AUTOIMMUNE CONNECTIVE TISSUE DISEASES

AUTOIMMUNE POLYGLANDULAR SYNDROME TYPE IV: COEXISTENCE OF THYMOMA, VITILIGO, MYASTHENIA GRAVIS, SYSTEMIC LUPUS ERYTHEMATOSUS, PEMPHIGUS VULGARIS AND ALOPECIA UNIVERSALIS

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Background: Autoimmune polyglandular syndromes (APS) type IV. The syndromes are defined by the coexistence of at least two autoimmune-mediated endocrinopathies and non-endocrine organs can also be affected. The exact pathogenesis remains unclear.

Observation: A 62-year-old female with a 16-year-history of generalized vitiligo presented to our clinic with erythema, blisters on scalp, trunk and upper extremities for 8 years and hair loss for 4 years. In 2002, multiple hypopigmented macules with circumscribed border had been found on her dorsal side of both hands and gradually affected her face, trunk and extremities. The diagnosis of vitiligo was made but she didn't receive any treatment. In 2010, several flaccid, thin-walled, easily ruptured blisters were found on her face and trunk. She also had arthralgia and multiple oral ulcers. Antinuclear antibodies were 1:1280 positive and anti-dsDNA was positive. She was diagnosed as systemic lupus erythema and she was treated with prednisone. The blisters showed remission. In 2014, hair from all hair-bearing regions were gradually falling off without any obvious reasons. In 2017, after she stopped taking prednisone, the blisters recurred. Biopsy taken from the blister revealed suprabasal cleft with acantholytic cells and a linear palisade of intact basal keratinocytes. Antibodies to desmoglin 1 and 3 were positive. Antinuclear antibodies were positive (1:640) and anti-dsDNA was positive.The rate of erythrocyte sedimentation was elevated to 41 mm/h The patient also had a history of thymoma (the thymoma was resected in 2002), myasthenia gravis, pure red cell aplasia and hypertension. The diagnosis of APS type IV: coexistence of thymoma, vitiligo, myasthenia gravis, systemic lupus erythematosus, pemphigus vulgaris and alopecia universalis was made.

Key message: We report an extremely rare case of 62-year-old female presented with coexistence of six autoimmune disorders successively.