

# Pattern of Non Infectious Vesiculobullous and Vesiculopustular Skin Diseases in a Large Tertiary Care Hospital

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## Abstract

**Pemphigus is an autoimmune blistering disease with in vivo bound and circulating autoantibodies against the cell membrane components of keratinocytes. It varies in incidence from 0.5 to 3.2 cases per 1,00,000 population per year showing equal proportion in both the sexes. We studied 600 cases of non neoplastic skin lesion out of which 30 cases of non infectious vesiculobullous and vesiculopustular skin diseases were included in the present study. Pemphigus vulgaris was the most common type of pemphigus in our study. Our findings correlated well with most of the other studies.**

## Introduction

### Pemphigus

It varies in incidence from 0.5 to 3.2 cases per 1,00,000 population per year showing equal proportion in both the sexes.

Acantholysis, the characteristic feature of the bullae of pemphigus was first demonstrated in 1943. Pemphigus<sup>1</sup> is an autoimmune blistering disease with in vivo bound and circulating autoantibodies against the cell membrane components of keratinocytes.

Pemphigus can be divided into 5 types

- a) Pemphigus vulgaris, with its reactive state, pemphigus vegetans.
- b) Pemphigus foliaceus, with its lupus variant, pemphigus erythematous, and its endemic variant, fogo selvagem.
- c) Drug induced pemphigus
- d) Ig A pemphigus
- e) Paraneoplastic pemphigus

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### Pemphigus vulgaris (Fig. 1)<sup>2</sup>

It is seen most commonly in the 4<sup>th</sup> and 5<sup>th</sup> decade. Large flaccid bullae involving oral mucosa, scalp, mid face, sternum and groin are present.

The bullae are in suprabasal location with basal keratinocytes firmly attached to the dermis like a "row of tombstones". Blister cavity may show acantholytic cells and eosinophils.

### Pemphigus foliaceus (Fig. 2)<sup>3,4</sup>

Early rupture of bullae results in formation of erythematous erosions that ooze and crust mainly involving face, scalp, chest and back.

Also known as superficial pemphigus because intraepidermal separation occurs in the granular layer leading to subcorneal bullae. Some times eosinophilic spongiosis may be seen.

### Bullous pemphigoid (Fig. 3)<sup>5,6</sup>

First described in 1953, it affects primarily elderly patients with large tense bullae. Subepidermal bullae arising at the dermo-epidermal junction are seen.

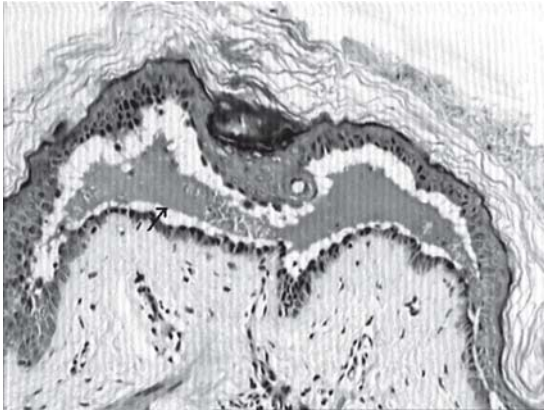


Fig. 1: *Pemphigus Vulgaris* (4X)-Suprabasal bulla.

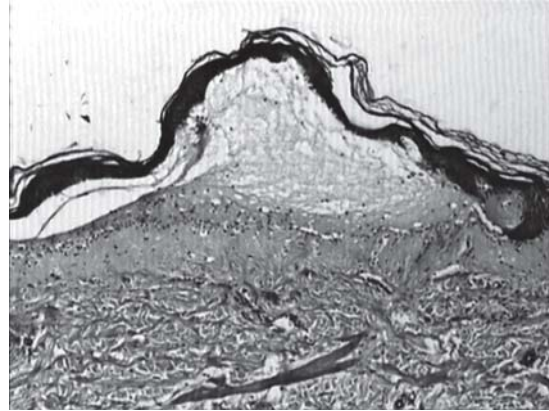


Fig. 3: *Bullous pemphigoid* (4X)-Subepidermal bulla.

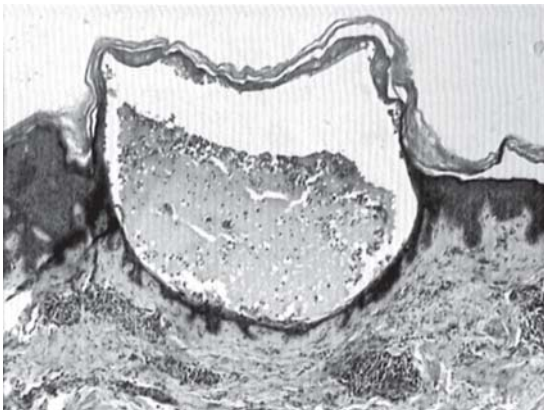


Fig. 2: *Pemphigus Foliaceus* (4X)-Subcorneal bulla.

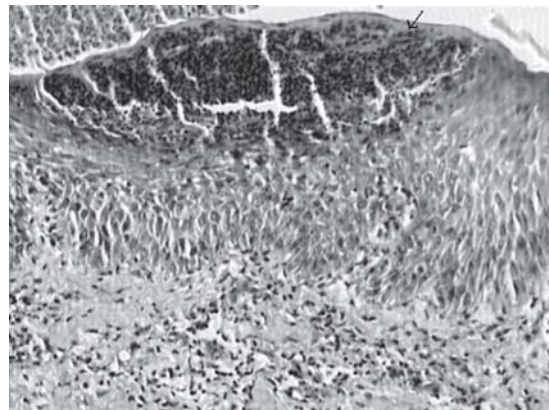


Fig. 4: *Subcorneal pustular dermatosis* (4X)-Subcorneal collection of neutrophils.

### **Bullous systemic lupus erythematosus<sup>7</sup>**

Most commonly seen in women, particularly black women. The lesions are either vesicles or bullae. Predominantly on the extensor surfaces of arms, elbows, or scalps.

Three histologic patterns can be seen.

- 1) Dermatitis herpetiformis like picture.
- 2) Vasculitis with subepidermal blister and pustule formation.
- 3) Basal layer vacuolisation, with subsequent blister formation.

### **Subcorneal pustular dermatosis (Fig. 4)<sup>8</sup> (Sneddon - Wilkinson disease)**

It occurs most frequently in persons over 40 years of age with a male to female ratio of 1:4. First described in 1956, it occurs as sterile pustules in the groin or other body folds of middle-aged women. A unilocular subcorneal pustule filled with neutrophils is the primary lesion. The granular layer beneath the pustule is at least partly intact, and the remainder of the epidermis show spongiosis.

### **Methods**

We studied total 600 cases of non neoplastic skin lesions, out of which 30 cases of non

**Table 1 : Age (in years) and sex wise distribution of non infectious vesiculobullous and vesiculopustular skin diseases (NIVVD) cases.**

Type	11-20	21-30	31-40	41-50	51-60	61-70	Total cases	%
	M F	M F	M F	M F	M F	M F		
Pemphigus Group	1 0	1 1	0 4	10 0		1 0	18	60.00
BP					2 3		5	16.67
SCPD		0 2	1 0	1 0	2 0		6	20.00
BSLE				0 1			1	3.33
Total							30	100

B.P. – Bullous pemphigoid; S.C.P.D. - Subcorneal pustular dermatosis (SCPD); B.S.L.E. – Bullous systemic lupus erythematosus

**Table 2 : Age (in years) and sex wise distribution of Pemphigus cases.**

Diseases	11-20	21-30	31-40	41-50	51-60	61-70	Total cases	%
	M F	M F	M F	M F	M F	M F		
Pemphigus Vulgaris			0 2	8 0			10	55.55
Pemphigus foliaceus	1 0	1 1	0 2	2 0		1 0	8	44.45
						18	100	

infectious vesiculobullous and vesiculopustular skin diseases were included in the present study.

The material used for the study were skin biopsy specimens from outpatient skin department of a large tertiary care hospital.

The present study was carried from 1<sup>st</sup> January 2005 to 30<sup>th</sup> June 2006.

The detailed history and informed consent of the patient was taken before doing the biopsy.

Skin biopsies were fixed in 10% formalin and processed for light microscopic examination. Paraffin embedded 5 micron thick sections were obtained and studied routinely with haematoxylin and eosin stain. The histological sections were scrutinized under scanner, 10x, 40x, and 100x magnifications.

## Results

In our study there were 30 cases (Table 1)

of non infectious vesiculobullous and vesiculopustular skin diseases (NIVVD) constituting 5% of all non-neoplastic skin lesions. Pemphigus vulgaris was the most common lesion constituting 33.33% of NIVVD occurring in 4<sup>th</sup> decade having a male to female ratio of 4:1.

There were 5 cases of Bullous Pemphigoid constituting 16.67% of NIVVD with most of the cases seen in 6<sup>th</sup> decade having male to female ratio of 1:1.5.

We found a single case of Bullous Systemic Lupus Erythematosus in a 49 year old female with active SLE constituting 3.33% of NIVVD.

## Discussion

### 1) Pemphigus

In our study pemphigus vulgaris was the most common lesion among pemphigus group. This was comparable to Salmanpour<sup>9</sup> *et al* series, Aboobaker<sup>10</sup> *et al* series, among Indians in South Africa, Wilson<sup>11</sup> *et al* series,

in New Delhi, India, and Adam<sup>12</sup> series, showing Pemphigus Vulgaris more common than Pemphigus Foliaceus. However Aboobaker<sup>10</sup> *et al* series, in people of South Africa and Wilson<sup>11</sup> *et al* series, in Oxford, U.K. showed cases of Pemphigus Foliaceus and Pemphigus Vulgaris in equal proportion.

In our study majority of cases of Pemphigus Vulgaris were in the 4<sup>th</sup> decade and that of Pemphigus Foliaceus in the age range of 2<sup>nd</sup> to 7<sup>th</sup> decade. This was comparable to Aboobaker<sup>10</sup> *et al* series, showing mean age of 48 years for Pemphigus Vulgaris and mean age of 43 years for Pemphigus Foliaceus.

Male to female ratio in our study was 4:1. This was comparable to Arya<sup>13</sup> *et al* series, and Ambady<sup>14</sup> *et al* series, showing male predominance. However Aboobaker<sup>10</sup> *et al* series, found predominance of females while Fernandez<sup>15</sup> *et al* series, found males and females in equal proportion.

Clinically bullae were present. Microscopy showed suprabasal bulla filled with pale eosinophilic fluid and few acantholytic cells in Pemphigus Vulgaris and a subcorneal bulla filled with faint eosinophilic fluid in Pemphigus Foliaceus. Eosinophilic spongiosis was not seen in any of the case in our study.

## 2) Bullous pemphigoid

In our study there were 5 cases of Bullous Pemphigoid constituting 16.67% of NIVVD. This was high as compared to Wong<sup>16</sup> *et al* series, showing 7.6/ million cases/year.

Most of the cases encountered were in 6<sup>th</sup> decade. However mean age in Wong<sup>16</sup> *et al* series, was 77 years.

Male to female ratio was 1:1.5 which was comparable to Wong<sup>16</sup> *et al* series, showing male to female ratio of 1 : 2.

Clinically tense bullae were present showing histologically a subepidermal bulla filled with faint eosinophilic fluid and few

eosinophils.

## 3) Subcorneal pustular dermatoses (SCPD)

Our study showed 5 cases of SCPD which constituted 20% of NIVVD. The age group varied from 3<sup>rd</sup> to 6<sup>th</sup> decade with male to female ratio of 1.5:1 which was comparable to Krog<sup>17</sup> *et al* series, showing SCPD in person over 40 years of age.

Clinically multiple pustules were seen.

Microscopic examination revealed subcorneal pustule filled with neutrophils. The granular layer beneath the pustule was intact with rest of the epidermis showing spongiosis.

## 4) Bullous systemic lupus erythematosus (Bullous SLE)

In our study there was a single case of Bullous SLE in a 49 year old female with active SLE, constituting 3.33% of NIVVD. This was low as compared to Wong SN<sup>16</sup> series, showing 3% and high as compared to Bernard<sup>18</sup> *et al* study, showing 2/100 cases of Bullous SLE amongst all the bullous lesions.

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**ANALYSIS OF A DENGUE FEVER OUTBREAK CAUSED BY DENV-1 IN MEDICAL COLLEGE CAMPUS, TRIVANDRUM, KERALA**

The major presenting features were fever (97.4%), myalgia (84.2%), periorbital pain (48.7%), backache (46%), giddiness (35.5%), abdominal pain (27.6%), polyarthralgia (21%), and syncope (15.8%). The average fever duration was 3.03 days. Major physical findings included petechiae (21.3%). IGM antibody was positive in 58 (44.1%), IgG antibody was positive in 36 (27.4%) and both were positive in 28 (21.3%). Virus isolation by RT-PCR was done in 64 samples, of which 40 (62.5%) were positive and all belonged to serotype 1 (DENV-1). Leucopenia (< 3000/ cumm) was present in 38 (28.9%) and 41 (31.2%) had thrombocytopenia.

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