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PHOTOLETTER TO THE EDITOR

Dermatitis herpetiformis co-localised with vitiligo in a patient with autoimmune polyglandular syndrome

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Abstract

We report a case of dermatitis herpetiformis co-localised with segmental vitiligo in a 37-year-old woman with a background history of autoimmune polyglandular syndrome type 2. We propose genetic mosaicism as a possible mechanism. There has only been one previous case report in which dermatitis hepetiformis co-localised in close proximity but not exclusively within vilitigo in a patient with autoimmune thyroiditis. To our knowledge, this is the first case report of dermatitis herpetiformis co-localised exclusively to segmental vitiligo in the presence of autoimmune polyglandular syndrome. (*J Dermatol Case Rep*. 2013; 7(3): 101-102)

Key words:

autoimmunity, Addison's disease, autoimmune hypothyroidism, coeliac disease, dermatitis herpetiformis, diabetes mellitus, Duhring's disease, vitiligo

A 37-year-old female presented with a 3-day history of a severe, pruritic, bullous eruption on the trunk and limbs associated with malaise. Her past medical history included Addison's disease, autoimmune hypothyroidism, type 1 diabetes mellitus, coeliac disease and vitiligo. Coeliac disease was diagnosed following distal duodenal biopsy which demonstrated subtotal villous atrophy. The patient reported strict adherence to a gluten-free diet. Vitiligo on the limbs, trunk, and neck was treated with moderate potency topical corticosteroids fourteen years prior to this acute presentation. Her non-identical twin sister also suffered from autoimmune hypothyroidism and vitiligo. On examination, multiple, well-demarcated, segmental areas of depigmentation were present on the forearms, neck, groin, and upper thighs. There were vesicles and bullae of up to 20 mm in diameter with adjacent crusted, erythematous papules exclusively within the depigmented areas but not within the pigmented patches (Fig. 1).

Indirect immunofluorescence, ANA, rheumatoid factor, and viral and bacterial swabs were unremarkable. Biopsy from the arm revealed a sub-epidermal vesicle filled with neutrophils and eosinophils with adjacent papillary microabscesses. The basal keratinocytes lacked melanin pigment. Granular IgA deposition at the dermal papillae was suggestive of dermatitis herpetiformis (DH) (Fig. 2). She was managed with a gluten-free diet and hydroxyzine 25 mg when required. The patient declined dapsone therapy due to her concerns regarding potential adverse effects. Her skin remained relatively well-controlled however she died 17 months later from a myocardial infarction.

Autoimmune polyglandular syndrome type 2 (APS-2) is defined by the association of autoimmune Addison's disease with either type-I diabetes mellitus, autoimmune thyroid disease, or both.[1] APS-2 may occur in many generations within the same family in an autosomal dominant pattern with incomplete penetrance, which may explain why the twin sisters were differentially affected in terms of disease burden.[1]

The most striking aspect of this case is the localisation of blisters exclusively within vitiligo. This phenomenon could be explained by Köbnerisation which has been reported in

both DH and vitiligo. Other cutaneous disorders have been reported to Köbnerise within vitiligo, such as psoriasis vulgaris.[2]

An alternative explanation is that of genetic mosaicism. Taïeb *et al*. hypothesised that segmental vitiligo occurred in Blaschko's lines due to genetic mosaicism.[3] The success of autologous melanocyte culture therapy in segmental vitiligo argues against a localised form of melanocyte autoimmunity and is in support of a local defective gene which can be "cured" by melanocyte donation.[3] The topographic co-localisation of both segmental vitiligo and DH could be explained by mosaic genetic variation, for example in the HLA-alleles, for both conditions.[4] There has been only one prior case report in which DH localised mostly, but not exclusively, within depigmented areas of vitiligo in a patient with autoimmune thyroiditis.[5] The authors of this report concluded that the topographic proximity of the two conditions was possibly coincidental.

We report a case of dermatitis herpetiformis arising exclusively within segmental patches of vitiligo, in the presence of autoimmune polyglandular syndrome type 2 and have postulated the causative mechanism of genetic mosaicism.

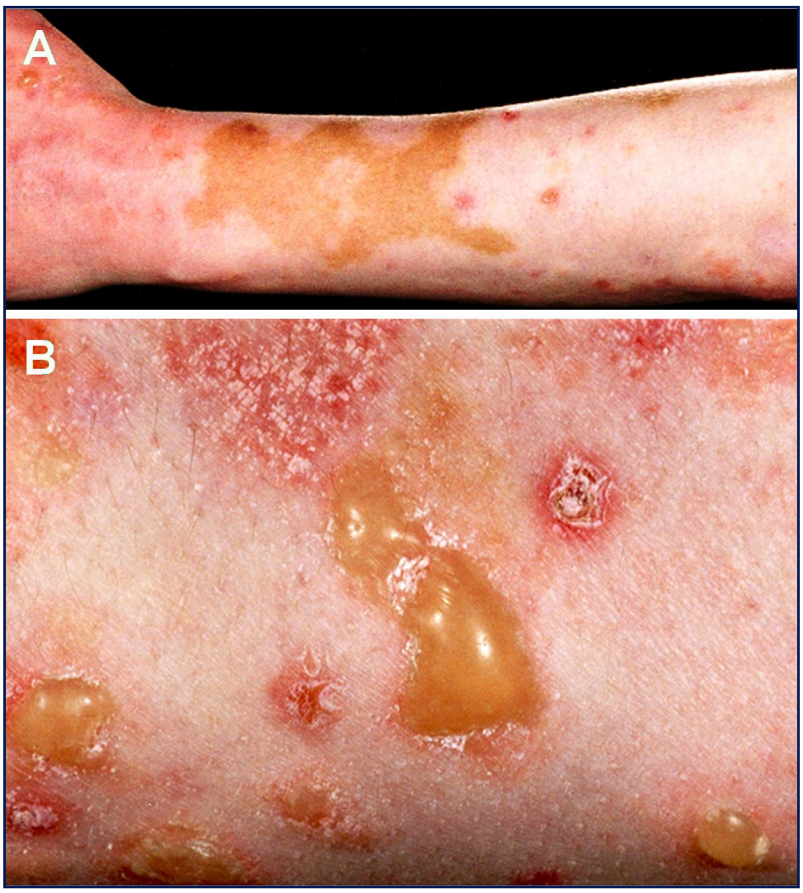

Figure 1

(A) Bullous and vesicular lesions located within segmental vitiligo on left lower arm and (B) a close-up image of representative bullae, vesicles and crusted papules.

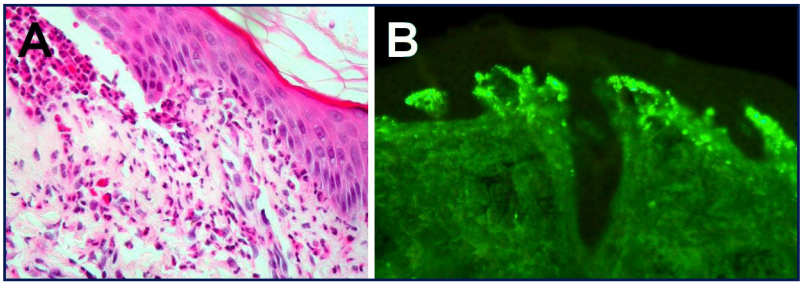

Figure 2

(A) Biopsy from the left arm demonstrating subepidermal blister formation (haematoxylin and eosin, original magnification: x200); (B) Direct immunofluorescence with granular IgA deposition in the basement membrane zone at tips of the dermal papillae.

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