Pathogenesis of Pemphigus Erythematous

S. Jabłońska, T. Chorzelski, M. Błaszczyk, and W. Maciejewski

Department of Dermatology, Warsaw School of Medicine, Koszykowa 82a, PL-02-008 Warsaw, Poland

Summary. Immunofluorescence studies were made by the indirect method in 54 cases of pemphigus erythematous, in 50 of which skin specimens from light-exposed and unexposed regions were investigated also by the direct IF method. IF Band was shown to be demonstrable in skin specimens from exposed regions in 81% of cases and from unexposed regions in 23%. ANA were found in some 31% of patients, though usually in titers below those of IC antibodies. There were 2 cases each of coexistence with myasthenia gravis and thymoma and with SLE. Virus-like particles, however, were found by electron microscopy only in 1 case with coexisting SLE. Detection of IF Band in skin specimens from a significant majority of patients with pemphigus erythematous, presence of ANA in some, and occasional coexistence of SLE suggest some relation of the disease with lupus erythematous.


Pemphigus erythematous (Senear-Usher syndrome) gave rise to much debate from the time it was first reported (1926); it was seen as a variety of lupus erythematous by some authors, and of pemphigus by others. Detection of acantholysis and in-vivo
bound circulating antibodies settled the argument in favour of the latter (Beutner et al., 1970). Detection of immunofluorescence (IF) Band in specimens of the facial and exposed skin and, in some cases, antinuclear antibodies (ANA) (Chorzelski et al., 1968) nevertheless suggested the coexistence of immunological abnormalities characteristic of lupus erythematosus. The findings were later confirmed by several authors (Bean and Lynch, 1970; Meneghini et al., 1970; Orfanos et al., 1971; Gianetti, 1973, 1974). In 4 of Gianetti's 6 cases (1974) there was indeed IF Band in skin specimens from the face, but ANA could be demonstrated in none. Preliminary electron-microscopic studies of 3 cases failed to reveal the virus-like particles characteristic of LE. He therefore sees no convincing immunological and electron microscopical grounds for considering the Senear-Usher syndrome a form of transition between lupus erythematosus and pemphigus.

In the present study it was intended to find out
a) whether IF Band in skin specimens from the exposed regions can be confirmed;
b) whether they can be found in specimens from the unexposed regions also;
c) the proportion of cases in which antinuclear antibodies can be demonstrated;
d) whether the virus-like particles characteristic of lupus erythematosus can be demonstrated in lesions that show IF Band; and
e) whether cases can be found in which pemphigus erythematosus and systemic lupus erythematosus genuinely coexist.

Material and Methods

Our material comprises 54 cases (25 men and 29 women) of pemphigus erythematosus, aged 22–88 years. Fifty four patients were checked for circulating intercellular and antinuclear antibodies (IC and ANA) by the indirect immunofluorescence (IF) method. In a total of 50 cases skin specimens from light-exposed and/or unexposed regions (32 and 48 patients respectively) were investigated by the direct IF method.

The procedure was as described by Beutner et al. (1973), and the parameters of the conjugates were as follows:
Conjugate goat anti-human IgG. U/ml 4, F:P molar 4.9, dilution for use 1/16 (1/4 U/ml).
Conjugate rabbit anti-human fraction II U/ml 8 F:P molar 2.4, dilution for use 1/32 (1/4 U/ml).

For detecting virus-like particles electron microscopy was used in 6 cases and, for comparison, in two of pemphigus herpetiformis.

The electron-microscopic technique was as follows:
Skin fragments cut into small pieces were fixed 2 h in 5% glutaraldehyde with 0.1 M phosphate buffer of pH 7.4, and after washing with buffer postfixed with 1% OsO4 in phosphate buffer during 2 h. Sections washed several times in buffer were dehydrated in rising concentrations of ethanol and propylene oxide, and embedded in Epon 812 (according to Luft, 1961).
Tissue blocks were cut with a glass knife on a Porter-Blum MT2 microtome into sections 30–40 nm thick.
The specimens were counterstained with uranyl acetate and lead citrate.
Stained sections were observed in an electron microscope (JEM 7).

Results

Immunoglobulins were invariably found in the intercellular spaces of specimens from exposed regions as well as, with the exception of 1 case, unexposed ones IF. Band