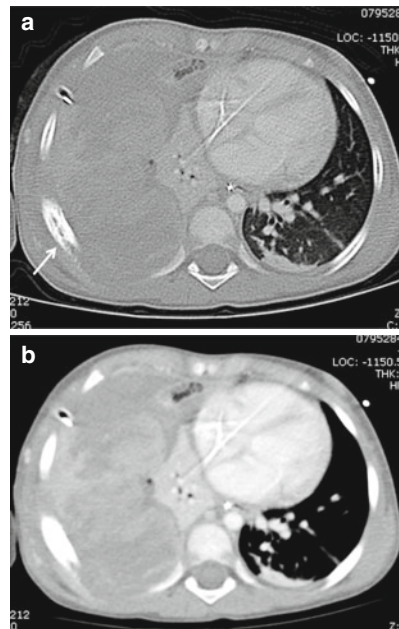


Fig. 2.8

of a presumed pneumonia. Previously, the patient had been treated as an outpatient for pneumonia at age 20 months. Subsequently, the findings became more widespread and in the very same location at 34 months, and kept worsening despite treatment, which prompted her admission.

Actinomycosis is an uncommon lower respiratory tract infection with aggressive features known to invade the chest wall and is associated with periostitis. Other aggressive pathogens which may be associated with chest wall invasion include tuberculosis or *Nocardia*. Particularly, if associated with empyema necessitans, other infections such as fungal (*Aspergillosis*, *Mucormycosis*, *Cryptococcosis*, etc.), *Staphylococcus aureus* (most common cause of pediatric osteomyelitis), *Streptococcus pyogenes*, etc.

Given the chronicity and resistance to treatment, two major differential categories should be considered: resistant infection with probable immune deficiency or tumor. Tumors considered are: Langerhans cell histiocytosis, Askin/PNET, primary bone tumor (Ewing more than osteosarcoma), rhabdomyosarcoma/other sarcomas, desmoid, lymphoma, or leukemia – often disseminated. Pleuropulmonary blastoma or metastatic tumors would be rare.

In our case, this patient tested negative for HIV with normal immunity but was prone to repeat infections from an undiagnosed non-cystic, non-cystic (solid) congenital cystic adenomatoid malformation. *Actinomyces israelii*, a slow-growing Gram-positive bacterium, often affects the gastrointestinal tract and head and neck. Pathologically, *Actinomycosis* and *Nocardia* behave similar to fungal infections causing solid peripheral mass, with *Actinomycosis* more prone to cause periostitis (like tuberculosis). Diagnosis is by biopsy and anaerobic culture.

Common imaging findings in *Actinomyces* infection are:

- Peripheral dense consolidation (non-segmental, often in the lower lobes) or mediastinal mass.
- Solid and slow progression consolidation. Cavitation and even fibrosis if left untreated.
- If the consolidation is untreated, it dissects along tissue planes with cutaneous sinus tracts with yellow thick pustulent drainage (with sulfur granules) and pleuritis with chest wall abscess.
- Pleural effusions and/or pericardial effusion.
- Multiple rib periostitis, +/- rib destruction, and severe rib eburnation.

(Fig. 2.5). Contrast-enhanced CT with solid parenchymal mass with subtle wall extension with posterior rib periostitis (arrows). Initial pneumonia at age 20 months (arrow) (Fig. 2.6a) appears resolved by age 29 months (Fig. 2.6b). (Fig. 2.7). Recurrence age 34 months (Fig. 2.7a), worsening and not responsive to antibiotics 38 months (Fig. 2.7b). Comparison with case of Ewing's sarcoma: note periostitis is confined to one rib (arrow) with more heterogeneous pleural mass and atelectasis right lung. No significant effusion (Fig. 2.8a, b).

Comments

Imaging Findings

Case 3: Non-Cystic Fibrosis Bronchiectasis

Estela Pérez Ruiz and Pilar Caro Aguilera



Fig. 2.9



Fig. 2.10

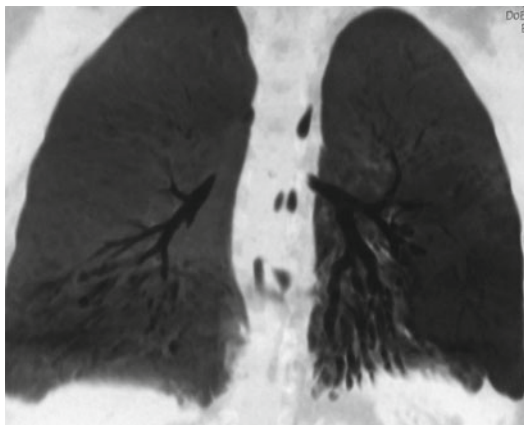


Fig. 2.11

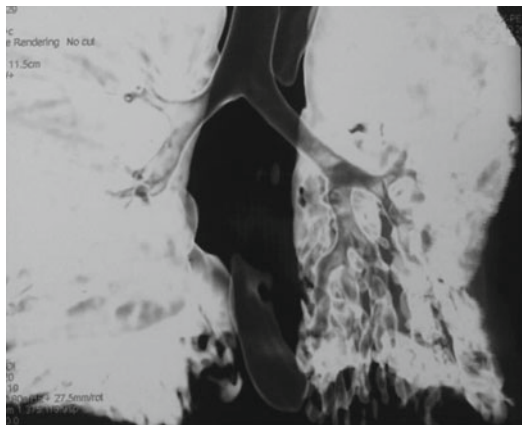


Fig. 2.12

An 8-year-old girl from the north of Africa presented with copious daily production of mucopurulent sputum. She had a history of previous pneumonia at the age of 2 years and recurrent respiratory tract infections since then.

Nowadays, an increase in the frequency of diagnosing non-cystic fibrosis bronchiectasis (NCFB) in children has been observed due probably to both heightened awareness of the disease and the wider availability of high-resolution computed tomography (HRCT). Moreover, NCFB remains an important cause of chronic suppurative lung disease in the developing world. Bronchiectasis is characterized by chronic airway inflammation that is driven by persistent infection and defined by the presence of permanent abnormally dilated medium-sized airways, with progressive destruction of the bronchial walls. Underlying causes have been described in 50–60% of NCFB. The most common underlying conditions include previous pneumonic illness, congenital and acquired immunodeficiency, primary ciliary dyskinesia, mechanical obstruction (inhaled foreign body, extrinsic compression, slow-growing tumor), and chronic aspiration. Fifty percent of cases keep without diagnosis. Postinfectious bronchial damage, particularly following adenovirus infection, still accounts for the majority of cases in childhood.

NCFB may be classified as a localized (one lobe) or generalized disease (multilobular) and also according to the pathological or radiographic appearance of the airways: *cylindrical*, dilated bronchi alone; *varicose*, focal constrictive areas between the dilated airways caused by defects in the bronchial wall; and *cystic*, airways ending in large cyst, saccules, or grape-like clusters.

In recent years, other terms have been proposed: *pre-bronchiectasis* (bronchial wall thickening on the HRCT scan, which may resolve entirely or progress), *HRCT bronchiectasis* (bronchial dilatation which may persist, resolve, or progress), and *established bronchiectasis* (the HRCT findings persist after 2 years, probably as an irreversible condition).

(Fig. 2.9). Copious daily mucopurulent sputum expectorated by the patient. (Fig. 2.10). HRCT (lung window setting): established bronchiectasis: mucous impaction, bronchial dilatation, and thickening with smaller adjacent pulmonary vessel (signet ring sign) in left lower lobe. (Fig. 2.11). Coronal reconstruction HRCT, mini-maximum intensity projection (miniMIP) reconstruction technique and (Fig. 2.12) tridimensional reconstruction HRCT provide a good depiction of established varicose bronchiectasis: bronchial dilatation (tram lines) and focal constrictive areas between the dilated airways caused by defects in the bronchial wall.

Comments

Imaging Findings

Case 4: Congenital Unilateral Pulmonary Vein Atresia

Estela Pérez Ruiz and Pilar Caro Aguilera

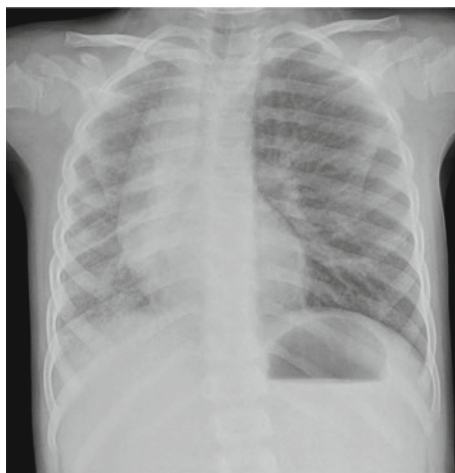


Fig. 2.13

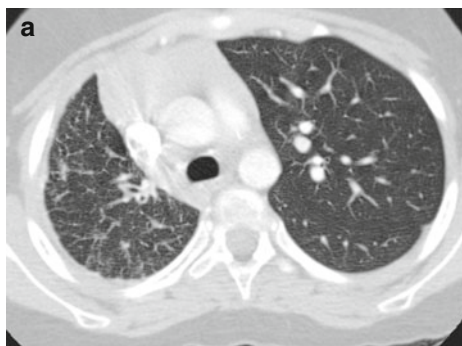


Fig. 2.14

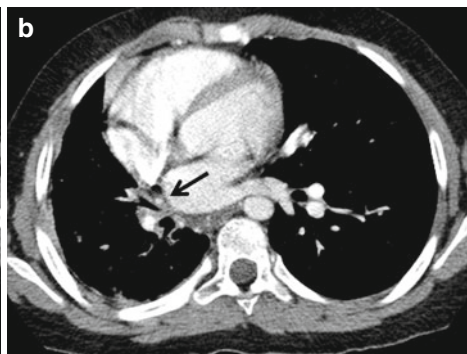
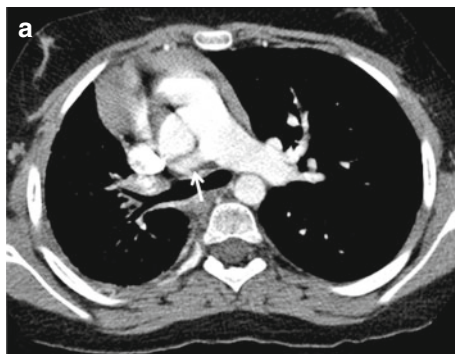


Fig. 2.15

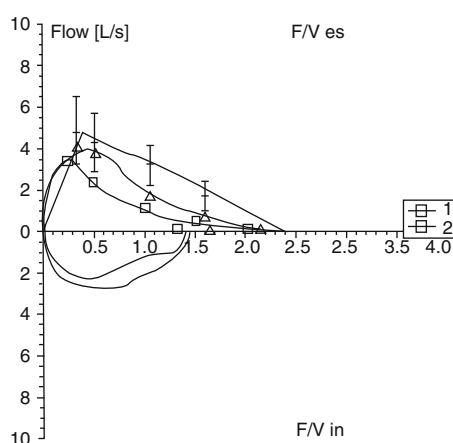
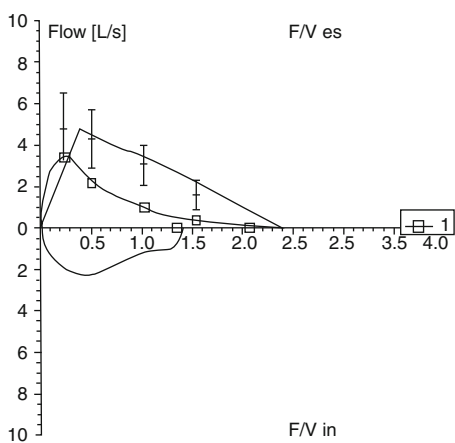


Fig. 2.16

A 3-year-old girl presents with history of both recurrent bronchitis and pneumonia since the age of 3 months. An asymmetric right hypoventilation was noted on thorax auscultation. She did not have a history of congenital heart disease nor evidence of bronchial obstruction at bronchoscopy. Clinical evolution has consisted of bronchial hyperresponsiveness, asthma-like symptoms, until the age of 11 years.

Unilateral pulmonary vein atresia without anomalous connection is a rare congenital abnormality that is usually present in infants, although some cases have been reported in adult patients. It results from failure of incorporation of the common pulmonary vein into the left atrium. It may occur in either lung, with no right- or left-side predominance. Pulmonary artery and parenchymal abnormalities are also present. Ipsilateral hypoplasia of the pulmonary artery is likely because of preferential pulmonary artery perfusion to the contralateral side and would account for the arterial systemic-to-pulmonary collateral vessels. Parenchymal manifestations include interlobular septal thickening, bronchial wall thickening, and ground-glass opacities, probably reflected in both pulmonary vein hypertension and engorged lymphatics. The most frequent presenting symptoms include recurrent infections in the hypoplastic lung, exercise intolerance, and hemoptysis due to the systemic collateral supply to the affected lung. Other associated congenital heart defects are found in approximately 50% of patients. Pulmonary artery hypertension may also be associated.

(Fig. 2.13). Anteroposterior chest radiograph reveals small right hemithorax with ipsilateral mediastinal shift and right diffuse interstitial infiltrate. (Fig. 2.14). CT scan (lung window setting): upper field section (a), lower field section (b). Septal pattern throughout right parenchyma with diffuse ground-glass attenuation and smooth thickening of the interlobular septa and bronchovascular bundles. The small right hemithorax is confirmed. (Fig. 2.15). Contrast-enhanced CT scan (artery phase): (a) Small right pulmonary artery (*white arrow*). (b) Absence of right pulmonary venous connection into left atrium, revealing a left atrial margin completely smooth at expected location of right pulmonary veins (*black arrow*). (Fig. 2.16). Flow volume curve with moderate degree of airway obstruction. The shape of the loop shows an intrathoracic obstructive ventilatory pattern (loop 1) with reversibility to bronchodilator test (loop 2). Bronchial hyperresponsiveness.

Comment

Imaging Findings

Case 5: Late Complications of Congenital Esophageal Atresia and Tracheoesophageal Fistula

Francisco Javier Pérez-Frías and Estela Pérez Ruiz



Fig. 2.17

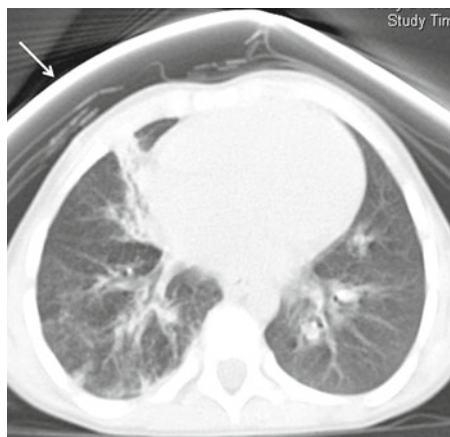


Fig. 2.18

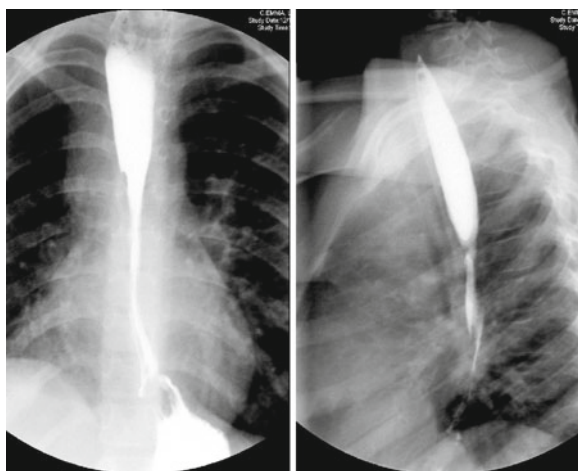


Fig. 2.19

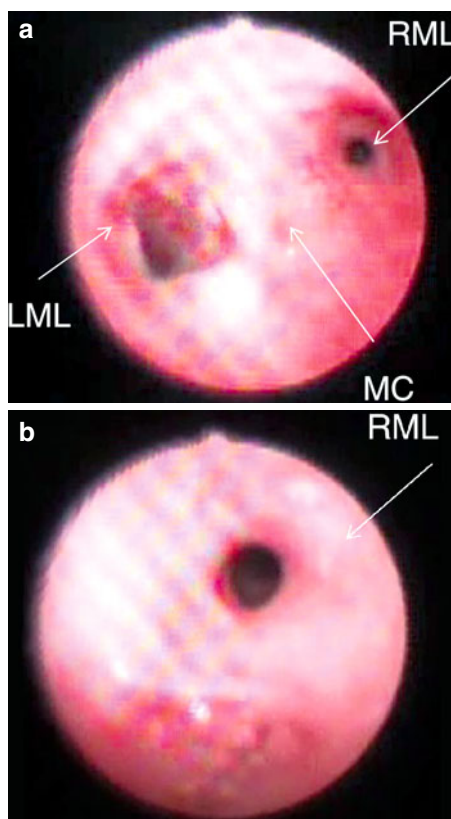


Fig. 2.20

An 8-year-old girl presents with recurrent bronchitis and pneumonia. She had a history of operated esophageal atresia (EA) with distal tracheoesophageal fistula (TEF).

Following EA and/or TEF surgery, patients have several long-term respiratory and esophageal symptoms during childhood, with a reported prevalence in up to 40% of cases which may persist life-long. In fact, “doctor-diagnosed asthma” has been described in 20% of cases. Late complications include tracheomalacia, a recurrence of the TEF, esophageal stricture, and gastroesophageal reflux (GR). Recurrent retrograde pulmonary aspiration can be due to either esophageal stricture or GR. Anastomotic stricture is especially common after repairing a gap of more than 2.5 cm, as an anastomosis under tension appears to increase the incidence of this complication, especially if a GR is present. The diagnosis of esophageal stenosis in the anastomosis site needs to be confirmed by esophagography or esophageal endoscopy. The esophageal stricture associated to GR origins recurrent airways aspiration which has been suggested to contribute to a persistent inflammation that may lead to several pulmonary diseases and other conditions, including chronic coughing, chronic hoarseness, posterior laryngitis, nocturnal choking, airway hyperreactivity (asthma), and recurrent pneumonitis, which may progress to obstructive and restrictive ventilatory defects. In cases of recurrent pneumonia, a bronchoscopy may evidence bronchial edema and hyperemia associated to a variable degree of bronchial stenosis, especially in the right bronchial tree. Performing a bronchoalveolar lavage also helps to investigate possible markers for GR disease and their relation to oxidation and inflammation and to correlate these with endobronchial biopsy findings. In conclusion, aspiration should be excluded in children and adults with a history of EA/TEF who present with respiratory symptoms and/or recurrent lower respiratory infections to prevent chronic pulmonary disease.

(Fig. 2.17). Axial thorax CT, lung window. Air entrapment in right upper lobe. Dilatation of proximal portion of the esophagus (*arrow*). (Fig. 2.18). Axial thorax CT, lung window. Bilateral diffuse bronchiectasis, predominantly in right lower lobe. Segmental atelectasis and bronchiectasis in middle lobe. Thorax protection with bismuth (*arrow*). (Fig. 2.19). Anteroposterior and lateral projection of esophagogram with oral contrast. Dilatation of proximal portion of the esophagus with straight stenosis in middle portion (at the anastomosis site). (Fig. 2.20). Flexible bronchoscopy: view of trachea at main-stem carina (MC). (a) Stenotic right main-stem bronchus (RML) compares to normal left main-stem bronchus (LML). (b) Hyperemia, inflammation, and stenosis of RMB secondary to recurrent pulmonary aspiration.

Comments

Imaging Findings

Case 6: Congenital Tracheal Stenosis with Tracheoesophageal Fistula

Pilar Caro Aguilera and Estela Pérez Ruiz

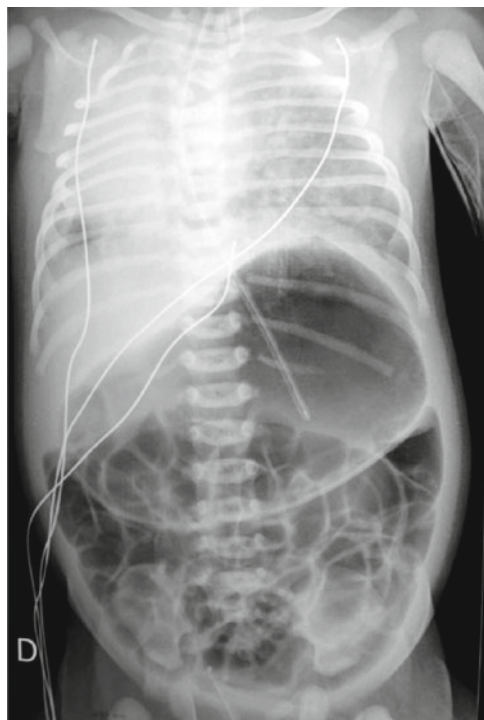


Fig. 2.21

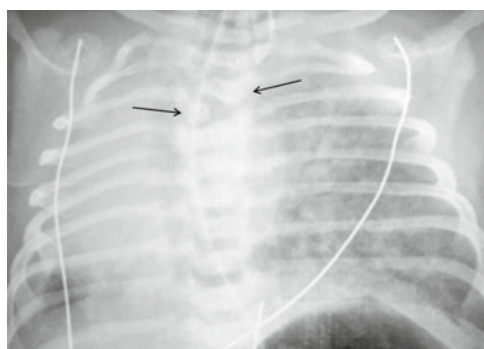


Fig. 2.22



Fig. 2.23

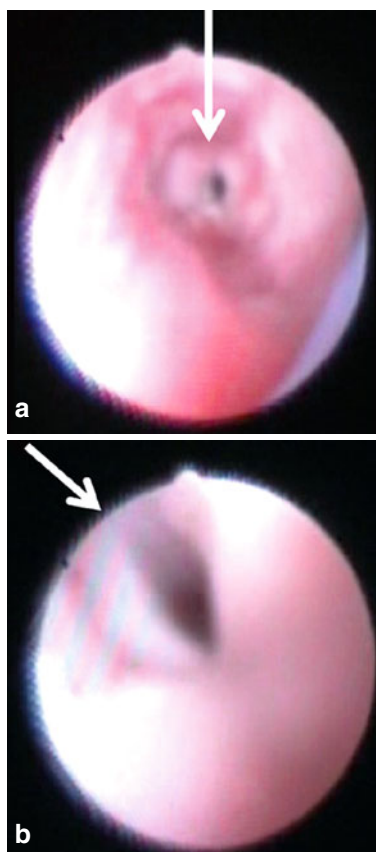


Fig. 2.24

An 8-day-old female newborn was admitted to the neonatal intensive care unit as she suffered from an acute respiratory insufficiency at birth. She needed endotracheal intubation and mechanical ventilation with severe ventilation problems.

Congenital tracheal stenosis (CTS) may be the result of an abnormality of the trachea itself or the effect of external forces compressing the airway. However, the common characteristic is narrowing of the trachea producing airway obstruction. Severe CTS and atresia of the trachea are rare malformations occurring with an estimated rate of two per 100,000 live births. Severe CTS, resulting in functional tracheal atresia, is frequently combined with other anomalies such as vertebral anomalies, anal atresia, cardiovascular anomalies, tracheoesophageal fistula (TEF), esophageal atresia, renal/radial anomalies, and limb defects (VACTERL association). Affected neonates present with severe respiratory distress immediately after birth. Without TEF, severe CTS is fatal and usually results in death within the first minutes of life. Survival is only possible with either a small tracheal lumen remaining, large enough for spontaneous breathing, or an emergency tracheotomy.

Prenatal magnetic resonance imaging (MRI) may provide a definitive diagnosis. Postnatal diagnosis is based on recognition of clinical signs in the newborn. The diagnostic should begin with thorax X-rays which may show unusual air distribution, deviation of the heart and mediastinum, and evidence of tracheal compression. Other diagnostic imaging studies may be used, including contrast esophagography, tracheobronchography, echocardiography, bronchoscopy (BC), and cardiac catheterization. Spiral sequencing or 3-dimensional reformatting CT scans and virtual BC are used to reconstruct the trachea and proximal bronchi. BC coupled with bronchography is a simple procedure that has excellent spatial and temporal resolution. Despite progress in surgical interventions, mortality remains high.

(Fig. 2.21). Thorax and abdomen X-ray. Central alveolar pattern, more evident in the left lung. Heart deviation to the right. Great gastric distension with bowel luminogram. Devices: endotracheal tube in D2, nasogastric tube in stomach. (Fig. 2.22). Detail of the thorax X-ray. Dorsal vertebral malformations (*arrows*) and dextrocardia. (Fig. 2.23) BC. TEF in a proximal and posterior position (*arrow* “A”) and the CTS distally (*arrow* “B”). (Fig. 2.24) BC. (a) Severe CTS in the middle/distal tracheal third with a punctate lumen (*arrow*). (b) Tracheal lumen at maximal obstruction (*arrow*); the small size of the trachea did not allow the pass of the 2.5-mm-diameter bronchoscope.

Comments

Imaging Findings

Case 7: Multidetector CT of the Central Airways in Children: 3D Imaging and Virtual Bronchoscopy

Isabel Gordillo Gutiérrez



Fig. 2.25

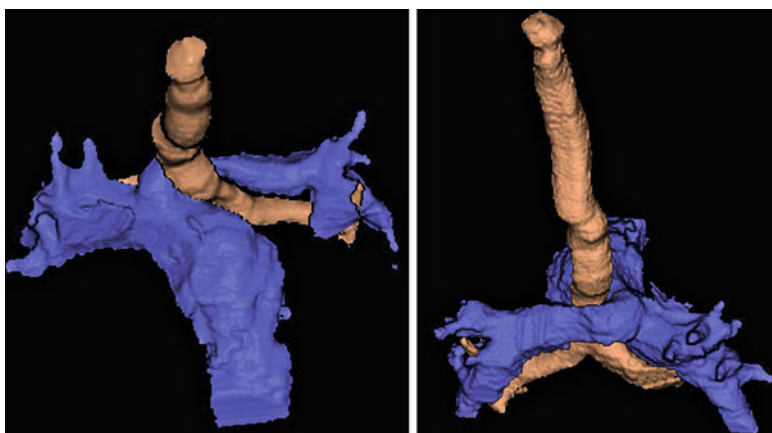


Fig. 2.26

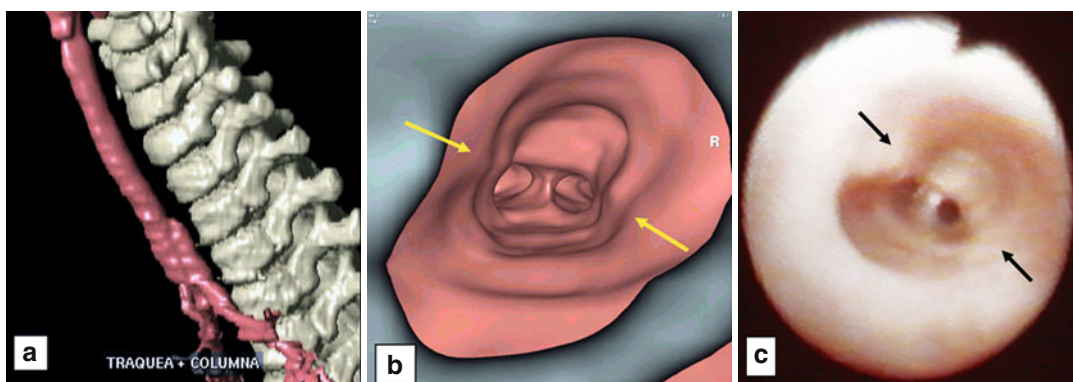


Fig. 2.27

Recent advances in multidetector computed tomography (MDCT) have revolutionized the noninvasive imaging evaluation of the central airways in pediatric patients.

Fiberoptic bronchoscopy (FOB) is the gold standard for diagnosis of tracheobronchial abnormalities. However, it remains an uncomfortable procedure and is of limited use in patients with severe stenosis of the bronchial lumen. MDCT-3D airway and mediastinal vascular reconstruction and virtual bronchoscopy (VB) is a good diagnostic tool in the evaluation of the large airways in children and presents information that renders abnormalities more obvious than they are on axial images.

Several limitations are associated with axial CT images: limited ability to detect subtle airway stenosis, underestimation of the craniocaudal extent of disease, difficulty displaying 3D relationships of the airways, and inadequate representation of the airways that are oriented obliquely to the axial plane.

Accurate identification and measurement of airway diseases are of paramount importance for assigning the appropriate diagnosis and planning the surgical procedures: this can be obtained with MDCT-3D and VB.

Advantages of this MDCT technology are:

1. Noninvasive procedure.
2. The creation of 3D reconstruction can help overcome the limitations of axial images.
3. Provides endoluminal, extraluminal, and extra-airway information that facilitates evaluation of spatial relationships.
4. VB is able to visualize areas beyond even high-grade stenosis.
5. It can give 3D road maps to surgeons and help in endobronchial treatments.
6. Can replace more invasive examinations such as FOB in selected cases (patients with no tolerance, high-risk patients and children).

Disadvantages are (1) it is not dynamic and is unable to show information such as pulsation from external pressure indicating vascular compression; (2) it does not evaluate mucosal surface; (3) motion artifacts, mucous plugs, or secretions inside the lumen can create false positives; (4) and it cannot provide samples for histological analysis.

MDCT-3D Imaging. VB and FOB are complementary techniques. Clinical applications of MDCT technology include assisting with diagnosis, helping in surgical planning treatments, and replacing invasive examinations such as FOB in selected cases.

Comments

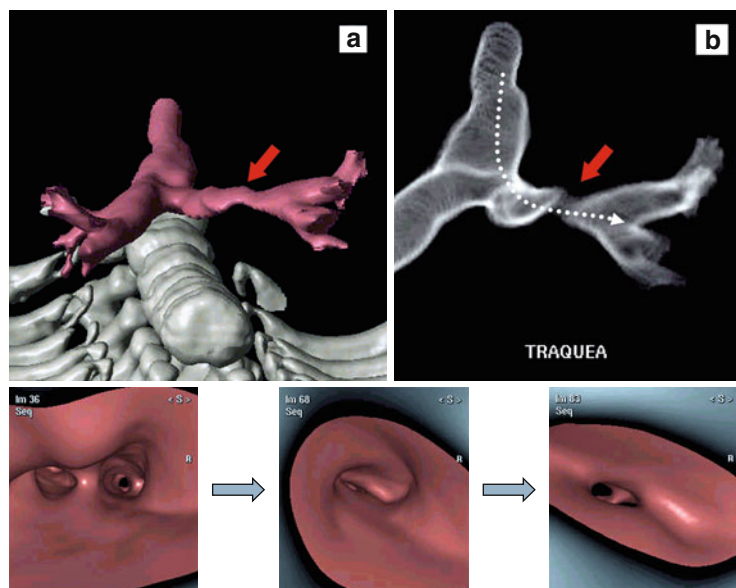


Fig. 2.28

MDCT-3D reconstruction in a girl with tracheal compression by an innominate artery (*arrow*) (Fig. 2.25) and in a boy with airway compression by a pulmonary sling (Fig. 2.26). Postsurgery exploration, 3D airway reconstruction (a), VB (b), and FOB (c) in a 2-year-old boy with tracheoplasty (*arrows*) (Fig. 2.27). 3D imaging (Fig. 2.28a) and VB (Fig. 2.28b) in a boy with left main bronchus stenosis (*red arrow*). FOB was not possible, but VB is able to visualize areas beyond even high-grade stenosis (Fig. 2.28). (Discontinuous line in b and gray arrows show the endoscopic way).

Imaging Findings

Case 8: Recurrent Spontaneous Pneumothorax

Pilar Caro Aguilera and
Francisco Javier Pérez-Frías

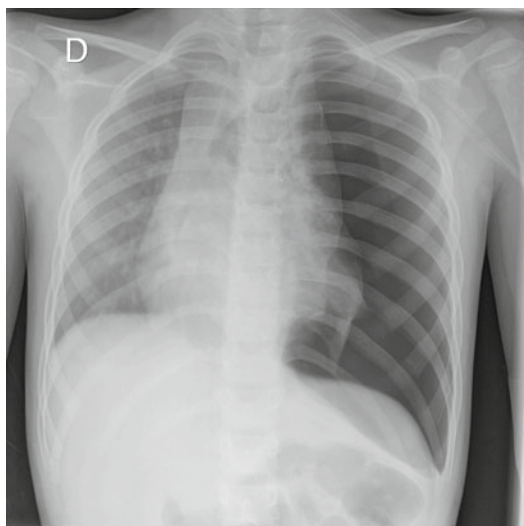


Fig. 2.29

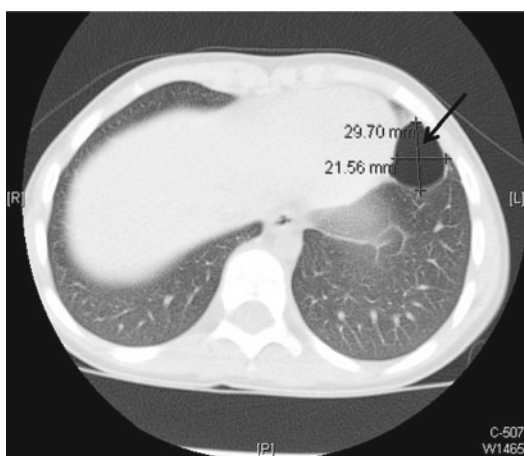


Fig. 2.30

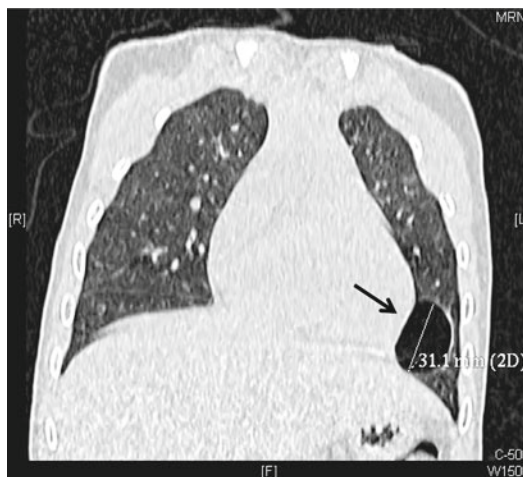


Fig. 2.31

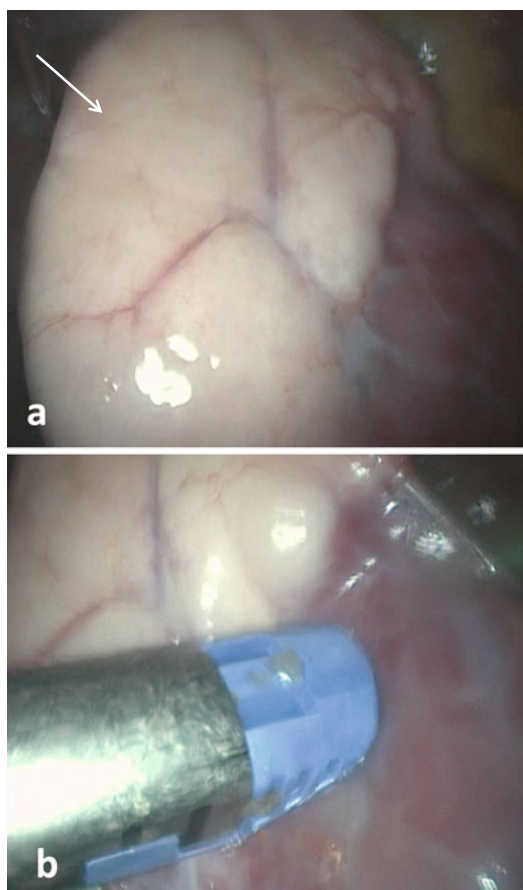


Fig. 2.32

An 8-year-old boy who suffered from recurrent spontaneous pneumothorax (SPT) at the ages of 5 and 7 years. He was admitted to the hospital as he had a severe chest pain. The auscultation showed hypoventilation in the left hemithorax.

Primary SPT is defined as a pneumothorax occurring in the absence of underlying lung disease and with no apparent pathology. SPT is a relatively rare condition in the pediatric population. The peak age of occurrence in this age group is bimodal, with most cases occurring either in the neonatal period or in late adolescence.

The risk of recurrent pneumothorax after a single episode of SPT with conservative therapy ranges from 16% to 52%, whereas following surgical management of SPT, it is less than 5%. The mechanism involved in this predisposition is thought to be related to the presence of blebs or bullae in the most apical portions of the lungs. In children, SPT is thought to be related to rupture of these apical subpleural blebs or bullae. The typical symptoms of pneumothorax, such as chest pain and dyspnoea, may be relatively minor or even absent in SPT so that a high degree of initial diagnostic suspicion is required. When the patient presents severe symptoms or signs of cardiorespiratory distress, tension pneumothorax must be considered. The diagnosis of SPT is usually confirmed by imaging techniques. Thorax X-ray must be the first step in the diagnosis of PST, although it has limitations, such as the difficulty in accurately quantifying pneumothorax size. Computed tomography (CT) is useful in the detection of small pneumothoraces and in size estimation. It allows identifying the presence of surgical emphysema and bullous lung disease or additional lung pathology.

Therapeutic management ranges from a conservative attitude in mild cases receiving supplemental oxygen, to active intervention – needle aspiration and chest drain insertion – or surgical intervention. It may include removal of underlying cysts or bullae thought to be responsible for the occurrence or persistence of the pneumothorax. Open thoracotomy or limited axillary thoracotomy and pleurectomy remain the procedures with the lowest recurrence rate, but video-assisted thoracoscopic surgery (VATS) with pleurectomy and pleural abrasion has good results as well and is better tolerated.

(Fig. 2.29). Thorax X-ray: left SPT.

(Figs. 2.30 and 2.31). CT image, axial and coronal reconstructions: large bullae ($29.7 \times 21.56 \times 31.1$ mm) on the left lower lobe (*arrows*).

(Fig. 2.32). VATS photographs: (a) Bullae as white structure (*arrow*) in the region of the left lower lobe. (b) Photograph during the resection of the bullae.

Comments

Imaging Findings

Case 9: Bronchotracheal Foreign Body

Pilar Caro Aguilera and Francisco Javier Pérez-Frías

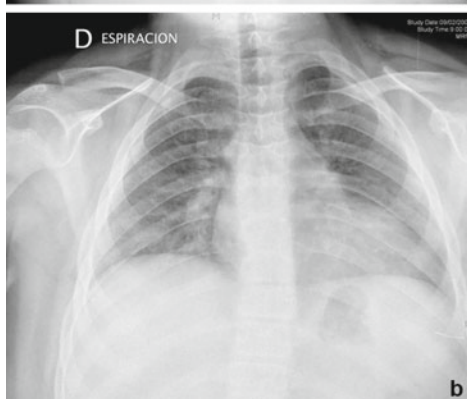
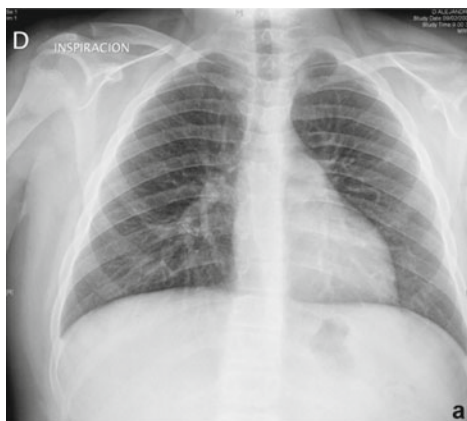


Fig. 2.33

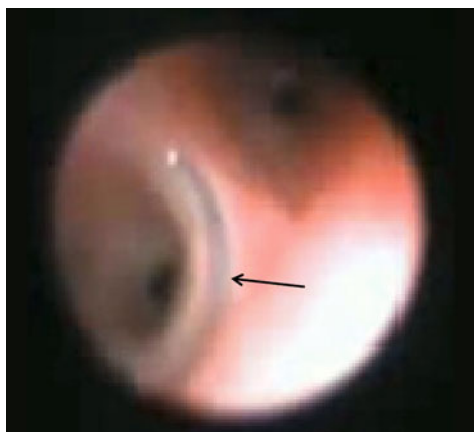


Fig. 2.34



Fig. 2.35

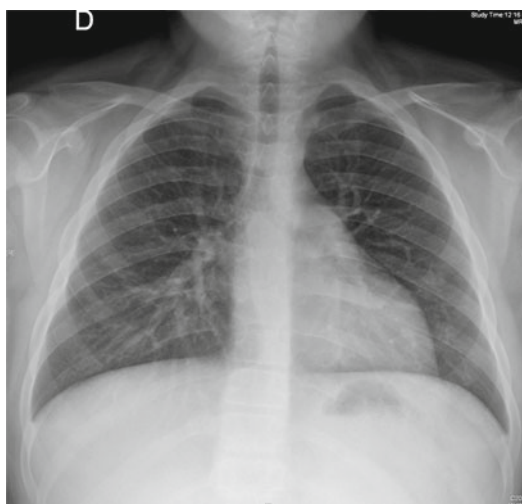


Fig. 2.36

A 13-year-old boy was referred from another center with a clinical history of foreign body aspiration and two negative flexible bronchoscopes. The child was without respiratory distress and non-pathologic signs during the exploration, except for moderate auscultatory signs of left hemithorax hypoventilation.

Foreign body (FB) aspiration is one of most frequent episodes in child respiratory pathologies. FB aspiration is a significant cause of morbidity and mortality in childhood.

The severity of the acute obstruction of the upper airway and the complications that FB creates in the lower airway require early diagnosis and treatment. Classically, the FB was previously removed by surgeons with a rigid bronchoscope under general anesthesia.

Nowadays, there are publications on removing FB with flexible bronchoscopy. At present, the discussion is which instrument is the best, a rigid or flexible bronchoscope. Flexible bronchoscopy is a minimal invasive procedure that allows making the diagnosis and locating the FB, which can only be removed in selected cases. Therefore, authors believe that FB removal is more effective with rigid bronchoscopy. It guarantees patient safety and the success of the procedure.

However, flexible and rigid bronchoscopes are complementary, so their combined use is the most appropriate choice. We report the management of this case of FB in pediatric patients with a combined procedure using flexible and rigid bronchoscopes.

Normal thorax X-ray in inspirations (Fig. 2.33a) and signs of air entrapment in the left lower lobe at the expiratory thorax X-ray (Fig. 2.33b).

Bronchoscope vision with rigid bronchoscope before extraction. The pen cap is in the left lower lobe bronchus and it seems a bronchial stenosis (*arrow*) (Fig. 2.34).

Plastic pen cap of a mechanical pencil (Fig. 2.35).

Final normal chest X-ray (Fig. 2.36).

Comments

Imaging Findings

Case 10: Endobronchial Lymphoma

María I. Martínez-León and
Antonio Martínez-Valverde

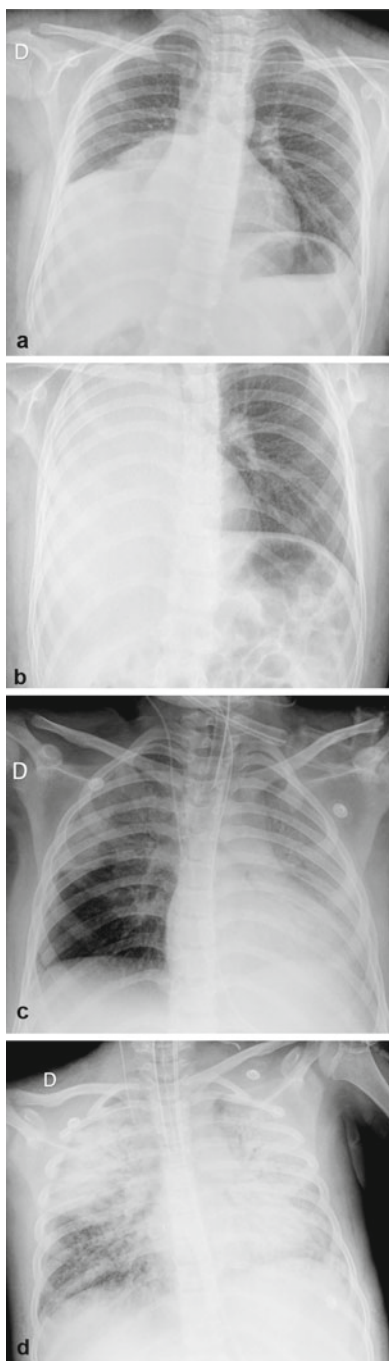


Fig. 2.37

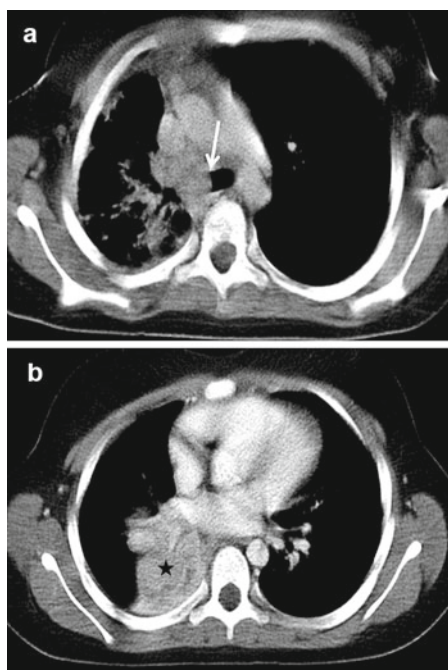


Fig. 2.38

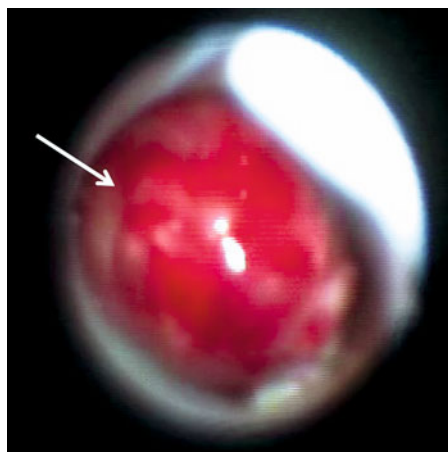


Fig. 2.39

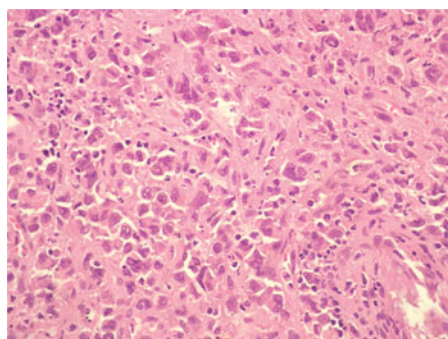


Fig. 2.40

A 10-year-old male patient was sent to the Traumatology Service of our hospital with a history of progressive right ankle pain with no response to treatment. No other symptoms were referred. A gammagraphy was performed, showing areas of hypercaptation in tibia and basal right lung. The bronchial lymphoma was diagnosed by chance while he was being investigated for a benign bone disease, due to the initial absence of respiratory symptoms.

This is a presentation of CD30+ anaplastic large-cell lymphoma (ALCL) with an unusual endobronchial polypoid mass. ALCL is a very rare disorder in childhood. The most common localization of this lymphoma is in the lymph node and skin, with endobronchial involvement being extremely rare among the pediatric population.

Clinical findings in the few reported cases of ALCL are nonspecific, including cough, dyspnea, hemoptysis, fever, shortness of breath, etc. Radiographic features include nodules, mass, air space consolidation, pleural effusion, cavitations, and cystic lesions, which are indistinguishable from a broad spectrum of disorders. All these reasons, together with the malignancy of the process, make an early diagnosis necessary in order to begin treatment as soon as possible.

This case illustrates that ALCL should be included in the differential diagnosis of endobronchial mass lesions.

(Fig. 2.37). (a) Initial chest X-ray shows right hilar mass with middle and inferior lobe atelectasis. (b) Chest X-ray 25 days later: white right lung with collapse and homolateral mediastinal deviation. (c) Chest X-ray after chemotherapy: there is no hilar mass, but now there is a white left lung. (d) Follow-up chest X-ray indicates an alveolo-interstitial pulmonary bilateral pattern with the need of intubation. It is compatible with acute adult respiratory distress syndrome.

(Fig. 2.38). A thorax CT without contrast (a) and enhanced (b) revealed a right hilum mass protruding into the carina (*arrow*) and a secondary collapse of the right lung (*asterisk*).

(Fig. 2.39). Flexible fiberoptic bronchoscopy revealed an endobronchial polypoid mass (*arrow*), with high vascularization, completely filling the lumen. This mass reached the lower third of the trachea, with its origin in the main right bronchus; partial resection with LASER was made, obtaining a tissue sample which gave the diagnosis.

(Fig. 2.40). Endobronchial ALCL pathology, Hematoxylin-Eosin.

Even though chemotherapy was started, the patient's outcome was unfavorable, and he died 18 days later.

Comments

Imaging Findings

Further Reading

Case 1: Postinfectious Bronchiolitis Obliterans

Book

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Case 2: Thoracic Rib Invasion from Actinomycosis

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Case 3: Non-Cystic Fibrosis Bronchiectasis

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Case 4: Congenital Unilateral Pulmonary Vein Atresia

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Case 5: Late Complications of Congenital Esophageal Atresia and Tracheoesophageal Fistula

Book

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Case 7: Multidetector CT of the Central Airways in Children: 3D Imaging and Virtual Bronchoscopy

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Case 8: Recurrent Spontaneous Pneumothorax

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Case 9: Bronchotracheal Foreign Body

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Case 10: Endobronchial Lymphoma

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