

Close Association of Dermatitis Herpetiformis and Chronic Tonsillitis in a Japanese Patient without Gluten Sensitivity

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Accepted August 10, 2001.

Sir,

Dermatitis herpetiformis (DH) is an intensely pruritic, papulo-vesicular dermatosis characterized by neutrophilic micro-abscesses and a granular deposit of IgA in the dermal papillae. In Caucasians, many patients with DH have gluten-sensitive enteropathy (GSE), which is important to DH pathogenesis (1). HLA studies have shown the high frequency of HLA-A1 B8 DR3 DQ2 in DH and patients with coeliac disease (2). Moreover, the skin lesions and IgA deposits disappear when patients are on a strict gluten-free diet and reappear following a gluten-containing diet (3). On the other hand, in Japanese patients neither the GSE nor the particular HLA haplotype is associated with DH (4–7). Therefore, a different pathogenesis of Japanese patients with DH has been expected (4, 5). We present here a Japanese patient with DH closely associated with chronic tonsillitis.

CASE REPORT

A 57-year-old Japanese man presented with a 5-year history of a pruritic microvesicles, erythema, excoriations, lichenified changes and post-inflammatory symmetrical hyperpigmentation involving the buttocks and elbows (Fig. 1a). The patient noticed that the pruritic eruptions had worsened in association with every acute inflammation of chronic tonsillitis. A biopsy specimen taken from an erythematous skin lesion on his buttocks showed subepidermal microabscesses composed of neutrophils in the dermal papillae. Direct immunofluorescence (DIF) of uninvolved skin revealed granular deposits of IgA and C3 at the top of the dermal papillae. Indirect immunofluorescence using normal human skin as a substrate showed neither IgA nor IgG classes of circulating antibodies to the basement membrane zone. Laboratory studies showed no obvious abnormalities, with the exception of elevated serum C3d-binding immune complex 15 mg/l (normal < 9.2). The patient's HLA was A11, A24, B60, C3, C7, DR9, DR15, DQ1, DQ3. Jejunal biopsy without gluten-free diet showed that there was no villous atrophy, and the count of intraepithelial lymphocytes per 1,000 epithelial cells was approximately 70 with an almost even distribution. A palatine tonsil biopsy showed the typical features of chronic tonsillitis. DIF revealed linear deposits of C3 on the basement membrane of his tonsil and an increase in the IgA-expressing lymphocyte population compared with IgG-expressing lymphocytes. Surgical intervention initiated new pruritic vesicles for the following 3 months after the tonsillectomy. Within 4 months after the tonsillectomy, the patient's DH skin lesions vanished spontaneously (Fig. 1b). There was a reduction in circulating immune complexes to within the normal limit. DIF did not demonstrate

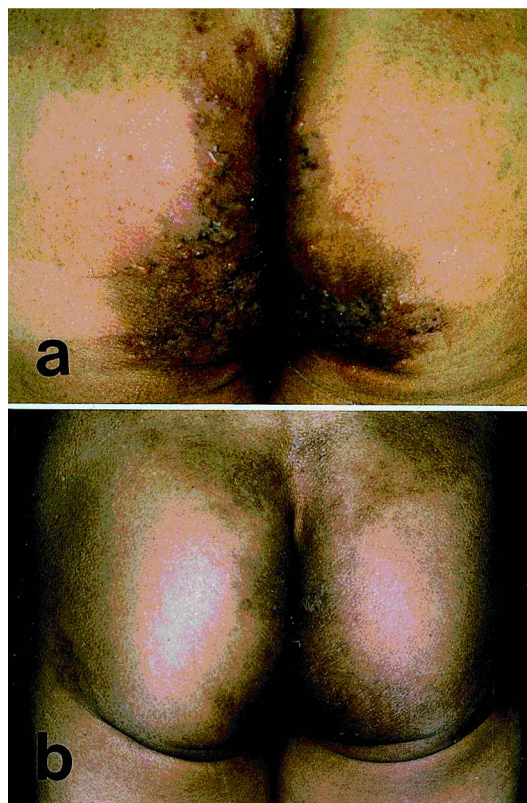


Fig. 1. (a) Vesicles, erythema, excoriations and post-inflammatory hyperpigmentations were present on the buttocks. Lesions are almost symmetrically arranged. (b) After the tonsillectomy, lesions of dermatitis herpetiformis completely disappeared without any other treatments.

any IgA and C3 deposits in the apparently normal skin on the buttocks and elbows 6 months after the tonsillectomy.

DISCUSSION

The present case has typical clinical, histopathological and immunohistological features of DH. GSE associated with DH is characterized by the histological features of the jejunum; i.e. a raised count of intraepithelial lymphocytes in mild GSE and villous atrophy in more severe cases (3). As for the former, Fry et al. definitely took the intraepithelial lymphocyte number below 200 per 1,000 epithelial cells, which covers 6 or more villi, to be within normal range (8). Because no jejunal villous atrophy was demonstrated and the count of intraepithelial lymphocytes was approximately 70 per 1,000 epithelial cells, we diagnosed the present case as DH without GSE.

The interesting point about this case was that the activity of DH was closely associated with the inflammatory state of chronic tonsillitis. Several lines of evidence support this conclusion: First, the eruption was clinically reproducible by surgical manipulation of the tonsils. Secondly, lymphocytes harbouring in the patient's tonsil expressed predominantly IgA. This finding is noticeable because the majority of lymphocytes in normal tonsils express IgG in much the same way as chronic tonsillitis without complications (9, 10). Under certain conditions, such as IgA nephropathy, which is possibly induced by tonsillitis, IgA expressing lymphocytes are predominant (10, 11). Thirdly, granular IgA deposits in the dermal papillae disappeared after tonsillectomy.

The current explanation for DH is that lymphocytes recruited in conjunction with GSE produce IgA autoantibodies that also react with dermo-epidermal junction (12). In the present case, the inflammatory reaction in the tonsils might be responsible for producing IgA class autoantibodies that react with the dermo-epidermal junction.

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Multiple Becker's Naevi: A Rare Presentation

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Accepted August 10, 2001.

Sir,

Becker's naevus (pigmented hairy epidermal naevus), a variety of epidermal naevus (1) is present in about 0.5% of young men (2). It is about 5 times more frequent in males than in females. Characteristically, it is a unilateral single lesion of the shoulder, upper arm, anterior chest or scapular region in males, appearing during adolescence. It may affect other sites, e.g. the lower limb (3, 4). It may rarely also be multiple and bilateral (5). We report here a rare case of multiple Becker's naevi with 7 distinct lesions present on different sites including the lower limbs.

CASE REPORT

A 28-year-old male consulted us for multiple, asymptomatic light to dark-brown macules that had been present for 15 years over both lower extremities, abdomen and front of chest, extending on to the left upper limb. At first, the patient noticed multiple asymptomatic light-brown macules over the left lower limb, which coalesced to form a larger patch and the pigmentation also grew darker. He noticed increased hair growth over the

lesion and gradually over the years he developed 6 similar lesions on the chest, abdomen and right lower limb. There was no history to suggest any neurological or any other systemic illness.

On cutaneous examination, 7 light-brown to dark-brown macules of sizes varying from 10 × 15 cm to much larger areas covering almost half of the lower limb with a "splash on" appearance at the periphery and central hypertrichosis in most of them. The lesions were situated on the chest, abdomen, back and right groin, extending on to the medial aspect of the right thigh, both knees and anterior aspect of both legs, left arm and forearm (Fig. 1). The lesions on the chest and anterior abdominal wall showed a sharp midline margin. Thorough physical examination did not reveal any neurological or musculoskeletal defect. Routine laboratory investigations including a haemogram, liver and renal function tests, urinalysis and examination of stool were all within normal limits. X-ray of the chest, spine and lower extremities did not reveal any pathological findings. Histopathological examination of the lesion from the chest showed mild acanthosis with increased pigment (melanin) in the basal cell layer, especially at the