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Tracheobronchial Involvement of Relapsing Polychondritis

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Abstract **Case Report**

Relapsing polychondritis is a rare disease that causes progressive and recurrent destruction of cartilage. We report a case of 38-year-old women with a medical history of asthma. A physical examination revealed a global expiratory wheeze and a saddle nose deformity. Chest CT showed marked thickening of the bronchial wall. The diagnosis of relapsing polychondritis is retained according to the criteria of Michet and Al and treatment was started with steroids. Airway involvement is the most severe and life-threatening aspect of the disease, and proves to be a therapeutic challenge.

Keywords: Relapsing Polychondritis (RP), Cartilage destruction, Autoimmune disease, Inflammatory disorder.

INTRODUCTION

Relapsing polychondritis is a rare multisystem disease of unknown etiology. However, the presence of fibrocartilagenous infiltration with CD4+ lymphocytes, immune deposits in tissue lesions, and elevated levels of autoantibodies against type II collagen suggest an immunologic mechanism [1]. The affected systemic cartilage and proteoglycan-rich tissues such as those of the ears, nose, larynx and trachea, articular cartilage, heart, blood vessels, inner ear, cornea, sclera and kidney, where recurrent inflammatory reactions occur. It usually begins in middle-aged individuals and is observed in all age groups [2]. The prevalence is approximately 4.5 cases per million, affecting all racial groups [3]. It is slightly more common in women, with a male-to-female ratio of 1:1.8 [4]. We report a 38-year-old woman with a diagnosis of early adult onset asthma.

CASE PRESENTATION

A 38-year-old woman with a past history of wheezing and stridor treated as asthma. Improvement was observed following treatment with steroids and antibiotics. Physical examination revealed global expiratory wheeze and saddle nose deformity (Figure 1). Further questioning revealed that she had been experiencing pain in her nose tree years ago.

CT of the chest revealed thickening and luminal narrowing of the wall of the lower third of the trachea extending to the main bronchi. This thickening respects the posterior wall (Figure 2). Bronchoscopy showed tracheobronchial wall inflammation and disappearance of tracheobronchial cartilage rings. A cartilage biopsy from nasal septum revealed non-specific inflammatory changes. CT of the sinuses showed left ethmoid-maxillary sinusitis without bone lysis and discreet thickening of the left lateral wall of the cavum. The pulmonary function test showed a severe irreversible airflow obstruction with a flattening of inspiratory limb. The diagnosis of relapsing polychondritis is retained according to the criteria of Michet and Al. The patient was treated with oral corticosteroids and Methotrexate 15 mg/week with good clinical progress.

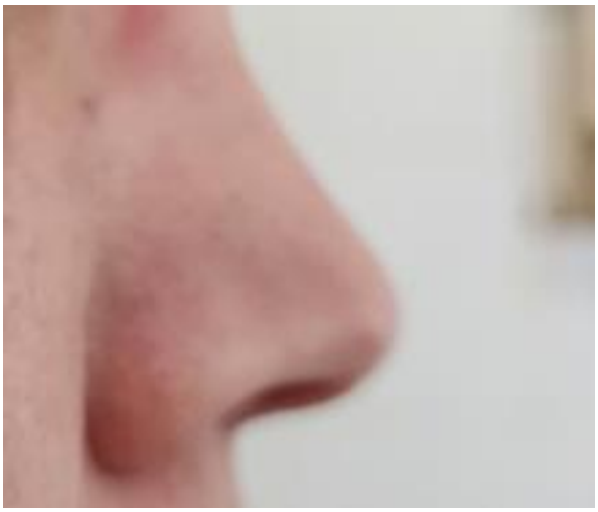

Figure 1

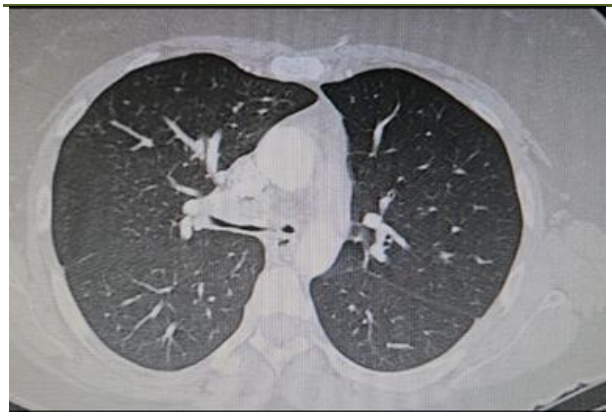

Figure 2

DISCUSSION

Relapsing polychondritis is a rare multisystem disease. Immune-mediated damage can spread to involve non cartilaginous tissues that are rich in proteoglycans, such as those in the eyes, inner ear, heart, blood vessels, and kidneys.

Up to 20-50% of patients will develop respiratory tract involvement, which is considered a poor prognostic factor. Airway involvement can cause airway edema and stenosis from airway inflammation, which can result in tracheobronchomalacia. It is responsible for a third of deaths among individuals with RP and the symptoms of this condition are dysphonia, inspiratory dyspnea, wheezing, and coughing [5].

There is no specific diagnostic test for RP. The diagnosis is usually based on clinical evidence and the elimination of differential diagnoses, primarily based on a combination of clinical features, radiographic findings, and/or biopsy of a cartilaginous site. Several clinical diagnostic criteria were developed for this disease. Hence, tissue biopsy is not always necessary if there is enough clinical evidence. McAdam *et al*., introduced the clinical criteria for RP in 1976 [6]. These clinical criteria were expanded upon by Damiani *et al*., in 1979 [7]. Michet *et al*., modified the criteria in 1986 [8].

Chest CT and pulmonary function tests (PFTs) are the primary modalities to detect airway involvement in patients with RP. The difference between relapsing polychondritis and other diseases is characterized by a saber-sheath-like morphology due to the lack of thickening of the posterior wall of the trachea, where no cartilage is present [9]. Pulmonary function tests and bronchoscopy should be indicated on an individual basis.

Positron Emission tomography with computed tomography (PET-CT) is a new diagnostic modality that seems to be helpful in early disease recognition especially in patients without organ involvement that is easily accessible for biopsy. This imaging modality also improves the assessment of the extent and disease activity during treatment [10].

The prognosis of the disease is related to the nature of organ involvement. Forms limited to inflammatory involvement of the cartilage of the nose or ears have a good prognosis. Involvement of the cartilage of the respiratory tract, cardiovascular involvement is detrimental to the prognosis of the disease.

Because of its rarity, randomized, placebo-controlled studies have not been conducted in patients with RP.

Systemic corticosteroid and immunosuppressive agents are administered as a standard treatment for RP; however, these medications can often be ineffective for airway involvement. Biologics such as infliximab and tocilizumab have been reported to be effective for RP, with the early use of biologics possibly preventing severe airway disorder [11]. TBM and laryngeal stenosis can be debilitating and life threatening, often requiring interventional procedures. Airway stenting has several critical complications, such as difficulty in expectoration, stent migration, and restenosis due to granulation.

CONCLUSION

RP is a rare and potentially fatal multisystemic autoimmune disease of unknown etiology, affecting primarily cartilagineous and proteoglycan-rich structures. To date, therapy of RP is still empiric, due to the lack of standardized guidelines on treatment, and is defined on the basis of disease activity and severity of organ involvement. Life-threatening or organ-threatening complications require high-dose corticosteroids and immunosuppressants. The early detection of airway abnormalities is important for the prevention of airway involvement in RP.

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