Erythema elevatum diutinum in association with dermatitis herpetiformis

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ABSTRACT

Erythema elevatum diutinum (EED) is a rare skin disease that initially presents as leucocytoclastic vasculitis and later resolves with fibrosis. Dermatitis herpetiformis is an autoimmune blistering disease characterized by granular deposits of immunoglobulin A (IgA) in dermal papillae. We report a rare association of these two disorders.

Key words: Dermatitis herpetiformis, erythema elevatum diutinum, immunoglobulin A

INTRODUCTION

EED is a localized, low-grade form of leukocytoclastic vasculitis of unknown pathogenesis characterized by persistent, symmetrical, red-purple papules, nodules, and plaques affecting the extensor surfaces of the extremities, the ears, trunk and buttocks. Dermatitis herpetiformis is an autoimmune blistering disease characterized by granular deposits of immunoglobulin A (IgA) in dermal papillae. The common link between these two disorders is the deposition of IgA antibodies, underlying gluten sensitivity in both, their association with celiac disease and response to treatment with dapsone. This is only the second case regarding the coexistence of these two disorders.[1]

CASE REPORT

A 50-year-old male presented with firm skin-colored and erythematous nodules over the knuckles, elbows, buttocks, knees, and lower legs which gradually increased in size in since 3 years duration [Figure 1]. Complete blood picture, liver function tests, urine routine, blood sugar were within normal limits. Biopsy of the nodules of the knuckles showed focal parakeratosis and mild spongiosis in the epidermis, perivascular neutrophilic infiltrate in the dermis, neutrophils in the vessel wall and leukocytoclasia [Figure 2]. Immunofluorescence showed linear IgA deposits in the basement membrane zone and weak granular perivascular C3 and fibrinogen deposits [Figure 3]. Four years ago itchy vesicles over the back and arms were detected in the patient. Biopsy of the vesicular lesions were consistent with dermatitis herpetiformis [Figure 4]. He was administered dapsone which leads to disappearance of lesions with recurrence after stopping treatment. With the clinical presentation, biopsy and immunofluorescence results, a diagnosis of erythema elevatum diutinum on a background of dermatitis herpetiformis was made.

DISCUSSION

Erythema elevatum diutinum (EED) is a rare cutaneous condition that initially presents as leukocytoclastic vasculitis (LCCV) of the skin and later resolves with fibrosis clinically characterized by persistent red-purple to yellow papules, plaques, and nodules. The symmetrical lesions typically affect the acral surfaces of the body, especially the extensor surfaces of the hands, but other areas such as the buttocks may also be involved.[2] Antecedent bacterial infections, collagen vascular diseases, myeloproliferative diseases, human immunodeficiency virus infection, cryoglobulinemia, Wegener granulomatosis, Crohn disease, systemic fungal diseases, IgA paraproteinemia, and other associations have been reported.[3,4] Histopathologically, EED initially presents as a leukocytoclastic vasculitis (LCCV) with polymorphonuclear neutrophils.
neutrophils, extravasated red cells, nuclear dust, and fibrin deposits in the walls of the small arterioles. Most of the lesions eventually resolve with fibrosis. Direct immunofluorescence usually demonstrates IgG and C3 deposits within vessels in the upper dermis.

Dermatitis herpetiformis (DH) is characterized by chronic, intensely pruritic, polymorphic, vesicles usually appearing on elbows, knees, buttocks, and scalp associated with gluten-sensitive enteropathy There are granular IgA deposits in the dermal papillae by the direct immunofluorescence in the perilesional skin of patients with dermatitis herpetiformis.\(^5\)

The close association of IgA with erythema elevatum diutinum and dermatitis herpetiformis is well known. Because of the association with IgA, the presence of erythema elevatum diutinum in a patient with chronic dermatitis herpetiformis is not surprising. The IgA in the skin may be an epiphenomenon or may represent deposition in the skin along with other immune complexes.\(^5\)

REFERENCES

1. Aftab MN, Dee A, Helm TN. Erythema elevatum diutinum arising in the setting of dermatitis herpetiformis. Cutis 2006;78:129-32.
2. Gerbig AW, Zala L, Hunziker T. Erythema elevatum diutinum: A rare dermatosis with a broad spectrum of illnesses. Hautartz 1997;48:113-7.
3. Sangueza OP, Pilcher B, Sangueza JM. Erythema elevatum diutinum: A clinical pathological study of eight cases. Am J Dermatopathol 1997;19:214-22.
4. Chow RK, Benny WB, Coupe RL, Dodd WA, Ongley RC. Erythema elevatum diutinum associated with IgA paraproteinemia successfully controlled with
intermittent plasma exchange. Arch Dermatol 1996;132:1360-4.

5. Collin P, Reunala T. Recognition and management of the cutaneous manifestations of celiac disease: A guide for dermatologists. Am J Clin Dermatol 2003;4:13-20.