RadioGraphics

Bronchial Atresia[1]

Matthew G. Gipson, MD • Kristopher W. Cummings, MD • Kyle M. Hurth, MD, PhD

History

A 22-year-old woman presented to the student health clinic with acute, sharp, left-sided chest pain that worsened with deep inspiration. Her symptoms had begun the previous night with shortness of breath. She sought treatment because of the persistent and progressive nature of the symptoms. A chest radiograph was obtained and showed a large left-sided pneumothorax. She was transferred to the emergency department, where she underwent placement of a thoracostomy tube. She reported receiving a diagnosis of a "congenital lung abnormality" several years earlier. Because the nature of the underlying lung abnormality was not clear from the patient history, repeat radiography and computed tomography (CT) were performed after thoracostomy drainage.

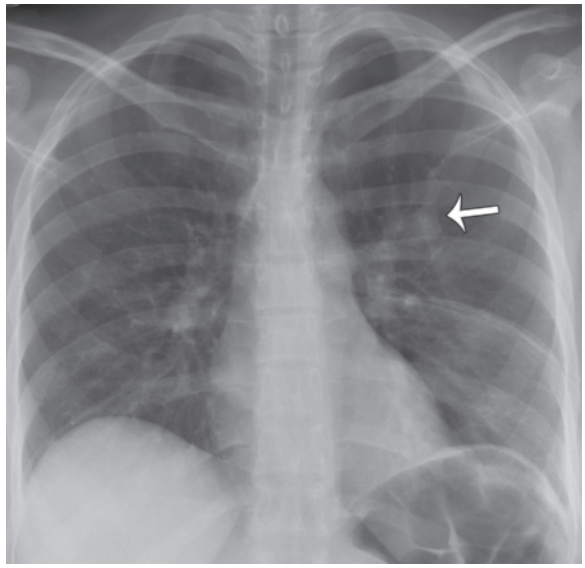

Figure 1. Radiograph shows a branching tubular area of increased opacity (arrow), which represents a bronchocele and extends from the left hilum into the left upper lobe. A thoracostomy tube was placed to treat a left-sided pneumothorax.

Imaging Findings

Chest radiography performed after thoracostomy drainage showed a focal tubular area of increased opacity in the left upper lobe and a small residual pneumothorax (Fig 1). Contrast material–enhanced CT of the chest was performed to further characterize the radiographic findings. Interval reexpansion of the left lung and a decrease in size of the pneumothorax were seen. A 3.3 × 1.5-cm nonenhancing tubular mass was seen extending cephalad from the left hilum, with surrounding hypoattenuation of the apicoposterior segment of the left upper lobe, a finding indicative of hyperinflation (Fig 2).

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[1]From the Departments of Radiology (M.G.G., K.W.C.) and Pathology (K.M.H.), Mallinckrodt Institute of Radiology, Washington University School of Medicine, 510 S Kingshighway Blvd, St Louis, MO 63110. Received December 11, 2008; revision requested January 6, 2009, and received January 15; accepted January 16. All authors have no financial relationships to disclose. **Address correspondence to** M.G.G. (e-mail: *gipsonm@mir.wustl.edu*).

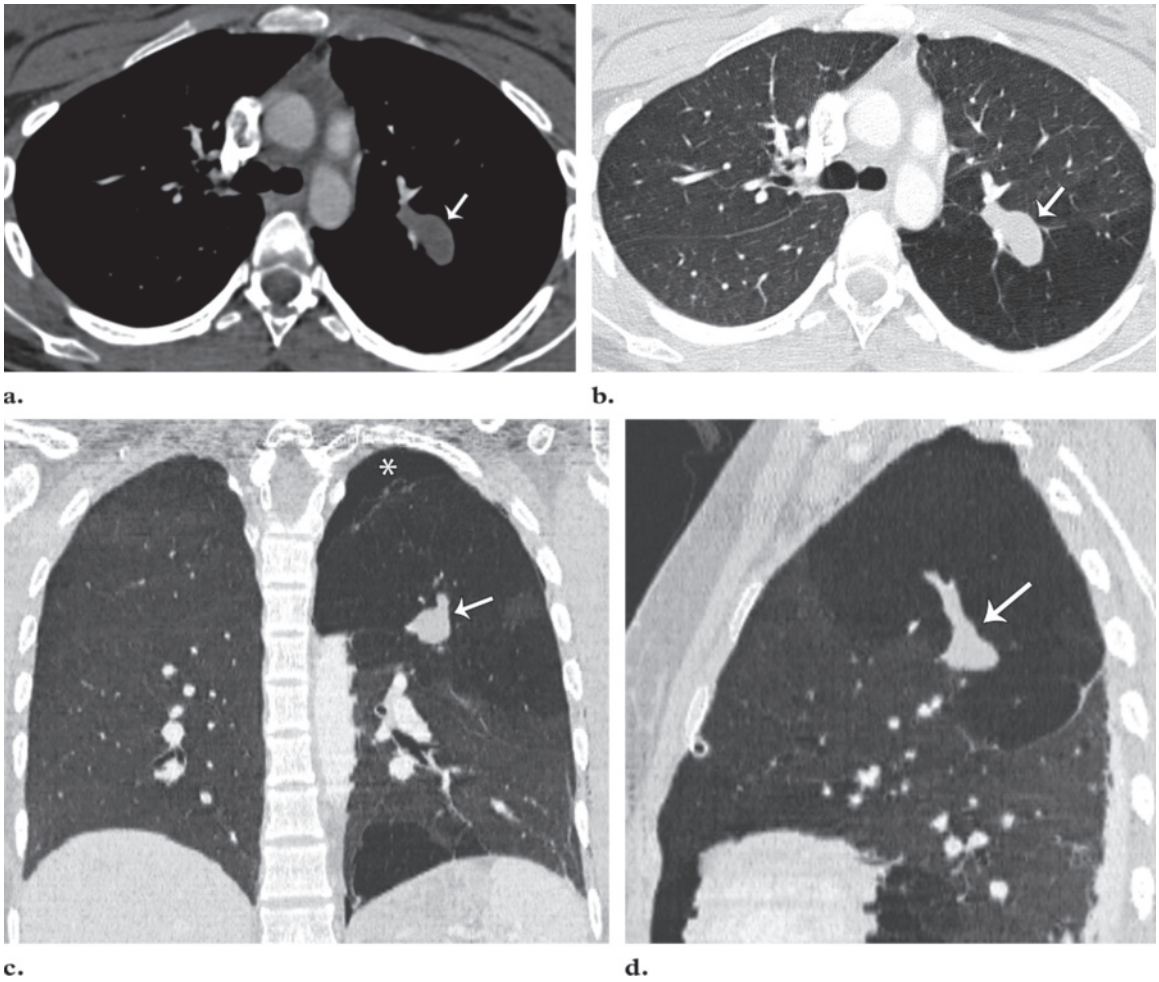


Figure 2. Axial soft-tissue window **(a)**, axial lung window **(b)**, and minimum-intensity-projection lung window coronal **(c)** and sagittal **(d)** contrast-enhanced CT images show a tubular nonenhancing lesion (arrow), a finding consistent with mucus impaction of the apicoposterior left-upper-lobe bronchus (bronchocele). Decreased attenuation is seen throughout the apicoposterior segment of the left upper lobe in **b–d**, a finding indicative of associated air trapping. These CT features are diagnostic of bronchial atresia. Note the small residual pneumothorax (∗ in **c**).

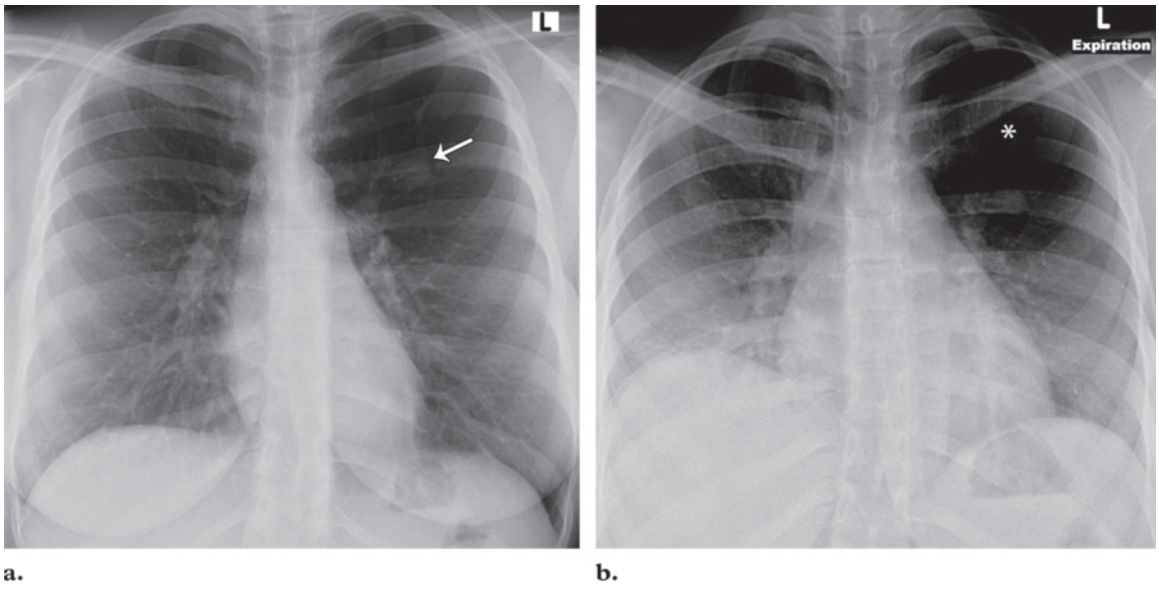


Figure 3. **(a)** Inspiratory radiograph obtained 2 years earlier shows a bronchocele in the left upper lobe (arrow). **(b)** Exhalation radiograph shows an area of hyperlucency (∗) surrounding the area of increased opacity, a finding indicative of air trapping.

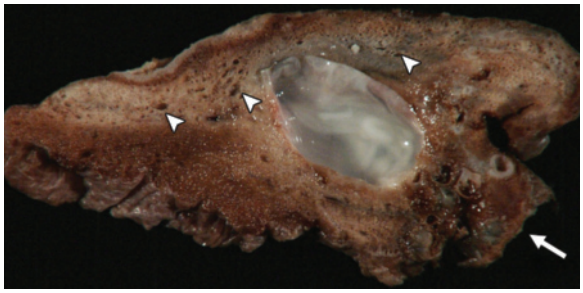
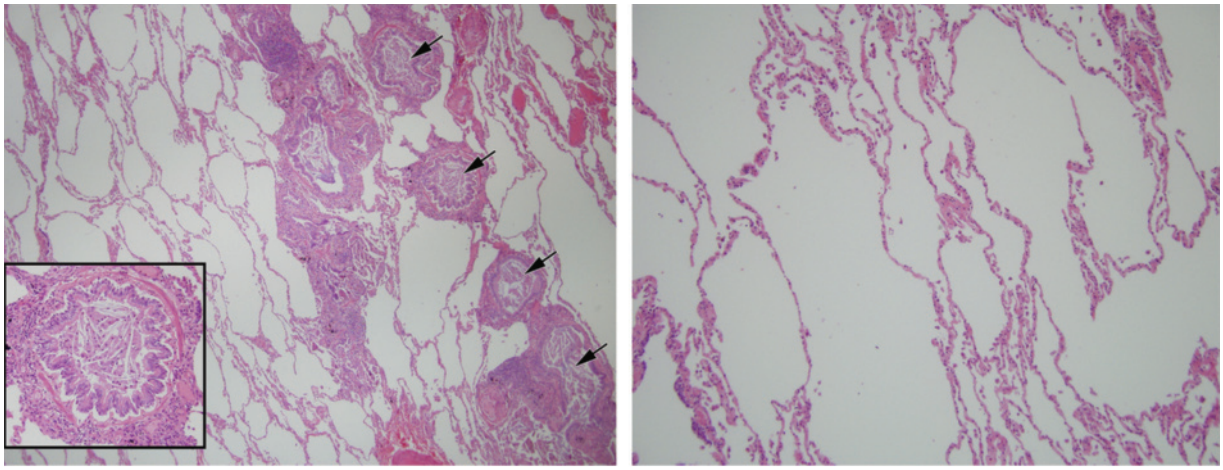


Figure 4. Photograph of the left upper lobectomy specimen shows a large bronchocele. The mucus plug was removed. Signs of hyperinflation are seen in the surrounding lung parenchyma (arrowheads). Arrow = bronchial resection margin.



a. b.

Figure 5. **(a)** Photomicrograph (original magnification, ×4; inset magnification, ×20; hematoxylin-eosin stain) shows respiratory bronchioles that are plugged with mucus (arrows) and surrounded by dilated alveoli. Inset shows a bronchiole lined with respiratory epithelium and filled with mucus, macrophages, and sparse neutrophils. Clefting within the mucoid debris is prominent and is secondary to dissolution of cholesterol and lipids. **(b)** Photomicrograph (original magnification, ×20; hematoxylin-eosin stain) shows alveolar dilatation. The septa are thin, fine, and nonfibrosed and interconnect with one another. There are no hyaline membranes present, and no significant inflammatory infiltrate is seen. This pattern is indicative of hyperinflation rather than destruction.

Earlier radiographs, including inspiratory and expiratory views, were acquired from an outside institution for comparison. They showed a tubular area of increased opacity that extended from the left hilum into the left upper lobe (Fig 3). On the expiratory view, there was persistent hyperinflation of the left upper lobe around the area of increased opacity.

Pathologic Evaluation

The patient initially was treated conservatively with chest tube placement, which resulted in prompt reexpansion of the lung. However, she developed a persistent pneumothorax secondary to a residual air leak, and the decision was made to perform a lingula-sparing left-upper-lobe segmentectomy. A soft pliable mass, which represented a mucocele, was palpated intraoperatively. The mucocele was surrounded by hyperinflated lung tissue, and the lung lobe appeared dilated. At gross examination of the resected left upper lobe, the bronchial resection margin was diminutive, with a cross section of 0.7 × 0.3 cm. Distal to the bronchial resection margin, the bronchus had an area of dilatation of up to 1.1 cm in diameter and was filled with a 2-cm long mucocele (Fig 4). Additional smaller mucus plugs were found in neighboring bronchi. The surrounding lung parenchyma was brown-tan and grossly unremarkable.

Microscopic analysis of the resected lung revealed airways plugged by mucus and prominent alveolar distention (Fig 5). Alveoli were enlarged and appeared intact, with minimal

loss of alveolar walls. Few floating septa and mild alveolar wall disruption were seen near the pleural surface along the periphery of the lung. Macrophages were sparse; however, there was no significant acute or chronic inflammatory infiltrate within either the bronchi or surrounding lung parenchyma, although bronchus-associated lymphoid tissue was seen. The respiratory epithelium lining the airways showed focal thinning and focal absence of pneumocytes but otherwise had a well-defined brush border with no signs of disruption or necrosis. Gross and microscopic findings were consistent with bronchial atresia uncomplicated by infection. These findings confirmed the radiographic diagnosis of bronchial atresia in the left upper lobe.

Discussion

Initially described in 1953, bronchial atresia is a congenital abnormality resulting from focal interruption of a lobar, segmental, or subsegmental bronchus with associated peripheral mucus impaction (bronchocele, mucocele) and associated hyperinflation of the obstructed lung segment (1). The apicoposterior segmental bronchus of the left upper lobe is most often involved, followed by segmental bronchi of the right upper, middle, and lower lobes (2,3).

The exact cause of bronchial atresia is unknown; however, focal bronchial interruption seems to occur before birth. Because the bronchial pattern is entirely normal distal to the site of stenosis, it has been suggested that the atresia is probably not a result of abnormal growth and development, but rather it is secondary to a traumatic event during fetal life (4). The airway develops systematically, with the lobar bronchi, subsegmental bronchi, and distal bronchioles appearing in the 5th, 6th, and 16th weeks of fetal development, respectively. One theory is that bronchial atresia is caused by intrauterine ischemia after the 16th week of gestation (5). Because other congenital lung anomalies that are known to develop earlier in embryogenesis occur with bronchial atresia, another possibility is that the lesion occurs earlier, during weeks 4–6 of intrauterine development. (6).

Gross and histologic findings vary depending on whether there is an infection distal to the atretic bronchus. When no infection is present, a characteristic mucocele usually is found just distal to the point of atresia. At gross inspection, the adjacent lung parenchyma may appear normal or hyperinflated. Microscopic analysis typically reveals distended alveoli, a finding indicative of lobar hyperinflation; however, in the absence of infection, destructive changes usually are minimal or absent. Thus, the term *emphysema*, when used to describe this and similar entities (eg, congenital lobar emphysema), is a slight misnomer because alveolar destruction is not a characteristic finding of bronchial atresia (in the absence of other causes). Gross and microscopic findings generally reflect nondestructive overexpansion of alveoli, a finding that is distinct from the alveolar destruction seen in emphysema (7).

A central, atretic bronchus leads to mucus impaction, which is characteristic of bronchial atresia (3). On radiographs and CT images, the hyperlucent or hypoattenuating lung surrounding the mucocele represents a combination of dilated air spaces (air trapping) and focal parenchymal oligemia, which is secondary to a combination of intrapulmonary vascular compression and hypoxic vasoconstriction (8). Distal hyperinflation is believed to be caused by collateral ventilation through intraalveolar pores of Kohn, bronchoalveolar channels of Lambert, and interbronchiolar channels (5,9,10). Some investigators believe that interbronchiolar channels, which connect terminal bronchioles from adjacent lung segments, may be the major conduit of collateral ventilation, because the pores of Kohn and channels of Lambert have not consistently been found in infants (9).

Bronchial atresia usually is benign and asymptomatic and is often discovered incidentally (11). If a patient is symptomatic, the clinical manifestations vary and may range from recurrent pulmonary infections to mild wheezing and dyspnea. Pulmonary function tests do not aid diagnosis (9). The mean age at diagnosis

is 17 years, and the abnormality is reported to be more common in men than in women (12). The differential diagnosis includes other abnormalities with mucus impaction, such as allergic bronchopulmonary aspergillosis, cystic fibrosis, or any lesion that causes bronchial narrowing and thus mucus impaction. The presence of a mucocele with adjacent hyperinflation helps narrow the differential diagnosis.

A classic radiographic finding of bronchial atresia is a branching tubular or nodular area of increased opacity that extends from the hilum with surrounding hyperlucent lung parenchyma. Images acquired during both inspiration and expiration may help confirm that a lung is hyperinflated. However, CT is the most sensitive imaging modality, and when findings are typical, they may be considered diagnostic in most cases. CT allows characterization of the lack of communication between the mucocele and hilum, can show smaller mucoceles not seen at conventional radiography, and is more sensitive in demonstrating segmental hyperinflation, associated mass effect, and possible calcification (5). CT and magnetic resonance imaging are useful in depicting the absence of vascularity and enhancement within the lesion and may help exclude a vascular cause (13).

Historically, treatment of bronchial atresia was controversial. Some physicians advocated performing surgery on all patients because a definitive diagnosis could be made only operatively (14). The majority of patients are asymptomatic, and therefore no treatment is necessary. It is currently thought that surgical excision should be reserved for patients with complications secondary to the atretic bronchus, such as infection or significant compromise of adjacent lung parenchyma. Lobar resection and segmentectomy have been used; however, the ultimate goal is to preserve as much normal lung parenchyma as possible to maintain pulmonary function. In our case, lingula-sparing segmentectomy was performed with excellent results, and the patient was discharged after an uncomplicated postoperative course.

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