Dermatitis herpetiformis (Duhring's disease)
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Dermatitis Herpetiformis
Dermatitis herpetiformis is an intensely pruritic, chronic recurrent dermatitis that has a slight male predilection. The disease is characterized by polymorphic lesions, which include urticarial plaques, papules, and pustules. It is associated with a high risk of gastrointestinal complications, particularly small bowel villous atrophy and celiac disease. An increased but rare risk of lymphoma is also reported in association with dermatitis herpetiformis.
Histopathology
The typical histologic features are best observed in erythematous skin adjacent to early blisters. In these zones, the dermal-epidermal junction is disrupted. Within 1 to 2 days, the rete ridges lose their attachment to the dermis, and the blisters then become unilocular and clinically apparent. At this time, the characteristic papillary microabscesses may be observed at the blister periphery. For this reason, the inclusion of perivesicular skin in the biopsy specimen is of
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utmost value. The papillary dermis beneath the papillae may have a relatively intense inflammatory infiltrate. Apoptotic keratinocytes may be noted above the papillary microabscesses.

**IF Testing**
In 1967, Cormane described the presence of granular deposits of IgA within the dermal papillae in both lesional and non-lesional skin. The presence of IgA in biopsy specimens from clinically normal skin immediately adjacent to areas of erythema, because false-negative results may occur when blistered or inflamed skin is evaluated. The presence of two appropriately selected biopsy sites is a strong indication that the patient does not have dermatitis herpetiformis.
Circulating IgA antibodies that react against reticulin, smooth muscle endomysium, the dietary antigen gluten enter the gut as substrate, IIF has been used to detect antiendomysial antibodies, which are present in 52% to 100% of patients.

Pathogenesis

Three important findings must be considered in the pathogenesis of dermatitis herpetiformis.
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Spruelike changes on jejunal biopsy. Patients with celiac disease develop IgA autoantibodies to tissue transglutaminase.
The IgA deposition results in activation of the complement system followed by chemotaxis of neutrophils.
Ultrastructural Study

The changes in dermatitis herpetiform is resemble those observed in the inflammatory bullae of bullous pemphigoid.
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