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## Autoimmune Bullous Dermatoses: Correlation between clinical and immunohistopathological diagnosis

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### INTRODUCTION

Autoimmune bullous dermatoses (AIBD) are a group of vesiculo-bullous disorders characterized by pathogenic antibodies directed against components of the desmosomes and other adhesion molecules at various levels. Histopathology and immunofluorescence are invaluable in confirming the diagnosis and for their appropriate management.

### METHODS

A prospective cross-sectional study was conducted from November 2022 to August 2024. Study population included patients suspected of AIBD. Patients of all age and either sex, willing to undergo biopsy for HPE and/or immunofluorescence were included while pregnant and lactating women were excluded from the study. Data was compared for concordance and discordance among the clinical, histopathological and immunopathological diagnosis.

### RESULTS

- Total cases = 63 (39 females and 24 males). M:F = 1:1.6
- Age = 3 – 84 years with mean age of 50.46 years
- Females (47.10 yr) had earlier age of onset as compared to males (55.92yr)
- Mean age for pemphigus vulgaris was 41.91 years
- Mean age for bullous pemphigoid was 61.71 years
- 31 patients belonged to pemphigus group (23 of PV & 8 of PF)
- 23 patients belonged to pemphigoid group (21 of BP & 2 of LPP)
- 2 cases of DH and 1 case of LABD.
- Overall there was 79.3% concordance and 11.11% discordance between clinical diagnosis and final diagnosis (confirmed by HPE and immuno-fluorescence)
- 80.5% concordance in pemphigus group
- 100% in pemphigoid group.
- HPE and Immunofluorescence findings aligned with clinical diagnoses in over 82% and 80% of cases respectively.

Final Diagnosis	Age range			Mean age			Clinical Diagnosis →	PV	PF	BP	LP-P	B-LP	LABD	DH	HHD	Final
	Female	Male	Total	Female	Male	Total										
PV	16 - 60	16 - 70	<b>16 - 70</b>	39.38	47.71	<b>41.91</b>	Final Diagnosis ↓	22							1	23
PF	35 - 68	45 - 63	<b>35 - 68</b>	51.80	53.00	<b>52.25</b>	PV	1	7							8
BP	35 - 80	60 - 84	<b>35 - 84</b>	58.21	68.71	<b>61.71</b>	BP	3		17				1		21
LP-P	40	17	<b>17 - 40</b>	40.00	17.00	<b>28.50</b>	LP-P				1	1				2
LABD	X	14	<b>14</b>	X	14.00	<b>14.00</b>	LABD						1			1
DH	X	65 - 70	<b>65 - 70</b>	X	67.50	<b>67.50</b>	DH							2		2
Negative/ Inconclusive	3 - 45	64 - 70	<b>3 - 70</b>	31.00	67.33	<b>49.17</b>	Inconclusive	3					1	1		5
	<b>3 - 80</b>	<b>14 - 84</b>	<b>3 - 84</b>	<b>47.10</b>	<b>55.92</b>	<b>50.46</b>	Non-blistering							1		1
							Clinical	29	7	17	1	1	2	5	1	63

Final Diagnosis	Female	Male	Grand Total	Disorder	Clinical diagnosis	Final diagnosis	Case diagnosed correctly	Concordance	Case diagnosed incorrectly	Cases missed
Pemphigus	21	10	<b>31</b>	Pemphigus	36	31	29	<b>80.5%</b>	4	2
Pemphigoid	15	8	<b>23</b>	Pemphigoid	18	23	18	<b>100%</b>	0	5
LABD	0	1	<b>1</b>	LABD	2	1	1	<b>50%</b>	0	0
DH	0	2	<b>2</b>	DH	5	2	2	<b>40%</b>	2	0
Negative/ Inconclusive	3	3	<b>6</b>	Other/ Inconclusive	2	6	-	-	-	-
<b>Grand Total</b>	<b>39</b>	<b>24</b>	<b>63</b>	<b>Total</b>	<b>63</b>	<b>63</b>	<b>50</b>	<b>79.3%</b>	<b>6</b>	<b>7</b>

### DISCUSSION

Possible reasons for Concordance	Possible explanation for Discordance
Typical skin lesion (blisters- flaccid/tense)	Improper history
Mucosal lesion in pemphigus	Partial treatment at time of presentation modifying the clinical picture
Typical distribution of lesions	Atypical lesions – tense lesions in pemphigus or flaccid lesion in pemphigoid
Age group- younger in pemphigus as compared to pemphigoid	Mucosal involvement in pemphigoid/ absence in pemphigus
Clinical signs- Nikolsky and Asboe Hansen sign	Secondary infections modifying the clinical picture
Biopsy of early lesion	Incorrect selection of lesion for biopsy
	False positive - Nikolsky
	Bulla spread sign – error in judgement(subjective)

### CONCLUSION

HPE was instrumental in differentiating between pemphigoid and pemphigus disorders, while direct immunofluorescence confirmed the deposition of immunoglobulins in various patterns. Based on this study we conclude that clinical, histopathological and DIF features be taken into consideration to arrive at final diagnosis as these methods may not be diagnostic individually in each and every case. This enables early and targeted treatment strategies in immunobullous disorders.

### REFERENCES

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