Tension Pneumothorax with Evolving Cysts in an Infant

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PRESENTATION

A 5-month-old previously healthy term male infant presents to a rural emergency department (ED) for a 1-week history of increasing congestion, poor oral intake, and a temperature of 103°F (39.4°C). He is being treated with amoxicillin for presumed pneumonia. His examination in the ED is significant for scattered rhonchi and mild dehydration. His chest radiograph reveals inflammatory changes without a focal infiltrate (Fig 1). However, because this is his fourth presentation to the ED during this illness, he is admitted for observation. During the next 2 days his respiratory distress and tachypnea progressively worsen. On day 3 of his hospitalization he begins having episodes of desaturation and worsening retractions refractory to oxygen via low-flow nasal cannula. His examination at this time is significant for diffuse rhonchi throughout both lung fields with decreased air entry at the lung bases. Because his clinical status is deteriorating, he is transferred to a higher level of care.

On arrival at the referral center he is lethargic, with decreased breath sounds and persistent desaturations. A respiratory swab polymerase chain reaction is positive for respiratory syncytial virus (RSV). His chest radiograph reveals a large right-sided tension pneumothorax (Fig 2). A pigtail chest tube is emergently placed, and his work of breathing improves. Within 2 days he is no longer requiring supplemental oxygen. The medical team is unable to successfully put the chest tube to water seal, however, because each time it is sealed there is a rapid re-accumulation of his pneumothorax. After several unsuccessful attempts, his chest tube is placed back to suction and he is airlifted, via helicopter, to a pediatric tertiary care center for further evaluation, now 11 days after his initial presentation. His chest radiograph just before transfer reveals a well-positioned chest tube with interval decrease in the size of the pneumothorax.

On arrival at the tertiary care center he is well appearing, with no increased work of breathing, and is saturating 97% to 100% on room air. His lung sounds are clear bilaterally, but an air leak is auscultated on inspiration. He has no crackles, wheezes, or rhonchi. Chest radiography is repeated to verify the position of the chest tube and incidentally reveals a large cystic-appearing lesion in his right lung that has not been previously described (Fig 3). Computed tomography of the chest is performed that reveals multiple air-filled cystic lesions in the right upper lobe of the lung as well as a persistent right-sided pneumothorax (Fig 4). Surgery and pulmonology are consulted for co-management. On day 17 of his cumulative hospitalization he undergoes wedge resection of the right upper lobe of his lung without complications. Samples of the cystic lesions are sent to the pathology laboratory to confirm the diagnosis.
DIAGNOSIS

The diagnosis of air leak secondary to rupture of a functional alveolar bleb, likely due to proximal mucus plugging in the course of RSV bronchiolitis, was confirmed by pathology (Fig 4).

DISCUSSION

Spontaneous pneumothorax in children outside of the newborn period is exceedingly rare, with an estimated prevalence of only 1 in every 10,000 hospitalized children. The bulk of reported cases are secondary pneumothoraces due to an acute infection, trauma, or congenital malformation. (1) In our case, we initially considered a congenital malformation such as congenital pulmonary adenomatoid malformation (CPAM), given the well-defined cystic lesions on imaging. CPAMs are the most common form of congenital parenchymal lung malformations, with an estimated incidence of 1 in 11,000 to 1 in 35,000 live births. (2) Most CPAMs are diagnosed prenatally during routine 20-week ultrasonography, and complications may include hydrops fetalis, recurrent infections in childhood, or malignant transformation. We reviewed prenatal ultrasonography of this patient, and there were no signs of lung lesions, and neither were these lesions visible on previous radiographs. In addition, spontaneous pneumothorax is a rarely described comorbidity of CPAM. This was definitively ruled out with histologic examination of the surgical specimen.

Also on the differential diagnosis was postinfectious pneumatocele. A pneumatocele is a thin-walled air-filled cyst in the lung parenchyma, sometimes seen in children secondary to a severe pulmonary infection, most commonly Staphylococcus aureus pneumonia. It develops when the necrotic airway forms a cystic lesion connected to the bronchial tree. Similar to the ball valve physiology described later herein, air gets trapped in this lesion, causing hyper-inflation of the cyst and resulting pneumothorax. (3) Histologically, samples obtained from these patients reveal necrotic debris and multinucleated giant cells. (4) Children with this condition are generally quite ill with hypoxic respiratory failure requiring aggressive management in an ICU setting. (5) Because this patient had a relatively short and mild preceding illness, pneumatocele was less likely, and it was ruled out by histologic analysis.

This case is remarkable for the abrupt change in the appearance of the radiograph before and after transfer to the tertiary care center. We considered the possibility that air transport contributed to the change. As explained by Boyle’s law, lower barometric pressure at high altitude causes...
volume expansion, which could unmask a more significant lesion than had been previously identified. (6)(7) Although helicopters travel at a lower altitude than fixed-wing transport (10,000 ft vs 20,000–30,000 ft), there are predictable changes in oxygen requirements and lung volumes at this altitude that could worsen an air leak. (7) However, histologic analysis revealed a thick wall surrounding the lesion, suggesting an inflammatory reaction rather than a simple air leak. In addition, the chest tube, set to suction, was functioning properly and should have prevented expansion of an intrapulmonary lesion. (7)

RSV infection was most likely the inciting event that led to his intraparenchymal cysts and persistent air leak. Bronchiolitis is the most common lower respiratory tract infection in children younger than 2 years. The incidence is estimated to be 11.4 to 19.6 cases per 100 children before age 1 year. (8) RSV is the most common cause. (9) Although case reports suggest that spontaneous pneumothorax secondary to viral lower respiratory tract infection occurs with an estimated prevalence of 0.6%, none were treated with surgical intervention. All reported cases resolved with either needle thoracentesis or chest tube placement. (1)(8)(9)(10)(11)

Histologic examination of the resected lung specimen identified an air-filled cavity bordered by reparative tissue (foamy macrophages) and foreign body giant cells (Fig 5). There was no epithelial lining in the cavity, adjacent smooth muscle wall, or satellite cysts, which definitively ruled out CPAM. Although these findings were nonspecific, the results point to a reaction that can be seen when the airway responds to a foreign body. Similar to a foreign body in the airway, mucus plugs can cause lung injury by a ball valve effect.

Figure 3. Anteroposterior chest radiograph on hospital day 11 shows a right-sided moderate pneumothorax with a pigtail chest tube in place. Circular luencies concerning for cystic/cavitary lesions are seen in the right lung.

Figure 4. Contrast computed tomography of the chest on hospital day 12 shows multiple air-filled cysts in the right upper lung with right-sided pneumothorax.

Figure 5. 1, A cavity (white space on left) lined by foamy macrophages (magnified in inset). 2, Immunostain for KP-1, a macrophage marker (brown areas positive), shows a thick band of positive cells, corresponding to the foamy macrophages, lining the cavity. 3, Immunostain for cytokeratin, a marker of epithelium, shows an inverse pattern to panel 2, with the band of macrophages staining negatively in contrast to the pulmonary parenchyma on the right. 4, Immunostain for smooth muscle actin shows that there is no muscular wall adjacent to the cavity. 5, Cavity (white space on the right) lined by foreign body giant cells.
During inspiration, the airway expands, allowing air to pass the obstruction. However, during expiration, the airway collapses around the obstruction and prevents gas from escaping. As the cycle repeats, the lung beyond the mucus plug expands and may lead to pneumothorax. (4)

Patient Course
Our patient had no postoperative oxygen requirement after he underwent thoracotomy with wedge resection of his right upper lobe. The chest tube was removed on postoperative day 5, and he was discharged home. He was seen in the clinic the following week and was well appearing, with clear breath sounds and no increased work of breathing. He has had no subsequent recurrences of his symptoms.

Summary
- Secondary air leaks and cavity formation as sequelae of respiratory syncytial virus (RSV) are exceedingly rare.
- Cavitary changes in the lung may develop secondary to RSV with mucus plugs and air trapping.
- RSV is a frequent cause of pneumonitis in infants that rarely requires surgical intervention, but it is important to recognize the possibility of such complications, which may occur even as the primary infection is resolving.

References
Visual Diagnosis: Tension Pneumothorax with Evolving Cysts in an Infant
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