Type II congenital pulmonary airway malformation in an esophageal lung

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A seven-month-old girl, born prematurely (birth weight 1000 g) from a twin pregnancy, was admitted to hospital due to recurrent pneumonia and tachypnea. She experienced cough and respiratory distress during feeding. The right hemithorax was smaller than the left, with diminished breath sounds and dullness. Chest x-rays revealed decreased lung volume and diminished breath sounds and dullness. On bronchography, the right lung was absent and the trachea only continued into the left main bronchus. Echocardiography and angiotomography revealed agenesis of the pulmonary artery right branch. The surgical finding was an esophageal right lung, which was removed; the histopathological diagnosis was type II congenital pulmonary airway malformation in an esophageal lung.

Key Words: Bronchopulmonary malformation; Chronic aspiration; Congenital cystic adenomatoid malformation; Congenital pulmonary airway malformation; Esophageal lung; Recurrent pneumonia

Learning objectives:
• To recognize clinical and radiological data suggestive of an esophageal lung, and to consider this condition in the differential diagnosis of persistent or recurrent pneumonia.
• To be aware that two lung malformations may occur simultaneously in the same patient.

CanMEDS competency: Medical Expert

Pre-test
• What clinical and radiological data lead to suspicion of an esophageal lung?
• What are the pathognomonic features of an esophageal lung in the barium swallow?

The esophageal lung or esophageal bronchus is characterized by an aberrant bronchus or lung that arises from the esophagus, with total absence of ventilatory function in the involved pulmonary tissue (1-5). The present report describes an unusual case of a type II congenital pulmonary airway malformation (CPAM) that occurred in an esophageal lung (6).

CASE PRESENTATION

The patient was a seven-month-old girl born after 34 weeks’ gestation in a twin pregnancy, with a birth weight of 1000 g. In the neonatal period, she developed pneumonia and required mechanical ventilation for 40 days, and was discharged eight days later. She was hospitalized on three more occasions due to pneumonias and was sent to the Hospital de Pediatría, Centro Médico Nacional Siglo XXI (Mexico DF) for evaluation and treatment.

On admittance, her weight was 2900 g, oxygen saturation was 92% on room air, with respiratory distress, asymmetrical thorax due to a smaller size of the right hemithorax. She arrived without an orogastric tube and, thus, previous oral feeding was assumed, but she experienced an episode of choking after trying to initiate by oral route.

A chest radiograph revealed a pencil-like tapering of her tracheal air column, and the right lung was smaller and with multiple radiolucent images compared with the left lung, which was overdistended (Figure 1). Bronchoscopy revealed 20% subglottic stenosis and progressive diminution of tracheal lumen and, thus, the carina could not be visualized. Endoscopy showed incompetent hiatus. In contrasted images, the barium swallow and a Vidalva manoeuvre revealed a three-branch ramifications emerging from the lower one-third of the esophagus, which ended in a cul-de-sac, establishing the diagnosis of esophageal lung (Figure 2). An echocardiogram showed probable agenesis of the right pulmonary artery, neoformation of blood vessels in the esophageal lung and venous return through pulmonary veins draining to the right atrium. Angiography with three-dimensional reconstruction clearly showed diminution of the tracheal diameter, which continued with the left main bronchus, and the esophageal lung emerging from the esophagus (with an orogastric tube inside); the right lung had

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lower volume than the left, with distortion of the normal architecture due to hypodense lesions (Figure 3). Additionally, this imaging modality also demonstrated venous drainage of the esophageal lung into the left atrium (Figure 4). Bronchography showed that the trachea was in continuation with the left main bronchus, and the right lung was excluded (Figure 5).

The intraoperative findings were right lung with main bronchus arising from the esophagus, with remnant veins in its hilum and lacking systemic arterial blood supply. A right pneumonectomy was performed. The histopathological analysis reported a type II congenital cystic adenomatoid malformation (Figure 6), corresponding to type II CPAM in the current nomenclature (6).

Figure 1) Chest radiograph showing dilation of the proximal trachea, with pencil-like tapering, right lung with diminished volume and multiple radiolucent images, and overdistension of the left lung.

Figure 2) Images of the esophageal lung. A Esophagogram showing three branches emerging from the lower one-third of the esophagus and ending in cul-de-sac. B The coronal plane of computed tomography shows the esophagus (black arrow) and the bronchus for the esophageal lung (white arrow).

Figure 3) Computed angiotomography showing diminution of the tracheal diameter, which continues with the left main bronchus (white arrow), and the esophageal lung emerging from the esophagus (with an orogastric tube inside, black arrow); the right lung had lower volume than the left side, with distortion of the normal architecture.

Figure 4) Computed angiotomography showing the venous drainage of the esophageal lung into left atrium (white arrow).

Figure 5) Bronchography revealing exclusion of the right lung. The trachea continues with the left main bronchus (white arrow), and abnormalities of the left tracheobronchial tree are present.

Figure 6) Histological section of the lung parenchyma. A A distorted architecture with multiple cystic spaces can be seen (original magnification ×10). B It is apparent that these cysts are covered by pseudostratified columnar epithelium (white arrow) and contain neutrophilic exudate (original magnification ×40).
DISCUSSION

To our knowledge, the present case is the first report of type II CPAM occurring in an esophageal lung. In this patient, the malformation corresponded to a group II classification of bronchopulmonary malformations communicating with the foregut (1,2,6).

Respiratory distress associated with feeding and persistent or recurrent episodes of pneumonia, as seen in the present case, are the most common clinical events in children with an esophageal lung. Bronchiectasis, hemoptysis, gastrointestinal hemorrhage and dysphagia can also be present (3-5). Diagnosis is made using esophagography, although other useful imaging modalities are computed tomography, magnetic resonance imaging and angiotomography. When an esophageal bronchus or esophageal lung is suspected, angiotomography shows parenchymal abnormalities and depicts venous and arterial blood flow with higher precision than other studies. Other complementary imaging modalities are fiberoptic bronchoscopy, digestive endoscopy and echocardiography. When the malformation and its vasculature cannot be delimited, arteriography is recommended (3-5).

An esophageal bronchus or esophageal lung is often unilateral. Malformed airways usually arise from the lower esophagus and the gastroesophageal junction, and histopathology shows squamous epithelium in the esophageal end of the communication and ciliated pseudostratified columnar epithelium on the pulmonary side (3).

Many malformations, such as pulmonary hypoplasia or agenesis, pulmonary sequestration, and malformations of the heart, kidneys, ribs, vertebral column and digestive tract have been associated with this entity (4). In this context, our patient presented other congenital-associated malformation such as agenesis of the pulmonary artery right branch and congenital tracheal stenosis.

REFERENCES

Surgical resection of the involved lung parenchyma is curative in the majority of cases. When a normal lung is demonstrated, more conservative surgery has been suggested in which an anastomosis of the esophageal bronchus is performed on the normal tracheobronchial tree (3-5).

CONCLUSIONS

An esophageal lung must be considered in children with cough or respiratory distress during feeding, recurrent pneumonia since birth or persistent opacities in chest x-rays. Other associated malformations, including CPAM as in the present case, must be investigated.

Post-test

• What clinical and radiological data lead to suspicion of an esophageal lung?

An esophageal lung must be suspected when respiratory distress associated with feeding (choking) and a persistent or recurrent pulmonary opacity in the chest x-rays are present.

• What are the pathognomonic features of an esophageal lung in the barium swallow?

In an esophageal lung, contrasted images with barium swallow and with Valsalva manoeuvre can reveal the branch ramification of a rudimentary or well-formed tracheobronchial tree emerging from the esophagus.

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