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Bronchoscopic observation of unusual deformities of bronchial cartilage and subsequent airway narrowing in respiratory relapsing polychondritis

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Summary

Background:	Relapsing polychondritis (RP) is a rare inflammatory disease characterized by recurrent chondritis and inflammation of other proteoglycan-rich tissues. An RP patient with co-existing respiratory tract problems could have a poor prognosis.
Case Report:	We reported a case of RP died with recurrent suffocation. At the early stage in this case, unusual deformities of bronchial cartilage were observed. Following systemic corticosteroid therapy, these deformities disappeared, and typical diffuse mucosal edema and dynamic collapse of airways developed.
Conclusions:	These bronchoscopic abnormalities could be the early stage of RP.
key words:	relapsing polychondritis • suffocation • deformity of bronchial cartilage

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BACKGROUND

Relapsing polychondritis (RP) is a rare systemic inflammatory disease that is characterized by recurrent inflammatory changes in proteoglycan-rich tissues in the body [1–5]. Approximately one-half to two-thirds of RP patients develop respiratory tract complications throughout their course [2,3,6,7]. Bronchoscopy is informative for respiratory RP, though it cannot be easily performed in all patients. The case of a patient with respiratory RP in whom bronchoscopy demonstrated bronchial cartilage deformity and subsequent destruction of the cartilage and mucosal edema, which caused airway narrowing, is reported.

CASE REPORT

A 71-year-old woman was admitted to our department because of recurrent episodes of dyspnea and suffocation. She developed cardiac tamponade following percutaneous coronary intervention for her angina four months before admission to our department. Open heart surgery was performed, and she was intubated at that time. During her stay in the intensive care unit, bilateral vocal cord palsy occurred, and she was temporarily tracheotomized. Thereafter, she was discharged with mild exertional dyspnea. Her dyspnea deteriorated twice, one and two months before admission to our department, and she was repeatedly tracheotomized with a diagnosis of infectious subglottic edema at the Department of Otolaryngology. Finally, since lower respiratory tract involvement was suspected, she was referred to respiratory medicine for further study and treatment. She had smoked a packet of tobacco everyday for 36 years and quit smoking two months ago. She had been a housekeeper throughout her life. She had no history of bronchial asthma or other allergic diseases. She did not have any other collagen disease. She appeared well with her tracheotomy. Systemic observation failed to find any cartilaginous structure abnormalities. Respiratory examination demonstrated absence of wheezing and crackles. However, forced expiration produced audible stridor. Laboratory examinations of the peripheral blood showed a slightly increased C-reactive protein level (1.1 mg/dl). Her chest X-ray was normal, while her chest CT showed some protuberances in the trachea (Figure 1). In accordance with the CT findings, bronchoscopy revealed that both ends of the bronchial cartilage crescent protruded into the bronchial lumen from the trachea to both main bronchi (Figure 2A,B). Bronchoscopic biopsy of the tracheal cartilage was performed from the protruding points of the trachea. On pathology, degeneration and necrosis of bronchial cartilage (Figure 3) suggesting the diagnosis of RP were seen. She had received systemic corticosteroids (methylprednisolone 125 mg/day for three days) for subglottic edema before bronchoscopy, which may explain why inflammatory cellular infiltration was absent. Further medical history taking revealed that she had experienced multiple arthralgia during the preceding two months, which improved following systemic corticosteroid therapy. She was started on oral prednisolone (PSL), tapering from 40 mg/day after the bronchoscopy. With therapy, the bronchial cartilage deformity disappeared, while diffuse edema of the bronchial mucosa and dynamic collapse of the major airway, typical features of respiratory RP, occurred (Figure 2C,D). Based on previous reports [8,9], methotrexate (MTX; 8 mg/week) was

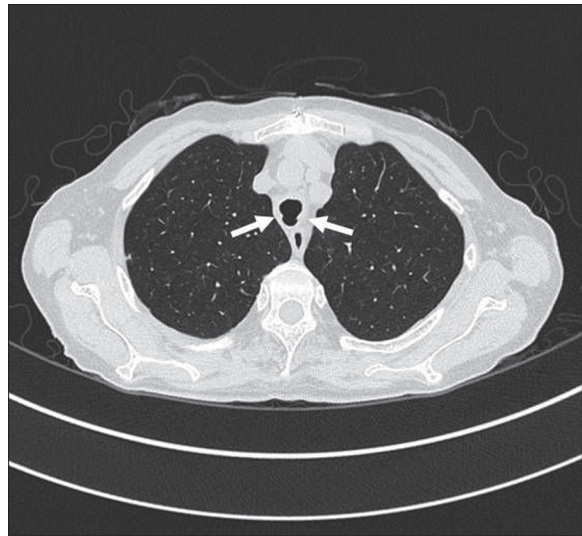


Figure 1. Chest computed tomography. Arrows indicate the abnormal prominence of the tracheal wall into the intratracheal space. These changes are associated with the bronchoscopic abnormalities of the tracheal cartilage of Figure 2.

added. Recently, she has been taking PSL 10 mg daily and MTX 8 mg/week. She experienced no further episodes of suffocation since the last tracheotomy. Nonetheless, her dyspnea is gradually deteriorating.

DISCUSSION

When the initial manifestation of RP is respiratory symptoms, clinical diagnosis is difficult and delayed. The diagnosis of RP is usually based on the modified criteria by Damiani and Levine [5], wherein one or more of the criteria of McAdams et al. [4] and pathological confirmation of chondritis lead to the diagnosis. In the present case, pathological and bronchoscopic confirmation of tracheobronchial chondritis and the history of multiple arthralgia led to the diagnosis.

About a half of RP patients experience respiratory tract complications, and 10% of RP patients die of this complication [6,7]. Thus, early diagnosis of respiratory involvement is critically important in RP with respiratory symptoms. Bronchoscopy is essential in identifying the exact site, nature, and severity of airway involvement, as well as making the pathological diagnosis, in RP. Nonetheless, bronchoscopy cannot be easily performed in all cases, since it does carry a risk of provoking dyspnea, collapse of the airways, hypoxia, and death [10]. Therefore, the characteristics of the early stage and the natural course of respiratory involvement in respiratory RP remain to be determined. In the present case, observation of the respiratory tree could be performed at the early stage through the tracheotomy hole. Bronchoscopy clearly revealed that inflammation developed predominantly in bronchial cartilage during the early stage of RP, followed by the destruction of bronchial cartilage and the development of edema in the bronchial mucosa, both of which caused dynamic collapse of the major airway.

The etiology and mechanism of RP are still unclear, but RP could be a collagen disease [1–3,11]. Anti-type II collagen

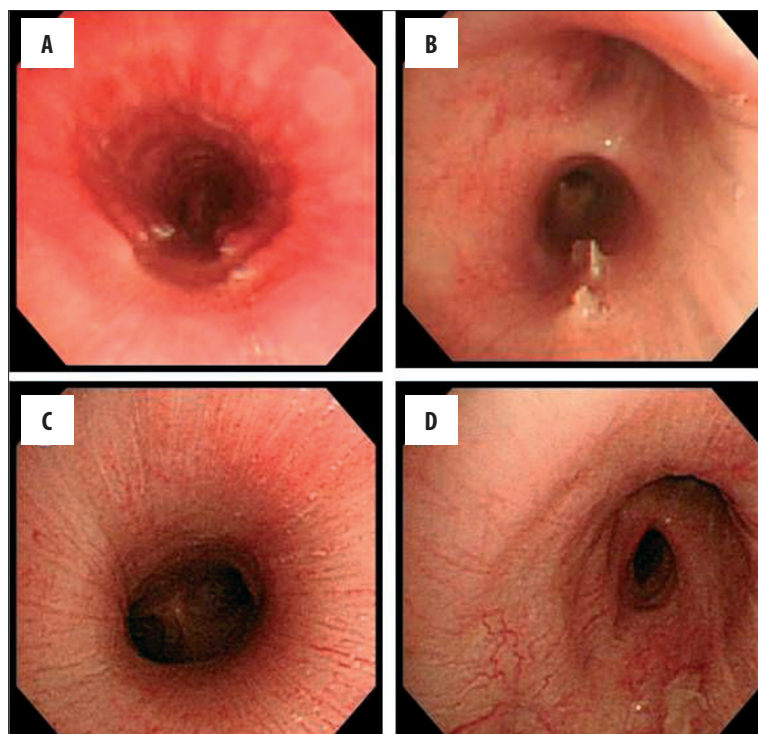


Figure 2. Bronchoscopic view of the trachea before (A, B) and after (C, D) systemic corticosteroid treatment. Characteristic deformities of tracheal cartilage are observed (A, B). These deformities disappear (A → C) (B → D) after systemic corticosteroid therapy, and diffuse mucosal edema develops.

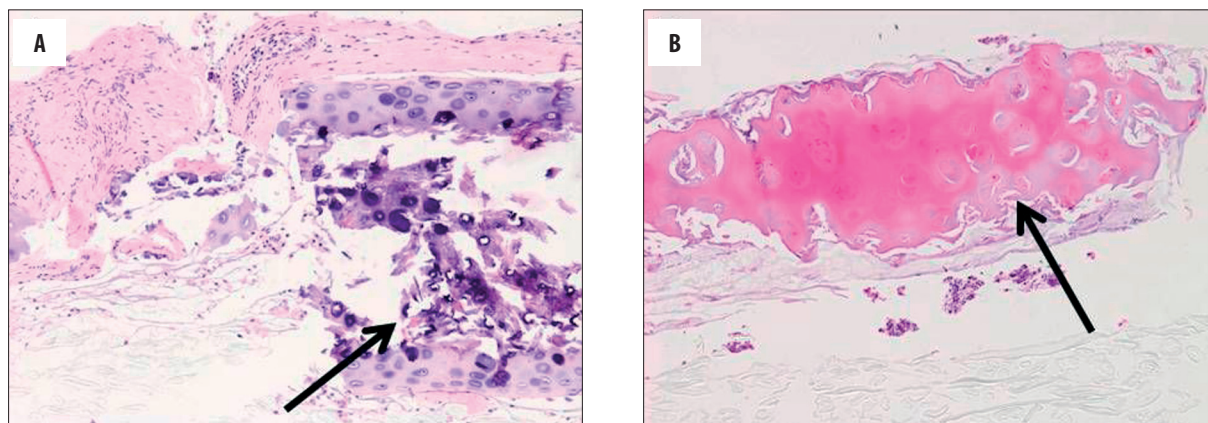


Figure 3. Pathological examination of the deformed part of the tracheal cartilage (hematoxylin-eosin stain; magnification $\times 100$). Deformity (A) and necrosis (B) of cartilaginous tissue show chondritis. Arrows in the figures show typical changes.

antibody helps make the diagnosis, and sometimes it indicates the activity of RP [11]. A rise in anti-type II collagen (84.6 EU/ml) was also observed in this case, which decreased to 19.9 EU/ml after treatment by PSL and MTX. It might also be possible that the preceding intubation for open heart surgery exacerbated the airway inflammation in the present case. Despite treatment with systemic corticosteroids and MTX, her symptoms are deteriorating. Addition of other treatment, such as immunosuppressive agents, tumor necrosis factor- α antagonists, and a bronchial stent, should be considered in the future [12].

CONCLUSIONS

A case of RP without deformity of outside cartilage but with unusual deformities of bronchial cartilage was described. Although the patient had a history of knee joint pain, at the initial stage, she only complained of respiratory symptoms, episodic dyspnea and suffocation. Physicians should

be aware of RP as a rare cause of respiratory symptoms, especially recurrent episodes of suffocation.

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