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Case Report

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Relapsing Polychondritis: A Case Report

Tekrarlayan Polikondrit: Bir Olgu Sunumu

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ABSTRACT

Relapsing polychondritis (RP) is an uncommon autoimmune disease characterized by repeated and progressive inflammation in cartilage tissue. The auricular cartilage is the location of the most common clinical findings and the most common site of initial involvement. Nasal chondritis is a manifestation present in 15% of patients. Progressive destruction of nasal cartilage leads to the characteristic flattening of the nasal bridge, resulting in saddle nose deformity. The case is, here, presented of a 38-year-old patient who developed nasal chondritis which resulted in saddle nose deformity, and was diagnosed with RP following rhinoplasty.

Keywords: Nasal chondritis; relapsing polychondritis; rhinoplasty; saddle nose deformity.

ÖZET

Tekrarlayan polikondrit kıkırdak dokularda tekrarlayan ve progresif inflamasyonla karakterize nadir otoimmün bir hastalıktır. Auriküler kıkırdak, en yaygın klinik bulgusu ve en sık başlangıç tutulum yeridir. Nazal kondrit hastaların %15'inde vardır. Nazal kartilajın yıkılması ile semer burun deformitesi gelişebilir. Burada, 38 yaşında nazal kondrit ile başlayan semer burun deformitesi gelişen ve rinoplasti sonrasında tekrarlayan polikondrit tanısı alan bir hasta sunuldu.

Anahtar sözcükler: Nazal kondrit; rinoplasti; semer burun deformitesi; tekrarlayan polikondrit.

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Relapsing polychondritis (RP) is an uncommon autoimmune disease characterized by repeated and progressive inflammation in cartilage tissue. It affects the ear, nose, respiratory tract and the proteoglycan-rich eyes, heart, and blood vessels.[1] The annual incidence has been reported as 3.5 cases per million[2] and it is generally seen in the 4th and 5th decades at equal rates in both genders.[3] Although the etiology is not fully known, autoimmune mechanisms against Type II collagen are thought to play a role.[4] Initial involvement of the disease is most often in the auricular cartilage, followed by joint involvement in the form of polyarthritis or oligoarthritis, and nose and eye involvement is observed later.[5] Nasal chondritis is a manifestation present in 15% of patients.[6] There may be concomitant systemic vasculitis, myelodysplastic syndrome, lymphoma, and other rheumatological diseases approximately in one-third of patients.[7] There may be concomitant systemic vasculitis, myelodysplastic syndrome, lymphoma, and other rheumatological diseases in a third of patients.[7] The case is, here, presented of a 38-year-old patient who developed nasal chondritis which resulted in saddle nose deformity and was diagnosed with RP following rhinoplasty.

Case Report

A 38-year-old female presented at our clinic with complaints of redness, pain, and swelling in the left ear. From the history, it was learned that there had been swelling in the left ear for approximately 1 month, and 1 week previously, there had been mild redness and swelling in the right ear (Fig. 1). There had also been pain and swelling in the nose on a total of three separate occasions, which had started 2 years previously. The patient

had been evaluated by the ear, nose, and throat clinic and had not responded to antibiotic treatment. Then, the bridge of the nose collapsed and saddle nose deformity developed. At that time, the patient was tested in respect of diseases such as granulomatosis polyangiitis, tuberculosis, and syphilis. Cultures were taken and the reports were negative. Rhinoplasty was performed on the patient. After the operation, swelling, pain, and redness again developed in the nose. There was nothing remarkable in the patient history, and no features in the rheumatological investigation. There was no arthralgia or arthritis. In the physical examination, there was edema, increased temperature, and sensitivity in the auricle. There was no involvement of the ear lobe. In the laboratory tests, sedimentation rate (79 mm/h) and C-reactive protein (15 mg/dl) were elevated. Other clinical parameters (urinalysis, thyroid tests, and liver function tests) were within the normal range. Antinuclear antibodies, antiphospholipid antibodies, antineutrophil cytoplasmic antibodies (ANCA), anti-Borrelia burgdorferi antibodies ımmunoglobulin (Ig)G/IgM, rheumatoid factor, anti-HIV, and VDRL tests were negative.

The tuberculosis cultures and hepatitis markers were negative, and the peripheral smear was normal. The thorax tomography results were normal and on magnetic resonance imaging, there was no laryngotracheal involvement. No ocular disorders were determined in the ophthalmological consultations. Transthoracic echocardiography and electrocardiography did not reveal any abnormalities. In the histopathological examinations made during the rhinoplasty operation, there was determined to be cellular infiltration by lymphocytes, neutrophils, and plasma cells, which was most evident in the cartilage-skin interface, and a reduced number of chondrocytes was seen in areas of cartilage destruction.

The patient was diagnosed with RP and treatment was started of 32 mg/day methylprednisolone (MP) and colchicine 1.5 mg/day. The patient used drugs for 1 week. As no response was obtained, pulse treatment with 250 mg/day. MP was started and applied for 5 days. The treatment was, then, continued with 64 mg MP and 15mg/week methotrexate (MTX) was added. After 2 weeks, the patient's complaints and acute phase responses improved (sedimentation rate: 33 mm/h and C-reactive protein: 4.5 mg/dl). The patient complaints and acute phase responses recovered. A schedule for tapering MP was given and to date, the treatment is ongoing at 4 mg/day MP and 15 mg/week MTX. Over a period of approximately 2 years, the patient has remained stable (Fig. 2).

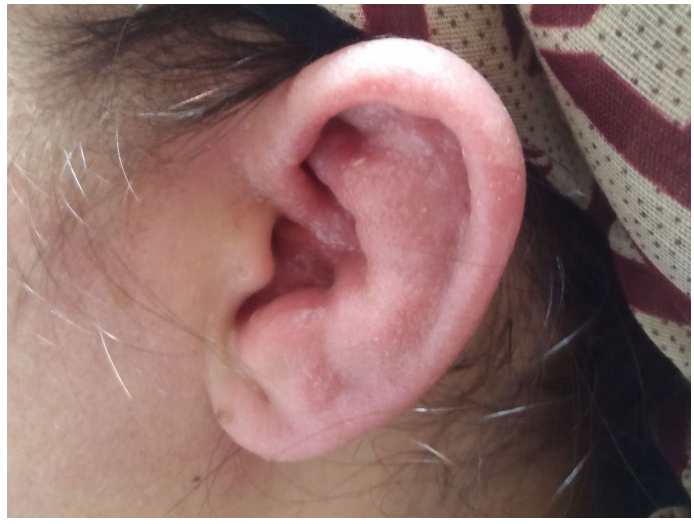

Figure 1. Auricular chondritis.

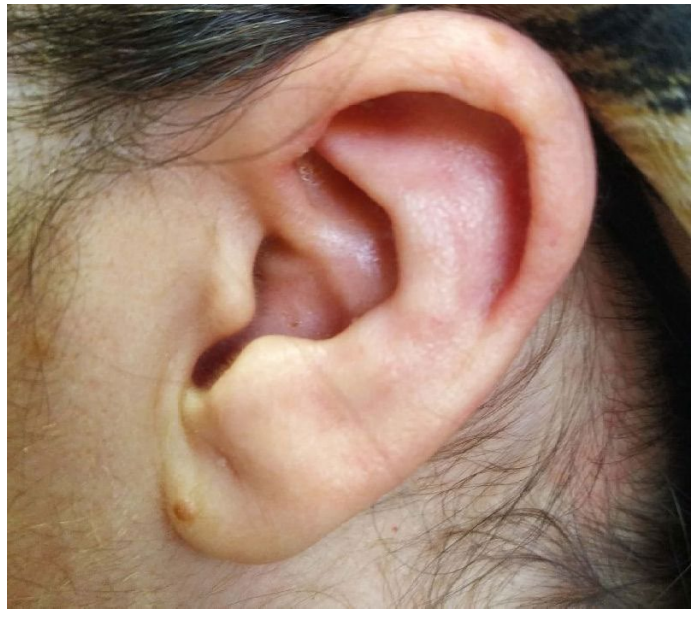

Figure 2. Auricula after treatment.

Discussion

RP is an uncommon but progressive autoimmune disease which can lead to fatal complications.[1] The most frequently involved site and the location of involvement at onset is the auricular cartilage. The ear lobe is not involved. Following attacks, cauliflower ear may develop associated with cartilage damage. The nose is a less involved location, but nasal cartilage involvement is painful. There may be a feeling of nasal obstruction, epistaxis, nasal discharge, and scabbing.[4] As in the current patient, saddle nose deformity can develop after repeated attacks. In this patient, auricular chondritis developed after saddle nose deformity and even after rhinoplasty.

Joint involvement is the common a manifestation of RP. Asymmetric, non-erosive, and seronegative oligo/polyarthritis may develop.[4] Our patient did not have joint involvement.

The presence of Behcet's disease findings such as oral and genital ulcers in addition to RP is called mouth and genital ulcers with inflamed cartilage syndrome.[4] Trauma, leprosy, syphilis, granulomatosis polyangiitis, RP, and Crohn's disease are included in the differential diagnosis of saddle nose deformity.[8] Before planning rhinoplasty, tests must be performed for these diseases. Diagnosis is made clinically and sometimes histopathological examination may be helpful. There is no specific laboratory test. Differential diagnosis with ANCA-related vasculitis is important.[4]

The treatment is generally symptomatic. There is no standardized treatment guideline. However, from experience gained from case series, the treatment is started with non-steroid anti-inflammatory drugs, colchicine, and dapsone in patients with mild symptoms, and steroids, azathioprine, MTX, and biological drugs can be used according to the severity of the patient. Infliximab has been reported to be of benefit in case series in patients with lung involvement in particular.[4] In the current patient, remission was achieved with steroids and MTX.

Mortality in RP is more than double compared with the general population, with the most frequent causes of death being respiratory, cardiac, and hematological involvement. Vasculitis, aortitis, anemia, male gender, uveitis, hearing loss, vestibular disorder, and the need for MP and cyclophosphamide infusion are criteria for a poor prognosis.[9]

Conclusion

RP is an uncommon, but progressive autoimmune disease which can lead to fatal complications. As in the current patient, especially in females, there may be presentation with saddle nose deformity and it must be emphasized that this should be kept in mind in the differential diagnosis.

Disclosures

Informed consent: Written informed consent was obtained from the patient for the publication of the case report and the accompanying images.

Peer-review: Externally peer-reviewed.

Conflict of Interest: None declared.

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