

# Relapsing Polychondritis-associated Refractory Airway Stenosis

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Relapsing polychondritis (RP) is a rare and debilitating rheumatic disease featuring recurrent and at times disseminated inflammation of cartilaginous structures, including auricles, eyes, peripheral joints, and airway<sup>1</sup>. Symptomatic airway compromise in RP is considered a common but severe cause of morbidity and mortality, which necessitates aggressive medical and bronchoscopic intervention<sup>2</sup>. We describe a refractory airway stenosis in a young patient with RP.

A 24-year-old man attended our clinic because of progressive pain and swelling in his left ear that had progressed to the right side with tenderness in his right eye for 1 year. He had been diagnosed with auricle chondritis with scleritis at a local hospital, and treated with oral azathioprine and elemental calcium. He was admitted to our hospital due to worsening symptoms of cough and hoarseness. Stridor and bronchial sounds could be detected upon examination. Chest radiograph showed no remarkable abnormalities (Figure 1). Subglottic stenosis of

70% was determined by an otolaryngologist. Laboratory evaluation was unremarkable except for elevated erythrocyte sedimentation rate (103 mm/h). RP was diagnosed based upon 3 separate anatomic sites of chondritis, negative antinuclear antibody, absence of vasculitis, intrathoracic neoplasm, and infectious disease. A chest computed tomography (CT) scan revealed thickening of the anterior and lateral walls of the airway from the vocal cord to bilateral main bronchi, associated with airway wall calcification and mediastinal lymph nodes (Figure 2). A 3-dimensional reconstruction confirmed these findings (Figure 3).

Because he had a poor response to pulse therapy (methylprednisolone plus cyclophosphamide) and maintenance therapy (low-dose azathioprine, cyclophosphamide, plus prednisolone daily), he underwent a rigid bronchoscopy and stenting of the left main bronchus (Figure 4A). A second examination by bronchoscopy showed collapse of the distal trachea and main carina (Figure 4B), and a moderate obstruction of the right main bronchus (Figure 4C). Collapsed left second carina and stenosis of left upper and lower bronchi were visualized (Figure 4D). In spite of intensive care, he subsequently died of pneumothorax, pneumonia, and septic shock at 5 months after his first visit.

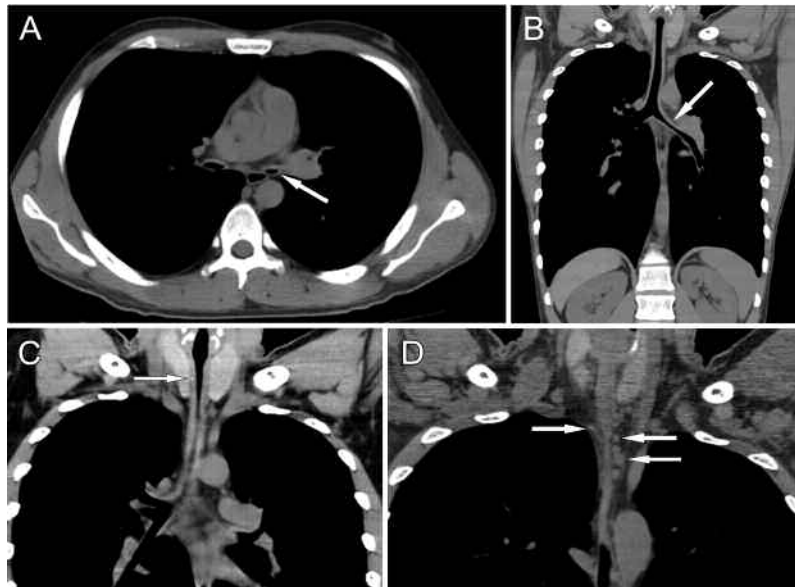
This case highlights that airway involvement in RP could become diffuse and rapidly progressive even if initial chest radiograph was normal. A further chest CT scan and subsequent 3-D reconstruction are valuable. Bronchoscopic balloon dilatation or stenting are optional modalities in medically refractory cases<sup>3</sup>.

## REFERENCES

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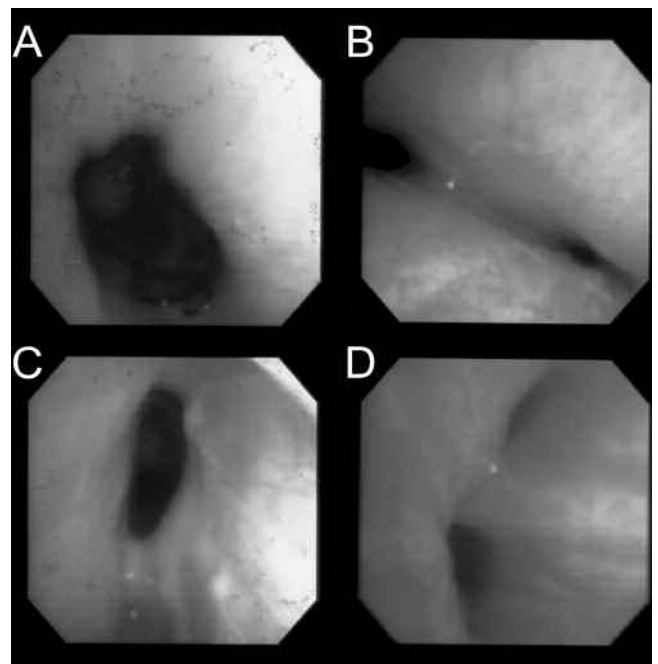
Figure 1. Initial chest radiograph showed no obvious abnormal findings.



*Figure 2.* Horizontal (A) and coronal (B) sections of a thoracic computed tomography scan revealed diffused thickening of the anterior and lateral walls with posterior wall spared, involving trachea, bilateral main bronchi, and right intermediate bronchus. Note the high-grade obstruction of the left main bronchus (white arrows). Airway wall calcification (C) and mediastinal lymph nodes with the largest diameter of 6 mm (D) are indicated by white arrows.



*Figure 3.* Three-dimensional reconstruction confirmed a long segment of high-grade stenosis in the left main bronchus (white arrow).



*Figure 4.* A non-covered self-expanding metal stent was implanted in the left main bronchus with a rigid bronchoscope (A). A second-look bronchoscopy showed collapsed distal trachea, main carina (B), and a moderate obstruction of the right main bronchus (C). Beyond the distal end of the stent, a moderate stenosis of left upper (lower) and a high-grade obstruction of left lower (upper) bronchi were observed (D).