

Multiple Autoimmune Syndrome in a Patient with Pemphigus Vulgaris

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Sir,

Multiple autoimmune syndrome (MAS) is an enigmatic aspect of autoimmunity defined as occurrence of at least three autoimmune diseases in the same individual. Since the first report by Humbert et al. (1) in 1989, various dermatological autoimmune disorders have been described in association with systemic autoimmune diseases as components of MAS (2). We report here a 58-year-old man who presented with a combination of three autoimmune diseases, i.e. alopecia universalis, insulin-dependent diabetes mellitus and pemphigus vulgaris. To our knowledge, occurrence of these three autoimmune disorders in the same individual has not been reported previously.

CASE REPORT

A 58-year-old man presented with multiple oral erosions and blistering over the scalp and trunk of 6 months' duration. He initially noticed painful erosions on buccal mucosa, shortly followed by development of blisters on normal-looking skin over the scalp and trunk, which ruptured spontaneously in 3–5 days, leaving raw areas and subsequent formation of crusts over them. Except for insulin, there was no history of any drug intake. He had also experienced loss of hair all over the body from the age of 11 years and diabetes mellitus since 32 years for which he was on regular insulin. There were no similar complaints in his family members.

On examination, there were multiple erosions of variable sizes with whitish overhanging edges over buccal mucosa and tongue. Multiple crusted erosions and a few intact flaccid bullae were noticed over the scalp, face and trunk. Bulla spread sign was positive. In addition, there was complete loss of hair all over his body (Fig. 1). With this clinical presentation and history, we made the diagnosis of pemphigus vulgaris with alopecia universalis. Tzanck smear and histopathological examination were consistent with pemphigus vulgaris. He was started on standard dexamethasone cyclophosphamide pulse therapy for pemphigus (3) with which his lesions subsided gradually.

DISCUSSION

The cutaneous autoimmune disorders can be divided into two categories: (i) organ-specific disorders like pemphigus vulgaris and bullous pemphigoid, where the autoimmune injury is confined to the skin and mucosa,

and (ii) organ non-specific disorders like systemic lupus erythematosus where the immunological injury occurs both in skin and internal organs (4). The most common dermatological diseases reported in MAS are vitiligo, alopecia areata, pemphigus, bullous pemphigoid and dermatitis herpetiformis.

The exact pathogenesis of MAS is not known. However, it has been stated that there is an increased tendency for development of a new autoimmune disorder in an individual with a previous history of an autoimmune disease (5). The reason for such a predisposition is not known. A possible explanation given for the development of MAS is the sharing of common genetic elements related to the different diseases (6). MAS is classified into three groups according to the prevalence of their association with one another (7).

In the literature, there are reports of association of pemphigus vulgaris with various autoimmune diseases like thymoma, myasthenia gravis, autoimmune thyroid diseases, alopecia areata and diabetes (8–10). However, the occurrence of alopecia areata (alopecia universalis), pemphigus vulgaris and insulin-dependent diabetes mellitus (IDDM) in the same patient has never been described before. Development of these three



Fig. 1. Loss of hair and crusted erosions on the scalp.

autoimmune diseases in the same individual can be partly explained by the common association of these diseases with HLA class II haplotypes, i.e. HLA DR4. Furthermore, the allele HLA DQB1* 0301 (DQ7), which is a marker for more severe long-standing alopecia universalis (11), is adjacent to the allele HLA DQB1* 0302, which has a strong association with diabetes mellitus and pemphigus vulgaris (DRB1*0402-DQB1*0302) (12). Interestingly, in both IDDM and pemphigus vulgaris susceptibility has been reported to be associated with amino acid substitutions at position 57 of the DQ beta chain (13). So there is a chance of coexistence of alopecia universalis with diabetes mellitus and pemphigus vulgaris. This unique MAS variant poses a therapeutic challenge to the treating physician, as the corticosteroids in the management of pemphigus can aggravate diabetes.

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