AN ATYPICAL PRESENTATION OF ERYTHEMA ELEVATUM DIUTINUM

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Revista HCPA. 2013;33(1):80-83

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ABSTRACT

Erythema elevatum diutinum is a rare chronic leukocytoclastic vasculitis of unknown etiology. Lesions are usually asymptomatic, although burning and itchiness can occur. The typical clinical presentation is characterized by persistent, symmetrical, papules and nodules that may coalesce to form larger nodules or plaques. It may be associated with various pathologies and the diagnosis is based on clinical and histopathological findings. Treatment is based on the use of dapsone and sulfonamides, first-line therapies, and other options such as niacinamide, tetracycline, colchicine, chloroquine and corticosteroids. We describe the case of a 65-year-old male patient that presented a single lesion on the dorsum of the hand, an unusual clinical presentation.

Keywords: Erythema elevatum diutinum; vasculitis leukocytoclastic; neutrophilic dermatosis

Erythema elevatum diutinum is a rare chronic leukocytoclastic vasculitis of unknown etiology, characterized by red to reddish brown or purple papules and plaques, predominantly on the extensor surfaces of the extremities. It may be associated with various pathologies and the diagnosis is based on clinical and histopathological findings. The treatment includes dapsone and sulfonamides, first-line therapies, and other options such as niacinamide, tetracycline, colchicine, chloroquine and corticosteroids. We describe the case of a 65-year-old male patient with an unusual single lesion.

CASE REPORT

A 65-year-old caucasian male smoker came to our clinic with a 3-month history of a pruritic lesion on the dorsum of the hand. Examination revealed a red-purple indurated plaque with crusts on the dorsum of the right hand (figure 1). A skin biopsy was performed and histology revealed chronic psoriasiform dermatitis with verrucous hyperplasia, papillary fibrosis and a predominantly eosinophilic and neutrophilic infiltrate (figure 2). These findings were consistent with erythema elevatum diutinum. The investigation for associated disorders or comorbidities was negative. Initially, the patient was treated with intralesional corticosteroids with low response. Therefore, oral dapsone 100 mg daily was begun, showing immediate satisfactory results, with only residual post-inflammatory hyperpigmentation (figure 3).

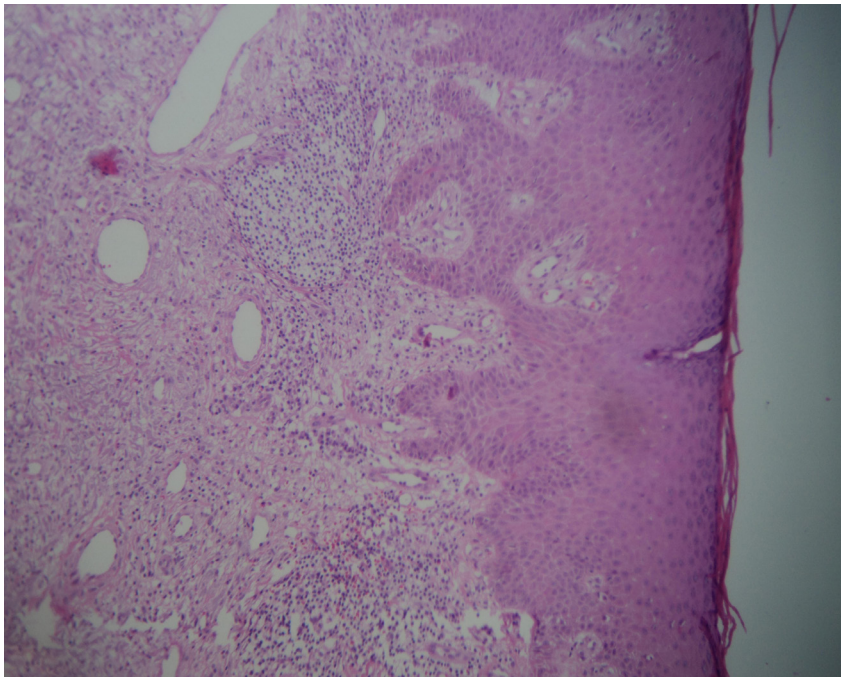


Figure 1: A red-purple indurated plaque with crusts on the dorsum of the right hand.



Figure 2: Histology revealing a chronic psoriasiform dermatitis with verrucous hyperplasia, papilar fibrosis and a predominantly eosinophilic and neutrophilic infiltrate.

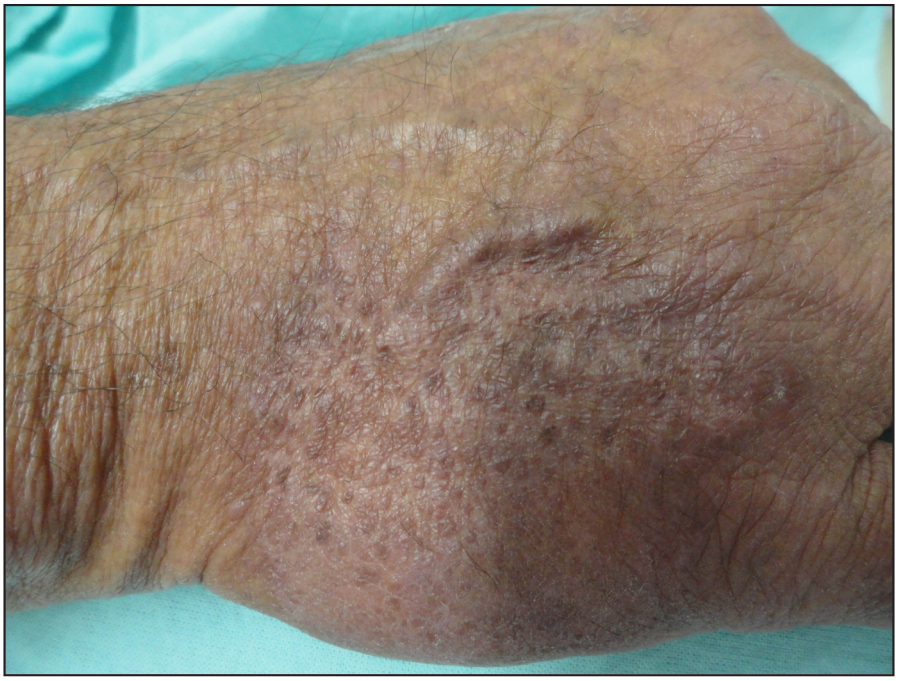


Figure 3: Patients skin lesion after oral dapsone 100 mg daily, showing satisfactory response, with only residual post-inflammatory hyperpigmentation.

COMMENTS

Erythema elevatum diutinum (EED) was first described in 1894. Since then, almost 250 cases have been reported in the literature (1). EED is a rare chronic leukocytoclastic vasculitis, which is more often seen in patients in the fourth through the six decade (2,3). It affects equally both genders and may be associated with many disorders. The cause of EED is unknown, but it is presumed to be related to vascular immune complex deposition. (2,4,5)

The typical clinical presentation is characterized by persistent, symmetrical, firm, tender, red to reddish brown or purple papules and nodules that may coalesce to form larger nodules or plaques, predominantly on the extensor surface of the extremities and near the joints (hands, elbows, knees) (2,5-7). Lesions are usually asymptomatic, although burning and itchiness can occur (3). Occurrences at atypical sites have been reported, including trunk, retro-auricular, palmar and plantar lesions (2,8). Joint pain is the most common systemic symptom, observed in up to 40% of the cases (8). Delay in diagnosis can occur in unusual presentations and atypical clinical forms are usually diagnosed only after histopathological analysis (10).

The differential diagnosis of EED is quite broad and includes Sweet's syndrome, granuloma annulare, sarcoidosis, pseudolymphoma, Wegener's granulomatosis and multicentric reticulohistiocytosis.

The histopathology of early lesions begin with leukocytoclastic vasculitis, polymorphonuclear cell infiltrate and fibrin deposition in the superficial and mid-dermis. Mature lesions demonstrate a combination of granulation response or skin healing combined with a proliferation of dermal spindle cells, with or without multinucleate giant cells (2). The diagnosis of erythema elevatum diutinum is based on the characteristic clinical lesion, histopathological findings and laboratory investigations (2).

In EED patients, it is important to exclude association with other disorders such as infectious diseases (*streptococci*, hepatitis B, HIV), certain malignancies (glioma, squamos cell carcinoma, breast carcinoma), autoimmunity (rheumatoid arthritis, Crohn's disease, Wegener's granulomatosis) and hematological abnormalities (myelodysplasia, multiple myeloma, lymphoma, paraproteinemias). For such purpose, a laboratory investigation should be performed: complete blood count, electrolytes, creatinine, liver function tests, rheumatoid factor, hemossedimentation rate, reactive C protein, urinary and serum protein

electrophoresis, Bence-Jones protein, hepatitis B, syphilis and HIV serologies, anti-endomysial antibody, anti-DNAse and antistreptolysin O (2,10). Even though screening is normal, long term monitoring is necessary to detect hematologic disorder early (1).

The treatment of EED is difficult because of its chronic and recurrent course. When possible, treatment of the underlying cause should be targeted. Dapsone and sulphonamides are first-line treatments for EED (2,11,12). Other employed therapies include tetracycline, colchicine, niacinamide, chloroquine and corticosteroids (2,9). With the regression of the lesions, residual hyperpigmentation with occasional atrophy is commonly seen (9).

The clinical findings in our case are unusual, since the case presented with a single lesion. The histopathological findings and the excellent response to dapsone are compatible with the diagnosis of EED. EED can be clinically confused with other disorders; an unusual presentation and its low incidence favor a delay in diagnosis and, consequently, an evolution to chronicity, when lesions are less responsive to medical therapy. This case reports an unusual clinical presentation with a single lesion, which has rarely been reported in the literature.

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Recebido: 24/09/2012
Aceito: 20/01/2013