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Case report

A case of congenital bronchial atresia with tracheobronchial stenosis caused by emphysema: Successful management with thoracoscopic surgery

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ABSTRACT

Introduction: Congenital bronchial atresia (CBA), as a rare developmental abnormality of the lung, is usually asymptomatic and is accidently discovered in most cases. Currently, no standardized guidelines for the treatment or management of CBA have been established.

Case presentation: A 22-year-old male soldier was referred to Shanghai Changhai Hospital, The First Affiliated Hospital of Naval Medical University due to chest tightness and shortness of breath after repeated strenuous activities. Contrast-enhanced computed tomography (CT) revealed an 18mm × 11mm solitary, well-circumscribed, and solid nodule with no enhancement in the right upper lobe (RUL), and emphysematous changes distributed throughout the RUL. A flexible bronchoscopic examination showed extrinsic compression stenosis in the bronchial opening of the right middle lobe (RML). After three-dimensional (3D) reconstruction CT and a multidisciplinary consultation, a diagnosis of CBA in the anterior segment (B3) of RUL was established. Subsequently, thoracoscopic right upper lobectomy was performed and resulted in an improved respiratory capacity 6 months after surgery. To date, the patient has good quality of life without any complication.

Conclusion: This study underscores the role of bronchoscopy, 3D reconstruction CT, and a multidisciplinary consultation in the diagnosis of CBA, and highlights that a thoracoscopic intervention should be considered in such case.

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1. Introduction

Congenital bronchial atresia (CBA) is a rare developmental malformation of the lung that was first reported by Ramsay et al. [1] in 1953. It has been hypothesized that CBA occurs after the sixteenth week of gestation as a result of intrauterine ischaemia. It is estimated that the prevalence of this disorder was 1.2 cases in 100,000 males [2]. Some authors have reported that the condition is more common in male than in female cases [3,4]. CBA is most often diagnosed in the neonatal period or in childhood, which is why the majority of studies reported are related to pediatrics [4,5]. Typically, this benign disease is discovered incidentally since about 2/3 of the CBA patients are asymptomatic. For symptomatic patients, the most commonly reported clinical manifestations are pneumonia, fever, chronic cough, pneumothorax, respiratory distress [5], and repeated lung infections [6], among which bacterium are the most common, and others include *Mycobacterium avium*, Aspergillus fumigatus and cytomegalovirus [6–8].

Given the rarity of CBA, bulk of data in related limited research are composed of case reports and small series with absence of systematic management, especially for the indications of operation. Currently, there are no current standardized guidelines for the treatment or management of CBA, and a consensus on asymptomatic cases has hitherto been undocumented. Therefore, the addition of case reports of this disorder would be extremely valuable if they provide insights into potential diagnosis and treatment options. Here, we present a unique case of CBA with tracheobronchial stenosis caused by emphysema who underwent thoracoscopic right upper lobectomy.

2. Case presentation

A 22-year-old male soldier with no remarkable medical history was referred to our hospital because of chest tightness and shortness of breath after repeated strenuous activities in October 2022. He was a lifetime non-smoker and had never undergone chest computed tomography (CT) scan before visiting Shanghai Changhai Hospital, The First Affiliated Hospital of Naval Medical University. On admission, his body temperature, blood pressure, heart rate, respiratory rate, and arterial oxygen saturation measured via pulse oximetry were 36.0 °C, 120/70 mmHg, 80 beats per minute, 12 breaths per minute, and 98 %, respectively. Furthermore, no significant finding was observed in the thorough physical examinations and laboratory examinations including 1,3-β-D-Glucan test, Galactomannan test, tuberculosis infection T cell.

The pulmonary function test showed a moderate reduction of respiratory capacity in which forced expiratory volume in 1 second (FEV1), forced vital capacity (FVC), and FEV1/FVC ratio were 1.91 L (46 % of the predicted value), 2.18 L (56 % of the predicted

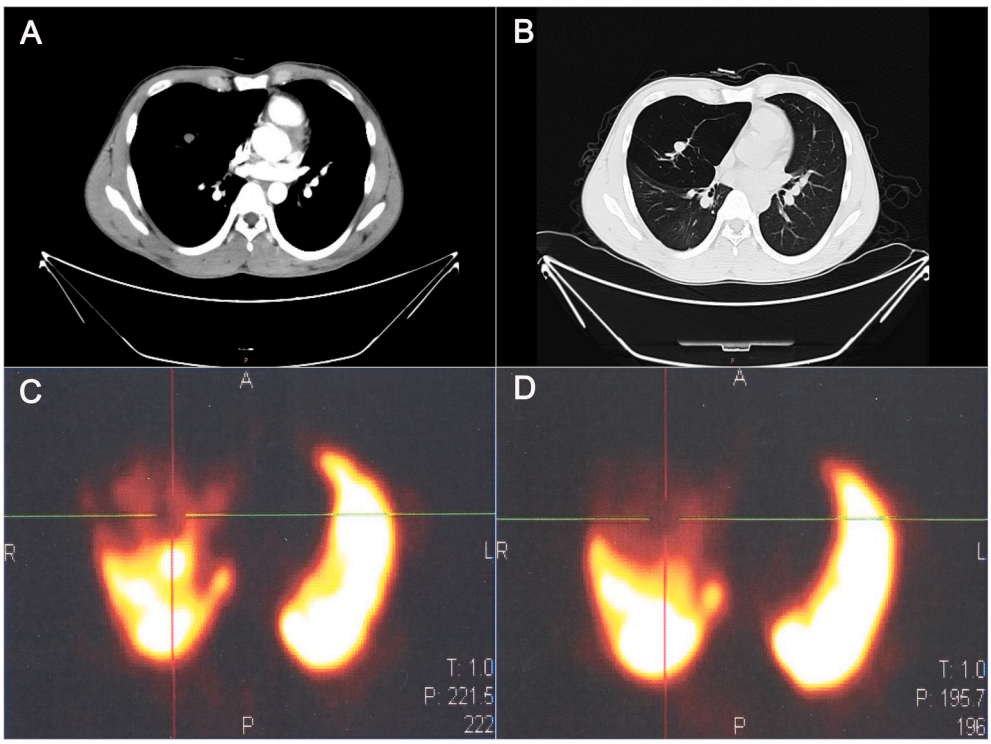

Fig. 1. Contrast-enhanced CT performed on admission revealing (A) an 18mm × 11mm solitary, well-circumscribed, and solid nodule with no enhancement in the right upper lobe (RUL), and (B) emphysematous changes (e.g. increased transparency and decreased broncho-vascular shadow) distributed throughout the RUL. Lung ventilation (C) and perfusion (D) scan indicating sparse/defective radioactive distribution area in the anterior segment of RUL.

value), and 71.3 %, respectively. Initial workup with contrast-enhanced chest CT revealed an 18mm × 11mm solitary, well-circumscribed, and solid nodule with no enhancement in the right upper lobe (RUL) (Fig. 1A), and emphysematous changes (e.g. increased transparency and decreased broncho-vascular shadow) distributed throughout the RUL (Fig. 1B), which were further substantiated by lung ventilation/perfusion scan indicating sparse/defective radioactive distribution area in the anterior segment of RUL (Fig. 1C and D). Additionally, a flexible bronchoscopic examination demonstrated normal structure of bronchus in the RUL (Fig. 2A) and extrinsic compression stenosis in the bronchial opening of the right middle lobe (RML) (Fig. 2B). Subsequently, three-dimensional (3D) reconstruction of the bronchial tree based on the contrast-enhanced chest CT was conducted and indicated absence of anterior segmental bronchus (B3) of his RUL (Fig. 3A). Also, post-processing of chest CT data for quantification was performed using 3D reconstruction, which showed that the ratio of RUL emphysema to right whole lung was 40 % (Fig. 3B).

Consequently, CBA in the anterior segment of RUL was suspected based on imaging findings. Given the rarity and complexity of CBA, a multidisciplinary consultation involving pulmonologist, anesthesiologist, thoracic surgeon, radiologist and nuclear medicine specialist was held, suggesting a diagnosis of CBA in the anterior segment of RUL and further surgery for this patient based on the following reasons: the bronchial opening of RML had presented with extrinsic compression stenosis and the mediastinum had shifted to the left due to the space-occupying effect of emphysema. Moreover, the patient works on the naval vessel with limited space, poor air circulation, inferior medical condition, and strong physical labor, which may lead to pneumothorax as a result of the progressive aggravation of RUL emphysema, and the recurrence of lung infection. Therefore, the patient underwent thoracoscopic right upper lobectomy on December 7, 2022 (Fig. 4A and B).

The blind-end of B3 with associated dilated distal segmental bronchi filled with mucus encased by hyperinflated lung parenchyma was identified in the specimen (Fig. 4C). Histopathologically, there was no proximal or central tracheal communication of B3, whereas distal to B3 there was a dilated mucus-filled bronchus (Fig. 4D). Furthermore, emphysematous changes in the distal side of the mucinous impaction were observed (Fig. 4E). These findings were compatible with B3 bronchial atresia. A follow-up pulmonary function test done 6 months after surgery showed an improved respiratory capacity in which FEV1, FVC, and FEV1/FVC ratio were 2.16 L (53 % of the predicted value), 2.28 L (59.9 % of the predicted value), and 82 %, respectively. Up to May 2024, the patient had good quality of life with smooth completion of daily work and training and did not develop any complication.

3. Discussion

Congenital bronchial atresia (CBA), as a rare separate entity of congenital lung malformations arising during the formation of bronchi, is typically characterized by segmental or lobar emphysema associated with the presence of mucoid impactions and/or a bronchocele and hyperinflated lung parenchyma of the involved lung segment [5,9,10]. This abnormality is usually observed in either the segmental or the subsegmental levels, and is rarely observed in the lobar level. Of note, CBA in the apicoposterior segmental bronchus of the left upper lobe seems to be most common [11]; however, the patient in the present study developed CBA in the anterior segment of RUL.

Chest CT scan is regarded as the diagnosis of choice for CBA, and the characteristic chest CT findings of CBA are mucocele and hyperinflation of the lung parenchyma [3,12]. However, similar findings are also reported in other serious disorders, such as lung cancer or bronchial adenoma [3,13,14]. It is well recognized that bronchoscopy has changed the course of pulmonary medicine and plays a pivotal role in the management of a variety of lung diseases [15–17]. Some publications have demonstrated that identification of the blind-ending bronchus via bronchoscopy is also helpful for CBA diagnosis [8,18]. Nevertheless, only 50 % of patients with CBA could be diagnosed by bronchoscopy, according to a retrospective study involving 12 CBA cases from Wang et al. [3], which could be attributed to the fact that the disrupted site was located too peripherally [8]. In the present case, we could not visualize and detect the blind end via bronchoscopy. Hopefully, confocal laser endomicroscopy that enables higher resolution and real-time microscopic analysis of tissue architecture has been demonstrated to play a role in multiple pulmonary diseases and has the potential to improve the

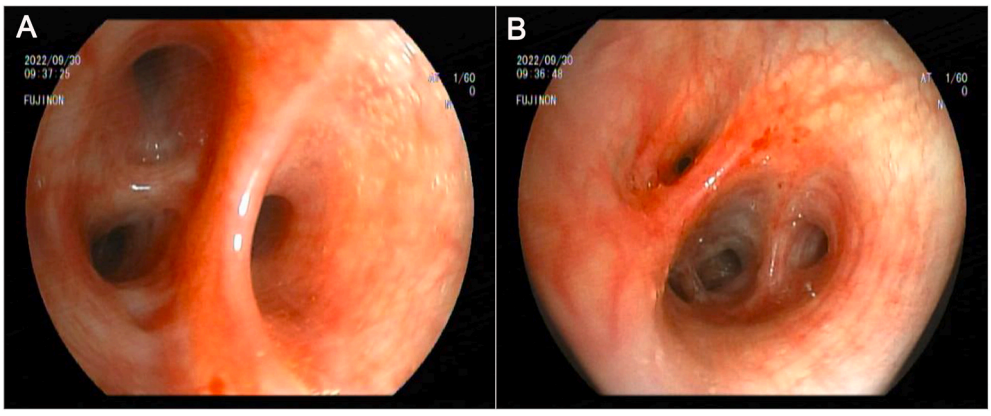

Fig. 2. A flexible bronchoscopic examination demonstrating (A) normal structure of bronchus in the right upper lobe, and (B) extrinsic compression stenosis in the bronchial opening of the right middle lobe.

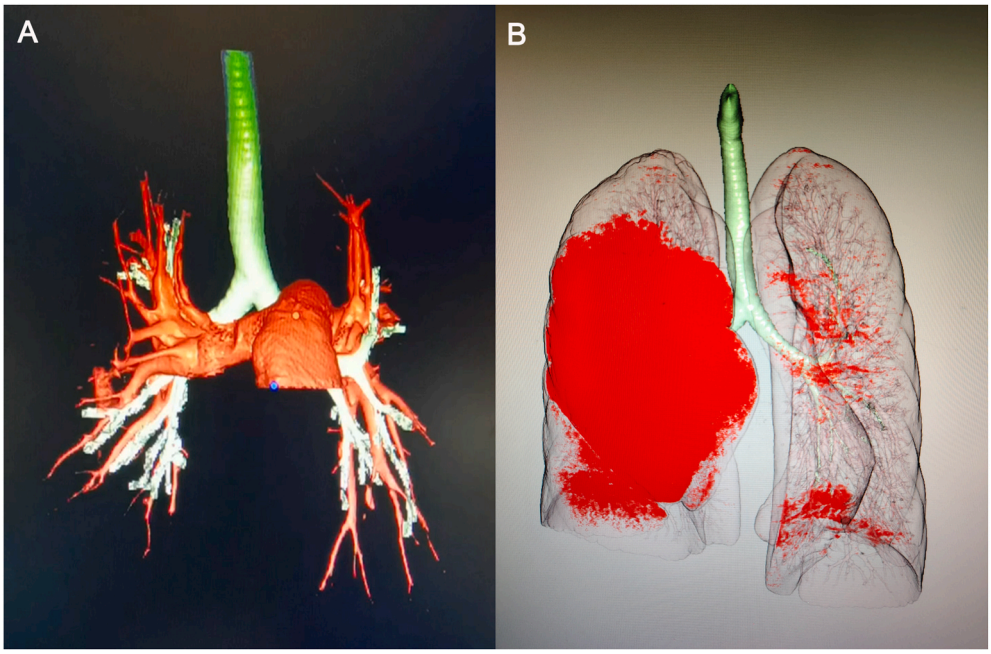

Fig. 3. 3D reconstruction CT showing (A) atresia of the B3 bronchial trunk of the right upper lobe (RUL) without communication with the right bronchial tree, and (B) the ratio (40 %) of RUL emphysema to right whole lung.

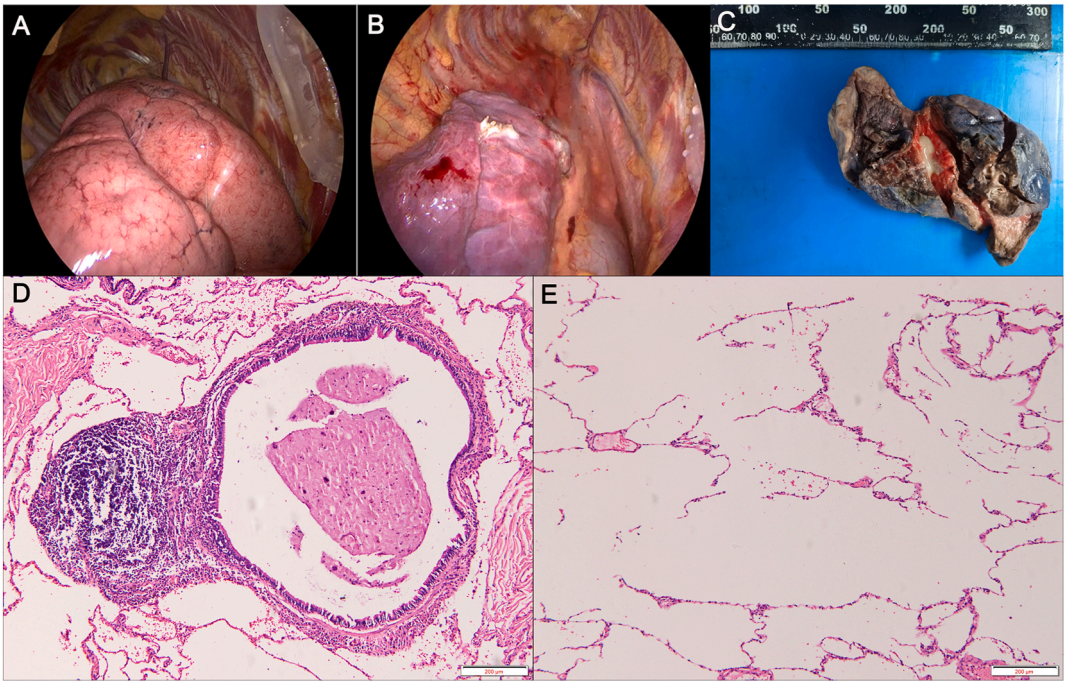

Fig. 4. Congenital bronchial atresia of the anterior segment (B3) of the right upper lobe. Thoracoscopic view before (A) and after (B) right upper lobectomy. C. Representative image of macroscopic extracted specimen demonstrating the blind-end of B3 with associated dilated distal segmental bronchi filled with mucus encased by hyperinflated lung parenchyma. Histopathological assessment (original magnification, × 200; hematoxylin-eosin staining) of the resected lung revealing the following: (D) there was no proximal or central tracheal communication of B3, whereas distal to B3 there was a dilated mucus-filled bronchus. (E) Emphysematous changes in the distal side of the mucinous impaction were observed.

detection of CBA [19]. Furthermore, 3D chest CT using multiplanar reconstruction and the volume rendering method could provide an additional help to the clinician, especially when the diagnosis of CBA via bronchoscopy is difficult [3,20]. Notably, histopathological assessment of lung tissue is necessary to make a definitive diagnosis of CBA [5]. It should be emphasized that a multidisciplinary consultation is worth to be recommended for CBA diagnosis due to rarity and complexity of this disorder itself.

There are no standardized guidelines regarding the treatment of CBA. It is accepted wisdom that surgical removal should be considered in patients who are presenting with symptoms, especially those with recurrent and severe infection symptoms, that cannot be treated effectively pharmacologically [3,4,11]. However, there is a significant debate regarding whether asymptomatic patients with CBA should be managed surgically or conservatively. Some clinicians prefer to perform surgical intervention on asymptomatic patients to avoid the extension of infection to the normal lung due to lung structural disruptions of the CBA cases caused by bronchodilation and emphysematous changes [4,21,22]. On the contrary, other experts suggest expectant management in which follow-up with regular chest radiographs should be performed to monitor asymptomatic patients with CBA [3]. The best way to address this debate is to invest enough resources into researching the natural history of CBA, envisioning prognostic tools, and building a global registry [5]. Considering patients with CBA are usually otherwise young and healthy with no comorbidity and good quality lung parenchyma, the thoracoscopic approach is therefore considered as a suitable minimally invasive surgery for treatment [3,4]. Interestingly, Masroujeh et al. [23] described another novel minimally invasive approach in a symptomatic case with CBA that used transbronchial needle aspiration under endobronchial ultrasound guidance for atretic airway opening. Obviously, the therapeutic applicability of this technique may be limited by structural integrity of distal to the atretic bronchus and can only be evaluated with further research. In terms of specific surgical manner, lobectomy is the most frequently reported resection since in the long term, the intact adjacent lung parenchyma compressed by the abnormal segment has increased risks of destruction [3,4]. In our case, because the bronchial opening of RML had presented with extrinsic compression stenosis and the mediastinum had shifted to the left due to the space-occupying effect of emphysema, thoracoscopic right upper lobectomy was performed.

4. Conclusion

In summary, we believe that this case report provides three important insights into CBA. First, 3D reconstruction CT could provide an additional help to the clinician, especially when the diagnosis of CBA via bronchoscopy is difficult. Second, a multidisciplinary consultation should be recommended for the management of patients with CBA. Third, thoracoscopic surgical resection should be considered in CBA patients with tracheobronchial stenosis caused by emphysema.

Ethical statement

This case was conducted ethically in accordance with the World Medical Association Declaration of Helsinki. The data collection was approved by the Ethics Committees of Naval Medical University (CHEC2019-061), and written informed consent was obtained from the patient for publication of this case report and any accompanying images.

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Data availability statement

Data included in article/supplementary material/referenced in article.

CRediT authorship contribution statement

Huan Lin: Writing – review & editing, Writing – original draft, Validation, Methodology, Investigation, Formal analysis, Data curation, Conceptualization. **Sen Tian:** Writing – review & editing, Writing – original draft, Investigation, Formal analysis, Data curation. **Chunlai Niu:** Writing – original draft, Validation, Methodology, Investigation. **Xiaping Shen:** Writing – original draft, Formal analysis, Data curation. **Mingming Wang:** Writing – original draft, Validation. **Changhong Wan:** Writing – original draft, Data curation. **Bowen Shi:** Writing – original draft, Formal analysis. **Hezhong Chen:** Writing – original draft, Supervision, Project administration. **Miaoxia He:** Writing – original draft, Formal analysis, Data curation. **Chong Bai:** Writing – review & editing, Writing – original draft, Supervision, Project administration, Conceptualization. **Haidong Huang:** Writing – review & editing, Writing – original draft, Validation, Methodology, Investigation, Formal analysis.

Declaration of competing interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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