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CASE REPORT

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Connected to the wrong pipe: esophageal bronchi mimicking bilateral bronchial atresia

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Key words

Esophageal bronchus; communicating bronchopulmonary foregut malformation; rare disease; neonatology.

ABSTRACT

Infants frequently present with respiratory symptoms, but diagnosing the underlying pulmonary condition is sometimes challenging. Here, we describe an infant presenting with respiratory distress due to a rare pulmonary condition. Different investigations eventually led

to the correct diagnosis and the patient successfully treated. Rare respiratory conditions need to be considered to ensure early and appropriate care.

A 2-week-old neonate was referred with a postnatal history of tachypnea, poor feeding and elevated plasma inflammation markers. The patient presented with mild episodes of coughing after feeding and diminished breath sounds of the right upper lung. A chest radiograph revealed bilateral consolidations. Whereas bronchoscopy was suggestive of bilateral bronchial atresia, computed tomography supported bronchial atresia of the right upper and middle lobe and a left-sided broncho-esophageal communication. Surprisingly, an upper gastrointestinal series revealed bilateral esophageal insertion of bronchi and the diagnosis of a communicating bronchopulmonary foregut malformation (CBPFM) was made. Two-stage lobectomy of the affected lobes and segments was performed at days 31 and 41 after birth. Histopathological examination exhibited hamartomatous lung tissue with purulent bronchopneumonia. At a follow-up examination after 4 years, the patient was asymptomatic and thriving well with oral feeds.

CBPFM are rare malformations. This case highlights the clinical challenge of diagnosing this rare condition. There is a need to raise awareness for such uncommon conditions and improve diagnostic accuracy. For optimal management a multidisciplinary approach is essential.

Introduction

Neonates presenting with pulmonary symptoms within the first days of life are not infrequent (1, 2). While the clinical presentation is usually non-specific, diagnosing the underlying pulmonary condition can be challenging (3). However, delayed diagnosis may result in considerable complications (4). Here, we describe an infant with prolonged neonatal respiratory distress, the investigations performed, how the correct diagnosis was made and the patient treated successfully. The aim of this case report is to show that the diagnosis of rare respiratory conditions is challenging, but a favorable long-term clinical outcome can be achieved when diagnosed and treated early.

Case presentation

The male patient was born via vaginal delivery as the first child of healthy parents. During his first two weeks of life, tachypnea, fever, poor feeding and elevated plasma inflammation markers were noted. Also, the patient presented with mild coughing episodes after feeding and diminished breath sounds of the right upper lung. A chest radiograph was performed revealing bilateral consolidations (**Figure 1A**). Late-onset neonatal sepsis (LONS) was suspected and an empiric, intravenous antibiotic treatment started. As the clinical symptoms persisted, bronchoscopy as well as high-resolution computed tomography (HR-CT) were performed during the same anesthesia. Whereas bronchoscopy was suggestive of bilateral bronchial atresia, HR-CT supported bronchial atresia of the right upper and middle lobe (**Figure 1B, C**) and a left-sided broncho-esophageal communication (**Figure 1D**, red arrow). Due to persisting coughing episodes after feeding, in a multidisciplinary team board meeting the need for further investigations were discussed. For the better visualization of the esophagus, it was decided to perform an upper gastrointestinal series. The investigation revealed a bilateral esophageal insertion of bronchi (**Figure 1E, F**) and the diagnosis of a communicating bronchopulmonary foregut malformation (CBPFM) was made. Two-stage lobectomy of the affected lobes and segments was performed at the age of 31 and 41 days. Histopathological examination exhibited hamartomatous lung tissue with purulent bronchopneumonia. The latest follow-up chest x-ray with 10 months of age showed a good surgical outcome (Fig. 2). At 4 years of age, the patient was asymptomatic and thriving well with oral feeds.

Discussion

Here, we report a patient that presented with non-specific respiratory symptoms tachypnea and coughing with feeding. In a multidisciplinary team meeting, different investigations were discussed. It was decided, that for this patient, the advantage of a better visualization of lung tissue and esophagus, outweighed the disadvantage of anesthesia as well

as extended radiation and CBFM as underlying condition was diagnosed. Successful surgery resulted in an excellent long-term outcome.

CBPFM is a rare congenital anomaly, that is defined by a patent congenital communication between the esophagus and the respiratory tract (5). The malformation is often labeled *esophageal bronchus* or *esophageal lung*; the latter if the main bronchus originates from the esophagus (6). No data on incidence or prevalence rates have been published, yet. The embryogenesis of CBPFM is not fully understood, but a focal mesodermal defect is suspected, making CBPFM and esophageal atresia variations of the same spectrum of malformations (5,7). CBPFM is reported to be more common in females and usually occurs unilaterally, while it is occasionally associated with other malformations, including cardiovascular anomalies, VACTERL association, skeletal malformation, anorectal malformation, or diaphragmatic hernia (6,8).

In accordance with earlier reports, the initial presentation of the patient described here was non-specific, and the diagnosis challenging. A recently published systematic review analyzed the clinical characteristics of CBPFM (5). Most children presented with respiratory symptoms after birth including respiratory distress, cough/ choking following food intake, recurrent respiratory infection, or hemoptysis. In almost two thirds of the cases, patients were diagnosed by upper gastrointestinal series. Initial misdiagnosis was common, while the reported mortality rate was high (13.1%).

In 1992, a classification system for CBPFM has been introduced: 1) combination with esophageal atresia and tracheoesophageal fistula, 2) absence of a main stem bronchus arising from the trachea as well as the total sequestered lung communicating with the lower esophagus, 3) communication of an isolated part of the lung, and 4) communication between a normal bronchial system and the esophagus (6). Of note, when CBPFM is combined with esophageal atresia, the bronchial malformation might be missed preoperatively, as no upper gastrointestinal series can be performed (16, 17).

There is the potential of compensatory alveolar growth following surgical resection of congenital thoracic malformations (CTM) performed in infancy that is believed to decrease with

age (9-13). However, data on long-term pulmonary outcome following the resection of CTMs are scarce and there is an uncertainty of the most appropriate outcome measurement (14). Most commonly used for the assessment of long-term pulmonary outcome is pulmonary function testing (PFT) (14) with the known limitations being age- and cooperation depending, reference values being based on testing results from healthy individuals that vary according to age, height, sex, and ethnicity, as well as the variability of individual measurements (15). Moreover, contradictory results of PFT results following the surgical resection of CTMs have been published (14) and there is a need for further studies investigating long-term outcome of patients with CTM after surgery.

Conclusions

CBPFM are rare malformations. This case highlights the clinical challenge of diagnosing this rare condition. It aims to raise awareness and improve diagnostic accuracy. For optimal management a multidisciplinary approach is essential.

Figures legends

Figure 1

Chest X-ray (A) showing consolidations in the upper two thirds of the right lung and behind the heart on the left side. Computed tomography (B and C) suggests atresia of the right upper and middle lobe bronchus, probably arising from a common bud. However, on the left side, all bronchi with the exception of the superior and the posterior-basal left lower lobe bronchi are patent. The course of the latter is suspicious of an esophageal bronchus (D, arrow). Upper gastrointestinal series shows esophageal insertion of a common right upper and middle lobe bronchus, as well as a common bronchus of segments VI and X on the left side (E and F).

Figure 2

Chest X-ray at 10 months of age showing a good surgical result.

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FIGURE 1

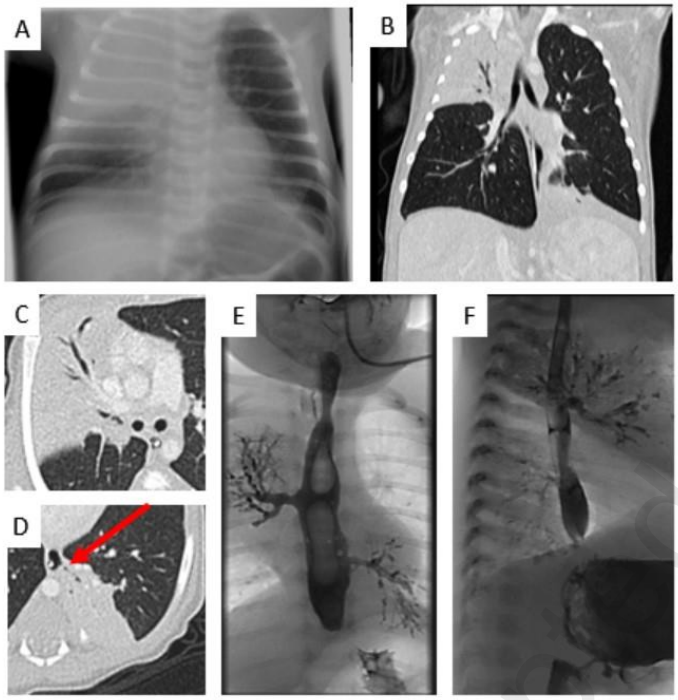

FIGURE 2