Case 2: The foreign body that wasn’t

A one-year-old previously healthy girl was referred to a paediatric emergency department with a two-day history of cough and nasal discharge. There was no history of wheezing, feeding difficulties or recall of foreign body aspiration. On physical examination, she was alert and afebrile. Her heart rate was 124 beats/min, her respiratory rate was 32 breaths/min and her oxygen saturation was 98% on room air. There was no evidence of increased work of breathing. She weighed 7.0 kg, which placed her below the third percentile for her age. Normal heart sounds, although louder over the right hemithorax, were heard on cardiac auscultation. There were no murmurs heard. The trachea was midline. Bilateral diminished air entry was present with no wheezes or crackles heard, although breath sounds transmitted on the right side were harsh in quality. A chest radiograph revealed a hyperinflated left lung with a mediastinal shift to the right (Figure 1). The hyperlucent lung suggested a ball-valve effect, raising the possibility of a foreign body aspiration in the left mainstem bronchus, but subsequent bronchoscopy did not reveal a foreign body. Further investigation established the diagnosis.

Figure 1) Posterior to anterior chest radiograph revealing hyperinflation of the left lung with a mediastinal shift to the right
CASE 2 DIAGNOSIS: SCIMITAR SYNDROME

Computed tomography imaging studies of the thorax with contrast indicated a hypoplastic right lung with total suprahepatic anomalous pulmonary venous return to the inferior vena cava (Figure 2) diagnostic of Scimitar syndrome. Scimitar syndrome was first described in 1836 and is a rare presentation of congenital cardio-pulmonary anomalies. In 1956, Halasz et al (1) drew a parallel between the shape of the anomalous right pulmonary vein and a scimitar, a short, curve-bladed Turkish sword. In 1960, Neill et al (2) conferred the syndrome with the name of its hallmark vein. It has an annual incidence rate of approximately two per 100,000 live births (3).

Scimitar syndrome is usually associated with right anomalous pulmonary venous return with right pulmonary hypoplasia of varying degrees. It can also occur on the left. Cardiac anomalies range from 63% to 75% and include dextroposition of the heart, right pulmonary artery hypoplasia, abnormal systemic arterial collaterals from the infradiaphragmatic aorta and secundum atrial-septal defect. Less commonly reported cardiac anomalies include hypoplastic left heart, coarctation of the aorta, patent ductus arteriosus, tetralogy of Fallot and persistent left superior vena cava. Scimitar syndrome is believed to be the outcome of failed connection between the right pulmonary veins and the left atrium during fetal development (3). Scimitar syndrome should be considered when the triad of respiratory distress, right lung hypoplasia and dextroposition of the heart are seen (4). Presentations range from asymptomatic manifestations in infancy to an incidental finding in adulthood. In patients presenting in infancy, symptoms include tachypnea, recurrent pneumonia, failure to thrive and signs of heart failure. These patients tend to have a poorer prognosis. Computed tomography with contrast and magnetic resonance imaging are the diagnostic investigations of choice due to their ability to delineate abnormal anatomy, although the diagnosis may also be made using echocardiography. Symptomatic patients may require surgical management. Other indications for operative intervention include the presence of an atrial septal defect accompanying the scimitar vein and stenosis of the abnormal right pulmonary venous trunk. Surgery for affected children may include occlusion of systemic arterial supply to the right lung, diversion of the right pulmonary venous return to the left atrium and repair of structural cardiac defects. Pneumonectomy has previously been indicated but ideally should be avoided because the right lung may contribute to some gas exchange and may increase in volume with growth (3).

The infant described in the current case was diagnosed serendipitously after presenting with upper respiratory symptoms and abnormal physical examination findings, which prompted a chest x-ray. Initial imaging studies directed clinical suspicion toward foreign body aspiration. While the history was not strongly supportive, caregivers will often not witness or recall an aspiration event. Emergency physicians rely on chest radiographs, including inspiratory and expiratory views. The expiratory view typically reveals air trapping due to the ball-valve effect in the lung on the ipsilateral side. The sensitivity and specificity of plain films with foreign body aspiration are low (range 40% to 60%). Bronchoscopy remains the test of choice for diagnosis and treatment of airway foreign bodies. Radiography demonstrated a hyperlucent lung in the infant. Differential diagnosis can be broken down into chest wall anomalies (Poland syndrome), lung parenchymal abnormalities, pulmonary vascular abnormalities, central airway abnormalities (foreign body aspiration or endobronchial tumour) or mediastinal abnormalities (foreign malformation or lymphadenopathy). Lung parenchymal abnormalities include, but are not limited to, bronchial atresia, congenital lobar emphysema, congenital pulmonary airway malformation and pneumothorax. Pulmonary vascular abnormalities include Scimitar syndrome, pulmonary agenesis, proximal interruption of the pulmonary artery and pulmonary artery sling (5).

Although presentations of congenital lung anomalies are infrequent in an acute setting, in contrast to infections such as pneumonia and bronchiolitis, the present case serves as an educational reminder to include congenital malformations in the differential diagnosis, especially in the context of an infant with an abnormal chest radiograph.

CLINICAL PEARLS

- It is important to consider a wide differential diagnosis when a hyperlucent lung is observed on chest x-ray.
- Scimitar syndrome is a rare congenital abnormality, which can present in infancy, can be associated with congenital heart malformations or remain undiagnosed until adulthood.

REFERENCES


Krishnan Paruwar MD
Western University
Rodrick Lim MD, Timothy Lynch MD
Department of Paediatrics, Western University
Children’s Hospital at London Health Sciences Centre
London, Ontario