

# Clinico Pathological Study of Autoimmune Vesiculobullous Disorders: A Case Series from a Resource-poor Rural Tertiary Care Center in South Tamil Nadu

Uma Selvaraj<sup>1</sup>, Madhavan Ramamoorthy<sup>2</sup>

<sup>1</sup>Senior Assistant Professor, Department of Dermatology, Government Theni Medical College, Theni, Tamil Nadu, India, <sup>2</sup>Senior Assistant Professor, Department of STD, Coimbatore Medical College, Coimbatore, Tamil Nadu, India

## Abstract

**Background:** Autoimmune vesiculobullous disorders are rare diseases that are characterized by blisters over skin and oral erosions. An accurate diagnosis is achieved by clinical examination, pathological correlation, and immunofluorescence. In resource-poor settings where immunofluorescence is not available, the diagnosis is made only with the available resources.

**Aims and Objectives:** To study the clinical and pathological features of autoimmune bullous disorders.

**Materials and Methods:** A total of 45 patients admitted to the Government Theni Medical College from 2005 to 2015 were analyzed retrospectively. Detailed clinical examination, Tzanck smear, and histopathological examination of skin biopsy were done for all patients. Immunofluorescence was not done due to lack of availability in our center.

**Results:** Out of 45 cases studied, 32 (71%) were pemphigus, 10 (22.22%) belonged to bullous pemphigoid group, 2 (4.44%) were chronic bullous dermatosis of childhood, one case (2.2%) was a bullous systemic lupus erythematosus. Out of 45 patients, 31 patients (68.8%) were in the age group of 41-60 years, 6 patients (13.33%) in the range of 21-40 years, 5 patients (11.11%) in the age range above 60 years, and 3 patients (6.66%) below 20 years. Out of 45 patients, 17 (37.77%) were males and 28 (62.22%) were females. All the bullous disorders showed a female preponderance. The clinical spectrum was consistent in all patients enrolled (100%) Tzanck smear was consistent in 95.5% of patients. Histopathology was consistent in 100% of cases.

**Conclusion:** Thorough clinical examination, aided by Tzanck smear and histopathology was helpful in arriving at a diagnosis of these vesiculobullous disorders in resource-poor center like ours where the gold standard immunofluorescence studies are not available.

**Key words:** Autoimmune bullous disorders, Bullous pemphigoid, Pemphigus

## INTRODUCTION

Autoimmune vesiculobullous disorders are a rare group of disorders where auto antibodies are directed against specialized structures essential for maintaining the integrity of skin leading to blister formation. Erosions involving skin and mucous membranes depending on the level of blister

formation. They are classified as epidermal and subepidermal. To arrive at a diagnosis, complete clinical examination, aided by Tzanck smear, histopathology of skin biopsy, and immunofluorescence studies need to be done. But in resource-poor centers like ours diagnosis could be achieved only with the available investigations. The aim of the study was to analyze the clinical and histopathological features of these disorders in our setup with the available resources.

## MATERIALS AND METHODS

A retrospective analysis of all vesiculobullous disorders admitted to our ward over a span of 10 years between 2005 and 2015 was analyzed.

### Access this article online



www.ijss-sn.com

Month of Submission : 05-2016  
Month of Peer Review : 06-2016  
Month of Acceptance : 07-2016  
Month of Publishing : 07-2016

**Corresponding Author:** Dr. Uma Selvaraj, 15/1/38A, Arjun Illam, Sivanantha Nagar, P.C. Patty, Theni - 625 531, Tamil Nadu, India.  
E-mail: drumashrikannan@gmail.com

Thorough clinical examination was done for all patients. Tzanck smear was done for all patients by derroofing an early vesicle and making a smear on a glass slide and staining with Leishman's stain and examined under light microscope. A skin biopsy taken from an early vesicle was subjected to histopathological examination after staining with eosin and hematoxylin.

All data regarding the patients' age, sex, duration of disease, reports of Tzanck smear, and histopathological findings were documented.

## RESULTS

Out of 45 cases studied, 32 (71.1%) belonged to pemphigus group. 10 cases (22.2%) belonged to bullous pemphigoid Group 2 patients (4.4%) belonged to chronic bullous dermatosis of childhood (CBDC) group and one patient (2.2%) was a bullous systemic lupus erythematosus (SLE) (Table 1).

Out of 32 cases of pemphigus, 28 were pemphigus vulgaris and 4 were pemphigus foliaceus. Predominant age group affected was 51-60 (16 patients) followed by 15 patients in the age group of 41-50. Youngest age incidence was seen in 3 patients in the range 0-10. The highest age range was seen in 5 patients in the ranges 61-70 years (Table 2).

Out of 45 cases, 28 were females (62.14%) and 17 (37.6%) were males (Figure 1) female predominance was seen in all the bullous disorders noted.

The clinical diagnosis could be arrived in all pemphigus patients with flaccid blisters and erosions, a positive

Nickolsky sign in pemphigus vulgaris and no mucosal involvement in pemphigus foliaceus (Figure 2).

All bullous pemphigoid patients showed tense blisters and erosions over trunk and extremities. Mucosal erosions were seen in only 4 out of 10 patients (Figure 3). Both cases of CBDC showed, tense blisters with few lesions showing "cluster of jewels" appearance (Figure 4). No mucosal involvement was seen in both cases. Bullous SLE showed tense blisters over extremities and with other cutaneous and lab findings diagnostic of SLE.

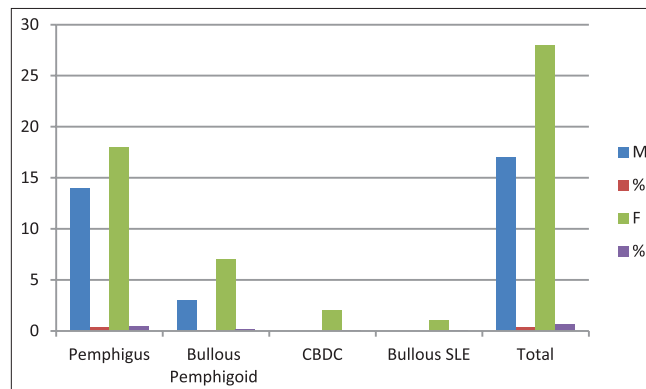


Figure 1: Sex wise distribution

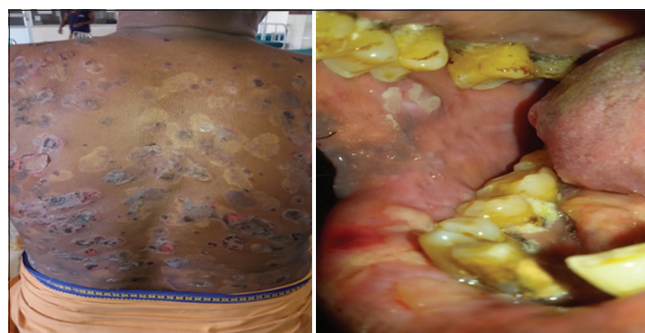


Figure 2: Flaccid blisters and erosions in pemphigus



Figure 3: Tense blisters in bullous pemphigoid

Table 1: Bullous disorders prevalence

Bullous disorder	Number of cases (%)
Pemphigus vulgaris	28 (62.2)
Pemphigus foliaceus	4 (8.8)
Bullous pemphigoid	10 (22.22)
CBDC	2 (4.44)
Bullous SLE	1 (2.2)
Total	45

CBDC: Chronic bullous dermatosis of childhood, SLE: Systemic lupus erythematosus

Table 2: Age wise statistics of bullous disorders

Type	0-10	11-20	21-30	31-40	41-50	51-60	61-70	Total
Pemphigus	0	0	4	2	13	10	3	32
Bullous Pemphigoid	0	0	0	0	2	6	2	10
CBDC	2	0	0	0	0	0	0	2
Bullous SLE	1	0	0	0	0	0	0	1
Total								45

CBDC: Chronic bullous dermatosis of childhood, SLE: Systemic lupus erythematosus

Tzanck smear was consistent in all cases of pemphigus with predominant acantholytic cells. 8 patients with bullous pemphigoid showed eosinophils in the smear and 2 patients showed mixed inflammatory cells. Two cases of CBDC and one case of SLE showed predominant neutrophils in the smear (Figure 5).

Histopathology of all pemphigus vulgaris showed suprabasal bulla with acantholytic cells in the bulla cavity and pemphigus foliaceus showed subcorneal bulla with acantholytic cells.

A biopsy was consistent with all cases of bullous pemphigoid with subepidermal cleavage with predominant eosinophils.

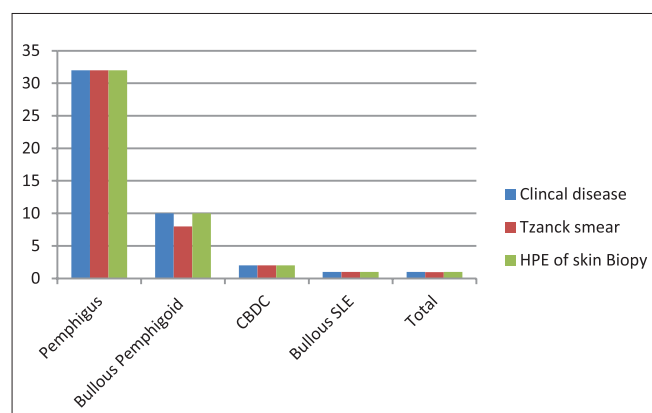
CBDC and bullous SLE showed subepidermal bulla cavity with predominant neutrophils (Figure 5).

## DISCUSSION

Autoimmune vesiculobullous disorders are characterized by antibody mediated destruction of structures essential for maintaining the integrity of skin leading to blisters and



**Figure 4: Chronic bullous dermatosis of childhood with “cluster of jewels appearance”**



**Figure 5: Clinical and histopathological correlation. CBDC: Chronic bullous dermatosis of childhood, SLE: Systemic lupus erythematosus**

erosions over skin and mucosa. They are categorized into epidermal and subepidermal blistering disorders.

Epidermal disorders include pemphigus vulgaris and pemphigus foliaceus. Subepidermal disorders include bullous pemphigoid, cicatricial pemphigoid, linear immunoglobulin A dermatosis, lichen planus pemphigoides, dermatitis herpetiformis, epidermolysis bullosa acquisita, and bullous SLE.

Thorough clinical examination aided by light microscopy and immunofluorescence would help us to make a definitive diagnosis of these bullous disorders.

Although immunofluorescence is considered as the gold standard, in resource poor settings where this facility could not be availed even in private labs diagnosis is based on the clinical and light microscopic findings only.

In our present study, pemphigus was the predominant bullous disorder in which pemphigus vulgaris was the most common. This was comparable with studies by Srinath *et al.*,<sup>1</sup> Arya *et al.*,<sup>2</sup> Huda and Afsar,<sup>3</sup> Zarea *et al.*,<sup>4</sup> Daneshpazhooh *et al.*,<sup>5</sup> and Micali *et al.*<sup>6</sup>

Predominant age group for pemphigus was noted in the range 41-60 which was comparable with other studies.<sup>1</sup>

Pemphigus and all other blistering disorders showed a higher incidence in females (62.14%) which was contradictory to studies by Srinath *et al.*<sup>1</sup> and Arya *et al.*<sup>2</sup>

Bullous pemphigoid was the predominant subepidermal disorder noted and was comparable with other studies.<sup>7,8</sup>

Predominant age group of bullous pemphigoid was between 41 and 70, with majority of patients presenting in the age group of 51-60. This was contradictory to studies by Srinath *et al.*,<sup>1</sup> Wong and Chua,<sup>8</sup> Bastuji-Garin *et al.*,<sup>9</sup> and Jung *et al.*<sup>10</sup>

All cases of pemphigus vulgaris (100%) showed suprabasal bulla and acantholytic cells on histopathology and all pemphigus foliaceus case showed subcorneal bulla which was comparable with studies by Srinath *et al.*<sup>1</sup>

Biopsy findings was consistent in all cases of CBDC and bullous SLE which was comparable with Srinath *et al.*<sup>1</sup>

## CONCLUSION

Although immunofluorescence was mandatory for definitive diagnosis of autoimmune bullous disorders, in resource poor settings where this facility is not available,

diagnosis could be arrived only with the available investigations.

## REFERENCES

1. Srinath M, Kambli, Madavamurthy P. Immunobullous disorders clinical histopathological and immunofluorescence study of 36 cases. *Mullers J Med Res* 2014;5:134-8.
2. Arya SR, Valand AG, Krishna K. A clinico-pathological study of 70 cases of pemphigus. *Indian J Dermatol Venereol Leprol* 1999;65:168-71.
3. Huda MM, Afsar MI. A clinicopathological study of pemphigus. *Indian J Dermatol* 2001;46:75-9.
4. Zaraa I, Kerkeni N, Ishak F, Zribi H, El Euch D, Mokni M, *et al.* Spectrum of autoimmune blistering dermatoses in Tunisia: An 11-year study and a review of the literature. *Int J Dermatol* 2011;50:939-44.
5. Daneshpazhooh M, Chams-Davatchi C, Payandemehr P, Nassiri S, Valikhani M, Safai-Naraghi Z. Spectrum of autoimmune bullous diseases in Iran: A 10-year review. *Int J Dermatol* 2012;51:35-41.
6. Micali G, Musumeci ML, Nasca MR. Epidemiologic analysis and clinical course of 84 consecutive cases of pemphigus in Eastern Sicily. *Int J Dermatol* 1998;37:197-200.
7. Bernard P, Vaillant L, Labeille B, Bedane C, Arbeille B, Denoeux JP, *et al.* Incidence and distribution of subepidermal autoimmune bullous skin diseases in three French regions. Bullous diseases French study group. *Arch Dermatol* 1995;131:48-52.
8. Wong SN, Chua SH. Spectrum of subepidermal immunobullous disorders seen at the National Skin Centre, Singapore: A 2-year review. *Br J Dermatol* 2002;147:476-80.
9. Bastuji-Garin S, Joly P, Lemordant P, Sparsa A, Bedane C, Delaporte E, *et al.* Risk factors for bullous pemphigoid in the elderly: A prospective case-control study. *J Invest Dermatol* 2011;131:637-43.
10. Jung M, Kippes W, Messer G, Zillikens D, Rzany B. Increased risk of bullous pemphigoid in male and very old patients: A population-based study on incidence. *J Am Acad Dermatol* 1999;41:266-8.

**How to cite this article:** Selvaraj U, Ramamoorthy M. Clinico Pathological Study of Autoimmune Vesiculobullous Disorders: A Case Series from a Resource-poor Rural Tertiary Care Center in South Tamil Nadu. *Int J Sci Stud* 2016;4(4):27-30.

**Source of Support:** Nil, **Conflict of Interest:** None declared.