A Report of Multiple Autoimmune Syndrome: Pemphigus Vulgaris Associated with Several Immune-Related Diseases after Thymectomy

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Sir,

The co-occurrence of three or more autoimmune disorders in an individual is called multiple autoimmune syndrome (MAS). According to the combination of diseases, it is divided into three types. Currently, its pathogenesis is still unclear, and environmental factors and genetic factors are related to the occurrence of this disease. Patients with a single autoimmune disease have a 25% risk of developing other autoimmune diseases. The coexistence of several diseases in an individual is rare. Here, we report a patient with thymoma who suffered from myasthenia gravis, vitiligo, pure red cell aplasia, systemic lupus erythematosus, lichen planus, alopecia areata, and pemphigus vulgaris after thymectomy. This is a novel combination of MAS. The presence of thymoma should alert the physician to the possible presence of MAS.

A 64-year-old female presented with an 8-year history of erythema and blisters on the scalp, face, trunk, and upper limbs. The patient had thymectomy 22 years ago, and she had been diagnosed with vitiligo, myasthenia gravis, pure red cell aplasia for 15 years, and systemic lupus erythematosus for nine years. She had been treated with oral prednisolone for both systemic lupus erythematosus and the blistering disorder and the blisters disappeared. During the process of prednisolone tapering, the blisters recurred. Her physical symptoms and past medical conditions were stable when the blistering disease recurred.

On physical examination, the hair on the scalp was almost completely lost [Figure 1]. Scattered erosive erythema with crusting and flaccid blisters were seen on the scalp, trunk, and upper limbs [Figure 2], with a positive Nikolsky’s sign. A number of irregular depigmented patches could be seen on her chest and the face [Figure 3]. The depigmented patches, when exposed to ultraviolet (UV) light, glowed blue [Figure 4]. Irregular longitudinal grooving and ridging of the nail plate, thinning of the nail plate, and shedding of the nail plate with atrophy of the nail bed were also present [Figure 5]. The oral cavity, vulva, and other mucous membranes were not involved. The muscle strength was normal.
Laboratory findings showed the erythrocyte sedimentation rate of 41 mm/h (0–20), lupus anticoagulant of 1.62 (≤1.2), antinuclear antibody (H-type) of 1:640 (<1:40), antidouble-stranded DNA antibody of 263 IU/ml (<100 IU/ml), antidesmoglein 1> 150 U/ml (<20 U/ml), and 91 U/ml (<20) of anti-desmoglein 3. No abnormalities in other blood parameters and urine, liver, and renal functions were detected. A skin biopsy from one of the blisters on her chest showed intraepidermal blister formation and acantholytic keratinocytes [Figure 6].

Based on these results, the patient was diagnosed as MAS—pemphigus vulgaris, myasthenia gravis, vitiligo, pure red cell aplasia, systemic lupus erythematosus, lichen planus and alopecia areata. The patient underwent treatment with methylprednisolone (24 mg/day). After treatment for 1 month, her skin lesions were resolved [Figure 7]. At the time of reporting, she was treated with 8 mg/day methylprednisolone, and the antidesmoglein 1 and 3 antibodies had dropped to 131 U/ml and 72 U/ml, respectively. Her condition was stable.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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