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Term Neonate with Respiratory Distress

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PRESENTATION

A female infant is born at 39 weeks to a 21-year-old primigravida with an unremarkable antenatal history and good prenatal care. The Apgar scores are 6, 7, and 8 at 1, 5, and 10 minutes, respectively. Physical examination reveals an appropriate-for-gestational age 3.3-kg term infant with grunting, tachypnea, and intercostal retractions. Decreased air entry is noted over the right side of the chest. A grade 2/6 systolic murmur is heard over the left sternal border with normal heart sounds and good peripheral pulses. The rest of the physical examination findings are within normal limits except for a right-sided small ear tag. The neonate requires positive pressure ventilation initially in the delivery room and then is placed on continuous positive airway pressure (CPAP) at 30% fraction of inspired oxygen. Chest radiography shows near-complete opacification of the right thorax with mass effect and normal lung markings on the left (Fig 1). Due to worsening respiratory distress, she undergoes intubation and is transferred to the regional perinatal center for diagnosis and management.

Differential diagnoses considered at this point for unilateral lung field opacity with mass effect include right-sided diaphragmatic hernia, congenital pulmonary airway malformation, pulmonary sequestration, or a space-occupying lesion like a tumor. Ultrasonography of the chest shows a large, solid mass in the right thorax with an intact diaphragm, an abnormality consistent with consolidated lung or a tumor (Fig 2).

DISCUSSION

Diagnosis

Computed tomography (CT) of the chest reveals the diagnosis (Figs 3 and 4). An aberrant left pulmonary artery (LPA) is seen arising from the right pulmonary artery consistent with an LPA sling. A severely stenotic right main bronchus at its origin and a possibly atretic right upper lobe bronchus are noted.

Case Progression

Given these airway and vascular anomalies, the infant is transferred to a tertiary center with expertise in cardiothoracic surgery. Repair of the anomaly includes reimplantation of the LPA to the main pulmonary artery and ligation of the ductus arteriosus. No atresia of the

bronchus is noted intraoperatively as the right lung inflated well once the sling was repaired. The neonate undergoes extubation on postoperative day 3 and is transferred back to the referring center on CPAP. She is subsequently weaned to room air by 4 weeks after birth. Genetic evaluation reveals no other anomalies. Microarray is offered and recommended but deferred by the family.

The Condition

LPA sling is a rare congenital vascular anomaly in which the LPA arises from the posterior aspect of the right pulmonary artery, coursing over the right main bronchus and then posteriorly between the trachea and esophagus to reach the left lung, forming a sling around the trachea. LPA sling is often associated with congenital tracheal stenosis.

First reported in 1897 by Glaevecke and Doehle, the term vascular sling was used by Contro et al in 1958 to distinguish it from vascular ring. (1)(2) Later, Berdon et al introduced the term “ring-sling complex” with description of the associated airway anomalies. (3) The first surgical correction was performed by Potts in 1958. (2) The exact incidence is not reported because of the rarity of the condition. Infants can present with varying degrees of respiratory distress within the first few weeks to months after birth. Although symptoms are noted early, they are usually attributed to other causes. In the current case, the near-complete opacification and mass effect of the right lung prompted appropriate imaging, which led to an early diagnosis. An LPA sling presenting as a unilateral lung anomaly may also be detected during antenatal ultrasound screening. (4) In our patient, antenatal ultrasonography did not reveal any anomaly. The signs and symptoms depend on the extent of airway involvement. Compression of the bronchus causes atelectasis or hyperinflation (ball valve mechanism). Tracheal compression can result in wheezing, stridor, or severe respiratory compromise. Occasionally, dysphagia and failure to thrive can be seen. Patients can be asymptomatic initially and present with symptoms in older childhood or as adults. LPA sling is associated with various cardiac and noncardiac abnormalities. Common cardiovascular associations include atrial septal defect, ventricular septal defect, tetralogy of Fallot, coarctation of aorta, and patent ductus arteriosus. Extra-cardiac anomalies include vertebral defects, anal atresia, tracheoesophageal fistula, renal anomalies, and limb abnormalities (VATERL), biliary atresia, absent gall bladder, Hirschsprung disease, and imperforate anus. (5)

The diagnosis of LPA sling is primarily radiologic. A chest radiograph can reveal right-sided hyperinflation or atelectasis, indentation of the trachea, tracheal narrowing, and right-sided tracheal bronchus. An esophagram is of limited value but can show an indentation anterior to the esophagus. Echocardiography can establish the diagnosis of pulmonary sling and delineate other associated cardiac anomalies. CT and magnetic resonance imaging (MRI) are the imaging modalities of choice and are useful for planning surgical management. CT has an advantage over MRI because of better visualization of lung parenchyma, faster scanning time, and lower requirement for sedation, but has the disadvantage of exposure to ionizing radiation. CT angiography gives the most information. (6) The degree of tracheal stenosis is best assessed with bronchoscopy.

Management

Management is based on the clinical symptoms and anatomy. Asymptomatic patients are followed clinically. Symptomatic patients undergo elective or emergent repair depending on clinical stability. The preferred approach for surgical repair is median sternotomy with cardiopulmonary bypass, thus allowing simultaneous repair of other intrathoracic anomalies if present. (7) LPA sling repair involves reimplantation of the LPA onto the main pulmonary artery. Another alternative approach involves trans-location of the undivided LPA anterior to the trachea at the time of tracheal resection for congenital tracheal stenosis. (8) Tracheal stenosis is the main comorbid condition that dictates the long-term prognosis in patients with LPA sling. Surgical repair for moderate-to-severe stenosis may include tracheal resection with end-to-end anastomosis, slide tracheoplasty, or patch tracheoplasty. Tracheal resection with end-to-end anastomosis is preferred if stenosis involves a short segment of the trachea (25%–40%). (9) Complications are fewer and the outcomes are better in patients treated with slide tracheoplasty. (10)

Lessons for the Clinician

- A left pulmonary artery (LPA) sling is a rare, congenital vascular anomaly that should be suspected in a neonate with a unilateral lung field opacification. Surgical correction is the treatment when indicated.
- The outcome and prognosis is usually favorable and depends on associated tracheobronchial, cardiac, and extracardiac anomalies.

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American Board of Pediatrics Neonatal-Perinatal Content Specification

- Know the clinical features of an infant with airway obstruction.

Figure 1.
Anteroposterior view radiograph of chest and abdomen at birth showing near-complete opacification of the right thorax.

Figure 2. Ultrasound scan of the chest, right thorax with A) a large mass on the right, consistent with a consolidated lung with an intact diaphragm, and B) liver.

Figure 3. Axial view of chest computed tomography scan showing A) main pulmonary artery, B) trachea, C) left pulmonary artery with an abnormal origin from the right branch instead of the main pulmonary artery looping around the trachea forming a “sling,” D) esophagus with enteric tube, and E) right pulmonary artery.

Figure 4. Coronal view of chest computed tomography scan showing A) trachea, B) left pulmonary artery with an abnormal course around the trachea at its bifurcation, causing stenosis of the right bronchus, C, D) areas of stenosis of the right main bronchus, and E) atelectatic upper lobe of the right lung.