PEMPHIGUS FOLIACEUS
(A case report)

RADHA RANI AGGARWAL * SUDARSHAN GUPTA † AND RAJ KUMAR GARG ‡

Summary

A case of pemphigus foliaceus clinically resembling dermatitis-herpetiformis is being reported. Patient did not respond to dapsone. Later on the basis of histology a diagnosis of pemphigus f liaceus was established. Cases having overlapping features of pemphigus foliaceus and dermatitis herpetiformis have been reported earlier by other workers.

Civatte[1] reported association of primary acantholysis with pemphigus and acantholysis has become the key to diagnosis of all types of pemphigus cases. Tzanck in 1948[2] described a very simple technique to confirm acantholysis in suspected cases of pemphigus. Rook AJ[3],[4] has also stressed the importance of primary acantholysis in the diagnosis of pemphigus. Presence of primary acantholysis is a must in all types of pemphigus. The diagnosis of bullous dermatitis herpetiformis is similarly diagnosed on histological grounds when subepidermal, nonacantholytic bullae are evident. However, Floden et al[5] believe that acantholytic process can exist in dermatitis herpetiformis. Biddle[6] has reported cases which started initially as dermatitis herpetiformis and later evolved into an eruption resembling pemphigus foliaceus and having malignant course. In Low's[7] opinion the two diseases were often interchangeable both in appearance and clinical course. Pemphigus vulgaris presenting as dermatitis herpetiformis has also been reported by Rattan Singh in 1973. Therefore a mixed clinical picture is occasionally seen and we should be aware of this.

* Asst Professor in Skin and VD
† Associate Professor, Pathology
‡ Senior Lecturer in Skin and VD
Received for publication on 1—5—1978

Case Report

A 60 years old male patient was admitted with the complaints of vesiculo-bullous lesions and intense itching for 4 months. Patient first developed few vesicles and bullae with moderate itching on the chest. Similar lesions then appeared on lower legs, buttocks, back and upper limbs. There was no history of involvement of muous membranes. After 2-3 days bullae used to rupture with oozing and crusting. Itching used to become intense after few days. There was no history of fever, allergy, drug ingestion, diabetes mellitus, anorexia or loss of weight.

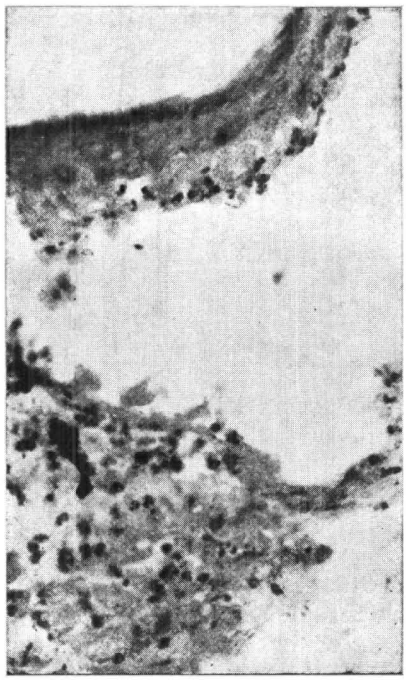
General and systemic examination were normal.

Dermatological examination showed grouped, crusted and vesiculo-bullous lesions on lower limbs, upper limbs, back, abdomen and chest. Bullae were flaccid in certain areas and tense in others. In some areas vesicles were seen to form circinate lesions with serpiginous margins. Post dermatitic hypopigmentation and a few scars were

present in healed areas. Flexural areas as well as palms and soles were uninvolved. Hair and nails were normal. Nikolsky's sign was positive.

Routine investigations were normal.

Histopathology was consistent with a diagnosis of pemphigus foliaceus (Fig).



Subcorneal bulla showing many acantholytic cells. Some acantholytic cells are attached to the roof of the bulla.

Discussion

Winklemann in 1960[8] reported two cases of bullous dermatoses which were diagnosed as dermatitis herpetiformis but clinically showed primary acantholysis on cytological and histological examination. Both patients responded to sulfonamides and dapsone. These cases illustrate the problem with regard to the nosology in these circumstances. Perhaps with more clinical reports these cases may emerge as a new disease entity. Although acantholysis is a fundamental part of the histological picture of pemphigus, yet it is seen in other diseases like chronic benign familial pemphigus, keratosis folli cularis, senile keratosis and viral vesiculobullous diseases.

References

1. Civatte A : Diagnostic histopathologique de la dermatitic polymorphe deuloureuse a maladie de Duhring - Brooq, Ann dermat et syph, 8 : 1, 1943.
2. Tzanck A : Le cytodiagnostic immediat en dermatologie, Ann dermat et syph. 8 : 205, 1948.
3. Rook A and Waddington E : Pemphigus and Pemphigoid, Brit J Derm, 65 : 425, 1952.
4. Rook AJ and Whimster IW : The histological diagnosis of Pemphigus, Brit J Derm, 62 : 443, 1950.
5. Floden CH and Centele H : A case of clinically typical dermatitis herpetiformis (Duhring) presenting acantholysis, Acta Dermat-venereol, 35 : 128, 1955.
6. Biddle AP : Pemphigus foliaceus or Dermatitis herpetiformis Duhring J Cutan Dis, 15 : 203, 1897.
7. Low RC : Pemphigus foliaceus, Brit J Derm. 21 : 101, 1909.
8. Winkelmann RK, Roth HL : Dermatitis herpetiformis with acantholysis or Pemphigus with response to sulphonamides, Arch Derm, 82 : 385 ,960.
9. Singh R, Pandhi RK, Pal D, et al : Pemphigus vulgaris presenting as dermatitis herpetiformis, Indian J Derm Ven, 39 : 84, 1973.