



AUTOIMMUNE BULLOUS DISEASES

IGA ANTI-TISSUE TRANSGLUTAMINASE AND IGA ANTIENDOMYSIAL ANTIBODY NEGATIVE DERMATITIS HERPETIFORMIS IN A FILIPINO PATIENT: A CASE REPORT

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Background: Dermatitis Herpetiformis (DH) is a chronic, relapsing cutaneous disease with associated gluten-sensitive enteropathy (GSE). DH is characterized by a symmetric, intensely pruritic, papulovesicular herpetiform eruption involving extensor surfaces of the body. It is believed to be rare among Asians and African-Americans. Diagnosis of DH relies on clinical presentation, histology, direct immunofluorescence, and serology. More rarely, seronegative DH occurs in 30% of patients with DH. Anti IgA anti-tissue transglutaminase and IgA antiendomysial antibodies are rarely expressed among the Japanese and may probably represent a variant of DH different from Europeans and other Caucasian races.

Observation: A 33 year-old Filipino female consulted because of a 3-week history of severely pruritic vesicles and crusts on the face, trunk, and arms. Patient noted no gastrointestinal symptoms on consultation. Skin punch biopsy revealed subepidermal blisters with collection of neutrophils at the dermal papillae. Direct immunofluorescence showed strong (+2) granular deposits of IgA at the suprapapillary tips of the epidermis in perilesional skin, thus validating the diagnosis. The patient's serum was negative for IgA anti-tissue transglutaminase and IgA antiendomysial antibodies. Patient was treated with topical corticosteroids and Dapsone 50 mgs OD with remarkable improvement.

Key Message: The unique features of "Japanese DH" are the following: (i) high frequency of deposition of fibrillar collagen in the papillary dermis; (ii) the rare occurrence of gluten-sensitive enteropathy; (iii) absence of HLA DQ2 and HLA DQ8; and (iv) rare association of autoimmune disease or lymphomas. This case of seronegative DH in a Filipino may represent a variant similar to the Japanese with no anti-transglutaminase antibodies and no specific HLA genes despite IgA deposits in the dermis.

