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Relapsing Polychondritis Case: An Important Diagnosis Not to Be Delayed

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1. Abstract

Relapsing Polychondritis (RP) is a rare disease characterized by inflammation of cartilage and connective tissues with destructive episodes. Although the pathogenesis is not completely known, there is an autoimmunity in which antibodies against mainly type II collagen play a role. In addition to chondritis of the ear, nose, and trachea; organs having proteoglycan structure such as eyes, the inner ear, heart, blood vessels, and kidneys can be affected, too. The diagnosis is based on clinical signs, laboratory examinations, imaging techniques and histopathological examination. There has been no standart treatment protocol but, corticosteroids are mainstay treatment of acute flares. Immunosuppressive drugs such as methotrexate, azathioprine, cyclophosphamide, and cyclosporine and biologics are used effectively. Here, a case with RP was presented. We would like to raise awareness of this rare disease and remind the fact that it causes mortality besides severe morbidities, makes the diagnosis of this disease more important.

2. Introduction

Relapsing Polychondritis (RP) is a very rare autoimmune disease characterized by recurrent destructive inflammation of the cartilage of the body. The most commonly affected cartilages are the auricular, laryngo-tracheobronchial, and nasal [1]. RP has many clinical manifestations with variable presentation among patients and may lead to death as the diagnosis of RP is difficult [2]. Around 30% of these cases are associated with other autoimmune diseases including all types of vasculitis, Behçet's disease, rheumatoid arthritis, ankylosing spondylitis; systemic lupus erythematosus, chronic inflammatory bowel diseases and hematological diseases such as myelodysplastic syndrome [3].

3. Case

A 38-year old Caucasian male presented to the dermatology department with bilateral redness and thickening localized on auricle for almost 2 months. The lesions were painful and, appeared suddenly on the left ear without any preceding trauma, then in time both ears became puffy. The patient received multiple courses of antibiotics that resulted in no improvement of lesions. The patient also told a history of chest pain and shortness of breath 4 months ago, the main cause was not detected then, and it improved spontaneously. He is an active smoker and has 20 pack-year smoking history. However, there was no history of rheumatological or medical anomalies including arthralgia or arthritis, nor remarkable family history or similar disorders. Dermatological examination revealed remarkable erythema, swelling, and tenderness of the bilateral antihelices of the ears with sparing of the ear lobes (Figure 1). Moreover, a remarkable saddle nose deformity (Figure 2) was noticed. He was consulted with ENT, rheumatology, ophthalmolo-

gy, and cardiovascular disease departments. A hearing assessment revealed mildly decreased remarkable hearing loss. A biopsy was performed from auricular cartilage which was consistent with chronic inflammation (Figure 3). Laboratory evaluation including complete blood count and complete metabolic profile including liver and renal function tests was unremarkable. Rheumatoid factor, anti-cyclic citrullinated peptide antibody, antinuclear antibody, antineutrophil cytoplasmic antibody, complement levels, viral hepatitis serologies were all negative. The initial CRP was 15.99 mg/L. Detailed investigation of underlying malignancy or cartilage involvement was detected by 18F-fluorodeoxyglucose Positron Emission Tomography-CT (PET-CT). There was no detected malignancy. The ophthalmological examination and echocardiography was normal as well.

His condition had been stable after induction with 40 mg/day prednisolone for 4 days and with a 32 mg/day prednisolone maintenance dose for a week, then it was further tapered to 8 mg every day and 20 mg of methotrexate weekly for the past few weeks. He is being followed-up by rheumatology department without any flares.

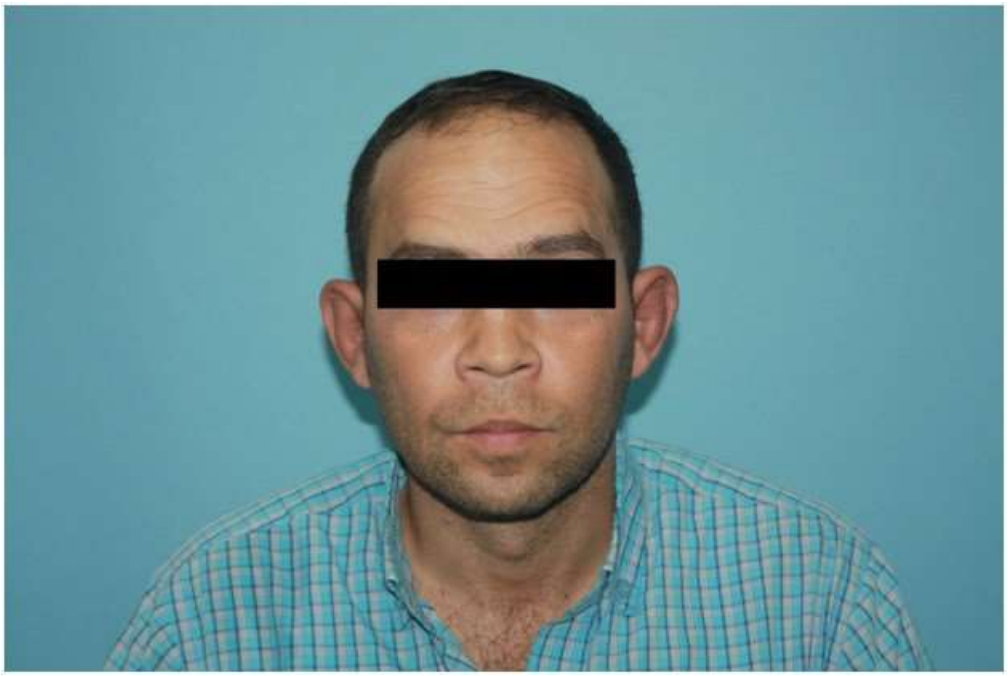

Figure 1: The appearance of auricular chondiritis of the patient with RP

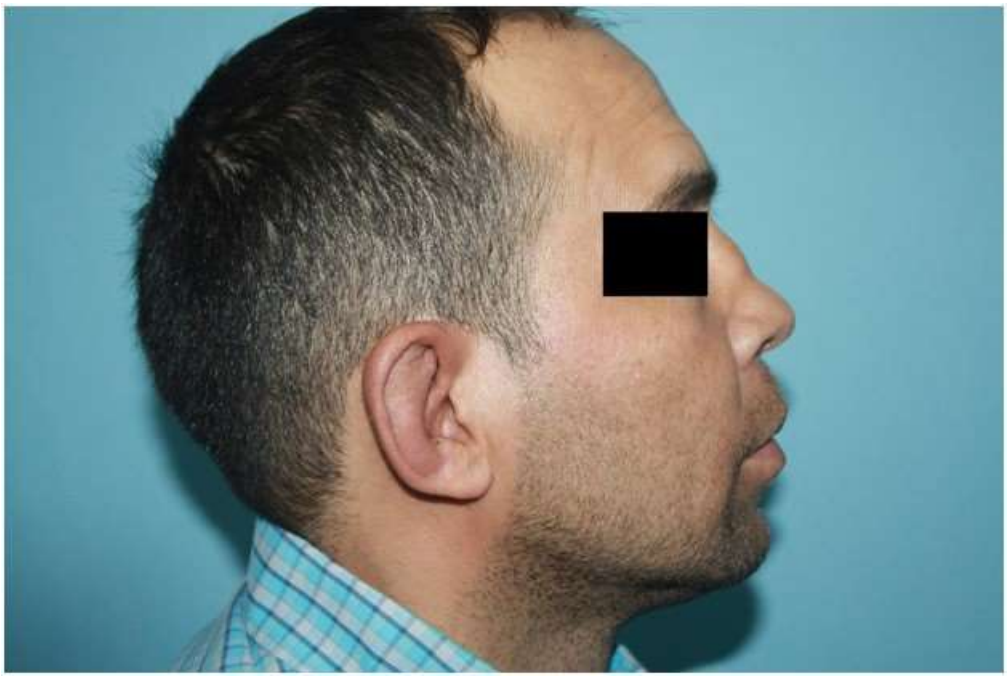

Figure 2: The appearance of nasal chondiritis of the patient with RP

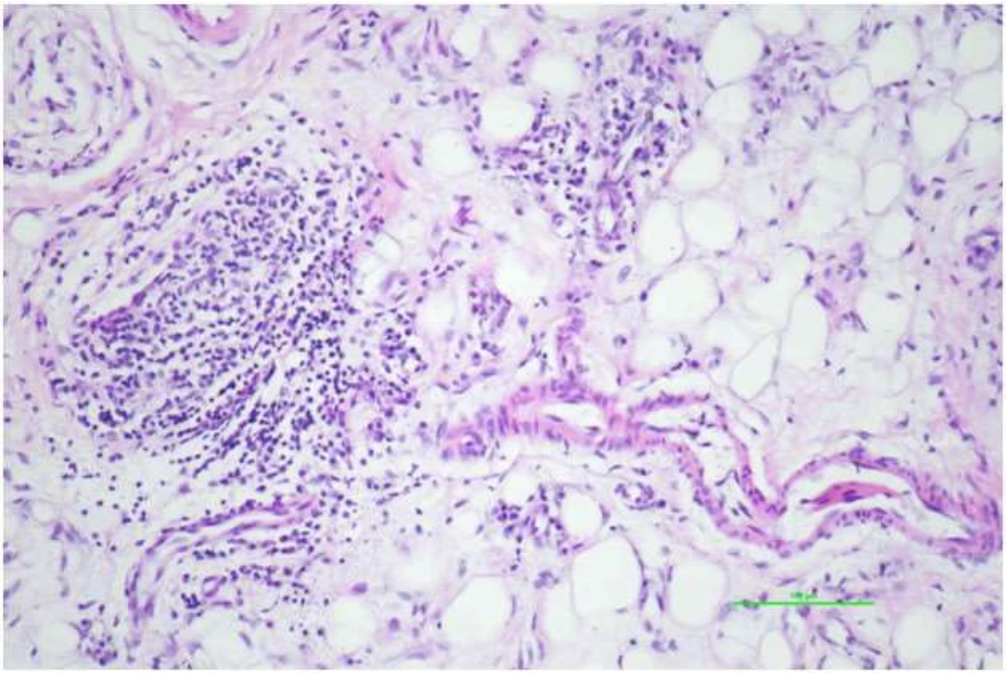

Figure 3: The histopathological findings showing the chronic inflammation in auricular cartilage of the patient with RP with H&E stain (x200)

4. Discussion

RP mainly affects middle-aged White men and women, without known sex predilection [4]. However, there have been reported RP cases in the pediatric population, too [5]. The incidence of RP is estimated to be 3.5- 4.5 cases per million people per year [2].

Auricular involvement is the most common feature of RP, but the auricular lobe, which has no cartilage, is typically spared. Other organs may also be inflamed such as costal cartilage, eyes, upper and lower airways, larynx, kidney, heart, and joints which include different types of cartilage structures that could be involved [6]. During the flares of RP, fever and fatigue could be seen [2]. PET-CT scanning may also show occult trachea-bronchial or aortic involvement and/or atypical clinical features of RP [7].

There are no specific laboratory tests for the diagnosis of RP. The diagnosis is based on some criteria including clinical findings, imaging methods, and occasionally a biopsy of the injured cartilage during acute flare. The original diagnostic criteria were the McAdam's criteria, in which three to six clinical features should be essential [3]. Later, Damiani and Levine proposed that the diagnosis of RP could also be made in patients meeting three of the McAdam's criteria even without histopathological confirmation or with one McAdam criterion if there is confirmed histological evidence of chondritis or when chondritis was found in two or more separate anatomic locations with the response to steroids and/or dapsone [8]. According to these criteria, in the present case, there was a confirmed histological result and chondritis in 2 separate anatomic locations with a apparent response to the steroid treatment.

There is no standard treatment of RP and it is mainly empirical, as the lack of randomized trials. The aim of treatment in RP is to reduce the frequency and intensity of flares and to prevent the development of irreversible damage [3]. Nevertheless, the corticosteroid is the mainstay treatment in acute flares and, immunosuppressive agents which have also known as steroid-sparing agents, such as cyclophosphamide, azathioprine, cyclosporine, methotrexate, and mycophenolate mofetil have been effectively used as maintenance treatments [3]. In our case, after the resolution of chondritis, prednisolone treatment was tapered and methotrexate treatment was started.

There are also case reports and case-series of RP patients treated with biological agents [3,9,10], but the number of clinical trials in this topic has not been enough. We would like to raise awareness of this rare disease and remind that RP could be a fatal disease rather than a cosmetic issue if there is a delay in establishing a diagnosis.

References:

1. Smylie A, Malhotra N, Brassard A. Relapsing Polychondritis: A Review and Guide for the Dermatologist. Am J Clin Dermatol. 2017; 18: 77-86.
2. Afridi F, Frosh S. Silent tracheobronchial chondritis in a patient with a delayed diagnosis of relapsing polychondritis. BMJ Case Rep. 2017; 2017: bcr-2017-220172.
3. Mathian A, Miyara M, Cohen-Aubart F, Haroche J, Hie M, Pha M, et al. Relapsing polychondritis: A 2016 update on clinical features, diagnostic tools, treatment and biological drug use. Best Pract Res Clin Rheumatol. 2016; 30: 316-33.
4. Hazra N, Dregan A, Charlton J, Gulliford MC, D'Cruz DP. Inci-

dence and mortality of relasping polychondritis in the UK: a population-based cohort study. Rheumatology (Oxford). 2015; 54: 2181-7.

5. Belot A, Duquesne A, Job-Deslandre C, Costedoat-Chalumeau N, Boudjemaa S, Wechsier B, et al. Pediatric-onset relapsing polychondritis: case series and systematic review. J Pediatr. 2010; 156: 484-9.
6. Lahmer T, Treiber M, von Werder A, Foerger F, Knopf A, Heemann U, et al. Relapsing polychondritis: An autoimmune disease with many faces. Autoimmun Rev. 2010; 9: 540-6.
7. Honne K, Nagashima T, Onishi S, Nagatani K, Iwamoto M, Minota S. Fluorodeoxyglucose positron emission tomography/computed tomography for diagnostic imaging in relapsing polychondritis with atypical manifestations. J Clin Rheumatol. 2013;19: 104-5.
8. Damiani JM, Levine HL. Relapsing polychondritis--report of ten cases. Laryngoscope. 1979; 89: 929-46.
9. Alqanatish JT, Alfarhan BA, Qubaiban SM. Limited auricular relapsing polychondritis in a child treated successfully with infliximab. BMJ Case Rep. 2019; 12: e227043.
10. Moulis G, Sailler L, Pugnet G, Astudillo L, Arlet P. Biologics in relapsing polychondritis: a case series. Clin Exp Rheumatol. 2013; 31: 937-9.