

ORIGINAL CONTRIBUTIONS

A CLINICO - PATHOLOGICAL STUDY OF 70 CASES OF PEMPHIGUS

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A clinicopathological study of 70 cases of pemphigus observed over a span of four and a half years from January 1992 to June 1996 at the Sir J.J. Group of Hospitals and Grant Medical College, Mumbai is reported. Pemphigus vulgaris constituted the single largest group of 43 cases, followed by pemphigus foliaceus (25 cases) and pemphigus vegetans (2 cases). Majority of the cases were seen in the age group of 21-60 years, with a slight male predominance. The youngest patient was 14 years while the eldest was aged 75 years. Mucosal involvement was seen in 31 cases of pemphigus vulgaris, as opposed to only 5 cases of pemphigus foliaceus. Flaccid bullae were present in 100% cases. Pruritus was complained of in 14 cases, though it was more common in pemphigus vegetans and vulgaris.

Salient histopathological features of pemphigus vulgaris observed were (1) intraepidermal suprabasal blisters (35 cases), (2) presence of acantholytic cells (40 cases), (3) "Row of tombstone appearance" (18 cases) and (4) acantholysis involving follicular sheath (20 cases). Main histopathological features of pemphigus foliaceus were (1) subcorneal blister (15 case), (2) acantholysis (24 cases) and (3) bulla cavity containing inflammatory infiltrate (12 cases). Both cases of pemphigus vegetans showed hyperkeratosis, papillomatosis and irregular acanthosis with intra-epidermal eosinophilic abscesses besides suprabasal lacunae.

Key Words : Pemphigus vulgaris, Pemphigus foliaceus, Pemphigus vegetans

Introduction

Pemphigus is a rare vesiculo-bullous disease having a grave prognosis. Acantholytic cells and clefts or bullae are formed in the epidermis as a result of interaction of antibodies with epidermal intercellular cement substance. In 1880, Auspitz recorded the histological findings of pemphigus blister and coined the term acantholysis.¹

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Lever has classified pemphigus into two categories based on the level of blisters. Pemphigus vegetans was considered a variant of pemphigus vulgaris with suprabasal clefts. Pemphigus erythematosus was considered a variant of pemphigus foliaceus due to a subcorneal bulla.

Though the disease is not uncommon in dermatological practice, only a few reports on this disease have been reported in Indian literature, especially from Northern India. The present study of 70 cases intends to highlight various clinicopathological aspects of the disease as it occurs in Mumbai region of Western Maharashtra.

Materials and Methods

This study comprises of 70 cases of pemphigus seen at the Şir J.J. Group of Hospitals and Grant Medical College observed over four and a half years (January 1992 to June 1996).

A detailed history with particular reference to the mode of onset, characteristics and distribution of the lesions was taken. In addition to the routine hematological and urine investigations, elliptical skin biopsy of early lesion and surrounding skin tissue of all 70 cases were processed by paraffin section technique and stained by haematoxylin and eosin. Multiple serial sections of the biopsy were studied.

Results

Out of the 70 cases of pemphigus studied, pemphigus vulgaris was the predominant type with 43 cases (61.4%) followed by 25 cases (35.7%) of pemphigus foliaceus and 2 cases (2.9%) of pemphigus vegetans.

Of the 43 cases of pemphigus vulgaris, 38 patients (88.4%) were between 21 and 60 years; and 2 cases (4.6%) were below 20 years of age. Only 3 cases (6.9%) were above 60 years. Out of the 25 cases of pemphigus foliaceus, 20 patients (80%) were in the age group of 21-60 years; while 2 patients (8%) were older than 60 years and 3 patients (12%) were below 20 years. Two cases of pemphigus vegetans were seen; both were between the age group 21-60 years.

There was a slight male preponderance in the ratio of 1.4:1 (25 males and 18 females)

Pemphigus vulgaris showed initial lesions involving mucous membranes in 37.2% cases and lesions including both skin and mucosae in 13.9% cases. Mucosal involvement at one time or the other was seen in 31 patients (72.1%). On the other hand, in pemphigus foliaceus, only

4% cases showed initial mucosal involvement and 12% cases showed involvement of both skin and mucous

Table I. Nature of Lesions observed during the course of Pemphigus

S.No	Type of Pemphigus	Pemphigus vulgaris	Pemphigus foliaceus	pemphigus vegetans
1.	Vesicle/ Bulla			
	● Flaccid	43 (100%)	25 (100%)	02 (100%)
	● Tense	-	-	-
	● Erythematous base	01 (2.3%)	08 (32%)	-
	● Non-erythematous base	42 (97.6%)	17 (68%)	02 (100%)
2.	Erosions	27 (62.8%)	11 (44%)	-
3.	Crusted lesions	16 (37.2%)	13 (52%)	-
4.	Vegetative lesions	02 (4.6 %)	04 (16%)	02 (100%)
5.	Erythematous plaques	02 (4.6%)	-	-
6.	Pustules	02 (4.6%)	02 (8%)	-

membrane involvement.

The nature of the lesions observed during the

Table II. Histopathological changes in pemphigus vulgaris

S.No	Histopathological Feature	No.of cases	Percentage
1.	Suprabasal clefts/ bulla	35	81.4
2.	Mid-epidermal vesicle	08	18.6
3.	Acantholysis	40	93.0
4.	Row of tombstone appearance	18	41.8
5.	Eosinophilic spongiosis	03	06.9
6.	Infiltrate in bulla cavity	23	53.5
7.	Dyskeratosis	-	-
8.	Basal layer budding	-	-
9.	Acantholysis involving follicular sheath	20	46.5ss

course of pemphigus is shown in Table I. Flaccid bullae were found in all cases. Blisters seen arising on non-erythematous skin were seen in 42 cases (97.6%) of pemphigus vulgaris, which spontaneously ruptured to give rise to erosions in 27 cases (62.8%). Crusted lesions, erythematous plaques, vegetations and pustules were present less frequently. In pemphigus foliaceus, blisters arising on erythematous skin were seen in 17 cases (68%),

crusted lesions in 13 cases (52%), erosions in 11 cases (44%) and pustules in 8% cases. Vegetations were seen in both cases of pemphigus vegetans.

Table III. Histopathological changes in pemphigus foliaceus

S.No.	Histopathological Feature	No.of cases	Percentage
1.	Subcorneal bulla	15	60
2.	Subgranular bulla	06	24
3.	Mid-epidermal bulla	04	16
4.	Dyskeratosis	02	08
5.	Hyperkeratosis	11	44
6.	Parakeratosis	11	44
7.	Acanthosis	09	36
8.	Papillomatosis	03	12
9.	Pigmentation	07	28
10.	Acantholysis	24	96

Distribution of skin lesions showed common sites of involvement in pemphigus vulgaris were generalised involvement in 16 cases (37.2%), face in 9 cases (20.9%), scalp in 16 cases (18.6%). On the other hand, in pemphigus foliaceus, there were lesions in generalised distribution in 8 cases (32%), face in 7 cases (28%), scalp in 4 cases (16%), trunk in 4 cases (56%) and involvement of extremities in 12 cases (48%).

Pruritus was seen in 14 cases (20%) of all pemphigus cases. Eleven cases (25.6%) of pemphigus vulgaris complained of pruritus. Both of our cases (100%) of pemphigus vegetans as compared to a solitary case (4%) of pemphigus foliaceus had pruritus.

Of the 36 cases of pemphigus vulgaris and 19 cases of pemphigus foliaceus in which history of Nikolsky's sign was provided, it was positive in 35 cases (97.2%) and 18 cases (94.7%) respectively. Both cases of pemphigus vegetans showed positive Nikolsky's sign.

Of the 43 cases of pemphigus vulgaris, 35 cases

(81.4%) showed intra-epidermal suprabasal vesicles and 8 cases (18.6%) showed mid-epidermal vesicles (Table II). Midepidermal vesicles were seen in old bullae, due to regeneration of the cells from the floor of the bulla. Acantholysis was seen in 93% cases as groups of cells or single cells within the bulla cavity. Dyskeratosis, basal layer budding and pseudo-epitheliomatous proliferation was not seen in any of the cases. Acantholysis affecting follicular sheath was seen in 46.5% cases. An inflammatory infiltrate was present in the bulla cavity in 23 cases (53.5%). Neutrophils were predominant in 9 cases (20.9%) and eosinophils 11 cases (25.6%). Eosinophilic spongiosis was seen in 3 cases (6.9%). Two of them were known cases of pemphigus and had developed new lesions after discontinuation of steroids. In 8 cases (18.6%), the epidermis was lost during the process. Five of these cases (11.6%) had been reported negative previously and could be diagnosed on the basis of the above histological features.

Of the 25 cases of pemphigus foliaceus studied, 24 cases (96%) showed acantholysis (table III). Subcorneal bulla was seen in 15 cases (60%), subgranular cleavage from middle epidermis in 6 cases (24%). Dyskeratosis, basal layer budding and pseudo-epitheliomatous proliferation were not seen in any of the cases. Acantholysis affecting follicular sheath was seen in 46.5% cases. An inflammatory infiltrate was present in the bulla cavity in 23 cases (53.5%). Neutrophils were predominant in 9 cases (20.9%) and eosinophils in 11 cases (25.6%). Eosinophilic spongiosis was seen in 3 cases (6.9%). Two of them were known cases of pemphigus and had developed new lesions after discontinuation of steroids. In 8 cases (18.6%), the epidermis was lost during the process. Five of these cases (11.6%) had been reported negative previously and could be diagnosed on the basis

of the above histological features.

Of the 25 cases of pemphigus foliaceus studied, 24 cases (96%) showed acantholysis (table III). Subcorneal bulla was seen in 15 cases (60%), subgranular cleavage from middle epidermis in 6 cases (24%). Dyskeratosis was seen in only 2 cases (8%). Inflammatory cells were seen in the bulla cavity in 12 cases (48%). In 10 cases (83.3%) the infiltrate comprised of polymorphs. Spongiosis was seen in 10 cases (40%) and exocytosis in 8 cases (32%). Acanthosis, hyperkeratosis, parakeratosis, papillomatosis and increased pigment formation were seen in a few cases.

Both cases of pemphigus vegetans showed hyperkeratosis, papillomatosis and irregular acanthosis with intra-epidermal eosinophilic abscesses. Suprabasal lacunae with a few acantholytic cells were seen. Both the cases were of Neumann type. No cases of pemphigus erythematosus, drug-induced pemphigus and paraneoplastic pemphigus were seen in our study.

Discussion

Our series is in accordance with other Indian series each of which has maximum cases of pemphigus vulgaris, followed by pemphigus erythematosus/ foliaceus and then by pemphigus vegetans.^{2,3} Majority (85.7%) of our patients were between 21-60 years, akin to Indian literature.^{3,4} Pemphigus vulgaris affects males and females equally,⁵ although in the present study, there was a slight male predominance in the ratio 14.:1, which probably reflects the higher male attendance in the out-patient department. Male dominance has been reported in the same ratio.⁶

Nature and distribution of lesions as well as mucosal involvement in different types of pemphigus in our series has followed the pattern seen in earlier studies.^{4,6,7,8,9} Flaccid bullae were seen in all the cases.

Mucosal involvement was seen in 31 patients (72.1%) of pemphigus vulgaris, and only in 5 patients (20%) of pemphigus foliaceus. Pruritus was seen in 25.6% of our cases of pemphigus vulgaris, slightly lower than reported before.¹² Nikolsky's sign was positive in 97.2% and 94.7% cases of pemphigus vulgaris and foliaceus respectively, in corroboration with reports in Indian literature.

The salient histological features of pemphigus vulgaris were (1) intraepidermal suprabasal blisters (2) presence of acantholytic cells (3) "Row of tombstone appearance" (4) Acantholysis involving follicular sheath.

Salient features of pemphigus foliaceus were (1) acantholysis (2) subcorneal cleavage (3) an inflammatory infiltrate.

Both of our cases of pemphigus vegetans were of Neumann type.

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