

Study of Clinico Pathological Correlation in Autoimmune Bullous Disorders.

Nageswaramma.S¹, D.S.S Srinivas Prasad², P.V.Sivaram Sandeep³

Department Of Dermatology, Venerology, Leprosy, Guntur Medical College, Guntur

Abstract

Background: Immune mediated vesicobullous diseases can be diagnosed by close attention to the clinical details, histological confirmation and the clinical setting of the blistering. Confirmatory studies such as immunofluorescence will help in diagnosis of cases where histopathological features alone are not diagnostic. To evaluate clinical findings in association with histopathological findings, to study the histopathological findings in various immunobullous lesions of skin, and to confirm with immunofluorescence wherever possible

Aim: The aim is to study histopathology of various Autoimmune vesicobullous disorders and its correlation with clinical evaluation in patients attending DVL department, Guntur medical college.

Material & Methods : A total of 80 patients of various bullous disorders attending to department of DVL in one and half year period from July 2015 to december 2016 were analysed. of these females (44), males (33), and (3) were children .The clinical features and histopathology were correlated for the diagnosis and direct immunofluorescence (DIF) was done whenever necessary.

Results: out of 80 patients we studied, females were 44(55 %), males 33(41.25%), children 3 (3.75%). 40 to 50 yrs age group were more commonly affected .we studied & correlated histopathology and DIF with clinical findings , of these 43 patients (53.7%) were pemphigus vulgaris. 18 (22.6 %) were bullous pemphigoid. 4 were (2.51%) Dermatitis herpetiformis, 2 (2.51 %) were pemphigus vegetans, 5 were pemphigus foliaceus(6.25%), 3 were CBDC (3.72%).

Discussion & Conclusion : Histopathology findings were correlated with clinical diagnosis in majority (95.5%) of autoimmune bullous disorders except few cases (5.25%) where diagnosis is done by immunofluorescence findings. To conclude, the correlation between clinical findings, Histopathology and Immunofluorescence plays a predominant role to confirm the atypical appearance of autoimmune blistering diseases.

I. Introduction

The bullous diseases have a history as old as that of medicine. Although blisters have drawn attention of medical caregivers throughout written history, only modern times have seen the origin of a clear classification of these diseases, based upon clinical and histological criteria. By close attention to the clinical details, histological confirmation and the clinical setting of the blistering a correct diagnosis can be achieved. Confirmatory studies such as immunofluorescence will help in diagnosis of cases where histopathological features alone are not diagnostic

II. Materials And Methods

The present study constitutes clinicopathological evaluation of primary vesicobullous lesions of skin. 80 cases of vesicobullous disorders where diagnosed in the department of DVL GGH Guntur, during the study period of one and half years.

A detailed history with particular reference to the mode of onset, characteristics and distribution of the lesions was taken. Histopathological evaluation of biopsies was done. Biopsy material was sent in 10% formalin. Immunofluorescence was done wherever possible.

Clinical data was recorded in the form of :

- Detailed clinical history .
- General & dermatological examination .
- Routine investigations.
- Tzanck smear for Acantholytic cells.
- After taking written informed consent of the patients, two biopsies were performed one from the fully developed vesicobullous lesion (lesional biopsy) for histopathological examination.
- The other from the perilesional skin area with in 2cm of diameter of lesion (perilesional biopsy) for Direct immunofluorescence whenever necessary.

III. Results

Table 1 . distribution of various immunobullous disorders.

SL No	Disorder	No. of Cases
1	Pemphigus Vulgaris	45
2	Bullous pemphigoid	19
3	Pemphigus Vegetens	2
4	Dermatitis herpetiformis	5
5	Pemphigus foliaceus	6
6	Chronic bullous disorder of childhood	3
Total		80

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out of 80 patients we studied, females were 44(55 %), males 33(41.25%), children 3 (3.75%).

Table 2 - Sex wise distribution of immunobullous disorders

TYPE	MALES		FEMALES	
	Number	Percentage	Number	Percentage
Pemphigus vulgaris	17	21.25	28	35.25
Bullous pemphigoid	9	11.25	10	12.52
Dermatitis herpetiformis	4	5.23	1	1.25
Pemphigus vegetens	0	0	2	2.52
Pemphigus foliaceus	3	3.75	3	3.75
CBDC	1	1.25	2	2.52
total	34	42.73	46	57.27

Table 3 – Age Wise Distribution Of Immunobullous Disorders

Type	0-10	11-20	21-30	31-40	41-50	51-60	61-70	>70	total
Pemphigus vulgaris			2	10	25	4	3	1	45
Bullous pemphigoid					5	7	3	4	19
Dermatitis herpetiformis			1	2	2				5
Pemphigus vegetens				2					2
Pemphigus foliaceus				1	2	2	1		6
CBDC	1	2							3
total	1	2	3	15	34	13	7	5	80

40 to 50 yrs age group were more commonly affected with immunobullous disorders.

Pemphigus vulgaris (25), bullous pemphigoid (5), dermatitis herpetiformis (2), pemphigus vegetens (2) cases was seen in our study. Increased cases of Bullous pemphigoid was seen in 51-60 years of age group. Pemphigus vulgaris was seen in 41-50 years of age group.

IV. Discussion

The diagnosis of autoimmune bullous disease was based on evaluation of Clinical findings with correlation to Histopathology The most frequent disorder in our study was pemphigus vulgaris followed by bullous pemphigoid ,Cases of limited number in our study were pemphigus foliaceus, pemphigus vegetans, CBDC, Dermatitis herpetiformis. Male to female ratio was 1:1.7 which is comparable to the findings of Shamim et al, Archana et al. 40 to 60 yrs age group were more commonly affected Majority of the lesions in pemphigus vulgaris were distributed in the oral cavity (44%) followed by trunk (38%) and extremities (33%). Most of them were flaccid bullae (90.9%) situated on a non erythematous base. The lesions in bullous pemphigoid were situated in the extremities in 62.5% followed by chest in 50% and were associated with itching in 55.4% of the cases. Most of them were tense bullae (90.9%) situated on erythematous base.

Out of 45 cases of pemphigus vulgaris studied clinically, 44 of them showed consistent results with biopsy features showing suprabasal bulla (94.4%) with neutrophils, eosinophils and acantholytic cells which has

been documented in many other studies. 1 out of 45 showed nonspecific features for which DIF was done and showed IgG positivity in ICS in fishnet pattern in 80%.

Bullous pemphigoid affects the elderly during 5th to 7th decade of life, with an average age of onset being 65yrs. In our study, out of 19 cases of bullous pemphigoid studied clinically, 18 cases showed consistent results with biopsy features showing sub epidermal split with eosinophils. 1 out of 19 cases showed nonspecific features for which DIF was done and showed homogenous linear deposits of IgG and C3 in the basement membrane zone. Pemphigus foliaceus cases in our study are 6, 5 (83.3%) cases showed consistent results with histology. One was diagnosed with help of immunofluorescence. Pemphigus vulgaris and foliaceus can be differentiated by histological features and DIF. Pemphigus vegetans cases are correlated successfully both clinically and histopathologically and are differentiated from pemphigus vulgaris.

Dermatitis Herpetiformis is 20-40 years and occasionally occurs in children. The histopathological findings noted were subepidermal bulla with neutrophils and a few lymphocytes. In our study total 5 cases are studied clinically and correlated with histological findings. 4 out of 5 (83.3%) consistent results are obtained. All the 3 clinically diagnosed CBDC cases, were histopathologically consistent with CBDC our study showed consistent results in correlation between clinical diagnosis with histopathological findings in majority of cases except in few cases where diagnosis was correlated with direct immunofluorescence. there was 95.5% correlation between clinical findings with histological features in our study with error of 4.5%. where immunofluorescence was done for the diagnosis of remaining cases.

Clinical photographs



Fig: 1- Pemphigus Vulgaris Showing Flaccid Bulla

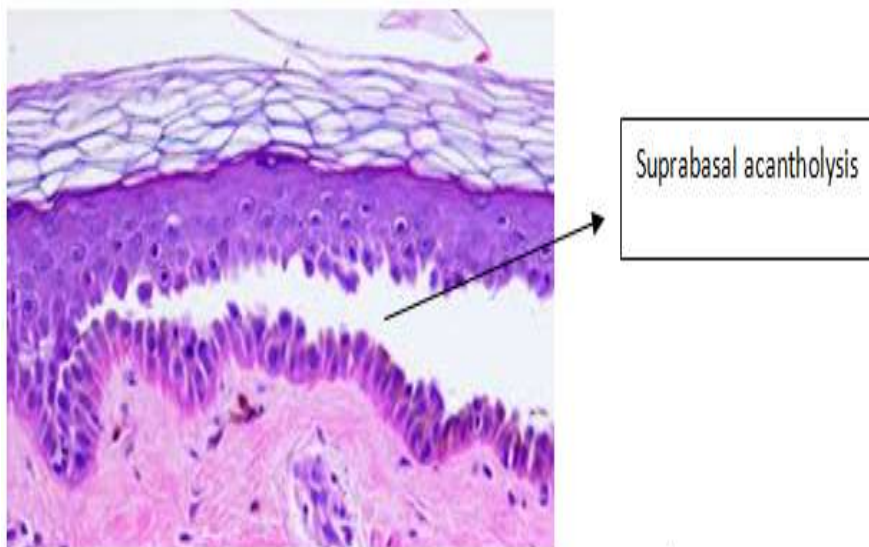


Fig:2 –Pemphigus Vulgaris Showed Intraepidermal Blister, Suprabasal Acantholysis

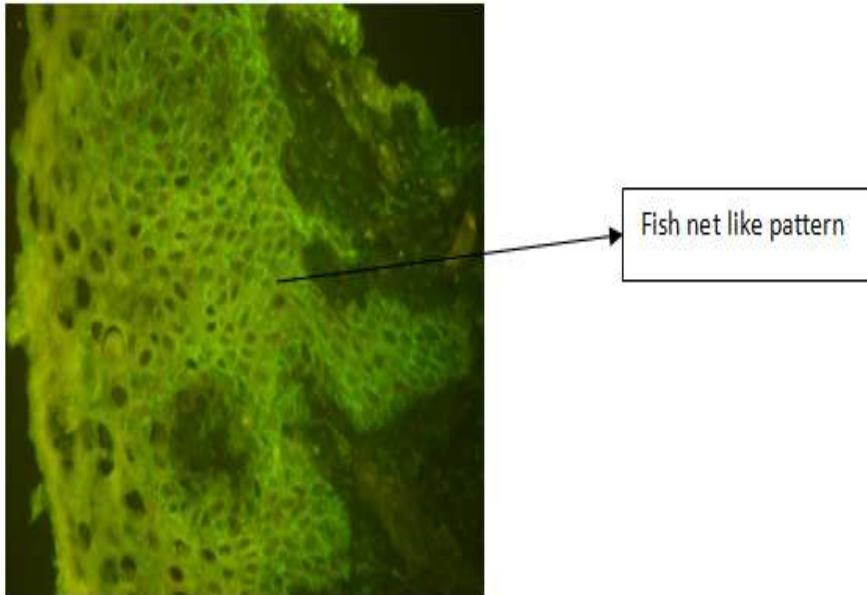


Fig 3: Direct Immunofluorescence Of Pemphigus Vulgaris Showed Lace/Fishnet Pattern Of IgG,Ics

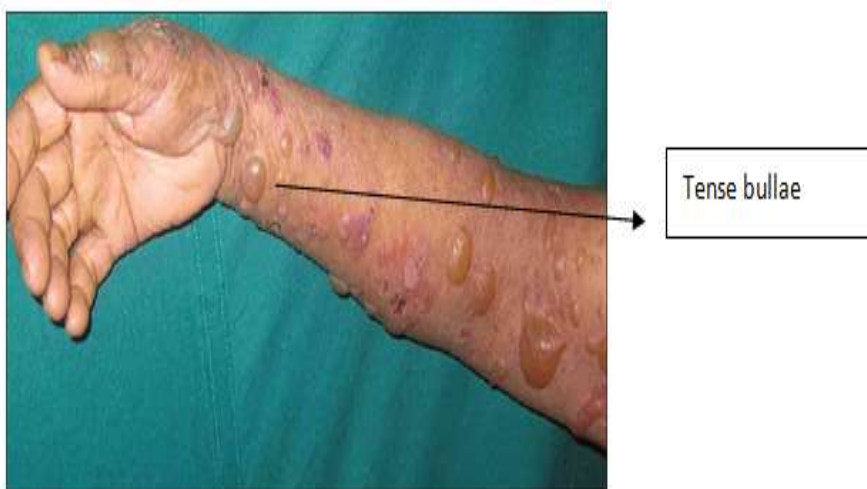


Fig : 4 – Tense Bullae In Bullous Pemphigoid Patients

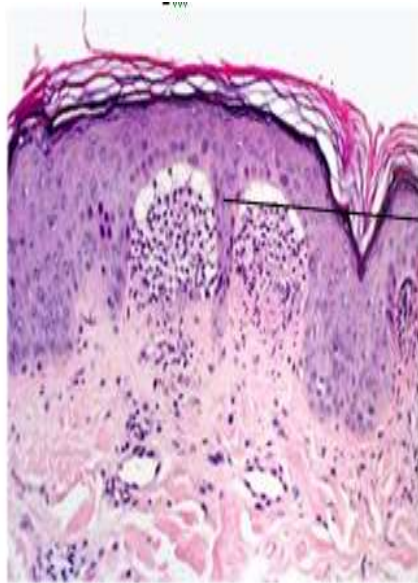


Fig:5 – HPE Showing Sub Epidermal Split



Multiple pruritic vesicles, bulla and erosions

Fig:6 – Vesicles And Bulla In Dermatitis Herpetiformis Patient



Subepidermal split with

Fig: 7- HPE Of Dermatitis Herpetiformis



String of pearl appearance of the vesicles

Fig: 8 – A child with chronic bullous disorder of childhood

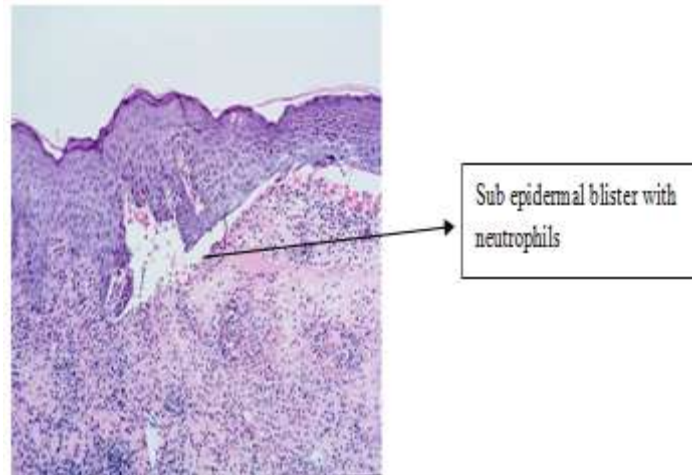


Fig: 9 – Hpe Of Cbd

V. Conclusion

The association of clinical and histopathological features in the study helped to arrive at the diagnosis in most cases with significant association. This increases the chance of making diagnosis with help of simple investigations without the need for expensive investigations like DIF. But Immunofluorescence study done in a few cases helped in confirming the diagnosis where histopathology and clinical features alone were inconclusive.

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