Atypical Pemphigus with Concomitant IgG and IgA Anti-Intercellular Autoantibodies Associated with Monoclonal IgA Gammopathy

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Key Words

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Abstract

A case is reported of a 60-year-old woman with acantholytic vesiculopustular dermatosis and IgA- λ monoclonal gammopathy. The histopathology of vesiculopustular lesions showed intraepidermal acantholytic and neutrophilic blisters. Direct immunofluorescence revealed intercellular (IC) IgG deposition with concurrent deposits of IgA and C3. Indirect immunofluorescence and immunoblotting studies revealed that the patient had circulating IgG and IgA anti-IC antibodies both of which recognized the 150-kD desmoglein that was pemphigus foliaceus antigen in a bovine desmosome preparation.

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Case Report

A 60-year-old woman was admitted to our hospital in February 1993 with a history of pruritic vesiculopustular eruption of 3 months duration. Her skin lesions were located mainly on the lateral sides of the trunk (fig. 1) and the dorsal parts of hands and fingers. They began as clear vesicles on both normal and erythematous skin and became purulent within a few days. The patient had hyperlipidemia and episodes of angina pectoris. Her family history was not contributory. Mucous membranes were spared. Nikolsky's sign was not elicited. Abnormal laboratory findings on admission included an erythrocyte sedimentation rate of 124 mm/h, serum triglyceride 480 mg/dl (normal 30-150 mg/dl) and serum IgA 1,074 mg/dl (normal 135-340 mg/dl). Immunoelectrophoresis showed a monoclonal IgA- λ gammopathy with no evidence of multiple myeloma. Nonactive hepatitis C was confirmed by serology.

A total of 3 biopsies were taken from the vesiculopustular lesions. A specimen taken from an early lesion showed an acantholytic bulla with a few neutrophils at the lower level than in pemphigus foliaceus.

Fig. 1. A vesiculopustular lesion on the trunk.

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A specimen taken from a relatively old lesion showed an acantholytic pustule in the superficial epidermis.

Direct immunofluorescence revealed intercellular (IC) IgG deposition with concurrent deposits of IgA and C3. Indirect immunofluorescence using normal human skin as substrate revealed that the patient had circulating IgG anti-IC autoantibodies and IgA anti-IC antibodies at titres of 1:160 and 1:40, respectively.

By immunoblotting studies using a bovine desmosome preparation [1], both of IgG and IgA anti-IC antibodies of this patient reacted with a 150-kD desmoglein which has been recognized as pemphigus foliaceus antigen. Immunoblotting studies using human epidermal extract revealed that the IgG antibodies of this patient reacted with the 140-kD protein (between the 150-kD human desmoglein and 130-kD human pemphigus vulgaris antigen), the nature of which is currently unknown.

Dapsone therapy was initiated at a dosage of 50 mg/day. Within a few days the skin lesions improved dramatically. While dapsone was continued, no new lesions appeared, but this therapy had to be stopped because of an increase in peripheral blood reticulocytes. At present the patient is controlled under 1 mg/day of betamethasone. The circulating anti-IC IgG and IgA antibodies remain positive. High levels of serum IgA and monoclonal IgA- λ gammopathy remain unchanged.

Discussion

This case is of particular interest in that both of the mid-epidermal and superficial acantholytic blisters with neutrophils were observed. In addition, indirect immunofluorescence and immunoblotting studies revealed that the serum from this patient contained both IgG and IgA anti-IC antibodies recognizing desmoglein (pemphigus foliaceus antigen) in a bovine desmosome preparation. At the present time, it seems likely that this case represents a rare example of the simultaneous occurrence of pemphigus foliaceus and intercellular IgA vesiculopustular dermatosis [2]. The association of monoclonal IgA gammopathy and dramatic response to dapsone therapy supported the diagnosis of the latter disease. The existence of the IgG anti-IC antibodies recognizing a 140-kD protein which is possibly a new antigen for pemphigus in human epidermal extract complicates the diagnosis of this case.

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