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A case of congenital bronchial atresia patient with subclinical infection who underwent lung resection

Saori Murata [a,1], Tomoyo Oguri [a,1], Shinji Sasada [a,*], Yumi Tsuchiya [a], Kota Ishioka [a], Saeko Takahashi [a], Shoji Kuriyama [b], Masahiro Kaji [b], Reishi Seki [c], Shigemichi Hirose [c], Morio Nakamura [a]

[a] Department of Respiratory Medicine, Tokyo Saiseikai Central Hospital, Japan
[b] Department of Thoracic Surgery, Tokyo Saiseikai Central Hospital, Japan
[c] Department of Diagnostic Pathology, Tokyo Saiseikai Central Hospital, Japan

A B S T R A C T

Congenital bronchial atresia, CBA, is rare and has often asymptomatic benign condition. The CBA condition usually arose during the formation of bronchi, but the CBA patients are able to live well into adulthood. This case highlights a potential surgical intervention for a CBA patient with subclinical infection.

A 55-year-old Japanese male had abnormal findings on his chest X-ray at an annual health check-up in March 2018. His chest computed tomography (CT) revealed bronchial stenosis and infiltrative shadow in the right inferior lobe. He was referred to our hospital for further investigation and was diagnosed CBA after a variety of examinations including bronchoscopy. His dilated bronchi were filled with mucus, the end of one of the bronchi had obstructive pneumonia, and subclinical infection in the CBA lesion was suspected. Also, the result of bronchoscopy disclosed intrabronchial infection with Gram-positive bacteria so we performed lobectomy onto the lower lobe. Although no protocol had been established, a surgical intervention would be necessary for this case.

1. Introduction

Congenital bronchial atresia (CBA) is a rare, asymptomatic condition characterized by a bronchial artery occlusion during the fetal period [1], with an estimate prevalence of 1.2 cases every 100,000 [2]. There is no guideline for the treatment or management of CBA, and there is no consensus on the treatment policy for asymptomatic CBA that is hitherto undocumented [3]. We report a case that a CBA patient with subclinical infection who underwent a surgical intervention.

2. Case presentation

A 55-year-old Japanese male patient had no history of any respiratory disease or of smoking. He took an annual health check-up in March 2018 and his chest X-ray showed pulmonary opacity, so he was referred to our hospital for further investigation. He was afebrile, and his body temperature was 36.6 °C and his SpO2 was 96% on arrival. His blood test results were WBC: 5800/μL; C-reactive protein: <0.30mg/dL; β-D-glucans: <6.0 pg/mL; interferon-γ release test: (−); antibodies to *Mycobacterium avium* complex: positive. His tumor markers were normal: cacinoembrionic antigen: 2.0ng/mL; cytokeratin: 0.82 ng/mL (19 fragments); progastrin-releasing peptide: 30.6 pg/mL. His pulmonary function test showed: vital capacity: 4.23L (111.9% of the predicted value); forced expiratory volume % in 1 sec: 74.5%; diffusion capacity for CO: 26.20 mL/min/mmHg (100.7% of the predicted value). His chest X-ray showed infiltrative shadow on the right hilar to inferior lunch mediastinum (Fig. 1A). His CT showed a globed finger sign along the broncho-vascular bundles, the formation of mucoid impaction, and the air density in the distal bronchus of the superior segment in the right lower lobe (Fig. 1B).

In his bronchoscopy result, the membranous portion of the right truncus intermedius was rotated about 90° to the right and no bronchial bifurcation in the region where the right B6 should be located (Fig. 2). The bronchial epithelium in the right inferior bronchus was smooth. Methicillin-susceptible *Staphylococcus aureus* (MSSA) was detected from the bronchial washing fluid from the right inferior lobe. Since the patient would have congenial CBA which could be associated to the infection and inflammation based on the findings, we performed lobectomy on his right inferior lobe. A yellowish, white area and a dilated bronchi filled with mucus was appeared in the specimen (Fig. 3A). Also, we observed foam cells, lymphocytes, and plasma cells filled with the alveolar space and there was some fibrosis developed in the distal part to

* Corresponding author.
E-mail address: ssasada@saichu.jp (S. Sasada).
[1] equally contributed author.

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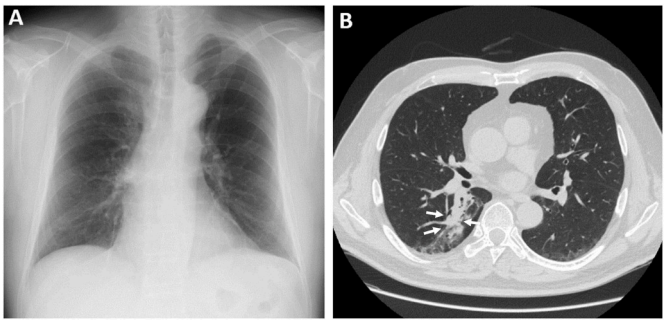

Fig. 1. Findings on chest radiography and computed tomography (CT) on admission. A. Chest radiography showed infiltrative shadow on the right middle field. B. Chest CT disclosed gloved finger sign along broncho-vascular bundles with the formation of mucoid impaction in superior segment of right lower lobe (white arrow).

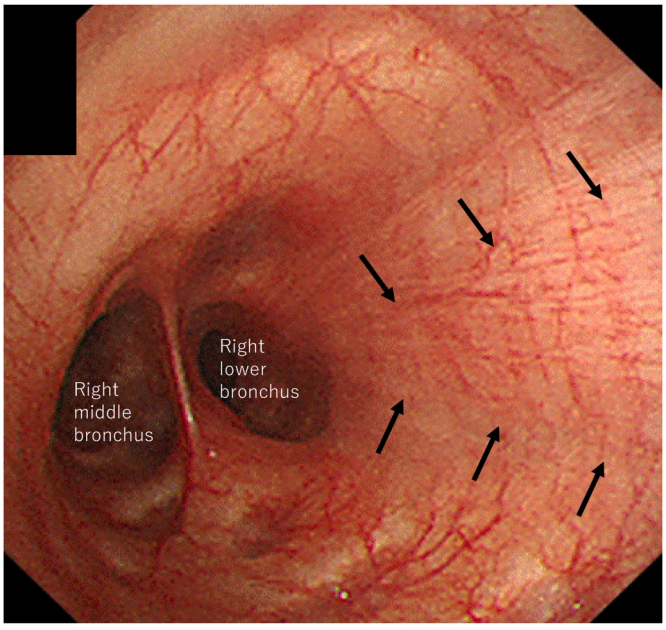

Fig. 2. Findings on the bronchoscopy. The membranous portion of right truncus intermedius was rotated about 90° to the right (black arrows), and bronchus of superior segment of right inferior lobe was absent.

the mucus. These finding indicated that the patient developed obstructive pneumonia (Fig. 3B). Also on Fig. 3C, the fine and fragile cartilage tissues were sparsely spread out, the supporting tissues were thin, and almost no bronchial gland was distributed. The patient was stable and didn't develop any complication for more than a year after the surgery.

3. Discussion

Congenital bronchial atresia was first reported by Ramsay in 1953 [4]. About two-thirds of the patients are asymptomatic and are often found in a chest X-ray in an annual health check in their 20s or 30s. One-third of the CBA patients may have symptoms, such as complain a cough, shortness of breath, recurrent infections, lesser extent wheezing, hemoptysis, chest pain, or pneumothorax [5,6]. Our patient was middle-aged, developed CBA in the right, inferior lobe, whereas the patients in Europe or U.S. have developed in the left upper lobe [7]. Some reports indicate that 50% of the patients was diagnosed by bronchoscopy and a 3D reconstruction CT could be used for a diagnostic purpose for the blind end of the bronchus [8,9]. We had a difficulty to visualize and detect the blind end since there was no branch in the bronchus of the superior segment of the right inferior lobe. Since CBA is often unaware and the patients live well into their late adulthood, a surgical intervention would be the first choice for the young, CBA suspected patients for diagnostic purpose. However, lobectomy was necessary for our case because it was difficult to differentiate between the abnormal and the normal sites and there was a risk that his bronchobpleural fistula would be collapsed by cutting the collateral path even partially. The lung structure of the CBA patients would be damaged by bronchodilation and emphysematous changes. This tendency causes an infection by acid-fast bacteria, such as *Pseudomonas aeruginosa* and Aspergillus genus. We frequently observe abscess and the infection spread to the adjunct regions [10]. Our patient developed obstructive pneumonia but no abscess formation. Although a CBA patient develops subclinical infection, we would be able to intervene medically before abscess is formed. A surgical resection should be considered because a CBA patient may experience disruption of lung structure, colonization of a drug-resistant bacteria, and the spread of the infection to the normal lung tissue without any symptom.

4. Conclusion

We have experienced a CBA patient with subclinical infection which required a surgery. We should decide the final decision of the course of the treatment based on maintaining the lung function and the whole body condition even though preventing formation of abscess would also be important to be consider – surgical intervention would be the primary choice especially among the young CBA patients.

Declaration of competing interest

The authors have declared that no conflict of interests exists.

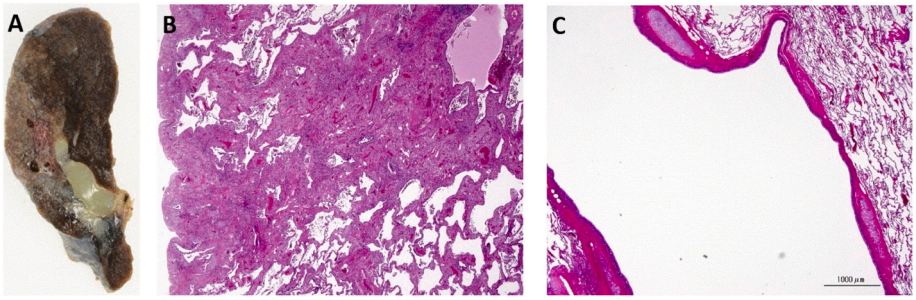

Fig. 3. Pathological findings. A. The gross appearance of hilar aspect. A dilated bronchus was filled with mucus. B. Microscopically, foam cells, lymphocytes, and plasma cells filled the alveolar space. C. Dilated bronchi seen with the minute and fragile cartilage tissue was sparsely distributed, the supporting tissue was thin, and there was almost no bronchial gland distribution.

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