Case report

Late diagnosis of relapsing polychondritis without auricular chondritis, initiating with tracheal chondritis: a case report

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Submitted: 13 March 2023
Accepted: 20 March 2023

Arch Med Sci Aging 2023; 6: e9–e12
DOI: https://doi.org/10.5114/amsa.2023.126517

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Abstract

Introduction: Relapsing polychondritis (RP) is a rare autoimmune disease of unknown aetiology that is characterized by recurrent inflammation of cartilaginous structures as well as other tissues.
Case report: We report the case of a 56-year-old male with recurrent airway infections for 5 years, with nasal and tracheal chondritis, in whom a late diagnosis of RP without auricular chondritis was made.
Conclusions: The clinical manifestations of RP are wide, and knowledge of these symptoms can decrease diagnosis delays and time to start treatment, thus avoiding tracheal involvement-associated morbidity.

Key words: relapsing polychondritis, autoimmune diseases, rare diseases.

Introduction

Relapsing polychondritis (RP) is a rare autoimmune disease (ORPHA: 728) characterized by recurrent episodes of inflammation and deterioration of cartilage tissue, as well as other tissues in the body. Its prevalence and annual incidence are unknown, but it is estimated to have an incidence of 3.5/1,000,000 per year [1].

Clinically, it is heterogeneous and generally has an onset in the fifth decade of life (40–55 years), usually with inflammation of the cartilaginous portion of the auricle without lobe involvement, and with nasal chondritis associated with saddle nose deformity. Less frequently, it can present with laryngotracheal compromise associated with hoarseness, stridor, dyspnoea, or airway collapse [2].

The first reported case of polychondropathy was reported in 1923 by Jaksh-Wartenhorst, who described a case of a 32-year-old man with fever, pain, and auricular chondritis [3]. Later, in 1976, McAdam established the diagnostic criteria for this disease, in which 3 or more of the following are required: bilateral auricular chondritis, non-erosive seronegative polyarthritis, nasal chondritis, ocular inflammation, respiratory tract chondritis, and/or audiovestibular damage. These are essential for the classification of patients because the diagnosis is only clinical and there are no specific serological markers [4].

In this article, we describe the case of a patient with relapsing polychondritis, who presented with laryngotracheal involvement without auricular involvement.

Case report

A 56-year-old African American man, a resident of southwestern Colombia and an itinerant salesman, had a history of recurrent upper and lower respiratory tract infections for more than 15 years. He also had progressive dyspnoea requiring tracheostomy for 10 years due to subglottic stenosis. He consulted our centre due to a 30-day history of symptoms characterized by discharge of purulent secretions through the tracheostomy, asthenia, and hyporexia, with a subjective perception of temperature rises. He had no pain, haemoptysis, joint or skin involvement, or visual impairment.

Upon admission, his physical examination showed vital signs within normal ranges, highlighting the tracheostomy and endocannula without purulent secretions, loss of the left nasal wing without involvement of the auricular cartilage (Figures 1, 2), normal cardiopulmonary auscultation, and no other compromise found in the osteoarticular examination.

The blood count, creatinine, and urinalysis were normal. Studies for chronic infections were negative (HIV, hepatitis B and C, syphilis, Mantoux test, and serial smear microscopy). The chest X-ray was normal. Autoimmunity studies included antinuclear antibodies (ANA) 1 : 160 nuclear fine granular pattern (AC-4). Other antibodies were negative (anti-extractable nuclear antigens, anti-dsDNA, and anti-neutrophil cytoplasm), and the C3 and C4 fractions were normal. The erythrocyte sedimentation rate was 23 mm in the first hour, with a C-reactive protein (CRP) of 64.72 mg/l.

Scans of the paranasal sinuses, neck, and chest showed diffuse thickening of the nasopharynx, trachea, and main bronchi, along with occlusion of the right Eustachian tube (Figure 3).

A previous clinical history was reviewed, in which a biopsy of the nasal septum was documented with fibrosis and chronic plasmacytic infiltrate with degenerative changes in the cartilage, and a nasopharynx lesion with a fibrotic scarring process. It was considered to rule out IgG4 disease; however, the findings and immunohistochemistry did not favour IgG4 disease, and serum levels of IgG4 were not available.

Given the coexistence of nasal and tracheal chondritis with characteristic recurrence associated with audiovestibular compromise, a diagnosis of RP was proposed. Treatment was initiated with prednisolone 20 mg orally every day, methotrexate 5 mg orally weekly, colchicine 0.5 mg daily, and chloroquine 150 mg daily. During the follow-up, favourable clinical evolution was seen, with remission of the nasal inflammatory involvement, improvement of dyspnoea, and diminished tracheal secretions.

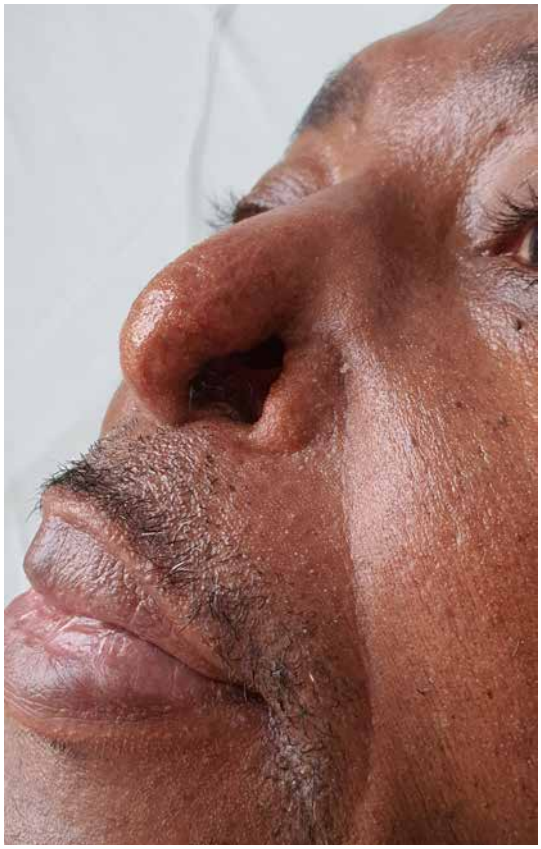

Figure 1. Deformation of the cartilage of the left nasal wing with septal perforation

Figure 2. Auricular pavilion without any inflammatory compromise

Discussion

RP is a rare systemic disease characterized by involvement of the auricular, nasal, and tracheal cartilage. Additionally, it may present with audiovestibular dysfunction, ocular inflammation, vasculitis, myocarditis, and seronegative arthritis [5]. Diagnostic criteria were established in 1970 by McAdam, which included 3 or more of the following findings: bilateral auricular chondritis, seronegative non-erosive inflammatory polyarthritis, nasal chondritis, ocular inflammation, cartilage inflammation of the respiratory tract, and/or vestibular or cochlear dysfunction [4].

The cardinal symptom in this patient was dyspnoea. Although there are no specific laboratory tests for diagnosis of RP, our patient presented 3 of the previously mentioned diagnostic criteria, as well as the typical histological findings [6]. Initially, our patient only had nasal and tracheal chondritis, without auricular involvement and with nonspecific symptoms such as fever and asthenia, resulting in a delayed diagnosis and treatment. Typically, a delay of approximately 10 years is reported in the literature, while in our case it was about 16 years [2].

There are 3 phenotypes described for RP, and the type 2 subgroup is characterized by predominantly lower airway involvement and less auricular involvement (43%), characterized by tracheomalacia (100%) and bronchomalacia (52%) [7]. Dyspnoea is a nonspecific symptom that is associated with a wide spectrum of diseases, and its origin is difficult to establish. Patients presenting with RP associated with dyspnoea may initially be diagnosed with asthma because they respond to short courses of steroids [8]. However, RP has a progressive course with relapses that worsen pulmonary function with laryngotracheal involvement, which increases morbidity and mortality, as in the case of our patient, who had been a tracheostomy user for 14 years, with multiple episodes of respiratory tract infection (pneumonia, tracheitis) [9]. Therefore, in patients with persistent symptoms of fever and dyspnoea, associated with multiple respiratory infections, without abnormalities in initial chest scans, RP should be considered among the differential diagnoses, because shortening the diagnosis time and early initiation of treatment avoids disease progression [10].

Pulmonary function tests were not performed at the hospital level due to institutional unavailability; however, it is important to evaluate the pattern of fixed airway obstruction in these patients. In addition, early dynamic computed tomography should be performed in these cases because it detects respiratory system complications [7].

Glucocorticoid therapy is a fundamental component in the treatment of RP and is a long-term indication for most patients. Depending on the magnitude of the involvement (multisystemic, severity), high-dose steroids may be required, along with additional immunosuppressive therapy [11]. Our patient was successfully managed with glucocorticoids, chloroquine, and methotrexate, which were started simultaneously to reduce the required doses of steroids, resulting in a subsequent improvement of the dyspnoea.

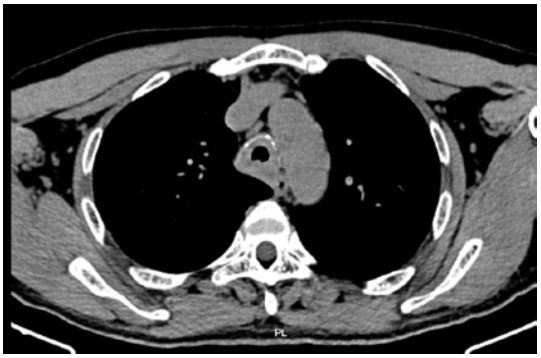

Figure 3. Tracheal chondritis with extensive compromise of the respiratory tract

Conclusions

We present the case of a patient with long-standing dyspnoea associated with recurrent respiratory tract infections, as well as subglottic stenosis that required tracheostomy. It was later determined to be the initial manifestation of relapsing polychondritis. Although auricular involvement is a common sign, according to the clinical phenotype of the patient, it may not be present. Therefore, physicians must be aware of the different clinical manifestations of this disease to achieve an early diagnosis and treatment.

Conflict of interest

The authors declare no conflict of interest.

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