SKIN

BRIEF ARTICLE

Relapsing Polychondritis Masquerading as Auricular Pseudolymphoma: Case Report and Literature Review

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ABSTRACT

Relapsing Polychondritis (RP) is a rare autoimmune disease associated with recurring inflammatory episodes predominantly affecting cartilage and other tissues throughout the body, including proteoglycan-rich structures. It is characterized by gradual deformation of tissue that leads to impairment of normal function. Areas involved include, but are not limited to, the respiratory tract, eyes, nose, joints, and vascular system. The inflammation targets cartilage, most commonly causing auricular and nasal chondritis but can involve cartilage throughout the body. Diagnosis of RP is often hindered by the vast variety of symptoms associated with this systemic disease and subtle symptomatology. In this paper, we demonstrate a case of relapsing polychondritis masquerading as pseudolymphoma and review recent literature relating to pathogenesis, diagnosis and treatment.

INTRODUCTION

Relapsing Polychondritis (RP) is an immune-mediated disease characterized by recurring inflammatory episodes predominantly affecting cartilage and other tissues throughout the body.[1] It is a rare diagnosis that is associated with the gradual deformation of the tissues' anatomical structures which then leads to the deterioration of normal function.[2] It is noted to affect less than 5,000 individuals in the United States and the peak incidence is in the fifth decade of life.[3,4] Areas involved include but are not limited to, the respiratory tract, eyes, nose, joints, and vascular system. Greater than 80% of patients with RP have auricular cartilage involvement.[5] Although it is thought to be genetically inclined, this condition occurs in association with other autoimmune diseases in about 30% of the patients affected, most commonly rheumatoid arthritis.[5] Clinical presentation is varied and can often be subtle, which can make diagnosis challenging. Patients often complain of unspecific symptoms including malaise, joint pain, and dyspnea or dry cough from airway involvement.[6]

Diagnosis can be determined based on finding a pattern of clinical signs and symptoms that are coherent with RP, but no one pattern is certain to be seen. These can

include inflammation or pain in cartilaginous areas, such as the ear.[7] Tinnitus and hearing loss can be found in patients with otologic manifestations.[8] Since there are no definitive laboratory tests to diagnose RP and is a diagnosis of exclusion. This includes assessing all related signs and symptoms, which can delay diagnosis. The differential diagnosis of ear lesions includes chondrodermatitis nodularis helicis, perichondritis, trauma, pseudolymphoma, and auricular pseudocyst. When systemic symptoms are present granulomatosis with polyangiitis and rheumatoid arthritis should be considered. After ruling out all other possibilities, patients with RP benefit from symptomatic treatments to help manage existing symptoms.[3] In this case, we report a subtle case of relapsing polychondritis in a patient with single auricular involvement and report current updates in pathogenesis, diagnosis, and treatment of the condition.

CASE REPORT

A 56-year-old Asian female presented to the dermatologist with a five-month history of itchy, painful, and swollen left ear. A previous skin biopsy demonstrated nodular lymphocytic infiltrate with concerns for lymphoma versus pseudolymphoma. The patient was seeking a second opinion due to a lack of clinical improvement with topical therapy and continued pain. The patient denied other systemic symptoms.

Physical exam revealed a flesh-colored plaque with diminishing skin lines on the left antihelix, concha, and helix (**Figure 1**). Biopsies were performed and histology showed plasma-rich nixed infiltrate with degeneration of marginal chondrocytes and fibrosis positive for relapsing polychondritis. Direct immunofluorescence had focal granular IgG, C3, and C5b-9 along the fibrocartilaginous junction consistent with relapsing polychondritis. Bloodwork slightly positive ANA at a titer of 1:80 with a mitotic intercellular bridge pattern. Other autoimmune antibodies were negative.

The patient was treated with prednisone 10 mg taper for 9 days. She also received 3 injections of 5 mg/1cc intralesional triamcinolone along the area of inflammation totaling 1cc. At one month follow-up the patient had significant relief of the pain and pruritus. In addition, she was started on oral doxycycline 50 mg daily for maintenance therapy. In her six-month follow-up, she reported significant improvement in her symptoms and decreased swelling of the area (**Figure 2**). At that time the doxycycline was discontinued, and she was instructed to follow-up if symptoms worsened.

DISCUSSION

Accurate clinical impressions provided on requisition forms can play a vital role in arriving at the correct histopathological diagnosis. The main findings of this study demonstrate that the use of the phrase “rule out eczema” by clinicians encompasses a wide array of conditions with varied etiologies, such as atopic dermatitis, nummular eczema, dyshidrotic eczema, contact dermatitis, neurodermatitis, seborrheic dermatitis, mycosis fungoides, psoriasis, and tinea infections. The breadth of this term’s usage underscores the importance of clearly indicating the clinical impression and differential diagnosis being considered before sending a biopsy to the dermatopathologist for interpretation. Imprecise terminology compromises patient care and may result in dermatopathologists rendering incorrect diagnoses.

Figure 1. Physical Examination. A flesh-colored plaque with diminishing skin lines on the left antihelix, concha, and helix.

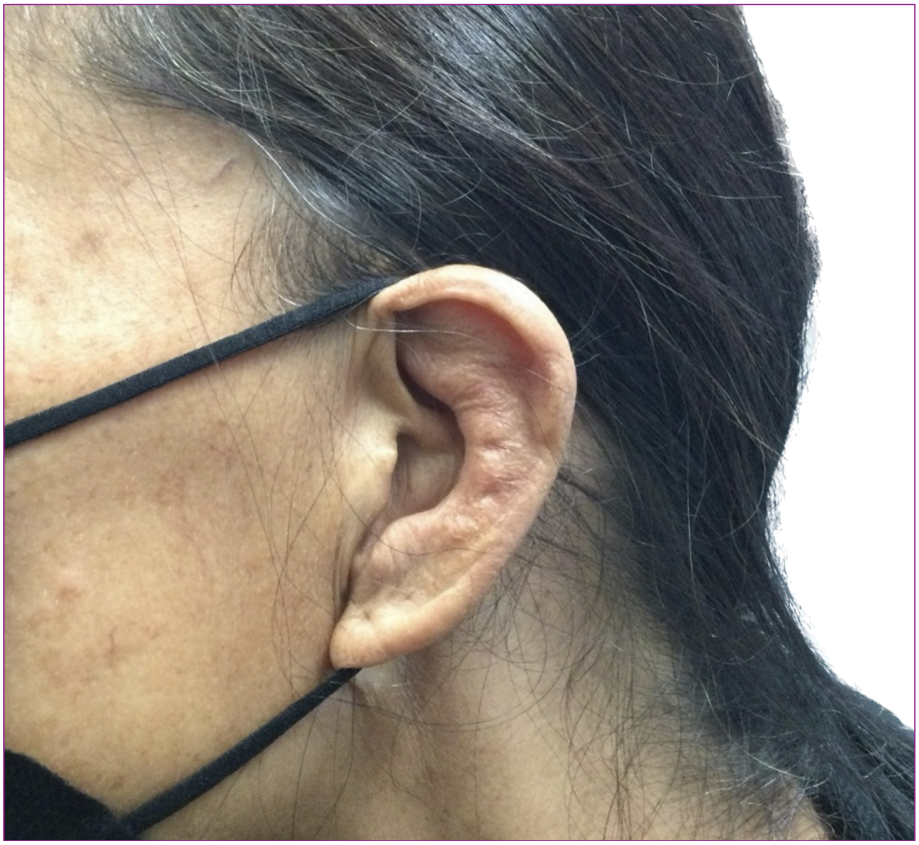

Figure 2. Six-month follow-up after systemic treatment reveals improvement in edema and inflammation. There is residual scarring present from biopsy sites.

"Eczema" is a descriptive morphological term rather than a specific condition and includes a variety of dermatological conditions that histologically present with spongiosis. It is frequently used interchangeably with "atopic dermatitis" since AD is the most common form of eczema. Despite the word "atopic", it is worth noting that approximately 60% of children who exhibit clinical signs of atopy do not show IgE-mediated sensitivity to allergens.[2] This discrepancy and resulting ambiguity prompted the World Allergy Organization to suggest a change in terminology, wherein "eczema" is used as a general term for skin conditions with certain clinical and genetic features, and "atopic dermatitis" is used for skin conditions with an IgE-associated process. Furthermore, eczema without signs of atopy is common, with studies reporting a prevalence of 45-64% in children and 40% in adults.[3] Therefore, even though the majority of respondents in our study include atopic dermatitis in their differential diagnosis, if eczema is colloquially used synonymously with atopic dermatitis, there may be a tendency to overlook other types of eczema with distinct etiologies that are not characterized by atopy, such as contact dermatitis or nummular eczema. The differentiation between AD and eczema is further complicated by the fact that ICD-9 and ICD-10 codes for AD are distinct from those for eczema, potentially leading to systematic coding errors that can impact billing, reimbursement, and medical research.[4]

In a 2013 survey study distributed among dermatologists and dermatology residents,[5] approximately one-third of the participants somewhat agreed with the statement that they were reluctant to add clinical information to requisition forms because they did not want to bias the dermatopathologist. Similarly, about one-third somewhat agreed that pathologists should make a diagnosis without clinical information. However, the requisition form serves as a vital document facilitating transition of care between clinicians and pathologists and carries significant implications for the accuracy of biopsy interpretations and clinicopathologic correlations. This is particularly evident in requisition forms sent to "rule out eczema," given that spongiosis is a histologic feature that is not specific to any single dermatosis. As such, histologic features alone may often be inadequate for a definitive diagnosis. The lack of specificity is especially problematic when the biopsy requisition form does not include accompanying clinical images, pertinent patient medical history, provider notes, or personal modifications to automated EMR phrases. In the absence of such clinical details, pathologists must rely exclusively on the information present in the requisition form. Unfortunately, the standardized format of many requisition forms may inadvertently replace the descriptive narrative that is often crucial for accurate diagnosis, especially in the absence of clinical photographs. The reasons for not including additional information might be linked to the time constraints faced by busy clinicians with high patient volumes,6 variability in the level of training or clinical experience among the personnel tasked with filling out the requisition form, or possibly a lack of awareness regarding the importance of providing a clear clinical impression or differential diagnosis on pathology requisition forms.

It should be noted that the findings of this study, which are based on self-reported data from a national sample of dermatology clinicians, may be prone to selection bias and may not be entirely representative, as the study did not include participants from every state. The validity of our results may also be impacted by non-response bias, considering potential differences between respondents

and non-respondents. Nevertheless, our findings highlight the importance of establishing an agreement on the proper nomenclature for eczematous or spongiotic dermatoses, especially with regards to enhancing communication between clinicians and pathologists. The use of non-specific terms such as “rule out eczema” on biopsy requisition forms can lead to broader differential diagnoses, which may increase the risk of misdiagnosis or diagnostic delays due to lack of specificity in the biopsy requisitions, thereby potentially delaying appropriate treatment and affecting patient outcomes.

Ambiguous phrases like “dermatitis unspecified,” often generated by EMR programs, offer limited value and thus should not be provided to clinicians when submitting biopsy specimens. Furthermore, the term “rule out eczema” is nonspecific, and conditions may not be readily distinguished based on histology alone. To enhance diagnostic accuracy, it is recommended that the phrase be discarded in favor of specifying which disorder the clinician is presumptively diagnosing clinically.

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