AUTOIMMUNE BULLOUS DISEASES

A CASE OF SUBCORNEAL PUSTULAR DERMATOSIS TYPE OF IGA PEMPHIGUS WITH MONOCLONAL GAMMOPATHYOF UNDETERMINED SIGNIFICANCE

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Background: IgA pemphigus is an exceedingly rare autoimmune blistering disorder.Based on clinical and histopathologicalcharacteristics it can be subdivided into subcorneal dermatosis (SPD) type and intraepidermal neutrophilic (IEN) type. Monoclonal gammopathyof undetermind significance (MGUS) is one of its complications with still unknown mechanism.

Observation: A 39-year-old woman was admitted to our hospital due to recurrent erythema, papule and scale on hertrunkand extremities for 2 years accompanied withpustulesfor 1 years. The lesion was first presented as erythema, scaly papules andpruritus on hertrunk and limbs 2 years ago. She was diagnosed as “allergic dermatitis” and given antihistamines and topical corticosteroid. The lesions regressed but relapsed irregularly. 1 years ago, pustules began to appear on the basis of erythema.Clinical examination revealed multiple erythema, papules on hertrunkand extremities, with pustules in both axillae, inguinal and mammary folds.
Histopathology showed subcorinal pustules with infiltration of neutrophils in the epidermis.Direct immunofluorescence revealed intracellular IgA deposition, while IgG, IgM, C3 were all negative. The serum autoantibodies against demolding 1 and 3 were both negative. Immunoelectrophoresis suggested that IgA and Kappa monoclonal immunoglobulin infiltration zones were intense positive. Bone marrow biopsy was negative.
A diagnosis of SPD-type of IgA pemphigus accompanied with monoclonal gammopathyof undetermined significance were made.The patient received systemic glucocorticoid medication to oral methylprednisolone 40mg per day and the lesions showed quick remission.There was no recurrence on tapering methylprednisolone within three months.

Key message: Coexistence of IgA pemphigus andMGUS is an uncommon conditionwith only a few reports in the literature.As the progression of MGUS to multiple myeloma or other related malignancies occurs at a rate of approximately 1% per year, theperiodic inspection and long-term follow-up is needed.