CASE REPORT

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

Philipp **Peters** [1], Elias **Seidl** [2,*], Jochen **Hubertus** [3], Dietrich **von Schweinitz** [4], Matthias **Griese** [1], Birgit **Kammer** [5], Karl **Reiter** [1]

*** Correspondence to:**

Elias.Seidl@ksipi.uzh.ch. ORCID: https://orcid.org/0000-0001-6610-3756

Doi

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[1] Department of Pediatrics, Dr. von Hauner Children's Hospital, University Hospital, LMU Munich, Munich, Germany

[2] Department of Respiratory Medicine, University Children's Hospital Zurich, Zurich, Switzerland

[3] Department of Pediatric Surgery, Marien Hospital Witten, Ruhr-University Bochum, Witten, Germany

[4] Department of Pediatric Surgery, Dr. von Hauner Children's Hospital, University Hospital, LMU Munich, Munich, Germany

[5] Department of Radiology, University Hospital, LMU Munich, Munich, Germany

ABSTRACT

Infants frequently present with respiratory symptoms, but diagnosing the underlying pulmonary condition is sometimes challenging.

A 2-week-old neonate was referred to a postnatal history of tachypnea, poor feeding and elevated plasma inflammation markers. The patient was 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 on 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.

IMPACT STATEMENT: This manuscript presents a complex case of a neonate referring to a postnatal history of tachypnea, poor feeding, and elevated plasma inflammatory markers. While initial diagnostics suggested bronchial atresia, further evaluation led to the diagnosis of a communicating bronchopulmonary foregut malformation. This case highlights the diagnostic challenges associated with rare neonatal conditions, emphasizing the need for increased awareness and improved diagnostic accuracy. Optimal management requires a multidisciplinary approach to ensure comprehensive care.

KEY WORDS

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

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 (C-reactive protein and erythrocyte sedimentation rate). 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 clinical symptoms persisted, bronchoscopy as well as high-resolution computed tomography (HR-CT) was 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 (**Figures 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 upper gastrointestinal series. The investigation revealed a bilateral esophageal insertion of bronchi (**Figures 1E, F**) and the diagnosis of a communicating bronchopulmonary foregut malformation (CBPFM) was made. A 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 (**Figure 2**). At 4 years of age, the patient was asymptomatic and thriving well with oral feeds.

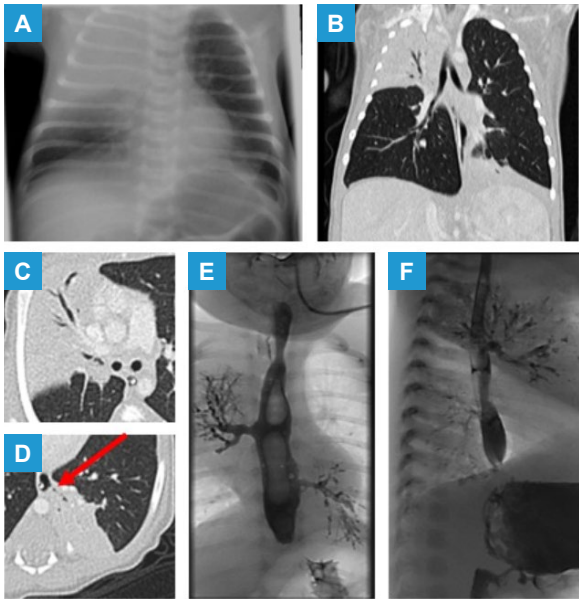

***Figure 1**. Chest X-ray (**A**) showing consolidations in the upper two thirds of the right lung and behind the heart on the left side. Computed tomography (**B** and **C**) suggests atresia of the right upper and middle lobe bronchus, probably arising from a common bud. However, on the left side, all bronchi except for 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). The 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**).*

DISCUSSION

Here, we report a patient who presented with non-specific respiratory symptoms, tachypnea and coughing during 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 disadvan-

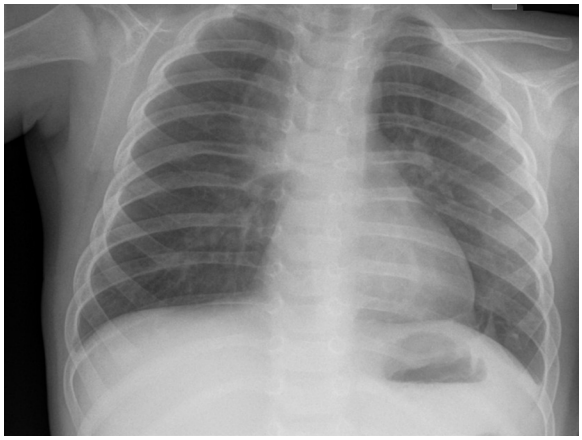

***Figure 2**. Chest X-ray at 10 months of age showing a good surgical result.*

tage 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 (6). No data on incidence or prevalence rates has 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 with 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 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.

COMPLIANCE WITH ETHICAL STANDARDS

Conflict of interests

The Authors have declared no conflict of interests.

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Author contributions

PP, ES, and KR developed the theoretical framework and performed the data interpretation. PP, ES, JH, DvS, BK, MG and KR were primary care physicians. KR supervised the project. ES took the lead in writing the manuscript. PP, JH, DvS, BK, MG and KR contributed to the manuscript providing critical feedback.

Ethical approval

Human studies and subjects

This case report was determined not to require Ethics Committee review.

Data sharing and data accessibility

Data are available upon motivated request to the Corresponding Author.

Publication ethics

Plagiarism

Authors declare no potentially overlapping publications with the content of this manuscript and all original studies are cited as appropriate.

Data falsification and fabrication

All the data corresponds to the real.

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