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An Incidental Finding

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PRESENTATION

A 26-year-old African-American man presented with lethargy. His girlfriend reported excessive daytime sleepiness that had worsened over the past several months. He had been referred for a sleep study to evaluate for obstructive sleep apnea but failed to follow up with this recommendation.

Assessment

He was afebrile with a blood pressure of 152/92 mmHg, a heart rate of 78 beats per minute, and a respiratory rate of 10. His room air saturation was 97%. He was difficult to arouse and morbidly obese, and the lung examination revealed distant breath sounds in the right middle and right lower lung zones. Arterial blood gas revealed a pH of 7.336 and a P_{CO_2} of 52 mmHg, which was consistent with pure hypercapnic respiratory failure.

Chest radiography in the emergency department revealed a hyperlucent right lung with tracheal shift to the left but no evidence of an effusion or infiltrate to suggest infection. Subsequent computed tomographic (CT) scans were obtained (Figures 1 and 2).

After bilevel positive airway pressure was administered for his hypercapnic respiratory failure, within several hours his mentation improved. The patient was admitted to the hospital for further observation. Once he was more awake, he revealed that he took multiple zolpidem (Ambien) to ''get a nap.'' When asked about his medical history, he denied previous respiratory problems.

Question: What disease process does the CT of the chest suggest?

A. Tension pneumothorax
B. Allergic bronchopulmonary aspergillosis
C. Congenital bronchial atresia
D. Cystic fibrosis

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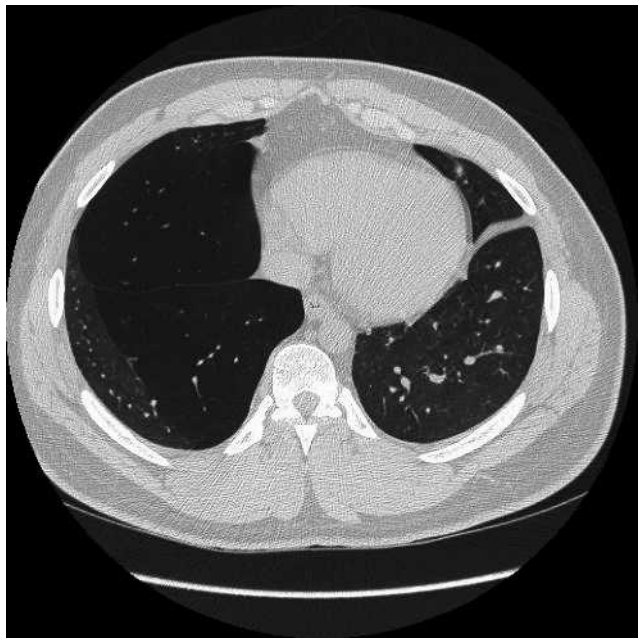

Figure 1. Axial view, computed tomographic scan of the chest.

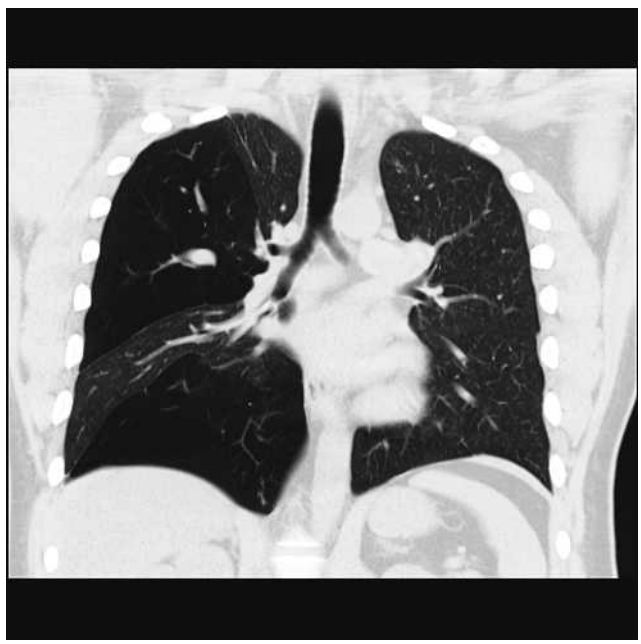

Figure 2. Sagittal view, computed tomographic scan of the chest.

DIAGNOSIS

The correct answer is C, congenital bronchial atresia, a congenital abnormality resulting from focal interruption of a lobar, segmental, or subsegmental bronchus with associated peripheral mucus impaction and associated hyperinflation of the obstructed lung segment.[1] Tension pneumothorax is incorrect, as the CT scan does not show an avascular collection of gas separating the visceral and parietal pleura. Additionally, the patient was hemodynamically stable and not hypoxic. Allergic bronchopulmonary aspergillosis is the most common cause of mucoid impaction but does not result in distal hyperinflation, as suggested by the CT scan. Cystic fibrosis is unlikely, as this diagnosis would manifest at a younger age, and CT findings are more likely to be bronchiectasis and peribronchial wall thickening, though mucus plugging can be present.

DISCUSSION

Congenital bronchial atresia was first described in 1953 by Ramsay in a series of patients with cystic lung disease.[1] The incidence is largely unknown, but it is thought to be a rare condition, with only 86 cases reported by 1986.[2] Any increase in the incidence since then could be explained by the increased availability of CT. The abnormality is an incidental finding in approximately 50% of cases, with most cases occurring in young males. Bronchial atresia appears on chest radiograph as a hyperinflated or hyperlucent area that may compress adjacent tissue and cause a shift of the mediastinum. The hyperinflation is thought to result from unidirectional flow through collateral airways (pores of Kohn, canals of Lambert).[1]

The etiology of congenital bronchial atresia is unknown but is theorized to be secondary to a traumatic event during fetal life, such as intrauterine ischemia, rather than a result of abnormal growth and development.[3] The tracheobronchial tree develops between days 24 and 36 of gestation. At 28 to 30 days, the primary bronchi are formed with outgrowths that lengthen into the segmental bronchi.[4] This period of development would seem to be most conducive to the formation of bronchial atresia. It would be interesting to know the incidence of congenital bronchial atresia among mothers who smoked during this period and those who did not. Unfortunately, there are no data on this relationship. In some cases, bronchial atresia may be acquired postnatally secondary to traumatic or postinflammatory insults to the bronchus, such as respiratory infections or surgery.[5]

Congenital bronchial atresia is usually benign and asymptomatic and is often discovered incidentally, as in our patient, whose hypercapnic respiratory failure was likely related to oversedation and not his congenital lung disease. The mean age at diagnosis is 17 years, and the disorder is more predominant in men.[1] Bronchial atresia is suggested on chest radiograph by a branching tubular or nodular area of increased opacity that extends from the hilum with surrounding hyperlucent lung parenchyma. However, CT, the most sensitive imaging modality, can be considered diagnostic in most cases. CT reveals a lack of communication between a bronchocele (a localized, mucus-filled dilatation of the bronchus) and hilum and is more sensitive in demonstrating segmental hyperinflation. The hyperlucent lung surrounding the bronchocele results from the combination of air trapping leading to dilated air spaces and focal parenchymal oligemia that is secondary to a combination of intrapulmonary vascular compression and hypoxic vasoconstriction.[6] Pulmonary function tests do not aid in the diagnosis.[7]

Gross pathologic findings include a blind-ending bronchus associated with a distal mucus-filled bronchocele but normal subsequent generations of bronchi.[7] Microscopically, the involved parenchyma consists of enlarged alveoli, which is indicative of lobar hyperinflation. Unless there is evidence of infection, destructive changes are usually minimal or absent.[1,7] This finding differentiates the disorder from emphysematous disorders such as congenital lobar emphysema.

The previous practice of performing surgery on all patients to make a definitive diagnosis has been largely abandoned because the majority of patients are asymptomatic and, therefore, no treatment is necessary. Because no communication with the normal tracheobronchial tree exists, infection is unusual. Lobar resection and segmentectomy have been used in patients with complications such as significant compromise of adjacent lung parenchyma or recurrent infection, but the ultimate goal is to preserve as much normal lung parenchyma as possible to maintain pulmonary function.[1]

Congenital bronchial atresia is a benign disorder with a significant radiographic appearance that is often discovered incidentally. Most patients do not develop symptoms related to the underlying lung defects and rarely need treatment. Our patient did quite well during his hospitalization and was discharged without any complications of his hypercapnic respiratory failure. He was again referred for a sleep study and counseled on the importance of weight loss.

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