

PEMPHIGUS : A CLINICAL STUDY OF 109 CASES FROM TRIPOLI, LIBYA

M Shafi, M L Khatri, M Mashina, M Ben-Ghazeil

From 1981 to 1992, 109 cases (90 females and 19 males) of pemphigus were seen in the Department of Dermatology, Central Hospital Tripoli, Libya. Age of onset of the disease varied from 17 to 85 years, while duration of the disease at the time of presentation varied from 3 days to 13 years. On the basis of clinical features and routine histological findings the cases were divided into various subtypes as : pemphigus foliaceus 65 cases, pemphigus vulgaris 34 patients, pemphigus erythematosus 5, herpetiform pemphigus 3 and vegetans type 2 cases. Three of our patients has diabetes mellitus preceding pemphigus, while 12 patients developed steroid-induced diabetes. Significant secondary bacterial infection occurred in all cases at some stage of the disease while oral candidiasis occurred in 15 cases. Eczema herpeticum was seen in 4 patients, while 2 had extensive tinea corporis. One of 4 patients of pemphigus vulgaris had complete shedding of nails and 1 female patient had alternate phases of pemphigus foliaceus and generalized pustular psoriasis. The features in our cases of pemphigus foliaceus were somewhat similar to Brazilian pemphigus foliaceus and we had more cases of pemphigus foliaceus, almost exclusively affecting females.

Key Word : Pemphigus

Introduction

Pemphigus is a group of blistering diseases, varying in clinical features and incidence in various population groups.

We have seen 109 cases of pemphigus since 1981 at the Central Hospital, Tripoli, Libya. An analysis of their clinical features is presented.

Materials and Methods

All patients were admitted for initial assessment and treatment and many of them were re-admitted several times with exacerbation of the disease. Details of history, physical findings, laboratory data, treatment, and follow-up were recorded in a protocol. The diagnosis was based on clinical features and histopathology. Immunofluorescence

studies could be arranged only in 7 patients.

Results

Of the 109 cases only 2 men were Indian while others were Libyan nationals, 90 (75.6%) were female and 19 (24.4%) were male. The age at onset of the disease ranged between 17 years and 85 years in general, in cases of pemphigus vulgaris between 20 and 85 (median age 40 years), in cases of pemphigus vegetans between 23 and 40 years (median age 31½ years), and in cases of pemphigus foliaceus between 17 and 85 years (median age 29 years). The most common age at onset was between 21 and 30 years (Table I).

Table I. Age at onset

Age group	Male	Female	Total
17-20 yrs.	-	10	10
21-30 yrs.	2	45	47
31-40 yrs.	2	18	20
41-50 yrs.	5	8	13
51 onwards	10	9	19
	19	90	109

From the Departments of Dermatology, Central Hospital and Faculty of Medicine, Al-Fateh University of Medical Sciences, Tripoli, Libya.

Address correspondence to : Dr M L Khatri
P O Box 13457, Tripoli.

The duration of the disease at the time of presentation to our department ranged between 3 days to 13 years. The onset of the disease process was gradual in 10² patients while it was acute in 7 patients.

The disease was asymptomatic in 81 (74.3%) but 28 (25.7%) patients complained of itching, mild in 15, moderate in 11, and severe in 2 patients.

The type of initial lesions and initial sites of involvement are given in Table II and III

Table II. Initial lesions noticed

Type of lesions	No. of Pts
Vesiculo bullous	47
Erosions	24
Both types	31
Vesicular erythematous patches	6
Papulo-Vesicular	1

Table III. Sites of initial lesions

Sites	No. of pts.
Face	4
Face & chest	5
Trunk	44
Extremities	5
Mouth	24
Mouth & trunk	6
Eyes	1

respectively. Out of 34 cases of pemphigus vulgaris 24 had lesions initially in oral mucosa and later developed skin lesions after an interval ranging from 15 days to 1½ years; one patient had initial lesions on the conjunctivae while 6 patients developed mouth and body lesions simultaneously and 3 developed skin lesions 1 week to 1 month earlier than the mouth lesions.

The nature of the various type of lesions observed during the course of the disease is given in Table IV. One female patient with pemphigus vulgaris had onychomadesis of all

Table IV. Nature of lesions observed during the course of the disease

Nature of lesions	No. of pts.	
Vesiculobullous		
Flaccid	109	109
Tense	3	
Erythematous base	43	
Without erythema	66	
Erosions		104
Crusted lesions		93
Vegetative lesions		3
Erythematous plaques		6
Annular lesions with vesicle in margins		14
Erythroderma		6
Onychomadesis		1
Grouped lesions		36

the nails during the initial acute phase but later complete regrowth of the nails was observed after the control of the disease. The details of distribution of the lesions is given in Table V.

Table V. Distribution of lesions

Sites	No. of pts.	Sites	No. of pts.
Scalp	56	Mouth	36
Face	77	Genital mucosa	2
Trunk	108	Anal mucosa	2
Arms	98	Nasal mucosa	1
Legs	97	Conjunctivae	2
Axillae	61		
Groins	71		

On the basis of clinical features and histopathologic findings the cases were divided in five sub-types (Table VI).

Table VI. Types of pemphigus

Types of pemphigus	Male	Female	Total
Pemphigus foliaceus	4	61	65
Pemphigus vulgaris	14	20	34
Pemphigus erythematousus	1	4	5
Herpetiform pemphigus	-	3	3
Pemphigus vegetans	-	2	2
Total	19	90	109

Other associated diseases present before and developing after steroid therapy are listed in Table VII. Eczema herpeticum developed in

Table VII. Other associated diseases

Disease	Before steroid therapy	After steroid therapy
Diabetes mellitus	3 pts	12 pts
Hypertension	5 pts	15 pts
Pulmonary tuberculosis (reactivation)		2 pts
Generalized pustular psoriasis		1 pt
Oral candidiasis		15 pts
Extensive tinea corporis		2 pts
Mycotic keratitis leading to panophthalmitis		1 pt
Eczema		4 pts

4 patients with pemphigus foliaceus while on high dose of steroid and 3 of them were on combination therapy of steroid and azathioprine. Variable degree of bacterial infection occurred in all cases at some stage of the disease. Pus culture of the infected lesions revealed growth of coagulase positive *Staphylococci* in 92, *E Coli* in 11, *pseudomonas aeruginosa* in 8, *Streptococci* in 5, and *Proteus mirabilis* in 4 patients.

Follow-up

One hundred and three cases could be followed for 6 months to 8 years, while 6 patients did not turn up after the first admission. Up to date follow-up was possible only in 34 cases. Six patients with pemphigus foliaceus went into remission ranging from 6 months to 2 years without treatment but later recurrence occurred in 5 while one of them is still in complete remission. Longest duration of the disease recorded during follow-up was 16 years in cases with pemphigus vulgaris. Three patients with pemphigus vulgaris died in hospital due to complications directly related to

the diseases and steroid therapy, 2 due to septicaemia and third due to cardiac arrest.

Comments

Although we could not estimate the exact incidence of pemphigus in Libya, it seems to be quite high. High incidence in Mediterranean population particularly in the Jewish race has been documented.¹ In the previous studies^{1,3} sex ratio has been recorded to be equal in male and female but in the present study there was predominance of female (F:M=90:19), moreover pemphigus foliaceus patients were all female except 3 male. In our study median age at onset of pemphigus vulgaris was 40 years, pemphigus vegetans 31½ years and pemphigus foliaceus 29 years, which is almost identical with previous reports.¹ However, in our cases of pemphigus foliaceus, the onset was mainly in young females. In 2 Indian patients with pemphigus vulgaris the age at onset was 26 years and 29 years. Early onset of pemphigus vulgaris is recorded in India.²

Itching was a significant complaint in 28 cases of pemphigus foliaceus in the present series while it has not been recorded as a prominent feature in the earlier studies.¹ The course of pemphigus foliaceus has been recorded previously as comparatively benign but in the present study majority of the patients had a fulminant course although 6 patients showed temporary remission without steroid therapy. One female patient had alternate phase of pemphigus foliaceus and generalized pustular psoriasis. This patient developed attacks of generalised pustular psoriasis while on high dose of methylprednisolone, which could be controlled on administration of weekly methotrexate therapy.

Taking into consideration, the early age of onset, initial sites, type of lesions and course of disease in our cases of pemphigus foliaceus, the similarity to Brazilian pemphigus foliaceus is striking. However there are some dissimilarities. In Brazilian pemphigus, sex ratio is equal but in this series the pemphigus foliaceus cases were exclusively seen in female except 3 males. Moreover we did not record any evidence of the disease in other family members, which is a prominent feature of Brazilian pemphigus foliaceus.³

It is concluded that the incidence of

pemphigus seems to be high in Libya, with high female predominance of the disease. For reasons, which need further study we have more cases of pemphigus foliaceus, almost exclusively affecting females.

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