

DERMATITIS HERPETIFORMIS



■ INTRODUCTION

Dermatitis herpetiformis (DH) is an immune-mediated blistering skin disease most often asymptomatic, and associated with gluten-sensitive enteropathy (GSE). DH occurs more frequently in individuals of Northern European ancestry and is rarely seen in Asians and African Americans. A slight male predilection has been reported. Typically, the onset of DH is in the second to fourth decade; however, all ages may be affected. The pathogenesis of DH is associated with the presence of GSE. There is increasing evidence that the auto antigen in DH is transglutaminase. This coupled with high gluten diet triggers granular deposition of IgA at the dermal-epidermal junction especially of the dermal papillae of the skin.

■ CLINICAL FEATURES

Clinical manifestations of DH are as skin-colored-to-erythematous vesicles appearing in a herpetiform pattern, symmetrically distributed over extensor surfaces including elbows, knees, buttocks, shoulders, and the posterior (nuchal) scalp. Rarely, if ever, are patients totally asymptomatic, although the degree of itching experienced by patients varies. Oral mucosa lesions occur infrequently in patients with DH.

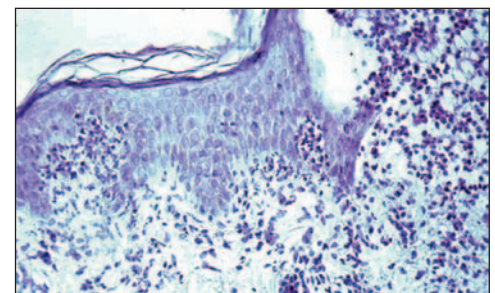
■ DIAGNOSIS

Light microscopy features are not diagnostic of DH, and direct immunofluorescence (IF) is required to confirm the diagnosis. Biopsies for direct IF should be performed on perilesional (normal-appearing) skin. H&E examination of lesional skin reveal neutrophils in the dermal papillae with fibrin deposition, papillary microabscesses, subepidermal vacuolization and vesicle formation. Granular IgA deposits in dermal papillae of perilesional skin on direct IF is the hall mark of DH.

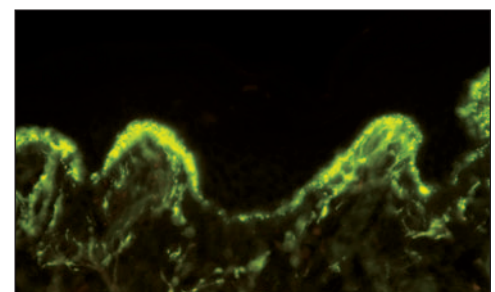
Other diagnostic markers include the presence of serum IgA endomysial (EMA) and tissue transglutaminase (tTG). (Our celiac disease profile II, Test # 121). The presence of EMA and tTG antibodies correlates with the extent of the gut involvement. Some patients with DH may be negative for EMA and tTG antibodies and some DH cases biopsies may be negative whereas positive for serum antibodies. Repeat biopsy studies in such cases can help to confirm the diagnosis of DH.



Herpetiform Vesicles on the skin.



Neutrophilic abscesses on the epidermis



Granular deposits of IgA at dermal papillae under Direct IF

OPTIMAL BIOPSY SITE FOR IMMUNOLOGICAL INVESTIGATIONS:

TISSUE	SITE	SPECIAL CONSIDERATIONS
Skin or Mucosa	Take biopsy from a noninflamed perilesional area, adjacent to active or new blister	Serum studies for EMA and tTG antibodies may help in situations when direct IF is negative.

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TREATMENT

Patient management includes medications, dietary avoidance of gluten, or both. A gluten-free diet is very difficult to achieve; however, limiting intake of wheat, barley, or rye products can lessen the symptoms. Dapsone and sulfapyridine are the primary

medications used to treat DH.

SUGGESTED READING

Karpati S. Dermatitis herpetiformis: close to unraveling a disease. *J Dermatol Sci.* 2004;34:83-90.

Hall RP. Dermatitis herpetiformis. *J Invest Dermatol* 1992; 99: 873-81.

Zone JJ, Meyer LJ, Petersen MJ. Deposition of granular IgA relative to clinical lesions in dermatitis herpetiformis. *Arch Dermatol.* 1996;132:912-8.

Kumar V, Zane H, Kaul N: Serologic markers of gluten-sensitive enteropathy in bullous diseases. *Arch Dermatol* 1992; 128: 1474-8.

Accetta P, Kumar V, Beutner EH, et al: Anti-endomysial antibodies. A serologic marker of dermatitis herpetiformis. *Arch Dermatol* 1986; 122: 459-62.

SPECIMEN SUBMISSION

Specimen collection kits are available free of charge. Please call (800) 537-8378 for an immediate shipment. Use appropriate tube(s) as follows:

Immunofluorescence:

Lesional biopsy: Red tube

Normal biopsy: Purple tube

H&E biopsy: Green tube

Serology: Orange tube

Specimens can be shipped by courier services, U.S. Postal Service and overnight carriers free of charge. Results are reported in two business days of specimen receipt via mail, fax and through IMMCO Online, a HIPAA-compliant web tool at www.immco.com.



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