Tracheobronchial Involvement in Relapsing Polychondritis Diagnosed on Endobronchial Ultrasound

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Abstract

Respiratory tract chondritis is not uncommon in patients with relapsing polychondritis (RP); however, diagnosing this condition remains problematic, especially in patients whose extrapulmonary manifestations do not predominate, as there are broad differential diagnoses of airway obstruction. We herein report the case of a 56-year-old man who presented with cough and dyspnea. Computed tomography of the chest demonstrated diffuse smooth thickening of the visualized tracheobronchial wall with a moderately narrowed lumen. Airway chondritis was diagnosed on endobronchial ultrasound following demonstration of thickening of the submucosal and cartilaginous layers in the anterior and lateral aspects of the bronchial wall, while the posterior region expressed less involvement. In conjunction with nasal and auricular chondritis, which were previously overlooked, RP was finally diagnosed.

Key words: relapsing polychondritis, endobronchial ultrasound


Introduction

Relapsing polychondritis (RP) is an episodic and progressive systemic inflammatory disorder involving cartilaginous structures, predominantly those of the ears, nose, laryngotracheobronchial tree and other connective tissue structures such as the eyes, kidneys, aorta, heart and skin (1). The clinical manifestations may vary due to differences in the affected organs, severity and duration of symptoms. Lower respiratory tract involvement has been reported in approximately 20% to 50% of patients and may sometimes be the presenting symptoms (2). Making a diagnosis of tracheobronchial involvement is a challenging task for clinicians. In addition, obtaining a histological diagnosis of tracheobronchial chondritis on bronchoscopic examinations remains difficult and is frequently overlooked. We herein report the case of a patient presenting with dyspnea and RP that was finally diagnosed on bronchoscopy with endobronchial ultrasound (EBUS).

Case Report

A 56-year-old man presented with a productive cough and dyspnea on exertion lasting for six months with a previous history of bilateral anterior uveitis four years earlier. His eye symptoms improved following the administration of 6-month prednisolone therapy without any recurrent episodes. He had a 60 pack-year smoking history and had already quit smoking four months previously.

On presentation, the patient was not tachypneic or dyspneic. On physical examination, he exhibited a slight loss of height of the mid portion of the nose, a so-called saddle nose deformity (Fig. 1A). In addition, his right pinna was observed to exhibit more swelling than its counterpart (Fig. 1B). However, the patient had not noticed these changes previously. On auscultation, central rhonchi were detected during the expiratory phase.

Chest radiography revealed narrowing of the distal trachea (Fig. 1C). Computed tomography (CT) of the chest demon-
Figure 1. (A) Image of the nose of the patient demonstrating a mild saddle nose deformity. (B) The right pinna showing mild auricular edema and a deformity sparing the ear lobule. (C) Chest radiograph demonstrating narrowing of the distal tracheal caliber (arrowhead). (D) Flow-volume loop of spirometry illustrating a pattern of upper airway obstruction.

Stratified diffuse smooth thickening of the tracheobronchial wall with a moderately narrowing lumen (Fig. 2A, B). A pulmonary function test also disclosed an intrathoracic upper airway obstruction pattern (Fig. 1D). In addition, a CT scan of the sinuses demonstrated swelling and depression of the cartilaginous portion of the nose, while the bony region remained intact. Serology testing was negative for antinuclear antibodies and antineutrophil cytoplasmic antibodies.

Bronchoscopy was performed to investigate the cause of the generalized airway wall swelling. On bronchoscopic examination, widespread bronchial wall edema was observed predominantly in the anterior and lateral parts of the bronchi that resulted in the disappearance of the cartilaginous tracheobronchial ring structure with preserved smooth muscle lines in the posterior aspect (Fig. 3A, B). Nevertheless, no dynamic airway collapse was observed during expiration. Thereafter, a radial scanning probe EBUS (UM-BS20-26R; Olympus Ltd.; Tokyo, Japan) linked to a processor (EU-ME1; Olympus) with a frequency of 20 MHz was applied to examine the airway wall. EBUS images showed diffuse thickening of the bronchial wall, continuing along the trachea into both main bronchi due to the thickened submucosal layer, while the cartilaginous layer also appeared swollen and ill-defined (Fig. 4A, B). A biopsy of the tracheal mucosa revealed only nonspecific inflammation.

Based on McAdam’s criteria (3, 4), RP was finally diagnosed and treatment was commenced with prednisolone at a starting dose of 30 mg/day (0.5 mg/kg/day). The patient’s respiratory symptoms, including dyspnea and coughing, markedly improved after two weeks of treatment. Subsequently, the prednisolone dosage was gradually reduced and maintained at 20 mg/day. Chest CT performed three months after treatment showed a significant decrease in the bronchial wall thickening, resulting in an increase in the airway luminal diameter (Fig. 2C, D). Cartilaginous rings remained absent on follow-up bronchoscopy, and a more patent airway with no evidence of tracheobronchomalacia was observed. EBUS also demonstrated improvement in the cartilaginous layer swelling (Fig. 4C, D).

Discussion

The differential diagnoses of diffuse airway narrowing on CT scans include granulomatosis with polyangiitis (GPA) (previously known as Wegener’s granulomatosis), RP, tracheobronchopathia osteochondroplastica, amyloidosis, papillomatosis, sarcoidosis and rhinoscleroma (5). Although bronchoscopy does carry a risk of provoking dyspnea, airway collapse, hypoxia, asphyxia and death (6), it remains the most appropriate procedure for diagnostically examining...
these disease entities in terms of defining the characteristics of endoluminal lesions and obtaining specimens for histopathology to reach a definitive diagnosis. Nevertheless, in some circumstances, transbronchial biopsies do not lead to a diagnosis because the pathological lesions occur in unapproachable sites such as the submucosa and cartilaginous layer where performing the biopsy procedure may result in anatomical damage. Open surgical biopsies are an alternative procedure to obtain tissue diagnoses; however, they are considered to be dangerous, especially in patients with airway compromise.

EBUS has proven its utility in evaluating the central airways (7-9). The ultrasonographic findings of EBUS demonstrate the structure of the airway wall layer-by-layer, which
Figure 4. (A) Endobronchial ultrasound image and (B) a diagram demonstrating the submucosal and cartilaginous layers of the left main bronchus showing marked swelling and degeneration of the cartilage layer as well as thickening of the submucosal layer. After treatment, marked improvement in the bronchial wall swelling of the left main bronchus was identified on endobronchial ultrasound (C, D). The cartilaginous layer was more clearly denoted.

has been shown to correspond with the histology. In this case, EBUS demonstrated thickening of the submucosal and cartilaginous layers in the anterior and lateral aspects of the bronchial wall, while the posterior portion expressed less involvement, indicating that the disease primarily affected the cartilage. Therefore, airway chondritis was diagnosed based on this EBUS finding.

A diagnosis of RP can be pinpointed if the patient meets at least three of the six McAdam’s criteria (3, 4): 1. bilateral auricular chondritis, 2. nonerosive, seronegative inflammatory polyarthritis, 3. nasal chondritis, 4. ocular inflammation, 5. respiratory tract chondritis, and 6. audiovestibular damage. Alternatively, the diagnosis can be made based on the presence of one McAdam’s criterion and histologic evidence of chondritis or two McAdam’s criteria and a response to corticosteroids or dapsone. In our patient, based on the presence of respiratory tract chondritis in conjunction with nasal and auricular lesions, a diagnosis of RP was ultimately made. In addition, the improvement in the respiratory tract chondritis after treatment with corticosteroids enhanced support for the diagnosis of RP.

Histologically, findings in the initial phase demonstrate cartilage inflammation leading to airway fragility (10). As the disease progresses, destruction of the cartilaginous support and progression of fibrosis are responsible for dynamic airway collapse. As the cartilage cannot be regenerated, the dynamic airway collapse is likely to be irreversible and can contribute to chronic respiratory insufficiency. At this stage, airway interventions such as tracheostomy, tracheobronchial stent placement and continuous positive airway pressure (CPAP) play an important role (2). Therefore, making an early diagnosis and providing prompt treatment in the early stage of the disease should be undertaken to prevent irreversible complications and achieve excellent outcomes.

Nevertheless, respiratory tract chondritis is not theoretically uncommon, and extrapulmonary manifestations are almost always the key to diagnosis. Unfortunately, extrapulmonary presentations are frequently overlooked due to the reduced number of symptoms and the need for skillful clinical recognition, as respiratory symptoms are often misinterpreted and diagnosed as other diseases (11). Careful physical examination, meticulous chest radiograph interpretation,
and recognition of unique pulmonary function test results are required to diagnose this challenging disease.

In conclusion, we herein reported a case of RP that presented with a cough and dyspnea. With the ability of EBUS to identify each layer of the bronchial wall, respiratory tract chondritis was diagnosed. Therefore, we would like to emphasize the utility of EBUS in the diagnosis of this disease.

The authors state that they have no Conflict of Interest (COI).

Acknowledgement
The authors thank Dr. Pensupa Raweelert for the constructive suggestions and English editing.

References