A 58-year-old man was referred to our clinic with an 8-month history of red-brown nodules on the dorsal surface of his left hand. He reported that he collected fish in his home aquarium as a hobby. On this basis, a prior diagnosis of nontuberculous mycobacterial infection was made, but systemic clarithromycin was ineffective. The patient also reported a constant painful sensation, which worsened in the evening and after exposure to low temperatures.

Dermatologic examination of the left hand revealed two large, tender, red-brown skin nodules on the extensor surfaces of the second and third metacarpophalangeal joint, one of which was partially covered by a hematic crust (Figure 1). There were no cutaneous or mucosal lesions elsewhere. Culture of the lesional tissue did not demonstrate significant mycobacterial, bacterial or fungal pathogens. Complete blood cell count, liver and renal function tests, serum protein electrophoresis and serological tests for autoimmune diseases were performed, and all results were within the normal range. A 5-mm punch biopsy was performed and histological examination revealed a leukocytoclastic vasculitis with neutrophilic perivascular infiltrates, dermal fibrin deposits and endothelial expansion. Special stains for mycobacteria and fungi were negative. On this basis, we confirmed the clinical suspicion of erythema elevatum diutinum (EED). The patient was treated with oral dapsone 50 mg daily. After 8 weeks of treatment, the lesions began to resolve leading to marked improvement. Complete resolution of the lesions was obtained after 14 weeks without any recurrence of the disease after 6 months follow-up.

EED is a chronic, recurrent form of cutaneous leukocytoclastic vasculitis. The etiology of disease is not well understood, but it is presumed to be related to vascular immune complex deposition in small vessel, possibly triggered by an unknown antigen. It may develop at any age but usually affects adults between the third and sixth decades.1–3
EED manifests as tender, persistent, red to reddish-brown papules and nodules, which may coalesce to form larger nodules or plaques. Lesions are distributed symmetrically on the extensor surfaces of the acral areas and joints. Truncal, axillary, facial, retroauricular and genital localizations are rarely described. The lesions may be asymptomatic or associated with a painful or burning sensation, which usually worsen in the evening or after exposure to cold temperatures. Histological findings are characterized by leukocytoclastic vasculitis and dense neutrophilic infiltrate in the early stages of the lesions and by fibrosis in the late stages.

EED may be associated with several hematological disorders (IgA gammapathy, multiple myeloma), infectious diseases (tuberculosis, streptococcal infection, HIV), immunological disorders (inflammatory intestinal diseases, rheumatoid arthritis) and certain malignancies (B-cell lymphoma, squamous cell carcinoma, breast carcinoma).

The differential diagnosis of EED includes several different entities, such as bacterial or fungal infections, granuloma annulare, Sweet syndrome, sarcoidosis, pseudolymphoma and Kaposi’s sarcoma.

EED usually improve with oral dapsone, as in our case. However, treatment is often ineffective due to the chronic nature of disease with a tendency for recurrence. Use of colchicines, tetracycline, cyclophosphamide and intralesional or topical high-potency glucocorticoids has also been reported.

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