Dermatitis herpetiformis (Duhring's disease)
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Dermatitis Herpetiformis
Dermatitis herpetiform is an intensely pruritic, chronic recurrent dermatitis that has a slight male predilection. The disease is associated with increased risk of gastroduodenal ulceration and an increased but rare risk of lymphoma. Dermatitis herpetiform is in association with SLE has also been reported.
Histopathology
The typical histologic features are best observed in erythematous skin adjacent to early blisters. In these zones, 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 nonlesional skin. However, some cases may present with only a granular or uniform deposition of IgA, making it difficult to confirm the diagnosis by direct immunofluorescence (DIF). In such cases, a repeat DIF is recommended. To rule out false-negative results, biopsies should be taken from clinically normal skin immediately adjacent to the erythema, because false-negative results may occur when blistered or inflamed skin is evaluated. The presence of IgA deposits in at least 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, and proteins of the intestinal villi have been identified in patients with dermatitis herpetiformis. These antibodies are present in the circulation and can be detected using immunofluorescence microscopy. Immunofluorescence (IIF) is a technique used to detect these 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. In advanced lesions, the lamina densa has been destroyed, as is noted in the "inflammatory bullae" of bullous pemphigoid.
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