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To Study the various Clinical Presentations-Age of Onset, Sex Preponderance, Course of Disease and Histopathological patterns of Autoimmune Vesiculobullous Disorders

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Abstract: The objective of the study was to observe the various clinical features, histopathological and immunofluorescence findings in patients with autoimmune vesiculobullous disorders. The overall M: F ratio of autoimmune vesiculobullous disorders in the study was 1: 1.4. Trunk was the most common site of involvement in all types of autoimmune vesiculobullous disorders. Overall trunk was the most common site of onset of lesions in autoimmune vesiculobullous disorders but in pemphigus vulgaris mucosal onset is more common than trunk and extremities. Incidence of autoimmune vesiculobullous disorder was 0.11% of the total number of patients attending the skin OPD. Pemphigus vulgaris constituted the most common subtype of autoimmune vesiculobullous disorder in this study, followed by pemphigus foliaceus. Pemphigus group of diseases were most common in the 4th decade while bullous pemphigoid was most common in the 7th decade. Histopathological examination showed features typical of each subtype of autoimmune vesiculobullous disorder.
Keywords: Histopathological, Autoimmune Vesiculobullous Disorders.

INTRODUCTION

The bullous diseases although uncommon, have a dramatic impact on the patient and their relatives and have severe economic consequences for the family and health services.

These diseases have been the subject of intensive investigation in recent years and the study of both the genetic and autoimmune diseases has made major contributions to our knowledge of the biology of the skin. The identification of the specific target antigens for the auto antibodies in the autoimmune bullous diseases has led to the discovery of many components of the desmosome and the adhesion complex linking the epidermis to the dermis [1].

The immunological diseases are classified largely by their clinical presentation, histopathology and immunopathology. The immunobullous diseases are characterized by pathogenic auto antibodies directed at target antigens whose function is either cell-to-cell adhesion within the epidermis or adhesion of stratified squamous epithelium to dermis or mesenchyme. These target antigens are components of desmosomes or the functional unit of the basement membrane zone known as the adhesion complex [1].

MATERIALS & METHODS

Source of data

This study was conducted in the Department of Dermatology, Venereology and Leprology, Government Medical College and Rajindra Hospital, Patiala. As it is a time bound study, all cases attending and referred from other department with an intact / ruptured, vesicle / bulla at the time of presentation were selected.

Method of collection of data

- Type of study- cross section observational
- The clinical data & histological findings

The clinical history like age, sex, demography, duration of the disease, site of lesion, any significant family history, any associated systemic disease, any history of drug intake, history of injury, risk factors and complications were noted in a pre-tested and pre-designed proforma after taking informed and written consent. Diagnosis was established by history and clinical examination. After this, biopsy was done under aseptic condition and with consent.

Inclusion criteria

All cases of autoimmune vesiculobullous disorder with fresh lesion, not on treatment attending the Dermatology Department irrespective of age, sex and associated diseases

Exclusion criteria

1) Autoimmune bullous disorders on steroids, immunosuppressive drugs
2) The following conditions are excluded from study –
   a) Infectious diseases-
      i. Viral- herpes simplex, varicella, hand-foot-mouth disease, herpes zoster.
      ii. Fungal- Candidiasis.
      iii. Bacterial- congenital syphilis, bullous impetigo, staphylococcal scalded skin syndrome.
   b) Inflammatory conditions: Bullous mastocytosis, erythema toxicum neonatorum, transient neonatal pustular melanosis, suckling blisters, vesicular form of Lepra reactions, pustular psoriasis, subcorneal pustular dermatosis.
   c) Genetic disorders: Epidermolysis bullosa, incontinentia pigmenti, bullous congenital icthyosiform erythroderma, Hailey-Hailey disease, Darier's disease.
   d) Metabolic: Acrodermatitis enteropathica, lead poisoning, Diabetic bullae, porphyria cutanea tarda.
   e) Drug Reactions: Bullous drug reactions, bullous fixed drug reaction, erythema multiforme, and Steven Johnson syndrome/TEN.
   f) Dermatitis- Allergic contact Dermatitis, Irritant contact Dermatitis.
   g) Traumatic conditions-Chemical injury, thermal injury, friction blister.
   h) Environmental-Phototoxic reactions, photoallergic reactions, insect bite reactions

RESULTS AND DISCUSSION

Incidence of autoimmune vesiculobullous disorders

The present study was conducted over a period of 24 months from November 2013 to October 2015 in the OPD of Dermatology, Venereology and Leprosy in Government Medical College and Rajindra Hospital, Patiala.

A total number of 48,750 patients attended the Dermatology OPD during this period out of which 70 were cases of autoimmune vesiculobullous disorders.

So the incidence of autoimmune vesiculobullous disorders among the OPD cases in two year works out to be 0.14%. Another study done by Kanwar AJ *et al.* [2] showed an incidence of 0.3% which is comparable.

However, it must be stated that ours is a hospital-based study and so the above number does not reflect the true incidence of autoimmune vesiculobullous disorders in the community.

In the present study, pemphigus vulgaris was the most common vesiculobullous disorder constituting 61.4% (43 out of 70 cases) followed by pemphigus foliaceus constituting 15.7% (11 out of 70 cases) of the total cases of autoimmune vesiculobullous disorders. Bullous pemphigoid constituted 11.4% of the cases. The frequency of bullous pemphigoid is less as compared to other studies.

The pattern of frequency of different types of autoimmune vesiculobullous disorders correlates closely to Arya *et al.* [3] study. The incidence of Linear IgA disease was 2.9% in our study which correlates closely with Kanwar AJ *et al.* [2] study. Dermatitis herpetiformis was present in 7.2% of cases.

Also there was a case of pemphigoid gestationis like in study of Solanki A *et al.* [4]. Other diseases like epidermolysis bullosa acquisita and subcorneal pustular dermatoses were not found during the present study like Kanwar *et al.* [74] study.

Age and sex distribution of vesiculobullous disorders

Table-1: Comparison of mean age of onset and sex distribution of vesiculobullous disorders in different studies

No.	Studies	Year	PV		PF		BP		DH		LAD	
			Age	M:F	Age	M:F	Age	M:F	Age	M:F	Age	M:F
1.	Micali G *et al.* [5]	1998	56	1:2.2	_	_	_	_	_	_	_	_
2.	Aboobaker J *et al.* [6]	2001	48	1:1.17	43	1:1.4	_	_	_	_	_	_
3.	Javidi Z *et al.* [7]	2007	40.64	1:1.4	44.39	1:1.2	_	_	_	_	_	_
4.	Deepti SP *et al.* [8]	2015	_	1:2.4	_	1:3	_	1:1.7	_	1:1	_	_
5.	Present study	2015	39.33	1:1.38	44.73	1:1.2	62.5	1:1.6	39.2	1:4	25.5	2:0

The mean age of onset for pemphigus vulgaris was 39.33 years and M: F ratio was 1:1.38 in the present study which is almost similar to the results of Javidi Z *et al*. [7] study. Pemphigus foliaceus presented in the 5th decade in the present study nearly similar to 5th decade in Javidi Z *et al*. [7] study.

Females had a slightly higher incidence of pemphigus vulgaris and pemphigus foliaceous compared to males in the present study. All of the above studies [6,7,5,8] also show similar results.

The mean age of onset of bullous pemphigoid was 62.5 years with a much higher incidence in females (M: F- 1:1.6) in the present study which is in concordance with Deepti SP *et al*.[8]. The mean age of onset of linear IgA disease was 25.5 years with both the patients being male patients in the present study.

Table-2: Distribution of types of vesiculobullous disorders

Type	Number of cases	Percentage
Pemphigus vulgaris	43	61.4%
Pemphigus foliaceus	11	15.7%
Bullous pemphigoid	08	11.4%
Linear IgA disease	02	2.9%
Dermatitis herpetiformis	05	7.2%
Pemphigoid gestationis	01	1.4%
Total	70	100

In the present study, pemphigus vulgaris constituted the most common vesiculobullous disorder constituting 61.4% (43 out of 70 cases), followed by pemphigus foliaceus constituting 15.7% of the cases. Bullous pemphigoid and dermatitis herpetiformis constituted 11.4% and 7.2% of the cases respectively. Linear IgA disease was found in 2.9% of cases and pemphigoid gestationis constituted 1.4% of total cases.

Table-3: Age distribution in present study

Age group(Years)	Number of patients	Percentage
0-10	01	1.4%
11-20	04	5.7%
21-30	10	14.3%
31-40	20	28.6%
41-50	14	20%
51-60	10	14.3%
61-70	09	12.9%
>70	02	2.8%
Total	70	100

In the present study, the youngest patient was 09 years old and the oldest patient was 75 years old. The maximum numbers of cases were seen in the age group 31-40 years (28.6%) followed by 41-50 years (20%), 51-60 years and 21-30 years (14.3%) which had equal number of cases. The mean age of the study population was 42.25 years.

Table-4: Sex distribution in autoimmune vesiculobullous disorders

Sex	PV	%age	PF	%age	BP	%age	DH	%age	LAD	%age	PG	%age	Total
Male	18	41.8%	05	45.5%	03	37.5%	01	20%	02	100%	00	-	29
Female	25	58.2%	06	54.5%	05	62.5%	04	80%	00		01	100%	41
Total	43	100	11	100	08	100	05	100	02		01		70
M:F ratio	1:1.38		1:1.2		1:1.6		1:4		-		-		1:1.4

In the present study, there were totally 29 males and 41 females giving a M: F ratio of 1:1.4 for autoimmune vesiculobullous disorders. Pemphigus group of disorders had female predominance, pemphigus vulgaris showing a M: F ratio of 1:1.38 and pemphigus foliaceus showing a M: F ratio of 1:1.2. Bullous pemphigoid had M:F ratio of 1:1.6. Dermatitis herpetiformis showed female predominance whereas Linear IgA disease showed male predominance.

Table-5: Distribution of subtypes of pemphigus group

Subtype	Number	Percentage
Pemphigus vulgaris	43	79.6%
Pemphigus foliaceus	11	20.4%
Total	54	100

In the present study, there were 43 cases of pemphigus vulgaris and 11 cases of pemphigus foliaceus out of 54 cases of pemphigus group. Pemphigus vulgaris (79.6%) was the most common subtype followed by pemphigus foliaceus (20.4%).

Table-6: Distribution of duration of illness in autoimmune vesiculobullous disorders

Duration (months)	PV	%	PF	%	BP	%	DH	%	LAD	%
1-6	10	23.26%	4	36.37%	5	62.5%	2	40%	0	-
7-12	8	18.60%	3	27.27%	1	12.5%	2	40%	1	50%
13-24	6	13.95%	2	18.18%	0	-	1	20%	0	-
25-36	4	9.30%	1	9.09%	2	25%	0	-	1	50%
>36	6	13.95%	1	9.09%	0	-	0	-	0	-

In the present study, most cases of pemphigus vulgaris (23.26%), pemphigus foliaceus (36.37%) and bullous pemphigoid (62.5%) had illness for 1–6 months before presentation. 40% of Dermatitis herpetiformis had illness for 1-6 months and 40% had duration 7-12 months. 50% of the cases of LAD had illness for 25–36 months and 50% for 7–12 months before presentation.

CONCLUSION

Incidence of autoimmune vesiculobullous disorder was 0.11% of the total number of patients attending the skin OPD. Pemphigus vulgaris constituted the most common subtype of autoimmune vesiculobullous disorder in this study, followed by pemphigus foliaceus. Pemphigus group of diseases were most common in the 4th decade while bullous pemphigoid was most common in the 7th decade.

Histopathological examination showed features typical of each subtype of autoimmune vesiculobullous disorder.

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