

Exacerbation of Relapsing Polychondritis After Renal Surgery: Case Report

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Abstract

Relapsing Polychondritis (RP) is a rare, progressive and multisystemic disease characterized by recurrent inflammation of cartilaginous tissues. In this report, we present a case of RP that consulted from a urologist due to complaints with bilateral hearing loss, tinnitus and bilateral auricular hyperemia three days after renal surgery. A physical examination revealed thickening of both external auditory canals, with edema and erythema of the auricles; bilateral nasal crusting and hyperemia of mucoperichondrium with pain were also present.

Keywords: Relapsing polychondritis, auricular cartilage, sensorineural hearing loss, differential diagnosis.

Renal Cerrahi Sonrası Şiddetlenen Tekrarlayan Polikondrit: Olgu Sunumu

Öz

Relapsing polikondrit; kırkırdak ve bağ dokuda inflamasyon ile karakterize, nadir görülen, ilerleyici ve multisistemik bir hastalıktır. Bu makalede renal cerrahiden 3 gün sonra başlayan bilateral işitme kaybı, tinnitus ve bilateral auriküler hiperemi şikâyeti ile yönlendirilen tekrarlayan polikondrit vakası sunulmaktadır. Fizik muayenesinde her iki aurikulada ödem ve

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eritem ile birlikte dış kulak yolunda kalınlaşma, burun kabuklanması, mukoperikondriumda hiperemi ve ağrı gözlenmiştir.

Anahtar Sözcükler: Tekrarlayan polikondrit, kulak kıkırdağı, sensörinöral işitme kaybı, ayırıcı tanı.

Introduction

Relapsing polychondritis (RP) is a progressive immune-mediated, inflammatory connective-tissue disease affecting the cartilage of the ear, nose, joints, tracheobronchial tree, cardiovascular system, skin, kidney, central nervous system, and various proteoglycan-rich tissues¹. RP usually manifests as a fluctuating and progressive course resulting in a significant shortening of life expectancy². There is no exact cause and etiology of RP; some triggering factors, such as cancer, drugs, viral infections, cartilage trauma are discussed in the literature³. In this paper, we report a case of RP that developed three days after severe renal surgery. The patient showed evidence of some criteria of RP plus bilateral severe-to-moderate sensorineural hearing loss (SNHL) and tinnitus.

Case History

A 45-year-old female was referred from a urinary clinic because of complaints of bilateral hearing loss with tinnitus and bilateral auricular hyperemia. It was her first attack. She also complained of nasal crusting, visual disability, and marked irritation with photophobia in both eyes. She had no arthralgia, cough, hoarseness, or fever. A physical examination revealed thickening of both external auditory canals, with edema and erythema of the auricles; bilateral nasal crusting and hyperemia of mucoperichondrium with pain were also present (figure 1,2,3).

Figure 1: Edema and erythema of the right auricle



Figure 2: Edema and erythema of the left auricle



Figure 3: Nasal crusting and hyperemia of mucoperichondrium



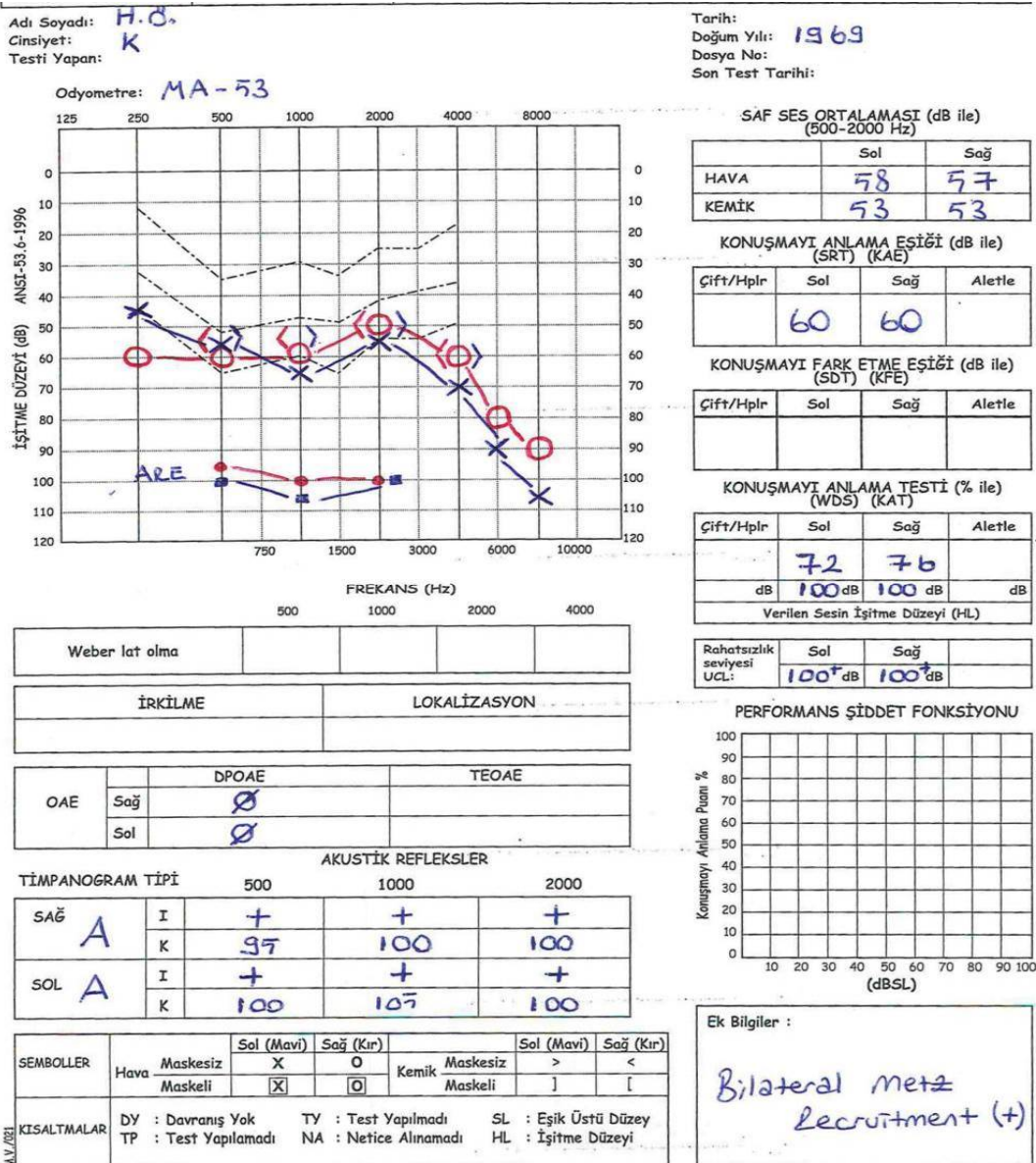
Ocular examination revealed posterior uveitis (posterior chamber activity with hypopyon in both eyes). Visual acuity was 40/100 in both eyes. Other ocular findings were normal. An endoscopic examination of the oropharynx and larynx was normal. Bronchoscopy, abdominal ultrasonography, thorax-computed tomography, and cardiological evaluations were normal. There was a bilateral moderate-to-severe SNHL involving all frequencies (figure 4).

All laboratory studies, including the erythrocyte sedimentation rate, C-reactive protein level, full blood count, renal and liver function tests, antinuclear antibody, rheumatoid factor, HLA-B27, HLA-B51, antineutrophil cytoplasm antibodies, and serological tests were normal. Under these examination findings, a diagnosis of RP was made on the basis of Mc Adam's criteria⁴, including auricle chondritis, nasal chondritis,

seronegative, ocular inflammation, and audiovestibular damage. She has no history of any other cartilage disease, such as systemic disease, and no history of musculoskeletal symptoms.

A high-dose prednisone (250 mg daily for five days) was administered. The patient responded well to the therapy after five days of treatment. All complaints, except hearing loss and tinnitus, were improved. She was discharged from the hospital one week after treatment.

Figure 4: Sensorineural hearing loss



Discussion

RP characterized by relapsing attacks of symptoms is very rarely seen in autoimmune disease involving cartilage and proteoglycan rich structures, such as the joints, eyes, auricula, inner ear, blood vessels, heart, kidneys, and tracheobronchial tree¹. Although the inflammation of the auricular cartilage, nose, joints, airways, and ocular conjunctiva are commonly seen, the cause of RP is not clear. However, some triggering factors, such as cancer, drugs, viral infections, cartilage trauma are discussed in the literature³. In this case, the RP symptoms were seen just after the surgical operation, and it was the patient's first attack. This suggests that surgical stress may be another triggering factor in these cases.

The disease appears with a wide spectrum of symptoms, depending on the involved tissue; therefore, the diagnosis of RP is very infrequent at the first attack unless there is cause to suspect. The mean time for diagnosis is nearly 2.9 years after the first attack⁴. There are some different diagnostic-defined criteria in the literature, such as McAdam's criteria⁴ (which is the first one described), Michet et al. criteria⁵, and Damiani and Levine criteria⁶. Others are modifications of McAdam's criteria, including the following clinical features where three of them are sufficient for a differential diagnosis 1) nasal chondritis, 2) bilateral auricular chondritis, 3) audiovestibular damage, 4) nonerosive seronegative inflammatory polyarthritis, 5) respiratory tract chondritis and 6) ocular inflammation. McAdam's criteria were used to diagnose our case, which presented four of six clinical features. Because of the sudden development of many clinical symptoms just after the surgical operation, the diagnosis of RP was an easy one. However, although polyarthritis is the most common symptom of RP, there was no sign of arthritis.

The treatment of RP is changeable, depending on the severity of the symptoms. In mild cases, low-dose oral corticosteroid and anti-inflammatory drugs are used. In severe cases, the patient is treated with a high-dose corticosteroid⁷. In cases of ineffective corticosteroid treatment, immunosuppressive drugs, including methotrexate, are the second choice of treatment⁷. Infliximab (a chimeric antitumor necrosis factor- α antibody), etanercept (a soluble tumor necrosis factor α receptor), and tocilizumid (an anti-interleukin-6 receptor antibody) are described by Caribon and Kavari^{8,9}. Our case responded well to high-dose corticosteroid therapy. Most of the symptoms were

improved, except SNHL and tinnitus. The severity of hearing loss and tinnitus did not change after treatment. This showed that, even though treatment modalities for RP are effective in many clinical features, high-dose corticosteroid therapy may not be effective if the inner ear is involved. The prognosis of the inner ear is not good.

Conclusion

RP is a very rare entity and difficult to diagnose at the first instance of an attack. RP is characterized by recurrent episodes of inflammation involving various cartilaginous structures. There are many suspected triggering factors that may exacerbate RP. Surgical trauma and stress may be some triggering factors in RP. Because of multiple organ involvement, various different, nonspecific manifestations are seen. Therefore, when examining patients with these different symptoms, we have to keep RP in mind.

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