Title
Mucoid impaction and lobar emphysema due to bronchial atresia associated with a bronchogenic cyst

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Bronchial atresia is associated with mucus impaction and hyperinflation distal to the lesion. Mucus impaction occurs because bronchial secretions cannot be cleared into the central airways. The lack of communication from the bronchus also leads to hyperinflation; although ventilation does not occur directly through the airway, distal alveoli become distended by collateral ventilation through the bronchiole-alveolar channels of Lambert and intraalveolar pores of Kohn. These collateral routes collapse during expiration, which leads to air trapping. Despite superficially resembling emphysema on thoracic imaging, the patients are asymptomatic, the abnormality is an incidental finding in about half of the cases.

The findings in this case were consistent with concurrent bronchial atresia to the left upper lobe and a bronchogenic cyst in the posterior mediastinum. The patient was treated for an associated infectious bronchitis, with some improvement in symptoms. However, she continued to have occasional exertional dyspnea. A cardiopulmonary exercise test was performed to exclude ventilator impairment secondary to localized dynamic hyperinflation, before consideration for surgical resection. As the results did not demonstrate any significant evidence of cardiopulmonary limitation, surgical intervention was deferred.

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imaging studies, hyperinflation from bronchial atresia is distinct in that the damage to the alveolar walls is minimal or absent.

Radiographs may demonstrate a branching, tubular mass surrounded by hyperinflated lungs with decreased vascular markings. A bronchocele appears as a rounded opacity radiating from the hilum and may contain an air-fluid level. If filled with mucus, the mucocele is often noted as a solitary pulmonary nodule and may be misinterpreted as an arteriovenous malformation, granuloma, or metastatic lesion. CT can demonstrate the lack of communication between the mucocele and the hilum and may reveal segmental hyperinflation distal to the mucocele. Hyperlucency results not merely from air trapping but also from parenchymal oligemia as a result of regional vascular compression and hypoxic vasoconstriction. Contrast-enhanced CT aids in excluding a vascular component which would be more typical of pulmonary sequestration. The differential diagnosis for mucus impaction with hyperinflation includes allergic bronchopulmonary aspergillosis, cystic fibrosis, interlobar sequestration, and bronchogenic cyst.

Bronchogenic cysts are also congenital abnormalities that originate from the primitive foregut with most occurring in the mediastinum and less commonly within the lung parenchyma. In the mediastinum, they may occur in the paratracheal, hilar, or most commonly, subcarinal regions. The cysts can also be found in the anterior and posterior mediastinum. These cysts account for about half of all congenital mediastinal cysts.

Like bronchial atresia, there is a slight male predominance. Although most patients are asymptomatic, symptoms can occur if the position and size of the cyst imposes on the trachea or bronchi, resulting in cough, wheezing, stridor, dyspnea, cyanotic spells, and pneumonia. Infection can occur when the cyst communicates with the tracheobronchial tree; however, such communication is atypical. The cysts contain mucoid material and usually do not demonstrate an air-fluid level unless manipulated. The walls typically contain smooth muscle and often cartilage.

Chest radiographs may show water density mass lesions consistent with serous or mucous contents. CT can further characterize the contents of the cysts based on the density as well as identify the structures adjacent to the cyst. Contrast enhancement of the walls would be suggestive of superimposed infection. If a solid component is detected, malignancy should be considered.

The respiratory system develops embryologically between weeks 3 to 4 beginning with the proximal airways. At the seventh week of gestation, the conducting airways begin to form; acinar development does not occur until around week 17. The aberration that leads to bronchogenic cysts occurs around the fifth week of gestation. The cyst forms when an aberrant bud erroneously separates from the tracheobronchial tree and fails to branch and differentiate.

There is controversy concerning the timing of when bronchial atresia is believed to occur. One theory is that bronchial atresia results from intrauterine ischemia around the 16th week of gestation after distal conducting airways have
developed. The interruption of blood supply results in infarction and repair of the bronchus with subsequent occlusion of the lumen and separation of the bronchocele. This mechanism is similar to the insult that results in congenital intestinal atresia.

The concurrence of a bronchogenic cyst and bronchial atresia in this case supports the second theory which suggests that atresia occurs earlier at around the fifth week of gestation, when bronchogenic cysts also develop. During this time period, multiplying cells at the distal tip of the bronchus disconnect from the bud but continue to branch independently, resulting in a separate airway that does not communicate directly with its central bronchus. In addition to being associated with bronchogenic cysts, bronchial atresia has also been associated with congenital cystic adenomatoid malformations and pulmonary sequestration, both of which occur earlier in embryonic life, between the fourth and 10th weeks of gestation. This case further supports the unifying hypothesis that a singular insult early in development led to both bronchogenic atresia and the bronchogenic cyst in our patient.

CONCLUSIONS

The coexistence of bronchial atresia with a bronchogenic cyst does not definitively resolve the debate over when bronchial atresia occurs during development. The occurrence of a bronchogenic cyst at the fifth week of gestation does not necessarily exclude a subsequent insult that leads to bronchial atresia after the 17th week. However, this case does provide more evidence favoring the notion that the insult producing bronchial atresia occurs earlier in development. Bronchial atresia and bronchogenic cysts are both rare anomalies; therefore, a theory of the genesis of these 2 anomalies that invokes the occurrence of 2 separate insults at disparate time points during development seems unlikely.

REFERENCES