

## Relapsing Polychondritis: Inflamed Joints and Ears

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**Background:** Relapsing polychondritis (RP) is an episodic and progressive inflammatory disease of the cartilaginous structures, including elastic cartilage of the ear and nose, hyaline cartilage of the peripheral joints, fibrocartilage at axial sites, and cartilage of the tracheobronchial tree. The spectrum of its presentations may vary from intermittent mild episodes of chondritis to occasional organ involvement or even life-threatening manifestations.

**Case Report:** We presented a 64 year-old male patient with bilaterally knee arthritis and discoloration of pinna.

**Conclusion:** There is lack of awareness about this disease due to its rarity. With this case presentation, our goal was to draw attention to this disease, which could be delayed for the diagnosis.

**Keywords:** Arthritis, chondritis, relapsing polychondritis

Relapsing polychondritis (RP) is an episodic and progressive inflammatory condition involving cartilaginous structures, predominantly those of the ears, nose, and laryngotraeobronchial tree (1). The spectrum of its presentations may vary from intermittent mild episodes of chondritis to occasional organ or even life-threatening manifestations. There is a lack of awareness about this disease due to its rarity. With this case presentation, our goal was to draw attention to this disease, which could be delayed for the diagnosis.

### CASE PRESENTATION

A 64 year-old male patient was referred to our department with bilateral knee pain. He also gave a several years history of episodic ear swelling, tenderness, tinnitus, and recurrent conjunctivitis. On physical examination, besides the bilateral knee arthritis, discoloration and deformity of the pinna were identified (Figure 1). In his medical history, similar recurrent arthritis, swelling, and erythema in his external ear were present. The test results were negative rheumatoid factor, anti-nuclear antibody (ANA), Antineutrophil cytoplasmic anti-

bodies (ANCA), and revealed elevated levels of erythrocyte sedimentation rate and C-reactive protein. A diagnosis of RP was made on this clinical basis and he was started on oral steroids. His score was evaluated as 21 according to the "RP disease activity index", which was developed recently by a worldwide panel of RP experts (2). He was also evaluated for myelodysplastic syndrome and other malignancies due to his weight loss. In his follow-up, methotrexate was added to the treatment regimen due to his poor response to steroids. Written informed consent was received from the patient for his medical data publishing.

### DISCUSSION

Relapsing polychondritis was first described by Jaksch-Wartenhorst in 1923, and the term "relapsing polychondritis" was first used by Pearson et al. in 1960 in their case series (3). Since then, the information on the clinical spectrum, pathogenesis, and management has grown considerably. It is a rare condition, with an estimated annual incidence as 3.5 cases per million in a certain area (1). The peak age of

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**FIG. 1.** Discoloration and deformity of the pinna and damaged cartilaginous structure due to repeated attacks

onset is the fifth decade of life, though cases have been reported in both extremes (3). Despite its unknown etiology, the pathogenesis seems to be an immunologic reaction to type II collagen, and studies have shown the association of both cellular immunity changes and abnormal autoantibody response in RP (1).

In RP, auricular and vestibular involvement is present in most patients. Due to the inflammation in the cartilaginous portion of the pinna, pain, discoloration, or tenderness in this area is frequently seen as the initial symptom. As in our patient, after repeated attacks, the cartilaginous structure of the ear is damaged and assumes a nodular or verrucous appearance, loses shape, and becomes soft and flabby (1, 3). Symptoms of vestibular dysfunction, as well as impaired hearing, occasionally occur. Joint pain is the second most common clinical feature of RP. Though any joint may be involved, metacarpophalangeal, proximal interphalangeal, and knee joints are the most commonly affected. In our case, knee joints were repeatedly affected during the course of the disease without any destruction. The joint involvement in RP is usually episodic, nonerosive asymmetric oligoarthritis, or polyarthritis and lasts for several weeks (1). Respiratory tract involvement is also seen frequently. Nasal chondritis may initially cause pain and tenderness, and the repeated inflammatory process might destroy the nasal cartilage leading to a saddle nose deformity (3). Laryngotracheal involvement by RP is a major cause of morbidity and mortality and is seen in about 50% of patients (1). Complications include destruction of the thyroid cartilage, acute upper airway collapse, and obstruction necessitating emergency tracheostomy. Cardiovascular involvement is seen in 24 to 52% of the cases, and is the second most frequent cause of death in patients with RP. The features vary widely and can include vasculitis, valvular heart diseases, and rhythm

disturbances (3). Although renal involvement is not frequent in RP, it is potentially lethal. It may be a part of an associated disorder (e.g. SLE or vasculitis) or may be primarily due to the disease itself. Ocular involvement (e.g. scleritis, episcleritis, and conjunctivitis) and dermatologic symptoms (e.g. erythema nodosum, erythema multiforme, panniculitis, purpura, livedo reticularis, and multiple neurologic abnormalities consistent with vasculitis) can occur during the course of RP (1). In around 30% of the cases, an association with other diseases especially systemic vasculitis or myelodysplastic syndrome is detected (3).

The diagnosis of RP needs to be based on clinical findings alone. The criteria described by McAdam et al. (4) have been used to confirm the diagnosis of RP (4). As the initial symptoms are often non-specific, the diagnosis may be delayed, with the mean delay from the first presentation to the time of diagnosis being estimated as 2.9 years (1). The role of laboratory investigations is purely supportive and to rule out other related or associated systemic diseases. Additionally, histopathological examination may not be useful since no biopsy finding is pathognomonic of RP.

Due to the relative rarity of RP, the treatment strategy is still largely empiric and needs to be based on case reports. Less severe symptoms, such as mild auricular or nasal chondritis and arthralgia, are generally treated with nonsteroidal anti-inflammatory drugs. Organ-threatening disease, including severe polychondritis, ocular or laryngotracheal involvement, systemic vasculitis, and glomerulonephritis, require systemic corticosteroids and different immunosuppressants, such as methotrexate, cyclophosphamide, azathioprine, cyclosporine, and mycophenolate mofetil (1, 3). After adding methotrexate (15 mg/week) to the treatment regimen due to the poor response to steroids in our patients, he was episode-free during his one-year follow-up. Given the autoimmune theory of etiopathogenesis of RP, a number of biologics are also being used increasingly in patients who do not respond to other medical therapy with favorable results (3). However, in a review on the biologics in RP, it was concluded that the experience with biologics in RP is very limited and its real efficacy and indications need to be better defined (5).

The prognosis varies according to involvements. The majority of patients with RP manifest a fluctuant course with intermittent inflammatory episodes. With the better management of the complications associated with RP, the survival has improved from 55% at 10 years in 1986 to 94% at the end of eight years in 1998 (1).

In conclusion, RP is a rare, multisystem, and potentially fatal disease. An increased awareness of the disease may have a considerable impact on the course of this relatively rare multi-system connective tissue disease.

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**Informed Consent:** Written informed consent was obtained from patient who participated in this case.

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