Association of Cutaneous T Cell Lymphoma and Bullous Pemphigoid

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Sir,

The association of cutaneous T cell malignant lymphoma (CTCL) and a vesiculobullous disease is extremely rare. Kaposi [1] in 1887 presented the first report of a pemphigus-like dermal lesion associated with mycosis fungoides. Since then a few associations of CTCL and other autoimmune bullous diseases have been published but according to the literature the association of CTCL with bullous pemphigoid has not been reported previously. We have diagnosed the joint occurrence of these two diseases in a 61-year-old woman.

Case History. Pyelonephritis and chronic uremia have been known for 5 years. In March 1987, there was an attack of herpes zoster on the right side of the trunk. The maculous dermal symptoms accompanying the pruritus have persisted essentially unchanged since then. in spite of oral antihistamine and local steroid treatment.

On admission the skin was dry and scaly. Pigmented brownish plaques, partly livid, irregular in size and shape, and slightly elevated, were seen more on the trunk. back and abdomen and less on the extremities. Their surface was scaly or eczematous and in some places the plaques were infiltrated. The patient was not taking drugs regularly. Other physical examinations revealed no pathological changes.

Laboratory studies showed moderately elevated BSR (20 mm/h). renal function tests (CN: 20.4 $\mu\text{mol/l}$, serum creatinine: 389 $\mu\text{mol/l}$) and 16% eosinophilia in the peripheral blood smear. All the routine laboratory findings were within normal range. In the urine *Escherichia coli* and *Proteus vulgaris* infections were detected which were treated with erythromycin (2.000 mg daily), on the basis of the antibiogram.

Because of the basic disease (CTCL) , a treatment with 90 mg/day of oral prednisolone and a single dose of 25 mg methotrexate (MTX) was initiated. On the 6th day following the MTX therapy, the patient began to complain of a burning sensation in the lips and of a mild erythema all over the body. Lentil-sized, tense vesicles appeared on the left flexor forearm, abdomen, waist and on the edge of the tongue. The Nikolsky sign was negative. Since the possibility of a drug side effect, but also of an autoimmune vesiculobullous disease arose as concerns the symptoms, a histology of the bullous lesion was performed too. The histologic picture of the plaque was interpreted as mycosis fungoides stage I–II.

The histologic sections of the bullous process were diagnosed as bullous pemphigoid. Direct immunofluorescence shows an intense linear band of IgG along the basement membrane zone (fig. 1). IIFtest was negative. Therapy: in response to an orally administered tapered dose of steroid, complete recovery was observed in the bullous dermal process within 1 month.

After 1 month she received PUVA therapy (44 J/cm²) and oral therapy with 25 mg MTX was repeated, given weekly for 3 months. The patient, who is currently still under care, is in good general condition. She receives 15 mg prednisolone alternately every second day. The bullous disease has not recurred, and the plaques are undergoing a progressive slight regression.

Discussion. Bullous pemphigoid was first differentiated from other bullous disease entities by Lever in 1953. Bullous pemphigoid is a disease of the elderly. In 80% of the cases it occurs in subjects aged above 60, primarily in men.

Cutaneous T cell malignant lymphoma was first described by Ali-bert in 1906. The pathomechanism of the disease can be explained by the conception of 'skin associated lymphoid tissue' [2].

Fig. 1. Direct immunofluorescence examination of the histologic section of the bullous process shows an intense linear band of IgG along the basement membrane zone.

We subsequently tried to establish why the two diseases are associated. Hodge et al. [3] observed that those cases of bullous pemphigoid, where circulating antibody cannot be detected in the serum, occurred as a paraneoplastic symptom. They found a tumor in 23% of such seronegative cases (our patient falls also in this category), whereas this was the situation in only 4% of seronegative cases. They explain the phenomenon in terms of antigen-binding capacity of the tumor.

It is also possible that autoantibody formation giving rise to bullous pemphigoid is initiated by T helper cells present in the CTCL infiltrate. In the course of the process the T helper cells recognize the bullous pemphigoid antigen located in the membrane of the keratinocytes as a foreign substance, and this induces autoantibody formation.

We regard this case as worthy of publication because of the particularly rare association.

References

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