CASE REPORT

Relapsing polychondritis: reversible airway obstruction or asthma

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ABSTRACT

We describe a patient who presented with poorly controlled asthma. Bronchoscopy showed collapsing airways, characteristic of RP (relapsing polychondritis).

Introduction

Relapsing polychondritis (RP) is a rare multi-organ disease characterized by recurrent inflammation of cartilaginous tissues (1). Respiratory tract involvement is seen in about 67% of patients throughout the course of the disease (1-9) and more than 75% of patients have a delay in diagnosis of more than one year (10). This case report aims at increasing the awareness of RP in a patient who presented with poorly controlled asthma, with a history compatible with RP. The airway obstruction in the initial stages of RP is due to airway inflammation rather than cartilaginous destruction and may respond to early, aggressive immunosuppressive therapy (1-9).

Case report

A 51-year-old non-smoking female with a background of adult onset asthma and recurrent anterior uveitis (iritocyclitis), presented with a 3-month history of hoarseness and worsening breathlessness. She had poorly controlled asthma unresponsive to multiple courses of steroids. Urgent bronchoscopy revealed severe dynamic collapse of intrapulmonary airways on inspiration. A retrospective physical examination revealed collapse of the nasal bridge, tenderness of the auricular cartilage and costochondral junctions and prolonged expiration wheezes. Flow-volume loop showed a very low flow rate during expiration due to severe expiratory flow obstruction (Fig. 1) and bronchodilator administration showed no improvement: forced vital capacity (FVC) was 3.40 L (110% of predicted); forced expiratory volume in 1 second (FEV1) was 1.65 L (60%); and FEV1/FVC ratio was 48%. Based on clinical features and bronchoscopy findings, a diagnosis of RP was made. Inflammatory markers and complement levels were normal. The immunological profile consisted of normal titers of antinuclear antibodies (ANA) and antineutrophil cytoplasmic antibodies (ANCA). CT scan of thorax showed small calibre trachea and proximal airways with bronchial wall thickening and a normal aortic root (Fig. 2). Our patient was initially treated with oral prednisone (1mg/kg/day), and was commenced on leflunomide. Currently, she is doing well with maintenance dosages of prednisone, 5 mg/d and leflunomide, 20 mg/d.

Discussion

Our patient had a history of poorly controlled asthma unresponsive to bronchodilators, with a history of inflammation of the cartilages of the ears and nose, and was previously diagnosed with recurrent anterior uveitis. Bronchoscopy revealed severe dynamic collapse of intrapulmonary airways on inspiration. Based on the history, clinical features, and bronchoscopy, our patient was diagnosed with RP according to the diagnostic criteria of McAdam et al. (2).

Relapsing polychondritis (RP) is a rare multisystemic disease characterized by recurrent inflammation of cartilaginous and non-cartilaginous tissues. When laryngotracheal or bronchial cartilages are involved, the disease can be life-threatening and needs aggressive treatment. There is no sex predominance, and the disease occurs at all ages, with a peak incidence between 40 and 60 years of age (2, 8).

The most common clinical manifestations are swelling and redness of the ears (88%) and arthralgia (81%). Episcleritis, iritis, hearing impairment, cataract, anemia, myocarditis, aortic valvular insufficiency, and glomerulonephritis have also been reported. Involvement of the larynx and trachea is present in 56% of the cases (1-9).

Respiratory tract involvement is seen in 14% to 38% of patients with RP at presentation and in 48% to 67% throughout the course of the disease (3-5). Respiratory symptoms represent the most serious manifestations. Inflammation of the laryngeal, tracheal and bronchial cartilages results in hoarseness, non-productive cough, dyspnea, wheezing, inspiratory stridor, and tenderness over the thyroid cartilage and anterior tracheal cartilage (4, 5). Occasionally, costochondral cartilage tenderness occurs.

Competing interests: none declared.
Effective cough is diminished because of cartilage collapse (4, 5). Airway collapse can lead to dynamic obstruction requiring emergency recognition and tracheostomy (11). Mortality from respiratory complications – mainly infections and tracheal collapse – has been reported to be between 10 and 50% (1, 4, 12). Lung function tests in laryngotracheobronchial involvement in RP reveal obstructive patterns typical of extrathoracic obstruction, intrathoracic obstruction, or both (13,14,15). Krell et al. showed that the intrathoracic obstruction was probably due to airway collapse as a result of bronchial cartilage destruction rather than to loss of elastic recoil (14).

Eye involvement is also frequent with approximately 20-30% of patients affected at the onset of disease and 50% affected during the course of the disease (16). All types of scleral inflammation, including necrotizing scleritis and posterior scleritis have been reported, however, diffuse anterior scleritis is the inflammation most commonly observed (16). Our patient had history of recurrent anterior uveitis.

Bronchoscopy is an essential diagnostic tool. It may reveal narrowing of the major airways or dynamic collapse and inflammation of the tracheobronchial tree (17). Fiberoptic bronchoscopy in our patient revealed severe dynamic collapse of intrapulmonary airways on inspiration.

CT is an important tool in the evaluation of patients with known or suspected RP. Lee KS et al. (18) reported that expiratory CT abnormalities were present in 94% of the patients with RP who were referred for airway imaging, yet only half of these patients demonstrated abnormalities on routine inspiratory CT scans. Thus, dynamic expiratory CT should be a standard component of imaging assessment in patients with RP. In patients with known disease, CT is useful for confirming intrathoracic involvement and for routine follow-up to assess progression of disease.

Early, aggressive immunosuppressive treatment of RP may delay or prevent irreversible cartilaginous destruction and airway collapse. There are no reports of complete reversal of cartilaginous destruction, and airway obstruction in response to immunosuppressive treatment of RP. Once the structural integrity of airway cartilage is lost, immunosuppressive therapy is inadequate, and airway stenting is needed to prevent total airways collapse.

In summary, RP is as rare as asthma is common. RP should be suspected in a patient who presents with asthma along with features of cartilaginous inflammation. Awareness of this condition is important to enable early diagnosis and therapy to reduce the risk of life threatening airway collapse.

Key message
Relapsing polychondritis may present as asthma unresponsive to bronchodilators.

References
6. RAPINI RP, WARNER NB: Relapsing poly-