Endobronchial Ultrasonography in the Diagnosis and Treatment of Relapsing Polychondritis With Tracheobronchial Malacia*

Yuka Miyazu, MD; Teruomi Miyazawa, MD, PhD, FCCP; Noriaki Kurimoto, MD; Yasuo Iwamoto, MD; Atsuko Ishida, MD; Koji Kanoh, MD; and Nobuoki Kohno, MD, FCCP

Relapsing polychondritis (RP) with tracheobronchial involvement has a poor prognosis, and a delay in diagnosis increases morbidity and mortality; however, the diagnosis is difficult to make. Endobronchial ultrasonography (EBUS) revealed changes in the tracheobronchial cartilage in two patients who met the criteria for RP, and facilitated the diagnosis. In these cases, EBUS revealed a poorly defined bronchial wall structure with two patterns of cartilaginous damage: fragmentation and edema. These cases were successfully treated by the implantation of nitinol stents, the sizes of which were determined by EBUS. EBUS was found to be useful in the diagnosis and treatment of RP.

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Key words: endobronchial ultrasonography; nitinol stent; polychondritis

Abbreviations: EBUS = endobronchial ultrasonography; RP = relapsing polychondritis

The diagnosis of relapsing polychondritis (RP) usually depends on a constellation of clinical and histologic features caused by chondritis.1–3 According to McAdams et al,1 if patients have at least three of the following six signs, the diagnosis is conclusive: bilateral auricular chondritis, nonerosive inflammatory polyarthritis, nasal chondritis, ocular inflammation, laryngotracheobronchial chondritis, and audiovestibular damage. The modified criteria was proposed by Damiani and Levine2: one or more of the criteria of McAdams et al1 with histologic confirmation or chondritis at least in two distinct locations with therapeutic response.

Endobronchial ultrasonography (EBUS) reveals the tracheobronchial wall structure and cartilaginous layer clearly.4,5 In this report, EBUS demonstrated changes in the tracheobronchial cartilage in two patients with RP who met the criteria of Damiani and Levine2 and facilitated the diagnosis.

CASE REPORTS

Case 1

A 67-year-old woman had undergone an emergency tracheotomy 5 years previously. Dyspnea initially improved following the tracheotomy, but then worsened over time and she was referred to our institution.

There was no evidence of auricular or nasal abnormalities. Flexible bronchoscopy demonstrated malacia of the tracheobronchial cartilage. EBUS revealed changes in the tracheobronchial cartilage, and facilitated the diagnosis.

Figure 1. Bronchoscopic views of stent placement before (top left, A) and after (top right, B) in a patient with RP (case 1). The openings for the left and right mainstem bronchi appeared dynamically collapsed before stenting. Two months after stenting, the airway is patent, and tracheal stent has been integrated into the tracheal wall. Bronchoscopic views before (bottom left, C) and after (bottom right, D) stent placement in case 2 show that the collapse of both mainstem bronchi is prevented and patency restored completely.
chial tree, with collapse of the airway on expiration (Fig 1, top left, A). Three-dimensional CT images demonstrated diffuse thickening of the tracheobronchial wall with a severely narrowed lumen. EBUS showed thickening of the bronchial wall due to submucosal edema, and the cartilage layer appeared ill-defined and absent in places; this continued along the trachea into both main bronchi (Fig 2, top left, A, and top right, B). Biopsy of the tracheal cartilage showed degeneration with fibrous changes and inflammatory cell infiltration. RP was diagnosed. The administrations of high-dose corticosteroids resulted in no improvement; therefore, it was decided to proceed with airway stenting.

Before stenting, we measured the diameter of the affected tracheobronchial tree using EBUS ensheathed with a balloon. Once water fills the balloon until it blocks the airway completely, EBUS provided a view of 360° so that we could evaluate the diameter. Through a flexible bronchoscope, we implanted four uncovered Ultraflex stents (Boston Scientific; Natick, MA) in the right mainstem bronchus (width/length: 10/20 mm), the left mainstem bronchus (10/40 mm), and the trachea (14/60 mm and 14/20 mm). After stenting, bronchoscopy showed that the stents were keeping the airway patent and were epithelized and integrated into the bronchial wall (Fig 1, top right, B). As a result, the patient reported improved dyspnea and returned to normal activities.

Case 2

A 61-year-old woman presented with sudden progressive dyspnea, necessitating emergency mechanical ventilation. After methylprednisolone pulse therapy, she was successfully extubated, but she remained dyspneic and required a tracheotomy. The severity of the dyspnea resulted in her being bedridden. Bronchoscopy showed severe narrowing of the trachea, extending into the mainstem bronchi and beyond (Fig 1, bottom left, C). EBUS revealed destruction of the normal cartilages, this continued along the central airway (Fig 2, bottom left, C, and bottom right, D). The hyperechoic third and fifth layers of the bronchial wall were indistinct, and the hypoechoic fourth layer was markedly swollen, indicating cartilage degeneration. Biopsy of tracheal cartilage confirmed chronic chondritis with plasma cell and lymphocytic infiltration. To maintain the airway, we implanted four uncovered Ultraflex stents in the right mainstem bronchus.
After Initiation of Anti-Tumor Necrosis Factor-α Therapy*

Chadi A. Hage, MD; Karen L. Wood, MD; Helen T. Winer-Muram, MD; Stephen J. Wilson, MD; George Sarosi, MD, FCCP; and Kenneth S. Knox, MD, FCCP

Pulmonary Cryptococcosis After Initiation of Anti-Tumor Necrosis Factor-α Therapy*

Chadi A. Hage, MD; Karen L. Wood, MD; Helen T. Winer-Muram, MD; Stephen J. Wilson, MD; George Sarosi, MD, FCCP; and Kenneth S. Knox, MD, FCCP

Many patients with rheumatoid arthritis are being treated with immunosuppressive regimens that include an agent directed at blocking tumor necrosis factor (TNF)-α. Although reportedly safe, tuberculous and fungal infections have emerged as significant complications of therapy. We report a case of pulmonary cryptococcosis soon after the initiation of therapy with the anti–TNF-α antibody, infliximab. A diagnosis was made early in the disease course, and the patient responded quickly to antifungal therapy. This case should alert clinicians to the increased incidence of pulmonary mycoses in patients receiving anti–TNF-α therapy.

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Key words: cryptococcosis; Cryptococcus neoformans; infliximab; tumor necrosis factor-α

Abbreviation: TNF = tumor necrosis factor

Pulmonologists often participate in the care of immunocompromised patients, as the majority of infections in this population involve the lungs. The immunosuppressive effects and infectious complications associated with the use of traditional cytotoxic agents are well known. The immunosuppressive effect of targeted anticytokine therapy, however, is just now being defined. Infliximab, a chimeric monoclonal antibody against tumor necrosis factor (TNF)-α, has revolutionized the care of patients with rheumatoid arthritis. Interestingly, the spectrum of infectious disease associated with the use of anti–TNF-α therapy is similar to that in patients with AIDS and includes tuberculosis,1 histoplasmosis,2 and cryptococcosis.3 This suggests that anti–TNF-α therapy produces a downstream defect in the T-helper type 1 arm of immunity. We report a case of pulmonary cryptococcosis after the initiation of infliximab. Symptoms resolved with discontinuation of infliximab and treatment with antifungal therapy.

Case Report

A 61-year-old man with a 6-year history of advanced rheumatoid arthritis was admitted to the Veterans Affairs hospital for shortness of breath and anemia. His medications included prednisone, 10 mg/d; methotrexate, 25 mg/wk; and leflunomide, 20 mg/d. He was started on infliximab and received three doses at 3 mg/kg ideal body weight, the last dose being administered 3 weeks prior to presentation.

He denied fever, chills, night sweats, chest pain, and weight loss. He denied any sick contacts or recent travel outside Indiana.

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