◇Case Report

〈本文p.1069～1072〉

A case of pemphigus vulgaris of gingiva

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Abstract
We described a 52-year-old female with a 3-month history of erosions in gingival margin. The histopathological findings of the gingival lesion showed acantholytic cells, a typical feature of pemphigus vulgaris. Direct immunofluorescence of a gingival biopsy showed intercellular staining with IgG and C3. Her serum antibodies against desmoglein-3 was positive. A diagnosis of pemphigus vulgaris was made, and gingival lesions were improved with oral prednisolone 0.5mg/kg daily. There is no recurrence of gingival erosions and no skin lesions of pemphigus vulgaris at present. In cases of intractable erosions of gingiva, pemphigus vulgaris should be considered as the differential diagnosis.

〈本文p.1073～1076〉

Pemphigus vulgaris with hyperkeratotic leisions on fingers

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Abstract
A 77-year-old woman developed pemphigus vulgaris with multiple hyperkeratotic lesions on her fingers. Histological examination of a biopsy taken from the lesions showed acantholysis and liquefaction degeneration. One year before the onset of pemphigus, edematous erythema was developed on her back. Histological examination of the erythema showed liquefaction degeneration and dermal mucinosis, indicating possible dermatomyositis. Although definite diagnosis of dermatomyositis could not be made, the hyperkeratotic lesions might arise from some damage to the basal layer with pemphigus.

〈本文p.1077～1080〉

A case of mucocutaneous pemphigus vulgaris

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Abstruct
A 55-year-old female with mucocutaneous pemphigus vulgaris (PV). Corticosteroid mono-therapy and combined therapy of other immunosuppressive agents were not effective. We treat by combination of corticosteroid, double filtration plasmapheresis (DFPP) and high-dose intravenous immunoglobulin (IVIG). These combined therapy was effective to our patient. Particularly we recognized that the additional IVIG treatment was useful for refractory mucocutaneous PV.

◇Case Report

〈本文p.1081～1084〉

A case of childhood pemphigus vulgaris successfully treated with oral prednisolone

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Abstract

A 7-year-old boy presented with a 3-year history of sore throat and erosion of the lip and oral cavity. Pemphigus vulgaris was suspected at previous hospital and referred to our out-patient clinic. He had severe mucosal lesions and pemphigus disease area index was 24. Anti-desmoglein 3 antibody ELISA was positive, and biopsy from the lip showed acantholysis in histologically and IgG deposition by direct immunofluorescence. He was treated with 1mg/kg/day systemic predonisolone, and erosions were relieved with 2 weeks. It led to complete remission in the seven months from the start of treatment. Because it is not easy to perform skin biopsy in children, serum test was considered to be useful.

〈本文p.1085～1088〉

Neutropenia induced by after intravenous immunoglobulin treatment on a pemphigus vulgaris patient

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Abstract

A 43-year-old man was referred to us with erythematous macules, blister formations, erosions on the scalp, face and trunk, and erosions on the gingival mucosa. Direct immunofluorescence showed positive reactions of IgG and C3 on the intracellular regions of the epidermal keratinocytes. The eruptions are diagnosed as pemphigus vulgaris. We applied intravenous immunoglobulin because of insufficient effectiveness of prednisolone. Immediate severe neutropenia occurred after first application. However, it did not occur after second and third application with other type of intravenous immunoglobulin. We should be aware of immediate severe neutropenia by intravenous immunoglobulin even though recurrences may not be induced.

◇Case Report

〈本文p.1089～1092〉

A case of pemphigus vulgaris with severe erosions in the oral mucosa successfully treated with high-dose intravenous immunoglobulin

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Abstract
A 81-year-old woman presented with multiple erosions in the oral mucosa. Results of histopathology, DIF and ELISA assay allowed us to make a diagnosis of pemphigus vulgaris. We started treating her with oral prednisolone and cyclosporine. Since these therapies were not satisfactory in the effect on the oral erosions, high doses of intravenous immunoglobulin (IVIG) therapy were applied. An improvement of her mucous lesions and a decrease of pathogenic anti-desmoglein 3 antibodies were obtained after IVIG treatment. This case suggested that IVIG is an effective and additive therapy for severe pemphigus.

〈本文p.1093～1096〉

A case of exacerbated pemphigus foliaceus at the site of radiation therapy for breast cancer

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Abstract
A 63-year-old woman with 3-year history of pemphigus foliaceus (PF), underwent postoperative radiation therapy for breast cancer. Approximately four weeks later, she developed scaly erythema, localized to the irradiated area of the left breast. A histological examination revealed a split in the subcorneal layer, and direct immunofluorescence showed the intercellular deposition of IgG. The re-elevation of anti-Dsg1 antibody levels, as confirmed by ELISA, supported the differential diagnosis of exacerbated PF, distinguishing it from radiation dermatitis. Subsequently, treatment with topical corticosteroids was deemed effective. This case suggests that radiation causes impairment of intercellular connections and interference with immunosurveillance and that it may promote the production of anti-Dsg1 antibodies.

〈本文p.1097～1100〉

A case of pemphigus foliaceus that showed eruptions of pemphigus herpetiformis during course

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Abstract
A 55-year-old woman. Erythema, blister and, erosions developed on her face, limbs, and the trunk in 2008. She was diagnosed as having pemphigus foliaceus from positive antidesmoglein 1 antibody and the results of the skin biopsy. In March, 2014, strong itchy erythema with vesicles appeared on her legs. On histopathology,intraepidermal blisters and eosinophilic spongiosis were seen. Direct immunofluorescence showed deposition of IgG and C3 on the cell surfaces of epidermal keratinocytes. Antidesmoglein 1 antibody was positive, but antidesmoglein 3 was negative. From the characteristic clinical features,histopathology and immunohistochemistry,we diagnosed this case as transition from pemphigus foliaceus to pemphigus herpetiformis.

◇Case Report

〈本文p.1101～1104〉

A case of pemphigus foliaceus with marked acanthosis

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Abstract

A 22-year-old male developed pruritic erythema on his face and trunk. Histopathologic findings showed parakeratosis and marked acanthosis with cleavage of the granular layer. Direct immunofluorescence studies revealed intercellular depositions of IgG and C3 in the epidermis. His serum level of anti-desmoglein 1 IgG was elevated. Based on clinical, histopathological and immunological findings, we diagnosed this case as pemphigus foliaceus with marked acanthosis.Clinical symptoms were improved with oral DDS application.

〈本文p.1105～1108〉

A case of pemphigus erythematosus

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Abstract

A 34-year-old male with history of pink scaling plaques on his face for more than 4 years visited our hospital. His chest and back were subsequently involved. Laboratory finding were positive for anti-desmoglein 1 antibody, but negative for anti-nuclear antibody. Histological findings disclosed vesicles in the upper epidermis and acantholytic cells. In addition, direct and indirect immunofluorescence showed intracellular deposition of IgG. Based on these clinical and histopathologic findings, the diagnosis of pemphigus erythematosus was made. The lesions were eventually healed in a month with an oral administration of prednisolone.

〈本文p.1109～1112〉

Herpetiform pemphigus induced by bucillamine

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Abstract

A 89-year-old woman who had been treated with oral bucillamine for rheumatoid arthritis for 7 months developed skin lesions. Physical examination revealed vesicles and annular exudative erythemas over her trunk and extremities. Histopathology showed typical eosinophilic spongiosis with intraepidermal blisters. Indirect immunofluorescence of human skin showed antibodies to cell surfaces. Enzyme-linked immunosorbent assays (ELISA) detected anti-desmoglein (Dsg) 1 antibodies, but antibodies to Dsg3 and BP180NC16a were negative. The diagnosis of bucillamine -induced herpetiform pemphigus was made. As skin lesions remained 3 weeks after discontinuation of bucillamine, prednisolone was administrated at a dosage of 30 mg/day, resulting gradual disappearance of skin lesions. ELISA indices of Dsg1 decreased to normal levels.

◇Case Report

〈本文p.1113～1116〉

A case of paraneoplastic pemphigus with acquired hemophilia

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Shimokawa, Mitsuhiro[1] Fujii, Kazuyasu[1] Higashi, Yuko[1] Aoyama, Yumi[2] Kanekura, Takuro[1]

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Abstract

A 76-year-old man presented with blisters and erythema on his limb and trunk. Histopathologically, there is a subepidermal blister with acantholytic cells. Direct immunofluorescence studies showed the deposition of complement in the epidermal basement zone. Indirect immunofluorescence studies showed the deposition of immunoglobulin G on stained sections of rat urinary bladder epithelium. A diagnosis of paraneoplastic pemphigus was made. There after, he had a bleeding in his mouse and nose. Hematological examination revealed prolonged activated partial thromboplastintime (APTT), decreased coagulation activity of factor Vlll., and Vlll inhibitor in his serum. The patient was given a diagnosis of acquired hemophilia.

〈本文p.1117～1120〉

A case of bucillamine-induced herpetiform pemphigus improved by drug discontinuation

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Abstract

A 77-year-old male with rheumatoid arthritis who had been treated by systemic bucillamine for 10 months developed wide-spread scaly erythema and erosion on his whole body. Histological examination revealed eosinophilic spongiosis and intraepidermal bulla without acantholysis. Direct immunofluorescence showed IgG deposition in the intercellular space of the epidermis. Anti-desmoglein 1 (Dsg1) antibodies were positive and drug lymphocyte stimulation test (DLST) was also positive for bucillamine. Based on these findings, we made a diagnosis of herpetiform pemphigus induced by bucillamine. After switching bucillamine to salazosulfapyridine, the eruption improved without additional systemic treatment accompanied with decreasing of the Dsg1 antibody titer. The eruption completely relieved and antibody titer became negative in 3 months.

◇Case Report

〈本文p.1121～1124〉

A case of pemphigus vegetans ― relation between clinical features and histopathological findings ―

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Abstract

A 66-year-old female had papillomatous plaques with itch on the left groin and the vulva. Histological examination revealed acantholysis and eosinophilic abscesses in epidermis at peripheral part of the eruption. In central part, the dermis showed edema in dermal papillae with eosinophil infiltration. Direct immunofluorescence demonstrated IgG deposition in the intercellular space in the epidermis, and the diagnosis of pemphigus vegetans (PVe) was determined. A biopsy taken from a healed lesion showed remaining papillomatosis with reduction of edema and eosinophils. Our results suggested that vegetating lesions in PVe consisted of acanthosis, papillomatosis, and dermal edema.