PEMPHIGUS VULGARIS PRESENTING AS DERMATITIS HERPETIFORMIS

RATAN SINGH, R. K. PANDHI, DHARAM PAL AND GYANESHWAR KALLA

Summary

A case of pemphigus vulgaris with clinical features of dermatitis herpetiformis, which showed acute exacerbations to potassium iodide and dramatic response to both di-amino-diphenyl sulfone and corticosteroids, is reported. Some of the other atypical features of this case are presented and discussed.

On the basis of histopathological criteria of intra-epidermal cleavage and acantholysis as laid by Civatte[1] and Tzanck[2], the diagnosis of pemphigus became very easy, which till 30 years ago was the best imbroglio in dermatology. In the majority of pemphigus cases, a reliable diagnosis can be made by combining the clinical features with histological appearance of an early bulla. However, considerable difficulty may be experienced in arriving at a diagnosis, particularly in some early cases of pemphigus foliaceus which masquerade as dermatitis herpetiformis (Floden and Gentele[3], Winkelmann and Roth[4], Doepfmer[5], and Sneddon and Church[6], Brocq[7]), regarded such cases as variants of dermatitis herpetiformis, while others (Hallopeau and Jousset[8], Low[9], and Senear[10]), believed that in certain cases, dermatitis herpetiformis may subsequently get transformed into pemphigus foliaceus.

In view of the fact that such case reports are rare in literature and that all such cases reported so far were pemphigus foliaceus, we would like to place on record, a case of pemphigus vulgaris which presented with clinical features suggestive of dermatitis herpetiformis.

Department of Dermatology and Venereology, Maulana Azad Medical College and Associated Irwin and G. B. Pant Hospitals, New Delhi-1

Received for Publication on 9—9—1972

Case Report

A 28 years old male was admitted to Skin ward of Irwin Hospital in November, 1970 with intensely itchy vesiculo-bullous eruptions, starting from the ankles and later involving the legs, arms, and trunk, with relapses and remissions for the last six years. Oral mucous membrane was never involved. He was treated variously with indigenous and allopathic systems of medicines but without much relief. The present episode started one month back.

Examination revealed an apparently healthy young man with oozing and crusted lesions mainly on the legs and thighs and a few on the upper arms and back. Few tense vesicular lesions were also present. Bulla-spread sign was doubtful. Some lesions on the thighs showed groupism. Few scattered pigmented macules were present at the sites of healed lesions. Oral mucous membrane showed pigmentation only but no ulceration was seen.

Investigations

Hb-12 gm%. Total leukocyte count 7500/cmm with Polymorphs 60%, Lymphocytes 35% and Eosinophils 5%. Urine and Stools-N.A.D. Smear examination from the floor of vesicle did not show any acantholytic cells or eosinophils.

Skin biopsy

An intact early vesicle was biopsied. It showed intraepidermal cleft in mid-epidermis containing essentially polymorphs. No acantholytic cells were seen. A repeat biopsy after one week showed identical features (Fig. 1).

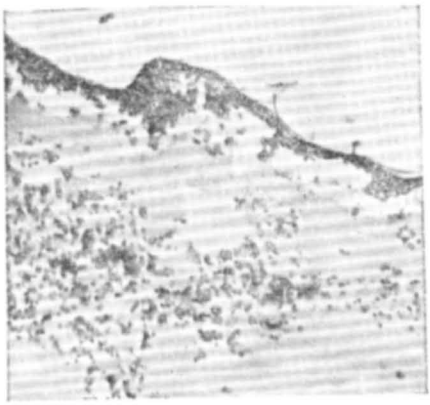

Fig. 1

Photomicrograph of an early intact vesicle, showing intraepidermal cleft in the mid-epidermis containing essentially the polymorphs and no acantholytic cells

Patient was given 3 doses of mist. Potassium iodide (½ G/dose). The following day, he developed multiple, intensely itchy vesiculo bullous lesions on the trunk. Bulla spread sign was positive. Acantholytic cells by Tzanck test[2], were seen. One of these early intact vesicles was biopsied. It showed suprabasal bulla with villi, containing acantholytic cells resembling pemphigus vulgaris. (Fig. 2).

Patient was put on di-amino-diphenyl sulfone, 100 mg twice a day. Condys' compresses and silver nitrate 1% was

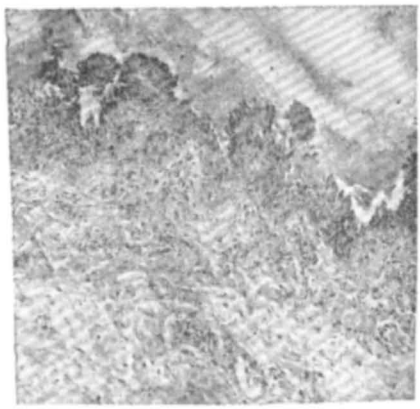

Fig. 2

Photomicrograph of a vesicle after giving potassium iodide. It shows supra-basal cleft with villi and acantholytic cells

given topically. Within two days, his skin condition showed dramatic improvement. He was discharged on 5-2-71 and was advised to continue di-amino-diphenyl sulfone, 50 mg daily but he discontinued it after 7 days. He remained alright for one month, when he relapsed and was hospitalised. He again showed severe exacerbations with potassium iodide and dramatic response to di-amino-diphenyl sulfone. Repeat biopsy showed intra-epidermal bulla, located at subcorneal and midepidermal sites, containing acantholytic cells (Figs 3 and 4 respectively). Patient was re-admitted for the third time, after a month with acute flare-up of the disease process. This time he was put on corticosteroids and showed dramatic response.

Comments

It has long been recognised that iodides dramatically provoke most patients with dermatitis herpetiformis. The present case also showed acute exacerbation on ingestion of potassium iodide though the bulla so produced was intra-epidermal with acantholysis, which is diagnostic of pemphigus. The two earlier skin biopsy specimens showed only intra-epidermal pustule with no

Fig. 3

Photomicrograph of a repeat biopsy after ingestion of potassium iodide showing subcorneal split

Fig. 4

Another section from the same biopsy, showing a mid-epidermal cleft

acantholysis. Similar observations have been made by Sneddon and Church[6] but in their cases subepidermal bulla was seen earlier and typical histology of pemphigus was seen subsequently. Like most patients of dermatitis herpetiformis, this patient also showed dramatic improvement with di-amino-diphenyl sulfone. Better response to di-amino-diphenyl sulfone and sulphamethoxy-pyridazine than to corticosteroids, in cases of pemphigus presenting as dermatitis herpetiformis has been reported by Winkelmann and Roth[4] and Doepfmer[5]. However, the cases reported by Sneddon and Church[6] did not respond to di-amino-diphenyl sulfone or sulphamethoxy pyridazine, but showed dramatic response to corticosteroids. During third relapse, this patient was treated with corticosteroids and equally gratifying results were obtained.

The third biopsy (Fig. 2) showed suprabasal bulla with villi and acantholytic cells, typical of pemphigus vulgaris. The authors are not aware of any earlier record of pemphigus vulgaris presenting as dermatitis herpetiformis. However, on re-admission, subcorneal bulla with acantholysis was seen (Fig. 3) Is it that pemphigus vulgaris is getting transformed into pemphigus foliaceus, an uncommon feature ; the evolution in the reverse direction is known. However, for pemphigus vulgaris, to remain benign in absence of any specific treatment and without involving the mucous membrane for the last six years is also unusual.

Acknowledgments

The authors are thankful to the Director-Principal, Maulana Azad Medical College and Associated Irwin and G. B. Pant Hospitals, New Delhi for permission to publish this case report. Our thanks are also due to department of Pathology, Maulana Azad Medical College, New Delhi, for histopathological studies of this case.

REFERENCE

1. Civatte A : Diagnostic histopathologique de la dermatite polymorphe douloureuse ou maladie de Duhring - Brocq. Ann Derm et Syph 3 : 1, 1943.
2. Tzanck A : Le Cytodiagnostic immediat en dermatologie. Ann dermat et Syph 8 : 205. 1948.

3. Floden CH and Gentele HE : A case of clinically typical dermatitis herpetiformis presenting acantholysis, Acta derm venereol (Stokh) 35 : 128, 1955.

4. Winkelmann RK, Roth HL: Dermatitis herpetiformis with acantholysis or pemphigus with response to sulfonamides. Arch Dermat 82 : 385, 1960.

5. Doepfmer R : Uber eine nosologisch unge Klarte bullose Dermatose (Pemphigus Chronicus Vulgaris oder Dermatitis herpetiformis). Hautarzt 12 : 452. 1961.

6. Sneddon Ian and Church Ronald : Pemphigus foliaceus presenting as dermatitis herpetiformis. Acta derm venereol 47 : 440, 1967.

7. Brocq L : Des rapports qui existent entre la dermatite herpetiforme et les affections dites a lheure actuelle Pemphigus foliace. Ann Derm et Syph (3rd series) 3 : 1245, 1892.

8. Hallopeau H and Jousset A : Sur Un noveau cas de Pemphigus foliace Consecutif a une dermatite herpetiforme. Ann Derm et Syph (3rd series) 7 : 370, 1896.

9. Low RC : Pemphigus foliaceus. Brit J Dermat 21 : 101, 1909.

10. Senear FE : Chronic pemphigus vulgaris. Arch Derm et Syph 65 : 429, 1952.

False

The immediate cause of greying was a great reduction in the number of, or total absence of normal melanocytes. Degeneration of pigment cells were shown as marked vaculation of the pigment cells with poorly organised subcellular constituents. Melanosomes, premelanosomes and mitochondria were reduced in number or structurally altered. Largerhans cells were not demonstrated. No alteration in pigment transfer was seen. The striking vacuolation in the cells is a morphologic expression of the frustrated attempt at maturation of pigmentary structures.

Reference : Arch Klin U Exper Dermat 236 : 368, 1970.