ORIGINAL RESEARCH PAPER

Dermatology

VESICULOBULLOUS DISORDERS-ETIOLOGY AND PATHOLOICAL PROFILE IN TERTIARY CARE CENTER, TAMILNADU

KEY WORDS:
vesiculobullous disorders, histopathology, Pemphigus, Pemphigoid

Jachak Aasawari* Post Graduate, Department of Dermatology, Venereology & Leprosy, Vinayaka Missions Medical College, Karaikal. *Corresponding Author

Sivakumar S. Professor, Department of Dermatology, Venereology & Leprosy, Vinayaka Missions Medical College, Karaikal

Sunjanaa Dhepa Assistant Professor, Department of Dermatology, Venereology & Leprosy, Vinayaka Missions Medical College, Karaikal

ABSTRACT

Background- Vesicles (blisters less than 0.5cm) and bullae (greater than 0.5 cm) occur in number of skin conditions. Vesicullobulous disorders are a heterogenous group of skin diseases in which autoantibodies are directed against cell adhesion molecules which are essential for the integrity of skin and oral mucosa.
Aim- to evaluate the etiological, histopathological of various immunobullous disorders of the skin.
Material & Method- A retrospective study was conducted on immunobullous disorder patients who attended the Outpatient Department of Dermatology, Vinayaka Missions Medical College, Karaikal, India, over a period of 21 months
Results- Of 83 patients, pemphigus vulgaris was the most observed disorder with 27 (44.57%) patients, followed by bullous pemphigoid with 20 (24.09%) patients, pemphigus foliaceus was seen in 12 (14.45%) patients. 6 (7.225) patients had vasculitis, 4 (4.82%) were having dermatitis herpetiformis. 2 (2.4%) of the patients had bullous SLE. Epidermolysis bullosa acquisita and lichen planus pemphigoides was seen in 1 (1.2%) patients each.
Conclusion – We recommend clinical, demographic and histopathological with immunofluorescence correlation for diagnosis of vesiculobullous disease.

INTRODUCTION-

Vesiculobullous disorders are a heterogenous group of dermatoses with a variety of frequently changing manifestations(3). They are an autoimmune disease which is rare with an incidence of 0.5 to 3.2 cases/100,000 population(1). It is a dermatological disorder in which the autoantibodies are directed against antigens present in epidermis or dermoepidermal junction(1) . Blisters include both vesicles and bullae which are cavities filled with fluid present either in or underneath the epidermis.

Vesiculobullous disorders can involve mucosal surface of oral cavity, conjunctiva, nasopharynx, oesophagus, urethra, vulva, cervix, scalp, chest, face and upper back. Lesions may also involve the flexor surfaces of the arms and legs, abdomen, axillae and groin(1).

Tzanck smear is used as a minimally invasive test for the diagnosis of the pemphigus group of AIBDs and was first used by Arnault Tzanck in 1947(4). Pathological evaluation involves systematic analysis, which includes the blister separation plane, the mechanism of blister formation, and the character of the inflammatory infiltrate, including its presence or absence(5).

This study was undertaken to evaluate the etiological, histopathological of various immunobullous disorders of the skin.

MATERIAL & METHOD

This cross-sectional, descriptive, chart-based, retrospective study was conducted on immunobullous disorder patients who attended the Outpatient Department of Dermatology, Vinayaka Missions Medical College, Karaikal, India, over a period of 21 months (between 1 January 2015 and 31 December 2018).

Institutional ethical clearance was obtained. Medical records of all patients with clinically suspected immunobullous disorders who attended the Outpatient Department of Dermatology were screened, after obtaining institutional ethical committee clearance. Nonimmune-mediated vesiculobullous lesions secondary to mechanical injury, infections, eczemas, and burns (chemical or thermal) were excluded from the study.

From these medical records, demographic data of patients, such as age, sex, provisional clinical diagnosis, tzank smear histopathological findings, and final diagnosis, were collected. In all patients, excisional biopsy from the lesional skin or oral mucosa, preferably including intact vesicle/bulla, was obtained for histopathological study . Histopathological diagnosis was made on the basis of the level of split, inflammatory infiltrate, altered keratinocytes and dyskeratotic cells, and pattern of arrangement of epidermal keratinocytes (e.g. dilapidated brick wall appearance and row of tombstone). On the basis of these features, the vesiculobullous diseases were divided into subcorneal [pemphigus foliaceus (PF), pemphigus erythematosus (PE), and subcorneal pustular dermatosis], suprabasal [pemphigus vulgaris (PV) and Hailey–Hailey disease], and subepidermal [bullous pemphigoid (BP), dermatitis herpetiformis (DH), and bullous systemic lupus erythematosis (BSLE)] blistering disorders.

Inclusion Criteria

All patients with suspected immunobullous diseases who had undergone Tzank smear and histo-pathological were included in the study.

Exclusion Criteria

All cases of nonimmune-mediated vesiculobullous disorders were excluded from the study.

RESULTS-

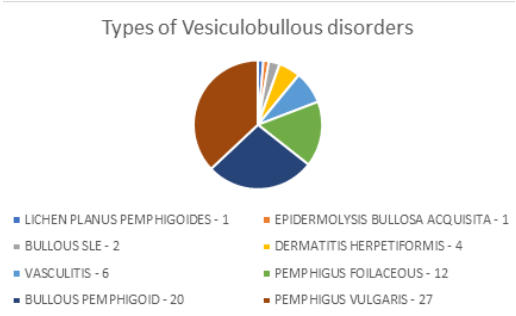
Of 83 patients, pemphigus vulgaris was the most observed disorder with 27 (44.57%) patients, followed by bullous pemphigoid with 20 (24.09%) patients, pemphigus foliaceus was seen in 12 (14.45%) patients. 6 (7.225) patients had vasculitis, 4 (4.82%) were having dermatitis herpetiformis. 2 (2.4%) of the patients had bullous SLE. Epidermolysis bullosa acquisita and lichen planus pemphigoides was seen in 1 (1.2%) patients each.

Male and females had almost similar incidence with 42 (50.60%) cases being females and 41 (49.39%) of the cases being males. Bullous pemphigoid group showed 12 (14.45%) of males and 8 (9.63%) females. Bullous SLE was seen only in female patients - 2 (2.4%). Dermatitis herpetiformis had a

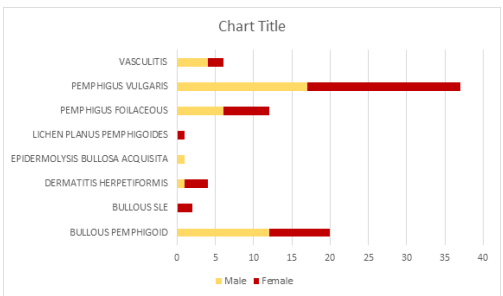
higher female prevalence compared to male, in this study, with 1 (1.2%) patient being male and 3(3.6%) patients being female. One case of epidermolysis bullosa acquisita was seen in male, where as single case of lichen planes pemphigoides was in female. Pemphigus foliaceus showed equal prevalence among male and female with 6 patients being male and 6 patients being female. Pemphigus vulgaris had a slightly higher prevalence among females with 20 (24.09%) cases where as males were 17 (20.48%) patients. Vasculitis group showed slightly male predominance with 4 (4.81%) cases being male and 2 (2.4%) cases being female.

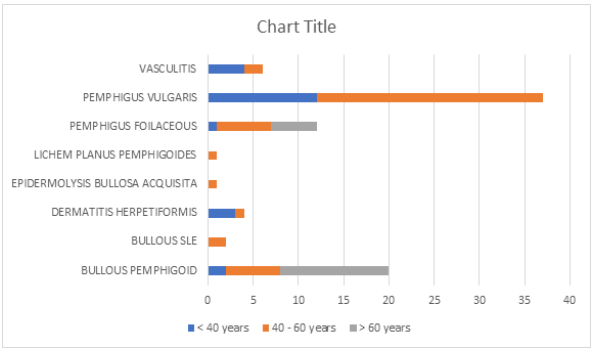
Of the 83 patients, the prevalence was high among patients aged 40-60 years with 44 patients falling into this category. Of these 44 pemphigus vulgaris had the most cases with 25 of 44 patients with the diagnosis. Deviating from this prevalance, bullous pemphigoid was observed to be highest in patients aged 60 years or above with 12 cases in that age group compared to 2 cases aged less than 40years and 6 cases in age group of 40 -59 years. Also dermatitis herpetiformis was observed to be more prevalent in age groups less than 40 years in comparison to 1 case in the age group 40-59 years and no cases noted above age 60 years, in this study.

	Number Of Cases	
LICHEN PLANUS PEMPHIGOIDES	1	1.2%
EPIDERMOLYSIS BULLOSA ACQUISITA	1	1.2%
BULLOUS SLE	2	2.4%
DERMATITIS HERPETIFORMIS	4	4.82%
VASCULITIS	6	7.22%
PEMPHIGUS FOILACEOUS	12	14.45%
BULLOUS PEMPHIGOID	20	24.09%
PEMPHIGUS VULGARIS	27	44.57%
TOTAL	83	



	Male	Female
BULLOUS PEMPHIGOID	12 (14.45%)	8 (9.63%)
BULLOUS SLE	0.0	2 (2.4%)
DERMATITIS HERPETIFORMIS	1 (1.2%)	3 (3.6%)
EPIDERMOLYSIS BULLOSA ACQUISITA	1 (1.2%)	0
LICHEN PLANUS PEMPHIGOIDES	0	1 (1.2%)
PEMPHIGUS FOILACEOUS	6(7.22%)	6 (7.22%)
PEMPHIGUS VULGARIS	17 (20.48%)	20 (24.09%)
VASCULITIS	4 (4.81%)	2(2.4%)





	<40 YEARS	40 - 59 YEARS	>60 YEARS
BULLOUS PEMPHIGOID	2	6	12
BULLOUS SLE	0	2	0
DERMATITIS HERPETIFORMIS	3	1	0
EPIDERMOLYSIS BULLOSA ACQUISITA	0	1	0
LICHEM PLANUS PEMPHIGOIDES	0	1	0
PEMPHIGUS FOILACEOUS	1	6	5
PEMPHIGUS VULGARIS	12	25	0
VASCULITIS	4	2	0

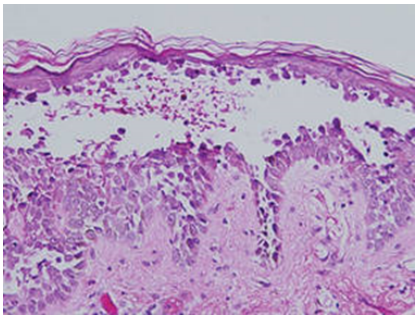


Fig 1(PV)

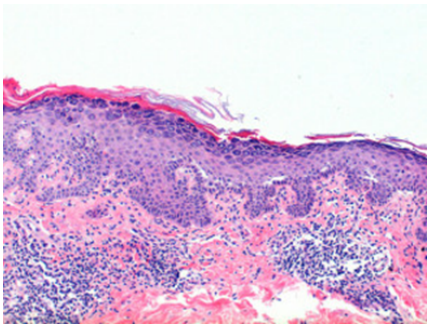


Fig 2 (PF)

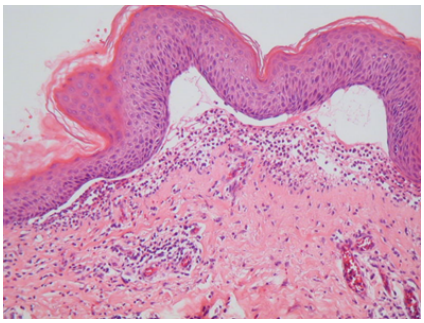


fig 3 (BP)

DISCUSSION:

Skin is the single largest organ of the body. It represents a window to the internal well-being. Various diseases along with its manifestations can commonly involve the skin and mucous membranes out of which vesiculobullous lesions form a predominant group[6].

Vesiculobullous diseases are a heterogeneous group of diseases, presenting with vesicle or bulla arising over the skin and/or mucous membrane. In spite of similar clinical presentations, they are remarkably histopathologically different from each other. Proper diagnosis is essential to prevent the fatal outcome if untreated.

Out of the 83 cases of pemphigus studied, PV was the predominant type with 27 cases (44.57%) followed by 20 cases (24.09%) of bullous pemphigoid and 12 cases (14.45%) of PF. This corroborates with an earlier study by Arya *et al.* where 61.42% of cases were PV.[7] Singh[8] and Chowdhury *et al.*[9] also found PV as the major group. Our findings were also consistent with Deepti S.P. *et al.* study and Arundhati *et al.* study.

In the present study, the prevalence was high among those aged 40-60 years with 44 patients (53.01%) falling into this category. Study conducted by Basu *et al.* included cases with age ranging from 17 to 85 years (mean 46.24 ± 15.48 years). This finding was also similar to the study done by Singh *et al.*[10] and Khannan *et al.*[11] A recent study of pemphigus by Chowdhury *et al.* showed the age at presentation to be from 16 to 76 years (mean 49.48 ± 16.51 years).[9]

There was a slight female predominance with male:female ratio of 1:1.02. This was similar to study by Basu *et al.* where ratio of 1:1.12 was found. It was also similar to that of a study by Chowdhury *et al.*, where female:male ratio was 1.9:1.[9] Zaraa *et al.*[12] and Deepti *et al.*[13] also documented similar findings. Bullous pemphigoid group showed 12 (14.45%) of males and 8 (9.63%) females, thus male:female ratio of 1.5:1 which was contrary to a study by Rokde et al. where the ratio was 1:1.44.

Our findings of pemphigus foliaceus, subcorneal pustular dermatoses and erythema multiforme were comparable with other studies. [14,15] Banu L et al [16] and Dipti SP et al [13] show 3 cases and 2 cases with subepidermal bulla and papillary microabscess in all cases of dermatitis herpitiformis.

CONCLUSION:

Clinical and histo-morphological study of vesiculobullous diseases can be used in confirming the diagnosis of diseases. Clinical data is essential for histopathological interpretation. Immunofluorescence study helps in confirming the diagnosis where clinical presentation and histopathology are inconclusive.

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