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Left interval thoracoscopic pneumonectomy for type II communicating bronchopulmonary foregut malformation in a 17-month-old girl

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ABSTRACT

INTRODUCTION: Communicating bronchopulmonary foregut malformation (CBPFM) is a rare congenital anomaly comprising a spectrum of airway anomalies connected to the oesophagus or stomach. Our management of a case of CBPFM is presented to improve knowledge of its treatment.

PRESENTATION OF CASE: A 17-month-old Japanese girl presented with fever (39.1 °C) and persistent cough. She was noted to be poorly developed (7.5 kg: −2SD). Chest X-ray radiography was suggestive of pneumonia involving the left lung; WBC was 41600/μL and CRP was 12.9 mg/dL. Computed tomography, upper gastrointestinal series, and bronchoscopy identified a fistula between the oesophagus and the left lung and severe left pulmonary artery hypoplasia, typical of type II CBPFM. Her left thoracic cavity was small with sclerotic lung tissue. We customised management by commencing a trial of intensive duodenal tube feeding without oral ingestion. She gained weight and her pneumonia improved enough to enable thoracoscopic left pneumonectomy through an unconventional dorsal approach, necessary because of the location of her CBPFM. Surgery was successful and tolerated well with unremarkable postoperative recovery. Currently she is asymptomatic and growing well.

DISCUSSION: We used minimally invasive surgery (MIS) to resect an anomalous bronchus and perform a left pneumonectomy after two weeks of intensive conservative management, including nil-by-mouth to optimise her condition for surgery.

CONCLUSION: Our case provides further evidence that CBPFM can be treated successfully by MIS (interval thoracoscopic pneumonectomy), especially after a period of intensive conservative management. Interval surgery should be considered actively prior to major surgery in smaller children if indicated.

1. Introduction

Communicating bronchopulmonary foregut malformation (CBPFM) is a rare congenital anomaly characterised by a communication between an isolated portion of respiratory tissue connected to the oesophagus or stomach [1]. Reports in the literature are sporadic and describe a spectrum of symptoms ranging from acute respiratory distress at birth, to chronic cough, recurrent pneumonia, and failure to thrive, with high risk for misdiagnosis if not investigated thoroughly.

Because of the communication between the lung and foregut, CBPFM must originate during early embryologic development when the tracheobronchial tree and oesophagus are closely associated. CBPFM can be classified into 4 main types [3], and our case is an example of type II, characterised by the absence of a main stem bronchus arising from the trachea and the presence of an underdeveloped lung with a mass in the unilateral chest cavity that originates from the lower segment of the oesophagus, which has also been called an “oesophageal lung”, first described in 1960 [4], with less than 30 cases described in the English literature [1].

Treatment for this condition has traditionally been resection of the anomalous pulmonary segment through an open approach, but as surgeons have become more experienced with minimally invasive surgery (MIS), thoracoscopic pneumonectomy is now a valid treatment option for CBPFM [5]. To date, two reports describing thoracoscopic surgery for CBPFM have been published; one is the case of a 23-month-old boy who had division of an anomalous bronchus and right upper lobectomy [6], and the other is the case of a 4-month-old boy who had division of an anomalous bronchus and left pneumonectomy [7]. In both cases, thoracoscopic

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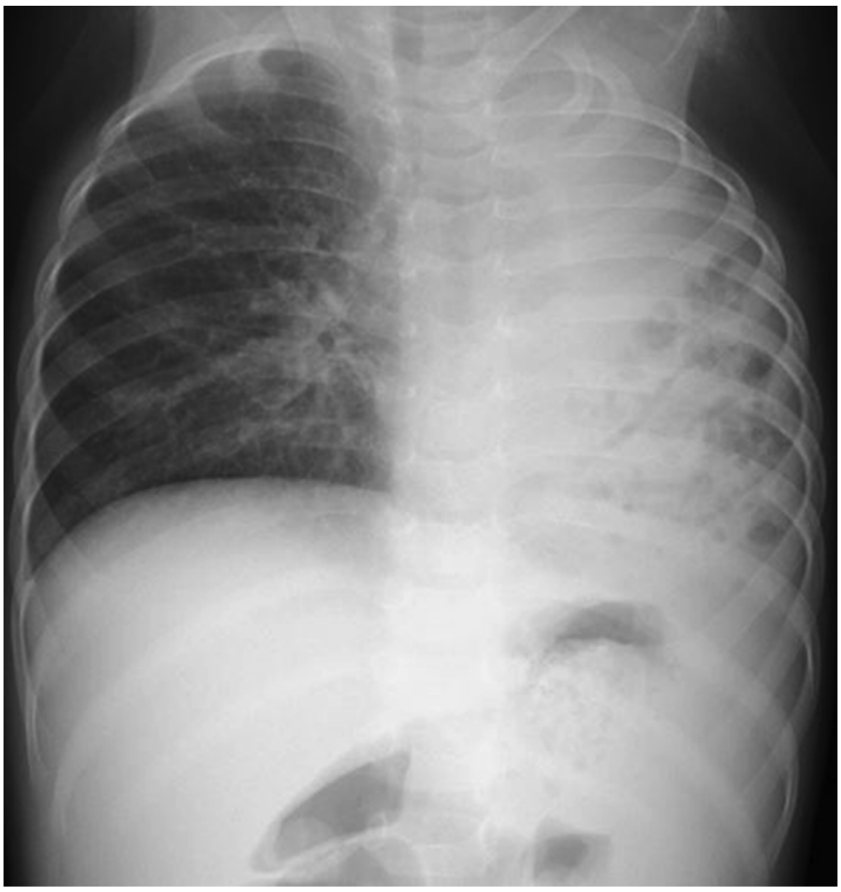

Fig. 1. Preoperative chest x-ray radiograph. Diffuse infiltration of the left lung.

pneumonectomy was performed as an emergency procedure with trocars positioned conventionally between the anterior and posterior axillary lines.

We present our original management of a case of type II CBPFM treated successfully by interval/elective thoracoscopic fistula transection and left pneumonectomy after intensive conservative management using an unorthodox dorsal approach.

2. Case report

A 17-month-old Japanese girl presented with a four-day history of fever (39.1 °C) and persistent cough; WBC (41600/μL) and CRP (12.9 mg/dL) suggested severe inflammation, and a chest x-ray (CXR) revealed a diffuse infiltration of the left lung (Fig. 1) consistent with a diagnosis of pneumonia. On presentation, she weighed 7.5 kg which was −2SD for her age according to standard Japanese height/weight charts for girls.

Our case had no drug history and no relevant family history. Her mother stated that she had never experienced any typical symptoms of pneumonia, such as a productive/congested cough or high fever prior to presentation, although her physical growth and development was poor. No patient perspective was possible given the age of our case.

Investigations were ordered to obtain more information about the pneumonia suspected from CXR on presentation. Computed tomography (CT) demonstrated a poorly aerated abnormal left lung with suspicion of a bronchopulmonary fistula (Fig. 2). An upper gastrointestinal series identified a fistula between the oesophagus and the left lung that was considered most likely be our case's left main bronchus embryologically (Fig. 2). On bronchoscopy, the trachea, right main stem bronchus, and right distal airways were normal; however, there was no evidence of bifurcation of the trachea and no left main stem bronchus (Fig. 3). Because of severe coexisting left pulmonary artery hypoplasia, there was no indication for preserving the left lung, and pneumonectomy was planned (Fig. 2).

Although clinically stable, our case's left thoracic cavity was small and thoracoscopy was contraindicated. However, she was also considered too frail to tolerate a thoracotomy, so a trial of nil-by-mouth with intensive duodenal tube feeding was commenced. After two weeks, CT showed improvement in her pneumonia (Fig. 4), WBC was 9300/μL, and CRP was 0.3 mg/dL, and thoracoscopic surgery was considered feasible. An unconventional dorsal approach was planned because the CBPFM fistula between the oesophagus and lung had displaced the left lung dorsally.

After induction of endotracheal general anesthesia, the patient was placed in the right lateral decubitus position with the left side at 30 degrees. A thoracoscopic left pneumonectomy was performed by a team comprised of a board-certified paediatric surgeon with 17 years' experience of MIS (including thoracoscopic surgery) and a board-certified general thoracic surgeon with 8 years' experience of thoracoscopic surgery in adults who was present for supervisory support.

A 5-mm trocar was placed in the 7th intercostal space at the border of the erector spinae muscle. The pleural cavity was insufflated to 4–8 mmHg, and a 5-mm 30-degree scope was introduced into the pleural space. Three additional 5-mm trocars were introduced; one in the 9th intercostal space at the border of the erector spinae, one in the 10th intercostal space posterior to the posterior axillary line and one in the 6th intercostal space posterior to the posterior axillary line. All trocars were placed between the border of the erector spinae and the posterior axillary line (Fig. 5). The

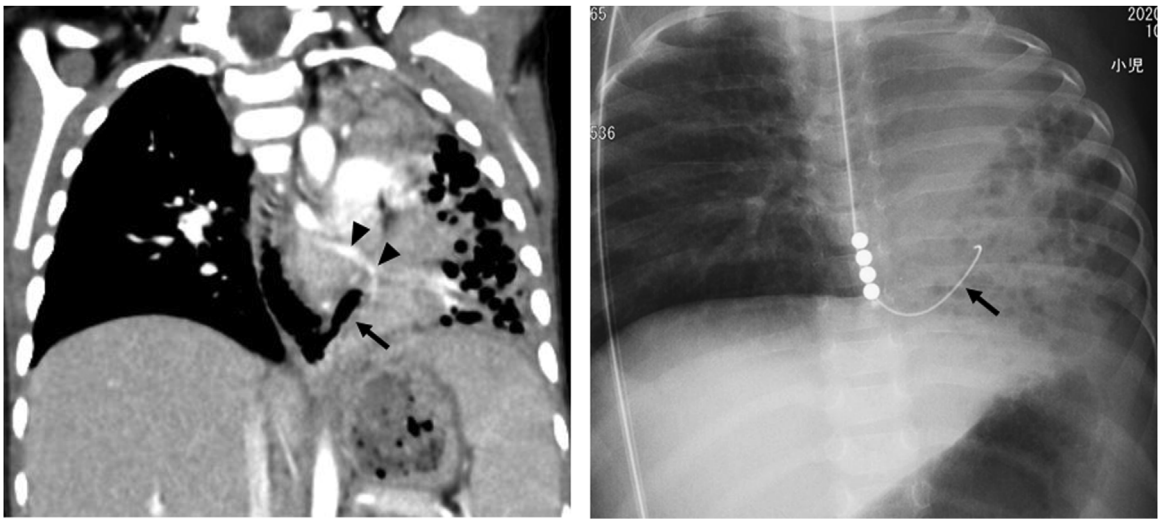

Fig. 2. Preoperative computerized tomography (lt) and upper gastrointestinal series (rt).
Poorly aerated abnormal left lung with the fistula between the esophagus and the lung (arrow), and atrophic/hypoplastic left pulmonary artery (arrowhead).
A guidewire has been passed through the fistula, from the esophagus to the left lung.

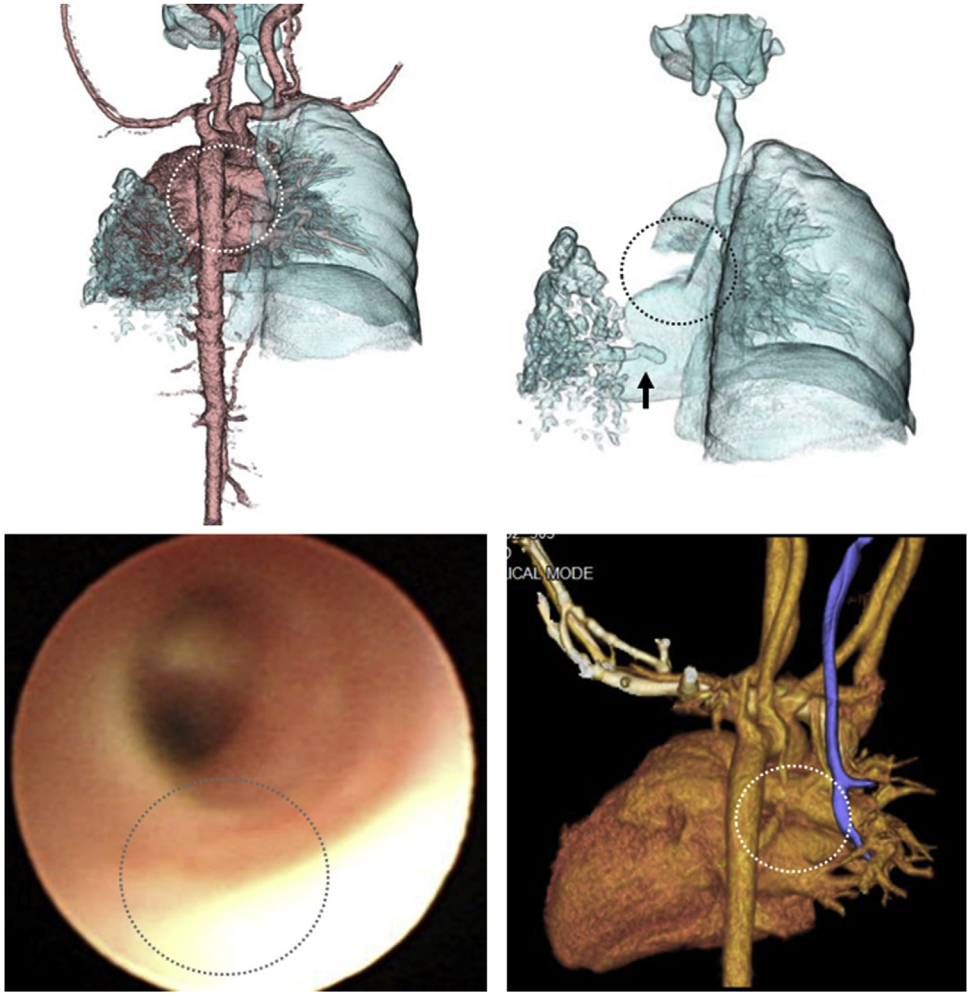

Fig. 3. Bronchoscopy and 3-D image from the computerized tomography.
Normal trachea, normal right main stem bronchus, and normal right distal airways, but no bifurcation of the left main stem bronchus (dotted circle). Arrow: Fistula.

inferior pulmonary ligament was taken down using monopolar hook cautery and bipolar forceps. The oesophagus was identified and dissected free from the pleura, and the oesophageal fistula was identified and dissected circumferentially (Fig. 6). The trocar in the 10th intercostal space was removed and a surgical stapling device (ENDOPATH® ETS stapler, Ethicon Endo-surgery; Johnson

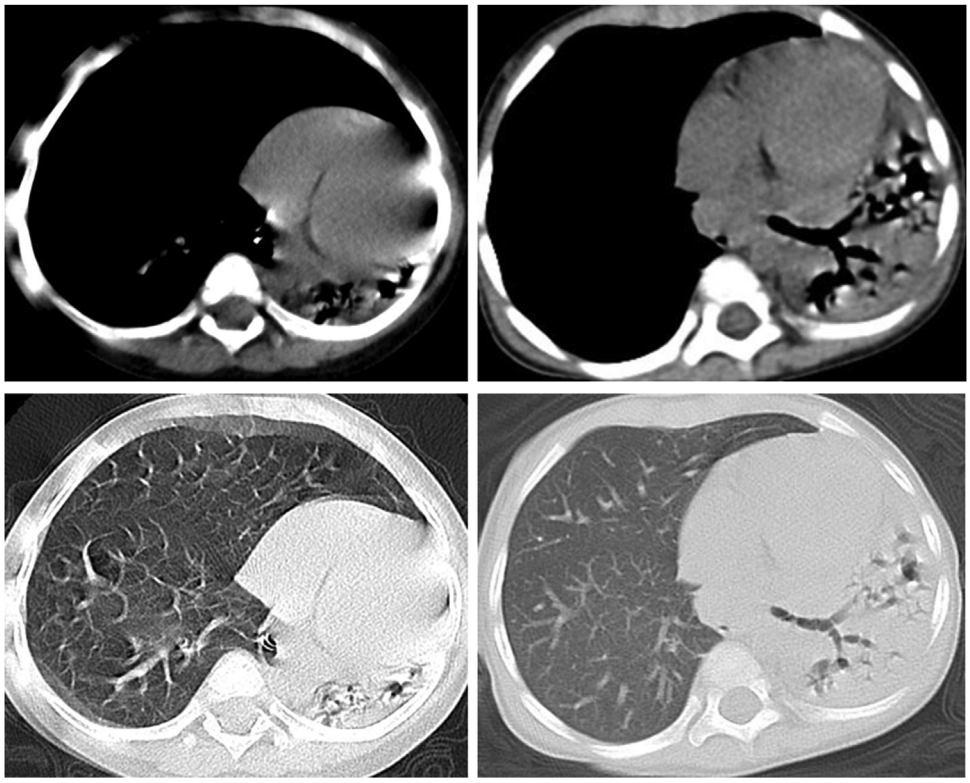

Fig. 4. Computed tomography pre and post conservative management.
Shrunken left lung after conservative management (lt) compared with its initial appearance (rt).

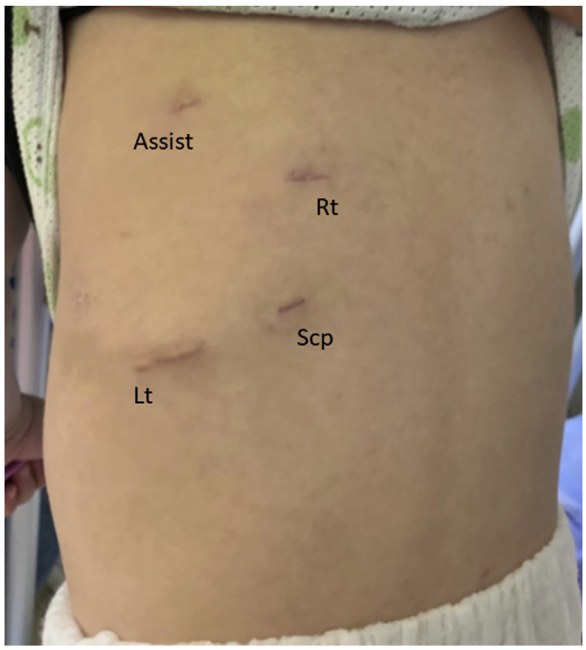

Fig. 5. Trocar positions for thoracoscopy.
All 4 trocars (5-mm) have been placed between the border of the erector spinae muscle and the posterior axillary line. Rt: Trocar in the 7th intercostal space for the surgeon's right hand. Scp: Trocar in the 9th intercostal space for the scope. Lt: Trocar in the 10th intercostal space for the surgeon's left hand. Assist: Trocar in the 6th intercostal space for the assistant. Note: The incision for the Lt trocar has been partially extended to remove the excised lung.

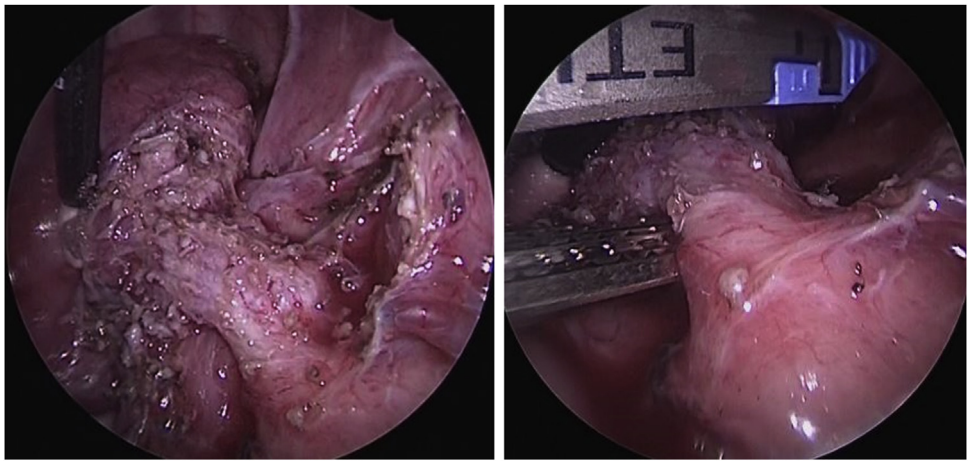

Fig. 6. Fistula between the esophagus and left lung (intraoperative findings).
The fistula was identified and dissected circumferentially (lt).
The fistula was clamped and transected with a stapling device (rt).

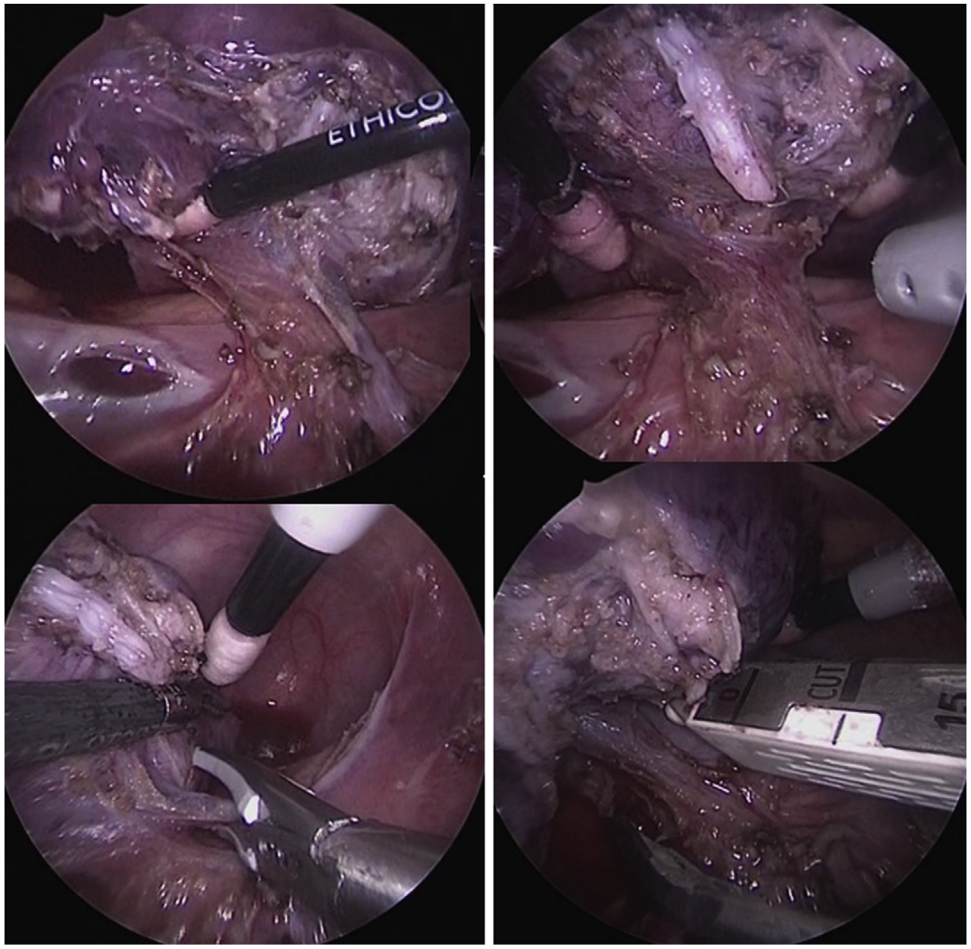

Fig. 7. Pulmonary vein and artery (intraoperative findings).
The inferior pulmonary vein was dissected free then transected (lt).
The pulmonary artery was clamped and transected with a stapling device (rt).

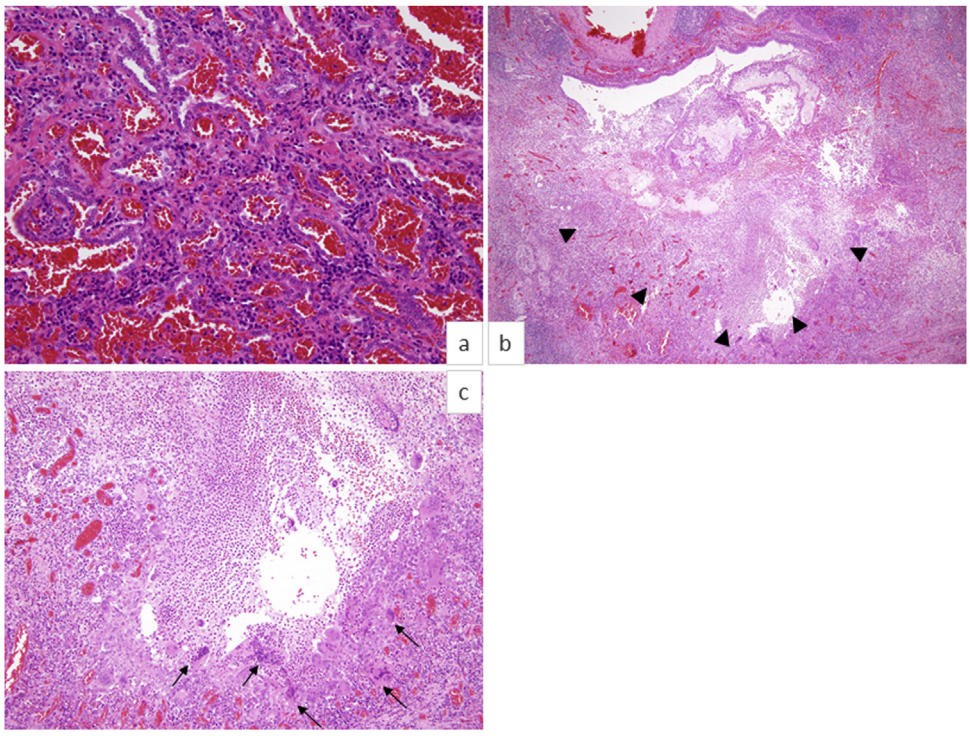

Fig. 8. Histopathology of Stoker type-3 congenital pulmonary airway malformation.
High power magnification showed cuboidal epithelium lining spaces (a). Histopathology of necrotizing acute bronchopneumonia (aspiration pneumonia). Low power magnification showing severe neutrophils accumulation and necrosis in bronchus and alveolar spaces (b: arrowheads). High power magnification showing acute inflammation surrounding foreign body giant cells (c: arrows).

& Johnson, Cincinnati, OH, USA) was introduced to transect the fistula directly (Fig. 6). There was no superior pulmonary vein present at the pulmonary hilum. A 3–4 mm inferior pulmonary vein was identified and dissected free circumferentially, then transected with a vessel sealing device (LigaSure™, Covidien, Mansfield, MA, USA) (Fig. 7). The pulmonary artery was identified and dissected circumferentially, noted to be 7–8 mm in size, clamped and transected with a different surgical stapling device (Echelon FLEX™ stapler, Johnson & Johnson, Cincinnati, OH, USA) (Fig. 7). The atrophic lung was placed in a 10 mm specimen retrieval bag and withdrawn through the 10th intercostal trocar site after partially extending the incision. No additional procedures were considered. The pleural cavity was irrigated, there were no signs of bleeding, and a chest tube was placed. The total operative time was 3 h. Histopathology showed Stoker type-3 congenital pulmonary airway malformation (Fig. 8).

The patient tolerated surgery well, and an oesophagogram performed on postoperative day 5 showed no evidence of leakage (Fig. 9). Oral intake was commenced and once a full normal diet was tolerated, she was discharged on postoperative day 7; total inpatient stay was 34 days. At her last outpatient clinic visit one month after surgery, she was well without any evidence of respiratory distress and had made satisfactory weight gain.

The work has been reported in line with the Scare 2018 criteria [2].

3. Discussion

Patients with CBPFM typically present with recurrent pneumonia and asphyxia, and the treatment of choice is dividing the fistula and resecting the anomalous lung tissue. Our patient presented with symptoms early in life, which is typical for CBPFM [8]. Symptoms of CBPFM may be nonspecific, but a persistent cough in a neonate should be investigated thoroughly to prevent misdiagnosis. Our case had no notable symptoms other than failure to thrive. Any child with failure to thrive and a chronic cough should have a CXR.

Although less than 30 cases of type II CBPFM have been reported in the English literature and no standardised guidelines exist for its management. Thoracotomy repairs appear to do well overall but there is little consistency because each case is unique. Thus, outcomes, re-hospitalisation for the management of complications and likely complications can vary. Although thoracoscopy has been used to treat two cases, it is contraindicated if there is insufficient pleural space. Anatomically, the fistula/bronchus originating from the oesophagus was located dorsally, so our approach also had to be dorsal/posterior. To achieve the best possible operative view for pneumonectomy in our case of CBPFM, fistula resection was performed first, followed by preparation of the pulmonary hilum.

4. Conclusion

We used interval thoracoscopic MIS to resect an anomalous bronchus and perform a left pneumonectomy in a 17-monnth-old girl with CBPFM, through an unconventional dorsal/posterior approach after two weeks of intensive conservative management to improve her clinical status after a history of pneumonia and failure to thrive. Our experience would indicate that thoracoscopy is not contraindicated in smaller patients if trocars can be inserted safely.

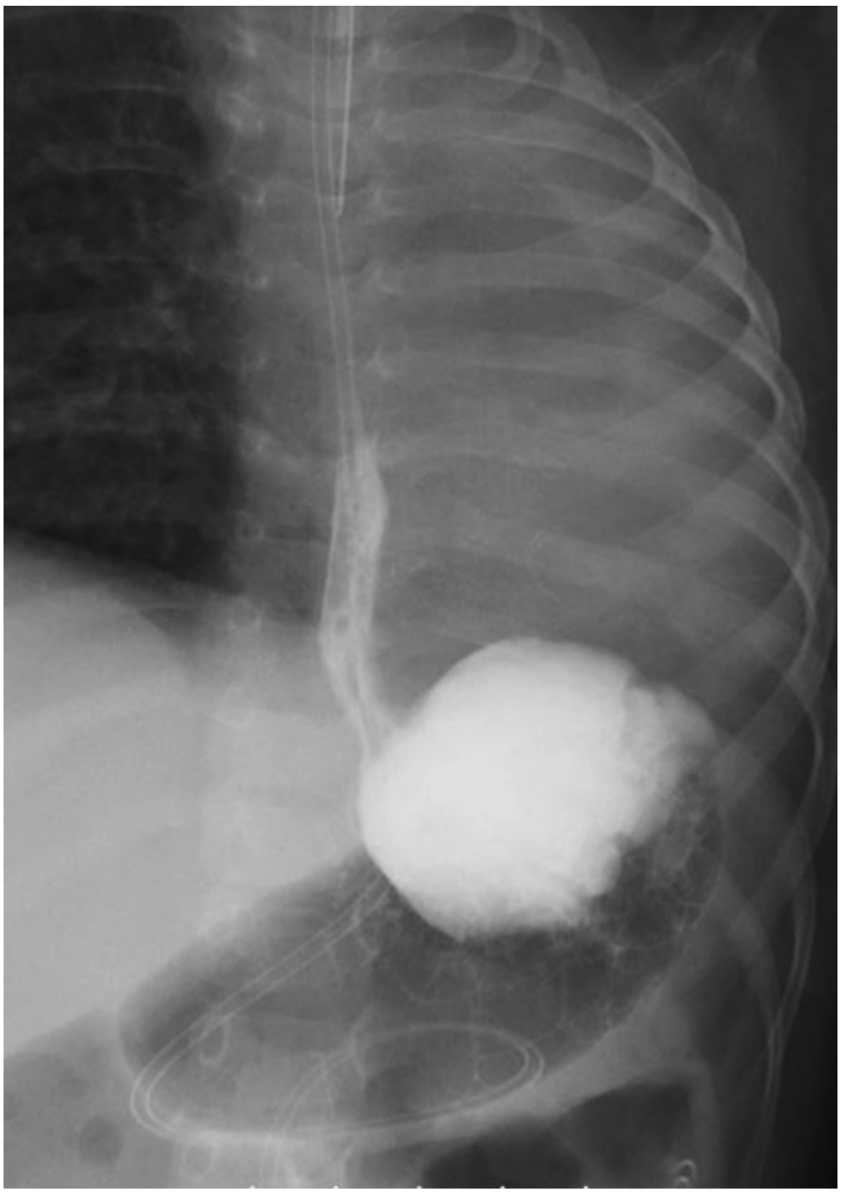

Fig. 9. Postoperative esophagogram.

Postoperative esophagogram showing no signs of leakage (rt).

Declaration of Competing Interest

The authors report no declarations of interest.

Sources of funding

There are no sources of funding for our research.

Ethical approval

12560506 (2020.02.21).

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Author contribution

Go Miyano: study concept and design, writing the paper.
Yukio Watanabe: data collection.
Takuo Hayashi: data analysis.
Geoffrey J. Lane: English review and revision of manuscript contents.
Kenji Suzuki: data interpretation.
Atsuyuki Yamataka: data interpretation.

Registration of research studies

1. Name of the registry: Go Miyano.
2. Unique identifying number or registration ID: 6150.
3. Hyperlink to your specific registration (must be publicly accessible and will be checked): This is a single case report, written informed consent was obtained from the patient for publication of this case report and accompanying images.

Guarantor

Atsuyuki Yamataka.

Provenance and peer review

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