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**Case presentation**

**An uncommon presentation of an uncommon disease: relapsing polychondritis overlap with systemic lupus erythematosus**

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**Abstract**

Relapsing polychondritis (RP) is a rare rheumatologic disorder in which recurrent episodes of inflammation result in destruction of cartilage of the ears and nose. The joints, eyes, audio-vestibular system, heart valves, respiratory tract, kidneys, and skin can also be involved. Skin involvement is most frequently linked to concomitant myelodysplastic syndrome and has rarely been associated with systemic lupus erythematosus.

A 47-year-old woman presented with violaceous, indurated, tender plaques on the bilateral cartilaginous ears with sparing of the lobes, consistent with RP. Further investigations revealed positive ANA and anti-Smith antibody, oral ulcers, a photo-distributed skin eruption, and biopsy-proven lupus nephritis, leading to a second concomitant diagnosis of systemic lupus erythematosus (SLE). The diagnosis of SLE associated with RP was made and the patient was started on oral prednisone and hydroxychloroquine.

This is a rare report of SLE associated with RP. It is unclear whether RP occurring in patients with SLE represents another clinical manifestation of SLE or a coexisting disease. However, a significant ANA titer in a patient with RP strongly suggests the presence of an associated autoimmune disorder. If immunologic abnormalities usually found in SLE are detected in patients with RP, it is important to monitor patients for the development of other manifestations of SLE.

**Keywords: Relapsing polychondritis, systemic lupus erythematosus, ANA**

**Introduction**

Relapsing polychondritis (RP) is a rare, episodic, and progressive inflammatory disease of presumed autoimmune etiology affecting the cartilage in multiple organs, including the ear, nose, larynx, trachea, bronchi, and joints. In addition, it can affect proteoglycan-rich tissues, such as the eyes, aorta, heart, kidneys, and skin [1]. Up to 50% of patients with RP can have tracheobronchial disease and serious sequelae from involvement of these tissues can include laryngeal, tracheal, and/or bronchial

obstruction. Other described sequelae associated with RP include an asymmetric non-deforming, non-erosive arthritis, scleritis or uveitis, and renal disease—from parenchymal lesions or vasculitis. Cardiac involvement can result in aortic aneurysms and valvular heart disease [1].

Additionally, RP causes many nonspecific skin disorders, and dermatologic symptoms are present in 50% of patients with RP [1]. The frequency of skin involvement in RP in the absence of an associated disease is approximately 35% but doubles in the presence of another systemic illness [1]. Aphthosis, nodules on the limbs, and purpura are the most common signs of skin involvement [2]. Skin involvement in RP is most frequently linked to myelodysplastic syndrome amongst other associated systemic illnesses [2]. However, RP has only been rarely associated with SLE [3].

## Case synopsis

A 47-year-old woman presented to the hospital with an 8 month history of daily fevers, malaise, and ulcerations along the hard palate and vermillion border of her upper and lower lips. The ulcers had been worsening during this time and increased oral bleeding was reported. Her past medical history was remarkable for symmetric polyarthralgias previously diagnosed as “rheumatoid arthritis (RA)” owing to a positive rheumatoid factor (RF), along with a persistent microcytic anemia. She also had a painful left ear draining serous fluid that had remained unresponsive to antimicrobial treatment.

Upon hospitalization, the cutaneous exam also revealed an erythematous and violaceous asymptomatic exanthem involving the back, chest, face, and proximal arms. She was febrile and coupled with a concern for hemoptysis, she had a CT scan of the chest, which demonstrated ground glass opacities. She was empirically started on vancomycin and piperacillin/tazobactam for possible pneumonia. She tested negative for active tuberculosis and an HIV test was negative.

Further history elicited symptoms of painful erythematous lesions on the palms, odynophagia, and chronic persistent xerostomia over the last 8 months.

She did not have any painful discoloration of finger or toes with cold exposure (Raynaud phenomenon), nor any skin tightening or thickening. The eruption on her chest and back was asymptomatic. She had noticed scarring alopecia on her scalp as well as easy loss of hair diffusely with gentle traction, brushing, and showering. She also had unintentional weight loss of 22 lbs. despite a good appetite and supplementation with high calorie nutritional shakes.

On physical exam, she was noted to have violaceous, indurated, warm, tender plaques with crusting on the bilateral cartilaginous ears with sparing of the lobes (Figure 1). She also had violaceous indurated thin plaques with scale crust on her eyebrows, cartilaginous nose (sparing the columella), and malar cheeks that did not blanch (Figure 2). On her chest and back there were erythematous, non-tender, edematous, blanching plaques most prominently on the sun-exposed upper chest, sparing her arms and legs (Figure 3). On her oral mucous membranes, there were ulcerations along the hard palate and vermillion border of her upper and lower lips. Her scalp had atrophic, crusted plaques with carpet-tack like follicular plugging at the right temporal/vertex scalp along with diffuse non-scarring alopecia with positive hair pull test.



**Figure 1.** Auricular chondritis of the left ear. There are violaceous, indurated plaques with crusting limited to the cartilaginous portion of the external ear, with sparing of the lobe. **Figure 2.** Nasofacial rash. There are violaceous, scaly plaques on her cartilaginous nose sparing the columella, and malar cheeks.



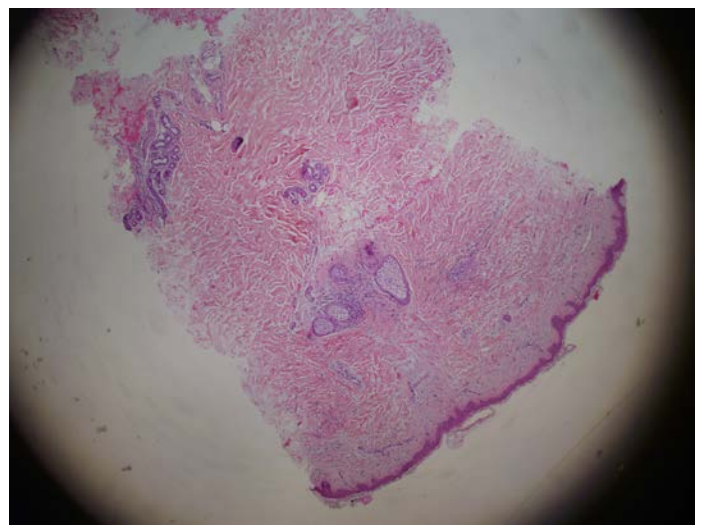
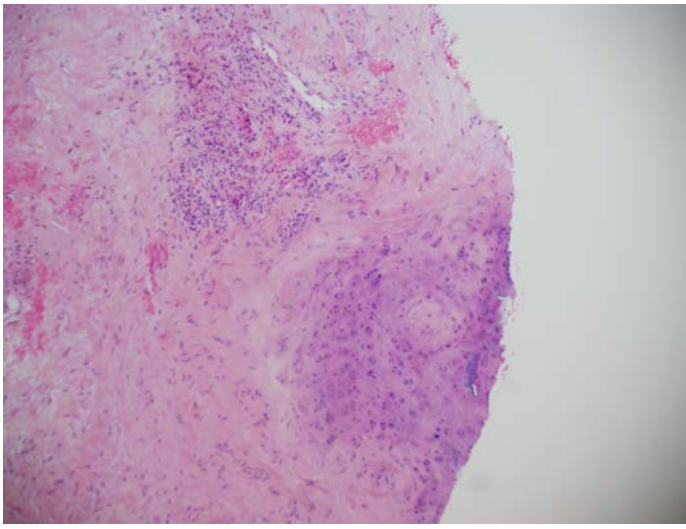
**Figure 3.** Photo-distributed rash. There are erythematous plaques on the sun-exposed upper chest.

It was noted that both of her ears were swollen and inflamed, involving the cartilaginous parts and sparing the lobe/pinna, prompting a dermatology consultation. Pulmonary evaluation and subsequent bronchoscopy did not reveal any airway inflammation or cartilaginous involvement.

Labs were remarkable for a high LDH, elevated ESR, and low complement levels (C3 and C4). Although the ANA (1:640), anti-SSA, anti-Smith, and anti-U1-RNP antibodies were positive, the anti-DS DNA (<1:10), anti-Scl70, anti-centromere, and anti-CCP antibodies were all negative. Urinalysis showed proteinuria 1.3 grams and microhematuria. Urine drug screen, Hepatitis B surface antigen, Hepatitis C antibody, and HIV antibody testing were negative. Biopsy of the ear showed a predominantly lymphocytic inflammatory infiltrate (with occasional histiocytes and rare eosinophils) adjacent to the ear cartilage, blurring the interface between soft tissue and cartilage--consistent with active relapsing polychondritis (RP) (Figure 4). Along with the chondritis, there was erosion of the epidermis along with superficial and deep perivascular and periadnexal lymphocytic inflammation. These findings suggested connective tissue disease concurrent with the chondritis.

Biopsy of the eruption on the chest supported the diagnosis of cutaneous lupus. Biopsy demonstrated a thinned epidermis, with a slightly thickened basement membrane, subtle interface dermatitis, interstitial mucin, and perivascular and periadnexal lymphocytic infiltrate consistent with connective tissue disease (Figure 5). Renal biopsy showed thrombosis, medullary tuft necrosis, mesangial deposits without mesangial hypercellularity or matrix expansion, consistent with grade 2 lupus nephritis.

The diagnosis of systemic lupus erythematosus (SLE) was met based on criteria from the American College of Rheumatology including: kidney involvement (grade 2 lupus nephritis per renal biopsy; proteinuria; microhematuria), positive ANA, positive anti-Smith antibody, oral ulcers, photosensitivity, and a photo-distributed discoid skin eruption. The diagnosis of SLE associated with RP was made and the patient was started on oral prednisone and hydroxychloroquine.



**Figure 4.** Chondritis, with lymphocytic inflammatory infiltrate adjacent to the ear cartilage, blurring the interface between soft tissue and cartilage **Figure 5.** Chest biopsy showing thinned epidermis, interstitial mucin, and perivascular and periadnexal lymphocytic infiltrate consistent with connective tissue diseases.

## Discussion

Relapsing polychondritis is a rare disease most commonly presenting as inflammation of the cartilage of the ears. Auricular chondritis is the most common initial finding. Acute involvement of the tracheal cartilage may cause collapse of the airway with obstruction and pulmonary infections. Other manifestations include audio-vestibular damage, asymmetric, non-deforming, non-erosive arthritis, scleritis or uveitis, heart valve disease, and renal disease.

Relapsing polychondritis may also be associated with other rheumatologic or autoimmune conditions. According to McAdam et al, 25-35% of patients with RP have a concurrent autoimmune disease [4]. However, RP has only been rarely associated with SLE [3,5]. Upon hospital admission, the patient in the present case was found to meet clinical criteria for SLE.

Although ANA positivity can be seen in RP patients without any associated disorder, the prevalence of a positive ANA in RP is low [6]. Finding a significant ANA titer (>1:100) in a patient with RP strongly suggests the presence of an associated autoimmune disorder. Associated conditions include acquired myelodysplasia, Sjogren's syndrome, mixed connective tissue disease, or SLE [6]. It is unclear whether RP occurring in patients with SLE represents another clinical manifestation of SLE or a coexisting disease. In our ear biopsy, the infiltrate at the cartilage was consistent with RP, whereas the superficial and deep perivascular and periadnexal lymphocytic infiltrate was consistent with cutaneous lupus. Along with the changes in the chest, the histology supports the clinical picture of both cutaneous lupus and co-existing RP. If immunologic abnormalities usually found in SLE are detected in patients with RP, it is important to monitor patients for the development of other manifestations of systemic lupus in addition to the other known complications of RP.

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