Pemphigus erythematosus (PE) is a variant of pemphigus foliaceus. PE patients exhibit the immunological features of both pemphigus and lupus erythematosus (LE), i.e. granular immunoglobulin (Ig) and C3 deposition in the basement membrane zone (BMZ), intercellular IgG and C3 deposition in the epidermis and the presence of circulating antinuclear antibodies (ANA) (1, 2). This report describes a case of PE in which skin lesions were induced by UVB irradiation.

CASE REPORT

A 62-year-old Japanese woman presented with a 6-month history of pruritic and slightly erythematous lesions with erosions on the nose, chest and extremities (Fig. 1a). The patient had no medical or family history and no history of any drug intake. A histological examination showed detachment of the stratum corneum (Fig. 1c). Infiltration of lymphocytes was observed in the upper dermis. Direct immunofluorescence (IF) demonstrated positive intercellular staining for IgG within the epidermis (Fig. 1d). Granular deposition of IgM and C3 was observed in the BMZ (Fig. 1e). Indirect IF revealed IgG deposition in the intercellular regions of the epidermis (Fig. 1f); however, no deposition of IgM or C3 was observed. The level of serum anti-desmoglein (Dsg) 1 antibodies was remarkably increased (> 150 U/ml). Anti-Dsg3 and anti-BP180 antibodies were negative. The levels of ANA, anti-SS-A antibodies and anti-RNP antibodies were elevated. According to these findings, the patient was diagnosed with PE and started on treatment with a topical corticosteroid. However, the skin lesions were found to be markedly exacerbated the day after exposure to sunlight on a summer day (Fig. 1b). Because this finding suggested that the patient had a photosensitivity, phototesting was performed. The minimal erythema dose (MED) for UVB was 50 mJ/cm², while a dose of 5 J/cm² of UVA irradiation did not induce erythema, indicating normal responses to UV light. After obtaining the patient’s written informed consent, UVB at a dose of 3 MED was irradiated on her back. A skin tissue specimen was taken from the UVB-induced skin lesion 24 h after the UVB irradiation and examined histologically. Acantholytic cells were observed in the upper granular layer, and lymphocytes had infiltrated the upper...
per dermis (Fig. S1a). Direct IF demonstrated positive signals of IgG in the intercellular region of the upper epidermis (Fig. S1b). Furthermore, granular deposition of C3 was observed in the BMZ (Fig. S1c). The patient was treated with oral prednisolone (30 mg/day) and intravenous immunoglobulin (400 mg/kg/day). The skin lesions gradually improved, disappearing within 3 months. The patient continues to receive treatment with 5 mg of prednisolone.

DISCUSSION

The effects of UV on autoimmune blistering skin diseases have been recognised since 1965 (3–5). Two cases of UV-induced PE have also been reported (3, 6). In most cases of pemphigus, UVB has been identified to be the wavelength responsible for inducing the skin lesions (5). Furthermore, it has been reported that photoprovocation of UVB can induce pemphigus skin lesions histologically and immunohistologically (7). On the other hand, photosensitivity is also known to be a major symptom of LE. Exposure to sunlight not only induces cutaneous LE lesions, but also aggravates systemic disease symptoms in LE patients (8). The action spectra observed in LE patients are commonly located in the UVB and/or UVA range, and irradiation of UV has been shown to be experimentally induce LE skin lesions within a few days (9).

In the present case, the skin lesions induced by UVB irradiation contained acantholytic cells and exhibited deposition of IgG in the upper epidermis, characteristics of PF. In addition, lymphocyte infiltration in the upper dermis and granular deposition of C3 in the BMZ were observed in the same skin tissue specimen. These findings are consistent with those of early-stage LE. Therefore, we speculate that the development of skin lesions due to UVB irradiation is associated with the immunological effects of both PF and LE in PE patients. Although further studies are required to obtain a better understanding of the pathomechanisms underlying the development of UV-induced PE, the present case contributes to extending the knowledge of PE.

The authors declare no conflicts of interest.

REFERENCES