Case Report

Efficacy of Nasal Continuous Positive Airway Pressure in Tracheobronchomalacia

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ABSTRACT: A 68-year-old woman was admitted to our hospital with a persistent productive cough of 1 year's duration and dyspnea on exertion. She had a history of chronic bronchitis during childhood and no history of smoking. Chest computed tomography (CT) images on expiration revealed diffuse mosaic attenuation in both lungs and airway collapse of the trachea and main bronchi. Fiberoptic bronchoscopy images showed a triangular tracheal lumen and marked narrowing of the main bronchi on expiration. Virtual bronchoscopy (VB) images showed subtotal stenosis of several segmental bronchi. In addition, three-dimensional (3D) images of the airways revealed excessive expiratory collapse of the trachea and distal discrete malacia. No abnormalities were seen in a surgical lung biopsy specimen. These findings indicated a diagnosis of tracheobronchomalacia (TBM). Because the airway lesions were diffuse, noninvasive ventilation with continuous positive airway pressure was started and resulted in long-term improvement in daytime clinical symptoms and arterial blood gas values.

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KEYWORDS: tracheobronchomalacia, mosaic attenuation, virtual bronchoscopy, three-dimensional images, fiberoptic bronchoscopy

Tracheobronchomalacia (TBM) results from weakness of the airway walls and/or supporting cartilage.[1)] It is classified as primary (genetic, congenital) or secondary (acquired), *e.g.*, chronic obstructive pulmonary disease or bronchial asthma.[2)] Although bronchoscopy and conventional computed tomography (CT) have been the standard techniques for diagnosing TBM, recent reports indicate that noninvasive virtual bronchoscopy (VB) and three-dimensional (3D) images of airways are also effective in diagnosis.[3,4)] Furthermore, TBM is associated with the presence of air trapping, which results in a mosaic attenuation pattern on chest CT images.[5)]

We report a rare case of primary TBM with mosaic attenuation on chest CT that was diagnosed by using VB and 3D images of airways to examine lesion distribution.

Case Report

A 68-year-old woman was referred to our hospital for a persistent productive cough of 1 year's duration and dyspnea on exertion. She had a past history of chronic bronchi-

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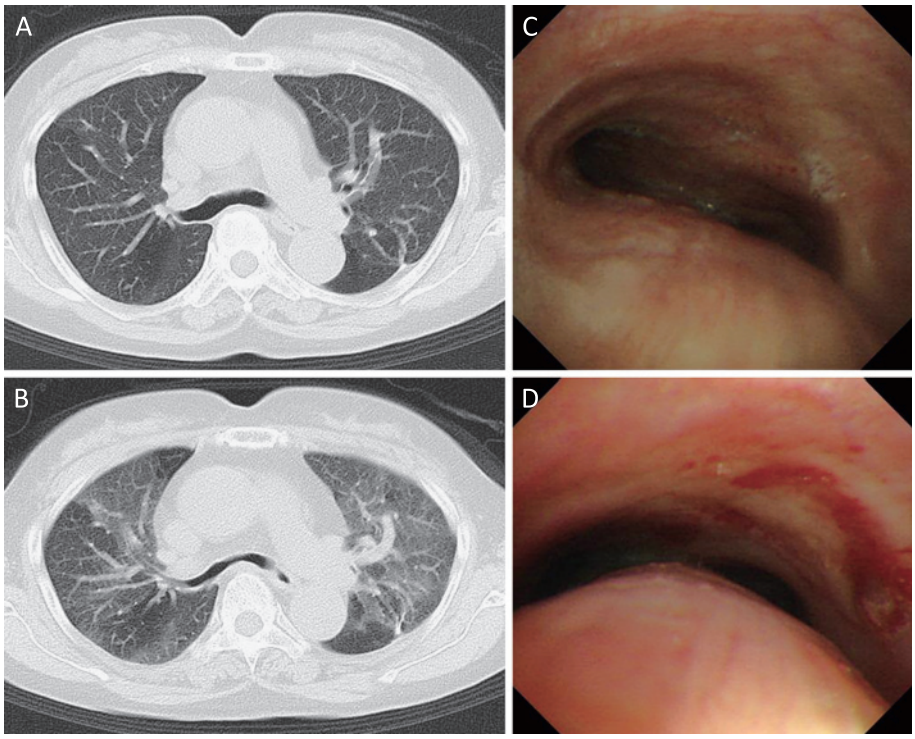

Fig. 1 Chest computed tomography (CT) images reveal normal tracheal appearance at the end of inspiration (A) and a diffuse mosaic perfusion in both lungs, and airway collapse of the trachea and bronchi, during expiration (B).
Fiberoptic bronchoscopy images show a triangular tracheal lumen and marked narrowing of the main bronchi on inspiration (C), as compared with findings on expiration (D).

tis during childhood and no history of smoking or dust exposure. Arterial blood gas analysis showed a pH of 7.42, $PaCO_2$ of 42.2 Torr, and PaO_2 of 64.0 Torr in room air. The results of a pulmonary function test—*i.e.*, vital capacity of 1.31 *l* (61.2% of predicted), forced expiratory volume in one second (FEV_1) of 0.85 *l* (58.6% of predicted), diffusion for carbon monoxide of 7.46 ml/min/mmHg (65.2% of predicted), residual volume/total lung capacity (RV/TLC) of 50.0%, and V50/V25 of 5.20—indicated mixed ventilation disturbance and impairment of the small airways. There was no evidence of airway reversibility. Chest CT images showed normal tracheal appearance at the end of inspiration (Fig. 1A) and diffuse mosaic perfusion in both lungs, with airway collapse of the trachea and bronchi, during expiration (Fig. 1B). Fiberoptic bronchoscopy images showed a triangular tracheal lumen, and the main bronchi were markedly narrower during expiration than during inspiration (Fig. 1C, 1D). VB images showed stenosis of the trachea and main bronchi and subtotal stenosis of several segmental bronchi (Fig. 2). Furthermore, 3D images of airways revealed excessive expiratory collapse of the trachea and distal discrete malacia (versus a healthy control) (Fig. 3). No fungal, bacterial, or mycobacterial pathogens were isolated from sputum or bronchial lavage fluid. Due to mosaic attenuation in the left lower lobe, the patient underwent video-assisted thoracoscopic surgery (VATS) as a diagnostic procedure. However, there was no evidence of chronic bronchiolitis, bronchiolitis obliterans, granulomas, or emphysematous change, and no other abnormalities were noted in the lung specimen. Consequently, the patient's condition was diagnosed as primary TBM. Noninvasive ventilation with continuous positive airway pressure (CPAP) was started at a pressure of 7 cm H_2O and resulted in long-term improvement in daytime clinical symptoms and arterial blood gas values. PaO_2 improved from 65.0 Torr to 78.7 Torr after CPAP was introduced.

Discussion

TBM is a characterized by excessive expiratory collapse of the central airways due to increased flaccidity of the membranous airway wall and/or weakness of the supporting cartilage.[1] The reported incidence of adult TBM was 4.1% among 78 nonsmokers with chronic cough[6], and Ikeda et al. found that TBM was present in 12.7% (542 of

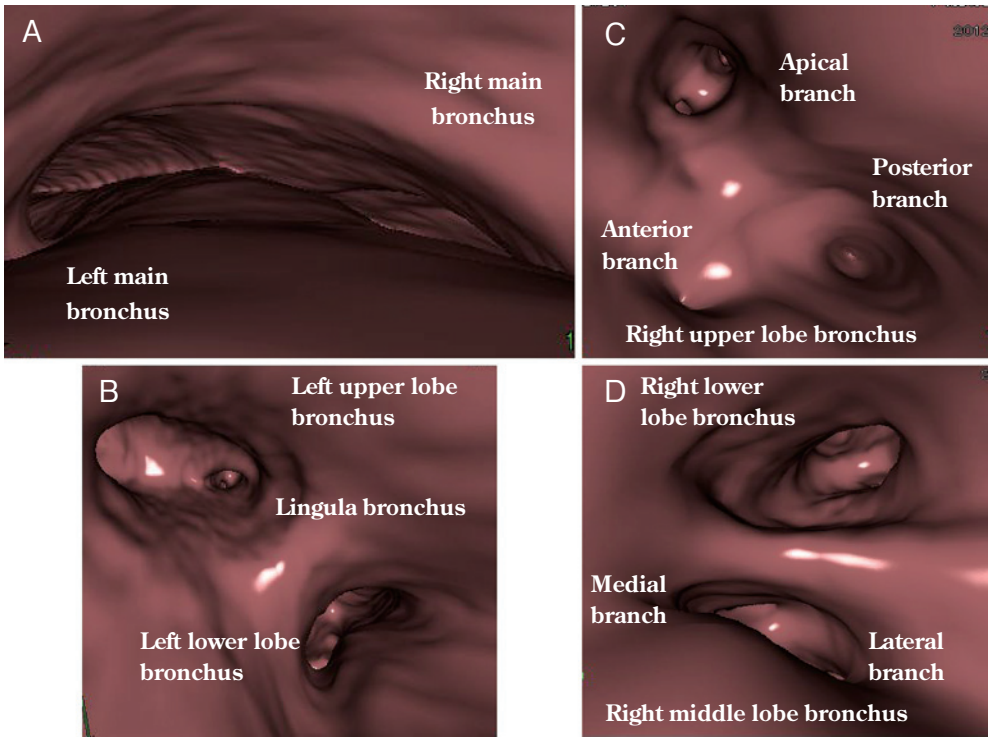

Fig. 2　Virtual bronchoscopy (VB) images show stenosis of the trachea and main bronchi and subtotal stenosis of several segmental bronchi.
(A) Tracheal bifurcation, (B) Left second carina bronchus, (C) Right upper lobe bronchus, (D) Right middle bronchial trunk

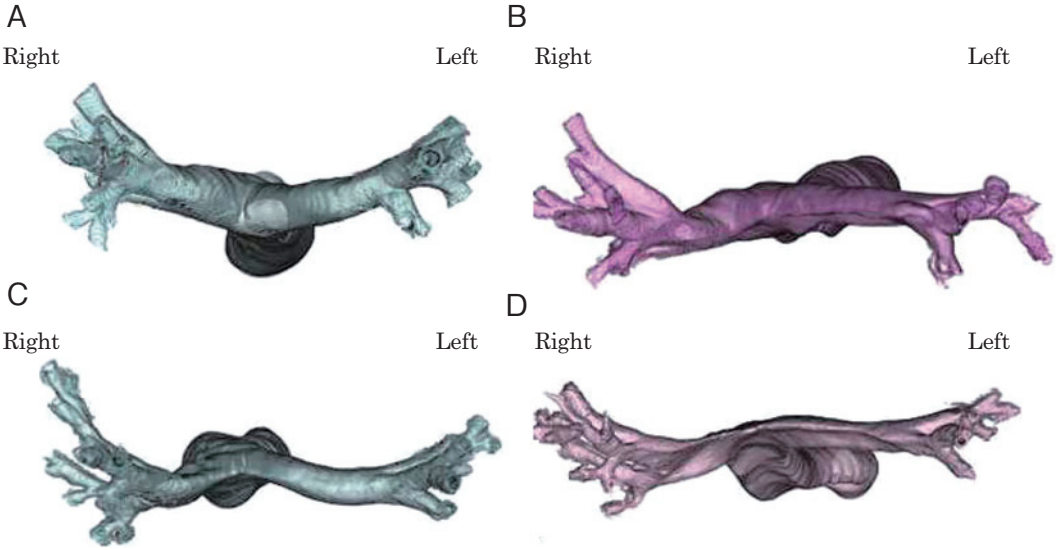

Fig. 3　Three-dimensional (3D) images of the bronchus show excessive expiratory collapse of the trachea and distal discrete malacia (compared with healthy control).
(A, B) Healthy control, (C, D) Present patient
(A, C) Inspiratory phase, (B, D) Expiratory phase

4283) of patients who underwent bronchoscopy.[7] TBM is classified as primary (congenital) or secondary (acquired), *e.g.*, chronic obstructive pulmonary disease or bronchial asthma, chronic inflammation (relapsing polychondritis), and posttraumatic, external compression of the trachea (due to tumors, cysts, abscess, or cardiovascular abnor-

malities).[2] The primary form can be idiopathic or result from congenital cartilaginous weakness, congenital syndromes, or other congenital anomalies such as tracheoesophageal fistula. In adults, secondary TBM is more common than primary TBM, but some adults have primary TBM that was undiagnosed in childhood. Our patient had no causes of secondary TBM, and no abnormalities were seen in lung parenchyma or bronchioli in a lung biopsy specimen obtained by VATS. These findings indicated a diagnosis of primary TBM. However, we cannot exclude the possibility of secondary TBM, as she had a past history of chronic bronchitis in childhood.

TBM should be considered in the differential diagnosis when unexplained symptoms such as persistent cough, dyspnea on exertion, or mosaic attenuation pattern on chest CT are found. Relapsing polychondritis is a rare multisystem disorder characterized by recurrent episodes of inflammation of cartilaginous structures of the external ear, nose, and peripheral joints.[8] There was no evidence of tracheobronchial wall thickening with calcification or elevation of serum anti-type II collagen antibodies in our patient, even though these are frequent findings in TBM.

Air trapping is caused by excessive retention of gas in the lung during expiration. Air trapping is visualized on end-expiration CT scans as parenchymal areas with a less-than-normal increase in attenuation and lack of volume reduction. A comparison of inspiratory and expiratory CT scans can be helpful when air trapping is subtle or diffuse. Zhang et al.[5] reported that, as compared with control subjects, patients with TBM had significantly more frequent and more severe air trapping. Furthermore, they speculated that air trapping in TBM patients reflects the presence of chronic disease of the small airways, which is induced by recurrent infection and impaired clearance of respiratory secretions.

In our patient, there was prominent mosaic attenuation on chest CT, indicating air trapping except for airway collapse. VATS was performed to assess the possibility of diseases of the small airways, such as bronchiolitis obliterans or hypersensitivity pneumonitis, but the histological findings showed no evidence of such disease.

Bronchoscopy has been the standard examination for diagnosing TBM, but recent techniques such as VB and 3D images of airways were shown to be feasible.[3,4] A major advantage of these techniques is that they noninvasively provide useful morphologic information on structural abnormalities. Moreover, VB can depict the passage through severe stenoses, which cannot be observed with conventional bronchoscopy.

Many adults with TBM do not require therapy when the condition is incidentally detected or they are asymptomatic.[2] However, there is no consensus concerning the treatment of symptomatic TBM. In patients with disabling symptoms, CPAP can keep the airway open and facilitate secretion drainage.[9] Ferguson & Benoist[10] reported that CPAP at a pressure of 10 cm H_2O reduced dynamic airway collapse in most patients with TBM. In our patient, introduction of intermittent diurnal CPAP resulted in improvement of her respiratory symptoms and oxygenation. Ernst et al.[11] found that, although silicone stent placement therapy improved clinical symptoms and functional status during the short term, complications such as stent migration, mucus plugging, and granulation tissue formation occurred in 90% of TBM patients. Recently, new surgical treatments such as tracheoplasty[12] have substantially improved outcomes in selected patients. These treatments are effective in the short term; however, evidence of long-term effectiveness is limited.

In conclusion, VB and 3D images of airways were useful in evaluating airway collapse in TBM. The long-term effectiveness of surgical treatments and noninvasive mechanical ventilation such as CPAP is uncertain, especially for cases of TBM with diffuse airways stenosis, as in our patient. Therefore, additional studies should attempt to ascertain outcomes for several potential therapies.

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Conflict of interest statement: All the authors declare that they have no conflict of interests regarding the present work.

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気管・気管支軟化症における経鼻的持続陽圧呼吸療法の効果

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要約：68 歳女性．1 年間持続する咳嗽，労作時呼吸困難を主訴に来院．喫煙歴なし．幼少時より慢性気管支炎を指摘．胸部 computed tomography（CT）ではモザイクパターンを呈し，呼気時に増強する気管の高度狭窄を認めた．Virtual bronchoscopy では，気管支末梢まで狭窄を示した．気道 three-dimensional（3D）再構築像では，呼気時に気管・気管支の著明な狭窄を認めた．胸腔鏡下肺生検では明らかな異常所見は認められなかった．以上より気管・気管支軟化症と診断した．気道病変がびまん性であったため，非侵襲的陽圧換気療法を開始したところ，臨床症状および動脈血液ガスの改善を認めた．

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索引用語：気管支軟化症，モザイクパターン，virtual bronchoscopy，気道 three-dimensional（3D）再構築像，ファイバー気管支鏡

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