AUTOIMMUNE BULLOUS DISEASES

LINEAR IGA BULLOUS DISEASE PRESENTING AS A HAILEY-HAILEY DISEASE ASSOCIATED WITH HASHIMOTO THYROIDITIS: A CASE REPORT

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Background: Linear IgA bullous disease (LABD) is a rare autoimmune subepidermal bullous disease characterized by linear deposits of IgA anti-basement membrane zone antibodies. Most cases of LABD are idiopathic and the etiology of the disease remains unclear, but some cases are occasionally induced by drugs, internal malignancies, and infections.

Observation: A previously healthy 17-year-old female patient presented with a 5-months history of vesicular eruptions. Physical examination revealed multiple vesicles and bullae on an erythematous base. First eruptions appeared in axillae and groins. The initial diagnosis was Hailey-Hailey disease. Topical treatment was established with gradual progression of the disease. The lesions spread on the lower trunk. back thigh surfaces, neck, facial skin around the mouth. The typical bullous eruption with a ‘string of pearls’ sign appeared on left forearm. Oral mucosa, eyes, and nails were spared. Direct immunofluorescence (DIF) demonstrated linear IgA-deposit pattern at the dermo-epidermal junction (DEJ) in the absence of other immunoglobulins and complement.

Key message: LABD is defined according to the three following clinical and histological criteria: vesicular or bullous eruption involving skin and mucous membranes (MMs), subepidermal blisters infiltrated predominantly by neutrophils in lesion biopsies and a linear IgA-deposit pattern at DEJ in the absence of other immunoglobulins on DIF.
In the described case, the symmetric distribution was confined to the areas typical of Hailey-Hailey disease. The adolescence-onset of disease was also similar to Hailey-Hailey disease.
Furthermore, the patient was diagnosed with Hashimoto thyroiditis which is also an autoimmune disease. Autoimmune diseases are common conditions which appear to develop in genetically susceptible individuals, with an expression of the disease being modified by permissive and protective environments.