CASE REPORT

Development of pemphigus vulgaris in a patient with vitiligo and Hashimoto’s thyroiditis

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ABSTRACT. Pemphigus vulgaris is an autoimmune blistering disease. An association of pemphigus vulgaris with vitiligo or Hashimoto’s thyroiditis has not been reported before. We reported a 38 yr-old female patient with Hashimoto’s thyroiditis and vitiligo who eventually developed pemphigus vulgaris on vitiliginous lesions. A genetic predisposition or a local event on vitiliginous skin may be responsible for the development of pemphigus in this patient.


INTRODUCTION

Pemphigus vulgaris is an autoimmune blistering disease of skin and mucous membranes which is rarely associated with other diseases, such as myasthenia gravis, thymoma and bullous pemphigoid (1, 2). On the other hand, vitiligo is an acquired disease characterized by well circumscribed milky white cutaneous macules and patches which may be associated with some organ specific autoimmune diseases such as autoimmune thyroid diseases, pernicious anemia, Addison’s disease, systemic lupus eritematosis, diabetes mellitus and inflammatory bowel disease (3). However, non-organ specific autoimmune diseases do not seem to be associated with vitiligo with any greater frequency than that found in the general population (4). Here we present a patient with pemphigus vulgaris associated with vitiligo and Hashimoto’s thyroiditis that may be a new autoimmune polyglandular syndrome (APS).

CASE REPORT

A 38-yr-old female patient was admitted to the hospital with ulcerations in her mouth and the body. Past medical history of the patient revealed that she had vitiligo for 10 yr and Hashimoto’s thyroiditis for 6 yr. Her family history did not reveal any autoimmune or cutaneous diseases. On the dermatologic examination, she had severe erosions and ulcerations in her mouth and limited number of ruptured bullae and erosions located in her body. Nikolsky sign was positive. Depigmented vitiliginous patches were present in her face, body and extremities together with acral areas. Overall 10% of the skin was involved, however, interestingly pemphigus lesions were located on the vitiliginous lesions and predominantly at the vitiliginous side of the normal-depigmented skin junction. No pemphigus lesion was detected on the normal skin of the patient. Histopathological and immunoflorescent examination were consistent with pemphigus vulgaris. On palpation thyroid gland mildly enlarged, firm and finely nodular. Routine laboratory examination, including complete blood count, blood and urine biochemistry, chest X-ray, was normal. She had not been receiving any specific treatment for Hashimoto’s thyroiditis except for thyroid replacement therapy with T4-Na 0.175 mg/day. Thyroid stimulating hormone (TSH) was 1.2 (ulU/ml) (normal range 0.35-4.95); T3: 154 ng/dl (normal range 95-190) and T4: 8.12 μg/dl (normal range 5-11). Antithyroid antibodies (antithyroperoxidase and antithyroglobulin) were positive. Thyroid ultrasound showed mildly enlarged thyroid without any nodule. For her pemphigus, therapy with high dose oral prednisolon (2 mg/kg/day) and oral azothioprine (3x50 mg/day) was started. The oral lesions rapidly responded to

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the treatment. However, skin lesions were resistant to the therapy and improved slowly.

DISCUSSION

The occurrence of three or more autoimmune diseases in the same patient is defined as APS. In these cases, common genetic etiologic links are shared by the related diseases (5). As the dermatologic component of this syndrome, vitiligo has an important place (4, 5).

This patient seems to have APS with the components of vitiligo, Hashimoto’s thyroiditis and pemphigus vulgaris. Coexistence of vitiligo and Hashimoto’s thyroiditis is a known association in APS (5). An association with vitiligo and pemphigus was only reported in a pediatric Indian patient before (6). In the present case, Hashimoto’s thyroiditis is also present. The exact mechanism explaining the association of pemphigus vulgaris with vitiligo and Hashimoto’s thyroiditis is not known, however a common autoimmune process may be responsible for the development of this association. In vitiligo, the presence of the human leucocyte antigen (HLA) DR4- DQ1 haplotype, predisposes for autoimmune diseases (7). The HLA DR4-DQ1 haplotype is also present in pemphigus patient (8). This haplotype may cause the production of antibodies which are responsible for the development of all three diseases. However, in the patient, localization of pemphigus lesions on the normal-depigmented skin junction cannot be explained only by autoimmune mechanisms. Some local events occurring in vitiliginous patches must have an additional role for the development of pemphigus lesions. In pemphigus, in addition to antidesmoglein 1 and 3 autoantibodies, cholinergic receptors on the keratinocytes play a role in the development of acantholysis. Cholinergic receptors regulate desmosomal adhesion of keratinocytes by altering the level of expression of both desmoglein 1 and 3 and the phosphorylation status of Dsg3 (9). In vitiliginous skin, decreased acetylcholine esterase levels and thus increased acetylcholine activity have been detected (10). The increased acetylcholine activity in this patient may enhance the appearance of pemphigus lesions. Furthermore, increased production of TNF-α has been detected in perilesional vitiligo patches (11). Similarly, TNF-α was detected as one of the mediators in the blistering process in pemphigus (12). TNF-α produced in vitiliginous patch may induce the development of pemphigus in this patient. Although the pathogenesis not clear, association of pemphigus vulgaris with vitiligo and Hashimoto’s thyroiditis is interesting and as more cases accumulate the mechanisms can be made clearer.

REFERENCES


Fig. 1 - Pemphigus vulgaris lesions located on vitiliginous plaques.