A case of Relapsing Polychondritis : Unmasking the Otitis Externa Mimic

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Case presentation

A 74 year old Caucasian male patient with past medical history significant for peripheral vascular disease and chronic right sensorineural hearing loss presented to clinic with complaints of right ear pain, swelling, erythema and tenderness for 14 days despite treatment with multiple antibiotics and NSAIDs. On detailed history taking he reported that he has been having these identical symptoms intermittently for almost seven months. He has been seen multiple times in urgent care and walk-in clinics where he was given alternating courses of treatments with topical, oral antibiotics, topical steroids and NSAIDs with only minimal resolution of symptoms.
He denied fatigue, arthritis, rash, or fever. There were no reports of trauma or abrasion.
He had chronic sensorineural hearing loss in the same ear.
No allergies or immunocompromised state.
Unremarkable family history.

Objective Data

- Pertinent findings on physical exam include stable vital signs, afebrile, not in distress, normal voice.
- He has diffusely erythematosus, tender, swollen right ear pinna and external canal sparing the lobe.
- Left ear, nose and eyes looked normal. Labs showed elevated
- CRP of 100 mg/L (normal range <3 mg/L) and ESR of 200 mm/hr (normal <20 mm/hr).
- ANA titer is 1: 160 with a homogenous pattern but negative autoantibodies.
- No red flags on CBC nor CMP.
- RPR is negative.

Diagnosis

Our patient was diagnosed with Relapsing Polychondritis as he has satisfied the diagnostic criteria per Damiani and Levine et al.,(Fig 2) with his auricular chondritis and sensorineural hearing loss accounting for two out of the six clinical features outlined by McAdam, plus a positive response to treatment with prednisone.

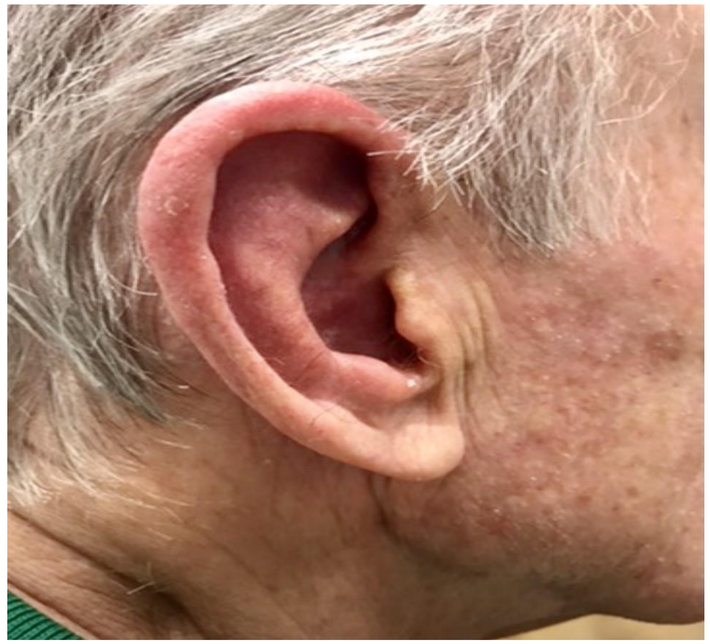

Figure. 1. Pathognomonic auricular swelling sparing the ear lobe manifested by the patient.

AUTHORS	DIAGNOSTIC CRITERIA
McAdam et al.	Positive for at least 3 out of 6 clinical features 1. Auricular chondritis 2. Nonerosive inflammatory polyarthritis 3. Chondritis of nasal cartilage 4. Ocular inflammation-Scleritis/uveitis/conjunctivitis 5. Inflammation of respiratory tract 6. Cochlear and/or vestibular damage-Conductive/ sensorineural hearing loss, tinnitus, vertigo
Damiani and Levine et al.	At least 1 out 6 features outlined by Mcadam et al. + Histological confirmation Or 2 out of 6 features outlined by McAdam et al. + Positive response to either dapsone or corticosteroids
Michet et al.	Inflammation confirmed in two out of three cartilages (auricular, nasal, or laryngotracheal) Or Inflammation has been proven in one of these cartilages, along with two additional minor criteria, such as hearing loss, ocular inflammation, vestibular dysfunction, or seronegative arthritis.

Figure. 2. Diagnostic Criteria for Relapsing Polychondritis.

Outcome and Follow-up

Prednisone 60mg daily was started as monotherapy. Rheumatology was also consulted. The auricular chondritis improved promptly. Steroid was slowly tapered then maintained on 10mg daily which prevented flare ups. Inflammatory markers subsequently trended down to normal after initiation of corticosteroids.

Discussion

RPC's exact etiology remains unknown, though up to one-third of patients demonstrate circulating antibodies against type II collagen, correlating with disease activity. Peak age of onset is 40 to 50 years with an estimated annual incidence of 3.5 cases/million people. It typically manifests in the fifth decade of life, with a slight female predominance. Auricular chondritis is a hallmark of the disease, often leading to progressive cartilage destruction and deformity if untreated. Both conductive and sensorineural hearing loss are also prevalent among RPC patients due to structural damage and inflammation of the ear. Diagnosis relies heavily on clinical criteria established by McAdam, Damiani & Levine, and Michet et al., which emphasise the presence of auricular chondritis, non-erosive inflammatory polyarthritis, nasal chondritis, ocular inflammation, respiratory tract chondritis, and audiovestibular damage.
Management of RPC is challenging due to the disease rarity and the limited clinical trials available. Treatment strategies are primarily derived from case reports. Low-dose glucocorticoids are commonly used for mild symptoms, while severe cases may require immunosuppressants like cyclophosphamide, methotrexate or cyclosporine combined with corticosteroids.

Conclusion

This case underscores the importance of early diagnosis and aggressive management to prevent irreversible damage in RPC. Continuous monitoring and adjustment of therapy are crucial to maintaining remission and minimizing the impact of this debilitating disease.