



A Characterization of Clinical Presentation and Outcomes in Patients With Oral Mucous Membrane Pemphigoid: A Retrospective Study

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Introduction

Background:

- Mucous membrane pemphigoid (MMP), is a chronic autoimmune subepithelial bullous disorder with involvement of mucosal surfaces (See Figures 1 and 2).¹
- Diagnosed by characteristic clinic manifestations, H&E, and direct immunofluorescence (DIF) microscopy that show linear IgG, IgA, and/or complement in the basement membrane zone (BMZ).¹
- There is limited data to guide initial therapy for MMP based on baseline severity or DIF results.

Objective:

- To evaluate and compare clinical outcomes in oral MMP patients with varying disease severity.

Methods

Design: Retrospective case series study.

Population: 53 oral MMP patients identified by immunopathology and EMR records that were seen by OHSUs Department of Dermatology.

Measurements: Disease severity was analyzed using our novel scoring system (See Table 1) and the Oral Disease Severity Score (ODSS) (See Table 2) at two time-points: Baseline visit and Final visit. Patients were then assessed for time to disease control, remission status, final effective medication, and initial and final severity based on immunoglobulin class.

Mild		Moderate		Severe	
Exam	Functional	Exam	Functional	Exam	Functional
Mild erythema 1-2 sites	1-3 pain score	Marked erythema 3-4 sites	4-6 pain score +Eating +Bleeding w/brushing	Marked erythema Erosions > 4 sites	7-10 pain score +Soft/bland food +Weight loss +Speaking/Dental

Table 1: Novel MMP Severity Scoring table

Site	Site Score	Activity Score
Outer Lips (1)		
Inner Lips (1)		
R Buccal Mucosa (1 or 2)		
L Buccal Mucosa (1 or 2)		
Gingivae ((1 each of 6 segments (Lower R/L/C, Upper R/L/C))		
Dorsum of Tongue (1 or 2)		
Right Ventral Tongue (1)		
Left Ventral Tongue (1)		
Floor of Mouth (1 or 2)		
Hard Palate (1 or 2)		
Soft Palate (1 or 2)		
Oropharynx (1 or 2)		
Total= Site + Activity		

Table 2: Oral Disease Severity Scoring (ODSS) table.²

Definitions

Disease Control:

- The time at which new inflammatory lesions cease to form and established oral lesions begin to heal.³

Time to Disease Control:

- The time interval from baseline to the control of oral disease activity.³

Partial remission:

- The presence of transient new oral lesions that heal without scarring within 1 week.³

Complete remission:

- The absence of new or established oral lesions for at least 2 months.³

Minimal Therapy:

- Dapsone ≤ 1.0 mg/kg/d, doxycycline ≤ 100 mg/d, mycophenolate mofetil ≤ 500 mg/d; mycophenolic acid ≤ 360 mg/d.³



Figure 1: Erythema and erosion on gingivae.⁴



Figure 2: Erosion on labial mucosa and moderate gingival erythema.⁴

Results

- Patients with baseline severe disease were more likely to require immunosuppressive drugs for partial or complete remission than their mild/moderate counterparts (OR = 7.51, 95% CI [2.09, 27.01])
- Patients with IgA involvement on DIF tended to be less likely to obtain "Complete" remission status than patients without IgA involvement (34% vs. 58%)
- Patients with IgA involvement on DIF required immunosuppressive for disease control than those without IgA involvement (OR = 3.69, CI [1.14, 27.01]).
- Baseline scores of the validated ODSS trended closely with the novel scoring system.
- Patients with IgA involvement on DIF tended to have Dapsone as their final effective medication less often than patients without IgA involvement (OR = 0.27, 95% CI [0.09, 0.86])

References:

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Results

		Baseline Novel MMP Severity Score		
		Mild/Moderate (35)	Severe (18)	Total (53)
Final Effective Medication	Anti-Inflammatory	23	4	27
	Immunosuppressive	9	13	22
	Topicals	3	1	4
DIF Results	IgG + IgA or IgA alone	18	11	29
	IgG	17	7	24
Final Novel MMP Severity Score	Clear	16	8	24
	Mild	13	6	19
	Moderate	6	4	10
Time to Disease Control	0-3 months	16	7	23
	4-6 months	7	7	14
	6+ months	12	4	16
Remission Status	Partial – No Therapy	1	1	2
	Partial – Minimal Therapy	17	8	25
	Partial – Long-term biologics	1	1	2
	Complete – No Therapy	5	1	6
	Complete – Minimal Therapy	10	3	13
Baseline ODSS Score	0-19	19	1	20
	20-29	12	7	19
	30+	4	10	14

Table 3: Baseline Novel MMP Severity Score vs. Outcome Measures

Discussion and Conclusions

- There is a strong correlation between severe baseline disease and the absence of complete remission status and the use of immunosuppressive medications. Perhaps it is necessary to begin with immunosuppressive medications earlier in clinical onset for better clinical outcomes.
- Contrary to some expert opinion our findings showed that patients with IgA were less successful on Dapsone than patients without IgA involvement.
- Limitations include the retrospective nature of the study and the use of unvalidated novel severity score system.
- Our novel scoring system needs to go through a process of validation through experts to determine its utility with MMP patients.
- The ODSS typically has a component of pain score that we were unable to include due to the retrospective nature of the study.
- Future work would need to be done in a prospective manner with standardized forms to determine best practices for management of oral MMP patients and creation of a novel severity score system for other mucosal sites than the mouth.