Recurrent Episcleritis as an Atypical Initial Presentation of Relapsing Polychondritis

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1 Introduction

- Relapsing polychondritis (RP) is a rare autoimmune condition causing recurrent inflammation of cartilaginous and proteoglycan rich structures.
- We present a case of RP with a rare first presentation of recurrent episcleritis.

2 Case Presentation

- A 35-year-old woman presented to the rheumatology clinic with recurrent episcleritis that began during pregnancy four years earlier. She was referred from an ophthalmology clinic for further evaluation of a suspected underlying rheumatic disease.

Clinical Features

- **Ocular Symptoms:** Episodes of redness, photophobia, and tearing occurring every six to seven months.
- **Auricular Symptoms**: Three episodes of painful ear swelling during pregnancy (after episcleritis), initially treated as otitis externa with antibiotics.

3 Clinical & Laboratory Assessments

Past Medical History	• Appendicectomy at the age of four, premature osteoporosis, hypertension.
Family History	• No history of autoimmune or connective tissue diseases.
Laboratory Tests	• Elevated ESR & CRP during episodes.
Imaging & Specialist Evaluations	• Ophthalmological Assessment: Confirmed recurrent nodular episcleritis. • Rheumatological Evaluation: Identified characteristic auricular chondritis, leading to the diagnosis of RP.



Figure 1: A close-up image showing redness and inflammation of the episclera, during one of the patient's episcleritis episodes.



Figure 2: Swelling of the auricular cartilage with sparing of the ear lobe.

4 Treatment Approach:

- **Initial Management**: The patient was treated with corticosteroids, under view to immunosuppression, but the therapy was discontinued due to steroid-induced glaucoma.
- **Steroid-Sparing Therapy**: She was transitioned to azathioprine, which provided sustained control of inflammation while minimizing adverse effects.
- **Long-Term Outcomes:** Over time, the patient improved with minimal flares, remaining active with no respiratory or systemic complications.

5 Discussion

Diagnostic Challenges

Our patient initially presented with episcleritis, and although ocular involvement occurs in up to 65% of RP cases, it is rarely inaugural. Chondritis remains the hallmark of RP, present at 60% of cases at onset, commonly affecting the pinna and nasal root.

- **Variability in Presentation:** Some patients initially present with non-auricular symptoms such as polyarthritis, bronchial stenosis, sudden deafness, or prolonged fever. Thus, a delay in diagnosis can continue until chondritis appears.
- **Diagnostic Approach:** RP diagnosis primarily relies on clinical features. Biopsies may also be used. Various clinical criteria have been proposed (see Table 1). A 2018 study comparing diagnostic criteria found the Damiani and Levine criteria (a modification of McAdam's) to have the highest sensitivity. Additionally, inflammatory markers and antibodies may support diagnosis; however, they lack both sensitivity and specificity.

Table 1 Diagnostic criteria

McAdam et al. [10]	(1) Recurrent chondritis of both auricles (2) Nonerosive inflammatory polyarthritis (3) Chondritis of nasal cartilages (4) Inflammation of ocular structures (5) Chondritis of respiratory tract (6) Cochlear and/or vestibular damage (3 out of 6)
Damiani and Levine [13]	(1) 3 out of 6 McAdam et al.'s criteria (2) 1 out of 6 McAdam et al.'s criteria + a positive histologic confirmation (3) 2 out of 6 McAdam et al.'s criteria + response to corticosteroid/dapsone (any of these)
Michet et al. [6]	(1) Inflammation in 2 out of 3 cartilages: auricular, nasal, and laryngotracheal (2) Inflammation in 1 of the above and meeting 2 other signs ocular inflammation, hearing loss, vestibular dysfunction, or seronegative inflammatory arthritis (any of these)

(Rose et al., 2018)

Table 1: Different diagnostic criteria proposed for relapsing polychondritis, highlighting key clinical manifestations and diagnostic thresholds.

Differential Diagnoses:

Differentiating RP from other conditions is critical. Unlike RP, auricular inflammation due to trauma or infection is typically unilateral and can involve the earlobe. However, conditions such as granulomatosis with polyangiitis (GPA) can mimic RP with bilateral auricular chondritis. In such cases, GPA-specific markers like anti-PR3 and MPO-ANCA help distinguish between the two.

Management and Treatment

Management of RP is often empirical, guided by disease severity and treatment outcomes.

- Mild flares can be managed using NSAIDs.
- Other cases may need corticosteroids or immunosuppressants like azathioprine.
- Outcome is then evaluated.eg; the aim in auricular involvement is to prevent deformity. The aim in respiratory involvement is to prevent complications like respiratory insufficiency and to consider the risk of medication-associated infection

Conclusion

- **Atypical Presentations Matter:** Recurrent episcleritis can be an early, uncommon sign of relapsing polychondritis (RP), warranting further exploration.
- **Recognizing RP:** Although RP is a rare condition it should be recognized across specialitites like opthalmology, otorhinolaryngology and Pulmonology given the variability of its presentations.
- **Improving Patient Outcomes :** Increased awareness and timely treatment can lead to better disease control and quality of life for affected individuals.

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