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DOI: [10.1159/000141960](https://doi.org/10.1159/000141960)**Relapsing Polychondritis in a Patient with Lupus Erythematosus: Another Manifestation of Neutrophilic Lupus?**

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Key Words

Polychondritis, relapsing · Lupus erythematosus, cutaneous · Neutrophilic dermatosis

Polychondritis is a rare feature in patients with lupus erythematosus (LE). We report the case of a man with chronic cutaneous lupus and relapsing chondritis of the ears, and suggest an integration of this association in the new concept of 'neutrophilic LE'.

Case Report

A 31-year-old Caucasian man had a 5-year history of red papules and plaques of the forehead, cheeks and neck, without hyperkeratosis or atrophy, exacerbated after sun exposure. A biopsy showed a lymphohistiocytic periadnexal infiltrate in the deep dermis, with a less marked perivascular involvement, and a nodular architecture but without granuloma. Neither epidermis nor dermo-epidermal junction were involved.

Moreover, the patient reported intermittent joint pain and swelling, and episodes of acute pain, redness and swelling of the ears which always spared the lobules. Sicca syndrome or Raynaud's phenomenon was not present.

Serological and blood tests revealed a fluctuant neutropenia (between 800 and 2,000 neutrophils/mm³), elevation of the erythrocyte sedimentation rate and positivity for antinuclear antibody with a speckled pattern (1/1,280). Anti-Ro/SSA and anti-La/SSB antibodies were positive on enzyme-linked immunosorbent assay, while antihistone and anti-dsDNA were negative. Complement testing showed low levels of CH50 and C4, related to a partial C4 deficiency (homozygous C4AQ0). There was no renal involvement.

A diagnosis of chronic cutaneous LE associated with relapsing polychondritis (RP) was established. Photoprotection and a treatment with hydroxychloroquine allowed complete healing of the skin lesions, without scars. The patient experienced frequent episodes of auricular chondritis, controlled by non-steroidal anti-inflammatory drugs. Dapsone (100 mg daily) was introduced but had only a slight effect on the frequency of chondritis. On a 6-year follow-up, he never had nasal, tracheal or ocular involvement, and the cartilage of his ears did not collapse.

Discussion

RP is a rare disorder which can occur in a primary form or can be associated with various other diseases. The reported frequency of association among patients with LE is less than 1%, and those

patients usually have a favourable prognosis of the chondritis [1, 2]. The pathogenesis of RP is not known, and it has been considered as an auto-immune disorder, on the basis of an association with HLA-DR4, and evidence of humoral and cellular responses to cartilaginous components [3]. However, histologically, the inflammatory cell infiltrate initially found in the cartilaginous matrix is predominantly constituted of neutrophils, which are only secondarily replaced by lymphocytes, plasma cells and histiocytes [4]. From an epidemiological point of view, RP shares with neutrophilic dermatoses (e.g. Sweet's syndrome or pyoderma gangraenosum) the same spectrum of associated diseases: acute and chronic myeloproliferative disorders, myelodysplastic syndromes, inflammatory bowel diseases etc. Besides, coexistence of RP and neutrophilic dermatosis is not rare, and RP is reputed to show clear sensibility to neutrophil-targeting drugs (i.e. dapsone or colchicine). Moreover, high levels of interleukin-8/CXCL8, a potent neutrophil chemotactic factor and a crucial mediator in neutrophil-dependent acute inflammation, are found in serum of patients with RP [5], consistent with an auto-inflammatory disease pathomechanism.

In a precedent issue, Antille et al. [6] reported a well-characterized case of amicrobial pustulosis of skin folds associated with undetermined connective tissue disease, a typical example of 'neutrophilic cutaneous LE'. Lipsker and Saurat [7] suggested in the same issue the concept of 'neutrophilic cutaneous LE' to include all lupus-associated neutrophilic dermatoses. We believe that lupus-associated polychondritis can be seen as another auto-inflammatory manifestation of this complex multi-factorial disease, and thus suggest adding this peculiar condition to the new concept of 'neutrophilic LE'.

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