AUTOIMMUNE BULLOUS DISEASES

DERMATITIS HERPETIFORMIS

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Dermatitis herpetiformis (DH) is a rare inflammatory skin disease that is considered the specific cutaneous manifestation of celiac disease (CD). Both DH and CD occur in gluten-sensitive individuals, share the same Human Leukocyte Antigen (HLA) haplotypes (DQ2 and DQ8), and improve following the administration of a gluten-free diet. Moreover, almost all patients with DH show typical CD alterations at the small bowel biopsy, ranging from villous atrophy to augmented presence of intraepithelial lymphocytes, and the generation of circulating autoantibodies against tissue transglutaminase (tTG). DH presents with polymorphic lesions, including papules, vesicles and small blisters, symmetrically distributed in typical areas including the extensor aspects of the limbs, elbows, sacral regions and the buttocks. Intense pruritus is almost the rule. However, many atypical presentations of DH have also been reported.
Recent evidences suggested that DH is changing. Firstly, some studies reported a reduced incidence of DH, probably due to early recognition of CD, so that there is not enough time for DH to develop. Moreover, data from eastern countries, where DH is very rare, highlighted the absence of intestinal involvement as well as of the typical serological markers of CD in most of the patients. Although occasionally, similar cases may also occur in Caucasian patients, complicating DH diagnosis.
The latter relies on the combination of clinical, histopathologic and immunopathologic findings. Detecting granular IgA deposits at the dermal-epidermal junction by direct immunofluorescence (DIF) from perilesional skin represents the most specific diagnostic tool. Further, assessing serum titers of autoantibodies against epidermal transglutaminase (eTG), the supposed autoantigen of DH, may also serve as a clue for the diagnosis. However, a study from our group has recently demonstrated that granular IgA deposits can also be found in celiac patients with non-DH inflammatory skin diseases, raising questions about the effective role of eTG IgA autoantibodies in DH and suggesting the need of a revision of diagnostic criteria, conceivably emphasizing clinical aspects of the disease together with DIF.
DH usually responds to the gluten-free diet. Topical clobetasol ointment or dapsone may be also applied to favor rapid disease control. Further research may provide the clue for future therapeutic options in patients with DH.