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 characterized by a rash of small, itchy blisters that often occur on the abdomen, arms, and legs. Dermatitis herpetiform is associated with celiac disease and an increased but rare risk of lymphoma. Dermatitis herpetiform is also reported to occur in association with SLE.
Histopathology
Dermatitis herpetiformis (Duhring's disease) = إرتياب الجلد الشحمي المزمن = دورينج داء

The typical histologic features are best observed in erythematous skin adjacent to early blisters. In these zones, 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 prime importance.
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The papillary dermis beneath the papillae may have a relatively intense inflammatory infiltrate of lymphocytes and plasma cells. 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. The diagnosis of dermatitis herpetiformis is confirmed by direct immunofluorescence (DIF) of skin. It is important to take biopsies from areas of erythema, because false-negative results may occur when blistered or inflamed skin is evaluated. The presence of IgA deposits in 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 the cryptic enterocyte of the intestinal epithelium have been detected in dermatitis herpetiformis. These antibodies are thought to play a role in the pathogenesis of the disease. Three important findings must be considered in the pathogenesis of dermatitis herpetiformis:

1. The disease is associated with celiac disease, which is characterized by a genetic susceptibility to gluten intolerance.
2. The disease is triggered by eating gluten-containing foods, which leads to an inflammatory response in the small intestine.
3. The inflammatory response results in the production of IgA antibodies that target various intestinal antigens, including reticulin and smooth muscle endomysium.

Pathogenesis

Three important findings must be considered in the pathogenesis of dermatitis herpetiformis.
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 resemble those observed in the inflammatory bullae of bullous pemphigoid.
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