Pulmonary Hypoplasia

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Key words: pulmonary hypoplasia, intrauterine development, environmental factors

IMAJ 2004;6:494–495

Pulmonary hypoplasia represents a spectrum of malformations characterized by incomplete development of the lungs. Abnormalities in the development of the lung tisssue can be classified into three groups: agenesis – a complete absence of one of both lungs; aplasia – a rudimentary bronchus that ends in a blind pouch, with no evidence of pulmonary vasculature or parenchyma; and a hypoplastic lung [1].

We describe a young patient with a hypoplastic lung who was admitted with a high grade fever and suspected left lower lobe pneumonia.

Patient description

A 25 year old man was admitted with high grade fever (38.5°C) accompanied by purulent cough and dyspnea (more than 30 respirations per minute). The cough started a week earlier and became worse 2 days before admission. On admission, the patient looked ill, had high fever with chills, and was breathing heavily with about 30–34 respirations/minute. On physical examination, no breath sounds were heard on the left lung; on the right lung there was an alveaolar respiration without any rhales or wheezes. The rest of the physical examination was unremarkable.

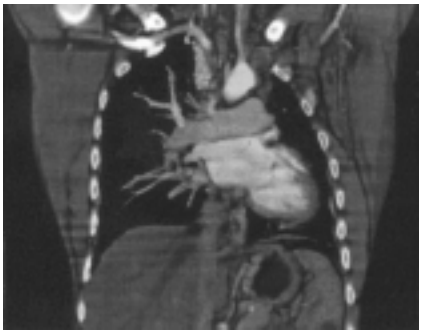
Blood results demonstrated 12,000 leukocytes with a strong left shift, without any other abnormalities. Blood cutures were negative, and a chest X-ray demonstrated diminished size of the left lung, without clear evidence of pneumonia. He was treated with intravenous antibiotics (cefalosporins) which led to a dramatic clinical improvement within 2 days. A chest computed tomography with angiography scan demonstrated a hypoplastic left lung with severe hypoplasia of the left pulmonary artery [Figure].

Comment

Hypoplasia of the lung is classified as primary (idiopatic) or secondary. Primary hypoplasia is an intrinsic defect in the process of lung development. The incidence has been estimated as 1–2 cases in 12,000 live births. Hypoplastic lungs are smaller and weigh less than normal lungs. There is a decrease in the number of alveoli and in the alveolar size, but the gross morphology is unremarkable. Abnormalities in the pulmonary artery have also been identified. The pulmonary artery is also hypoplastic.

The variation in the morphologic findings depend on the cause of the hypoplasia and on the timing: if the insult occurs in the early weeks of intrauterine development, the number of airways is abnormal and the blood supply comes only from the systemic circulation; if the insult occurs in late pregnancy or early infancy, the pulmonary artery is present, the bronchial artery distribution is normal and its volume increased, and the number of airways is also normal.

The radiologic manifestations are characterized by reduction in size of the treated lung in one hemithorax. The reduced volume is indicated by approximation of the ribs, elevation of the ipsilateral diaphragm and shift of the mediastinum. In most cases the contralateral lung is overinflated. CT of the chest shows a patent bronchus, a hypoplastic pulmonary artery and a small lung with a shift of the mediastinum. CT scan is necessary to differentiate between hypoplasia from other conditions such as atelectasis or Swyer-James syndrome [2]. Clinical findings depend on the severity of the abnormality. Patients – whether children or adults – can be asymptomatic or present with respiratory symptoms such as dyspnea and wheezing with infection of the tracheobronchial tree or pneumonia [3].



CT 4-D reconstruction showing a small left lung with lack of the left hillus.

Secondary hypoplasia occurs in association with enviromental factors or other congenital anomalies. Several mechanisms have been implicated in secondary pulmonary hypoplasia, and decreased volume of the ipsilateral hemithorax is the most frequent association. The most common cause is a space-occupying mass like congenital diaphragmatic hernia. The diameter of the pulmonary artery in a fetus with congenital diaphragmatic hernia correlates with the outcome [4]. Other mechanisms include thoracic neuroblastoma and a sequestred lung. A variety of musculoskeletal deformities of the chest and abdominal wall are associated with hypoplastic lung. Also associated with

secondary hypoplasia are anomalies of the kidney and the urinary tract. The most frequent syndrome is Potter's syndrome (renal agenesis, abnormal fascies, limb abnormalities and pulmonary hypoplasia). Decreased pulmonary vascularity can decrease the size and number of alveoli. This defect was seen in some patients with tetralogy of Fallot [4].

The prognosis of patients with primary or secondary hypoplasia depends on the severity and the associated anomalies. Patients with milder forms of hypoplasia can survive until adulthood and their pulmonary function tests show abnormalities, e.g., an increased residual volume or hyper-responsivness to metacholine. These anomalies predispose to respiratory infections that can be severe.

References

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Capsule

Sildenafil citrate (Viagra) in men with erectile dysfunction and chronic stable angina

Studies have shown that sexual activity does not lead to exaggerated heart rate or blood pressure responses, and that stable angina patients are not at greater relative cardiovascular risk during sexual intercourse. Men with cardiovascular disease are more likely to have erectile dysfunction (ED) than the general population. Sildenafil citrate is an oral treatment for ED and a potent and selective inhibitor of cyclic guanosine monophosphate (cGMP)-specific phosphodiesterase type 5 (PDE5). Using a treadmill as a validated surrogate for sexual activity, Fox et al. investigated a) the safety and tolerability of sildenafil in patients with ED and stable angina not requiring nitrates and, in light of the mechanism of action of sildenafil suggestive of its potential as an antianginal agent, and b) the effect of sildenafil on the time to the onset of effort angina. They provided direct evidence that sildenafil does not cause coronary steal or reflex tachycardia and is even likely to improve the anginal threshold, and concluded that these patients can safely receive sildenafil treatment for their ED.

Eur Heart J 2003;24:2206

A. Roth

Capsule

Antimicrobial resistance incidence among *Helicobacter pylori*-infected persons

Duck et al. probed the question of *H. pylori* antimicobial resistance. *Helicobacter pylori* is the primary cause of peptic ulcer disease and an etiologic agent in the development of gastric cancer. *H. pylori* infection is curable with regimens of multiple antimicrobial agents, and antimicrobial resistance is a leading cause of treatment failure. The Helicobacter pylori Antimicrobial Resistance Monitoring Program (HARP) is a prospective, multicenter U.S. network that tracks national incidence rates of *H. pylori* antimicrobial resistance. Of 347 clinical *H. pylori* isolates collected from December 1998 through 2002, 101 (29.1%) were resistant to one antimicrobial agent, and 17 (5%) were resistant to two or more antimicrobial agents. Eighty-seven (25.1%) isolates were resistant to metronidazole, 45 (12.9%) to clarithromycin, and 3 (0.9%) to amoxicillin. On multivariate analysis, black race was the only significant risk factor ($P < 0.01$, hazard ratio 2.04) for infection with a resistant *H. pylori* strain. Formulating pretreatment screening strategies or providing alternative therapeutic regimens for high risk populations may be important for future clinical practice.

Emerg Infect Dis 2004;10:1088

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