RESIDENT’S FORUM

Multiple hypertrophic purpuric to erythematous papuloplaques on four limbs of a 38-year-old man

Case report

A 38-year-old healthy man presented with a 6-month history of mildly pruritic skin lesions on joints of four limbs. Initially, the itchy erythematous papules developed from extensor aspects of the joints, then the papules gradually progressed to violaceous plaques. He denied any trauma history or chronic scratching on the cutaneous lesions. On physical examination, there were multiple purpuric to erythematous hypertrophic, oval-shaped, and well-demarcated plaques with varying size on the extensor aspects of the left second and third metacarpophalangeal joints, left elbow, and bilateral knees (Figure 1). The initial clinical diagnosis was lichen planus or lichen simplex chronicus. Skin biopsy revealed marked hyperkeratosis, hypergranulosis, moderate acanthosis, and thickened dermal vessels infiltrated and surrounded by many neutrophils, and scattered eosinophils and lymphocytes. Nuclear dusts and focal mild extravasation of red blood cells with some vessels surrounded with fibrin deposits were seen (Figure 2). No bacilli are detected by Gram and Warthin-Starry stains.

Figure 1 (A) There are two hypertrophic, well-defined, and violaceous plaques on the dorsum of the left hand; (B) arrangement of the dusky red papuloplaques on the bilateral knees is symmetrical.

Figure 2 (A) Histopathology showed hyperkeratosis, hypergranulosis, moderate acanthosis, and infiltrates of mixed type of inflammatory cells in upper dermis (hematoxylin-eosin staining; 40×); (B) thickened dermal vessels infiltrated and surrounded by many neutrophils, scattered eosinophils, and lymphocytes. Nuclear dusts, extravasation of red blood cells, and perivascular fibrin deposits are seen (hematoxylin-eosin staining; 400×).
Diagnosis

Erythema elevatum diutinum (EED).

Discussion

EED is a rare type of chronic cutaneous vasculitis, which usually involves adults. The clinical manifestation showed symmetrical, firm, and well-demarcated purpuric to erythematous papuloplaques on extremities, especially surrounding the extensor aspects of joints. Atypical presentations, such as vesiculobullous, hemorrhagic, and ulcerative lesions, are also reported. Pruritus, tenderness, or burning sensation may develop on the cutaneous lesions. Nevertheless, some lesions are asymptomatic. The clinical differential diagnoses contain Sweet's syndrome, pyoderma gangrenosum, granuloma faciale, fixed drug reaction, erythema multiforme, lichen planus, porphyria cutanea tarda, fibrous histiocytoma or dermatofibroma, bacillary angiomatosis, Kaposi's sarcoma, xanthoma, and necrobiotic xanthogranuloma.

Histopathologic findings of EED are associated with the stage of the lesions. In the early lesions, the findings are compatible with leukocytoclastic vasculitis, inclusive of nuclear dusts, extravasation of red blood cells, perivascular infiltrates of mixed type of inflammatory cells, and fibrin deposition of vessels in superficial and mid-dermis. The late-staged lesions demonstrated fibrosis, proliferation of vessels, lipid deposition in dermis, and less inflammatory cells.

The exact pathogenesis of EED remains elusive. However, EED is related to viral infection (especially among patients with HIV), bacterial infections, rheumatic diseases, and hematologic diseases. Immunoglobulin A monoclonal gammopathy was also reported to be correlated with EED. Furthermore, type 3 hypersensitivity reaction may play an important role in the pathogenesis of EED. Deposition of immune complexes on the vessels can trigger the chain immunologic reaction to damage the vessels.

The effective treatment strategy is to treat the underlying diseases, which are related to EED. It is sometimes hard to search for the true underlying disease associated with EED. Moreover, sulfonamides or dapsone is the first choice of treatment. There are other alternative treatments that include niacinamide and tetracycline, colchicine, corticosteroids (oral, topical, or intraleisional type), and chloroquine. Recurrence rate of EED is still high if the underlying triggering factors are not under control. Long-term follow-up is necessary among this group. In our case, the patient received topical potent steroids only without receiving further survey after skin biopsy. However, poor response was noted after 3 months of treatment.

We report this case to emphasize the importance of including EED in the differential diagnosis of multiple hypertrophic purpuric to erythematous papuloplaques that develop on four limbs.

Yung-Yi Lee, Ju-Hung Ko, Mei-Ching Lee, Jheng-Wei Lin
Department of Dermatology, Chang Gung Memorial Hospital, Taipei, Taiwan
Chang Gung University College of Medicine, Taoyuan, Taiwan
*Corresponding author. Jheng-Wei Lin, 199, Tun-Hwa North Road, Taipei 105, Taiwan. Tel.: +886 2 27135211 3397; fax: +886 2 27191623. E-mail address: s81095@gmail.com (J.-W. Lin)

References


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