Acta Scientific Otolaryngology

Volume 3 Issue 6 June 2021

Research Article

Mucous Membrane Pemphigoid with Different Manifestations- A Cross Sectional Study

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Received: February 19, 2021

Published: May 11, 2021

Abstract

Background: The study aimed to assess the different manifestations of MMP among patients diagnosed as MMP.

Methodology: The study was conducted as a cross sectional study at PMCH, Patna for a period of 3 years on 30 patients diagnosed as a case of MMP in the departments of Otorhinolaryngology, dermatology and ophthalmology. Detailed history regarding clinical features, mode of presentation, and duration of symptoms was obtained and entered in questionnaire. Statistical analysis- Data was compiled using Ms Excel and analysed using IBM SPSS 20. Categorical data was expressed as frequency and percentage.

Results: Majority of patients with MMP belonged to 61 to 80 years of age (83.3%) and majority of cases were females. The most common site of involvement was oral mucosa (96.7%), followed by ocular conjunctiva in 63.3% and laryngeal mucosa (46.7%). About 55.8% patients with oral lesions presented with oral ulceration and about 47.4% cases with ocular lesions presented with non-specific conjunctivitis. In cases with laryngeal involvement, 28.6% cases presented with laryngeal stenosis with breathing difficulty. Nasal mucosal erosions, localized erythematous plaque, dysphagia and odynophagia were predominant manifestations in cases with nasal, skin, pharynx and oesophageal involvement respectively.

Conclusion: Mucous membrane pemphigoid is a rare disorder which most commonly involve oral mucosa. However, ocular, nasopharynx, larynx, oesophagus involvement can also be observed. Depending upon site of involvement, patients may present with varied manifestations such as oral erosions and ulcers, desquamative gingivitis in case of oral lesions, cicatrizing conjunctivitis in case of ocular involvement.

Keywords: Mucous Membrane Pemphigoid; Oral; Ocular; Pseudo Membrane; Airway

Introduction

Mucous membrane pemphigoid (MMP), also called cicatricial pemphigoid or Ocular or Oral-Gingival Pemphigoid is a rare, chronic autoimmune disorder which is characterised by subepithelial blisters involving mucous membranes or occasionally the skin [1,2]. The disease may involve mucous membrane of conjunctiva, oral cavity, larynx, nasopharynx, oesophagus, genitourinary tract, and anus. Most common site of mucous membrane pemphigoid is oral mucosa followed by the conjunctiva [3]. Oral mucosal lesions may present in milder form as chronic erythematous lesions to severe erosions due to bullae rupture covered by fibrinous slough [4]. As ocular involvement is also common in these patients, ophthalmic examination must be conducted in presence of oral lesions [4].

The exact incidence of MMP is unknown, however, in a study in UK, the estimated incidence of MMP was 0.8 per million population [5]. Females are predominantly affected as compared to males and the disease mainly occurs in the elderly population belonging to age range of 60 to 80 years [6,7].

MMP is characterised by separation of epithelium from the connective tissue due to genetic defect or alterations in anchoring proteins. Diagnosis of MMP can be made by clinical features and diagnostic histopathological characteristic. Histology of MMP reveal presence of subepithelial blisters and direct immunofluorescence has typical feature of binding of IgG and C3 to the basement membrane zone (BMZ) [8].

As the disease is very uncommon, it is important to identify the spectrum of clinical manifestations in MMP. The present study was thus conducted to study the different manifestations of MMP among patients diagnosed as MMP at our institute.

Methodology

The study was conducted as a cross sectional study at PMCH, Patna for a period of 3 years from 1st December 2017 to 30th November2020. All the 30 patients diagnosed as a case of MMP in the departments of Otorhinolaryngology, dermatology and ophthalmology in PMCH, Patna during the study period and giving consent were included. Diagnosed cases not willing to participate in the study were excluded. After obtaining clearance form Institute's ethical committee, written consent was obtained from all the patients willing to participate. Data regarding sociodemographic variables such as age gender, socioeconomic status was obtained. Detailed history regarding clinical features, mode of presentation, and duration of symptoms was obtained and entered in questionnaire. History regarding autoimmune disease and family history of MMP or other autoimmune disease if any was also documented. All the patients were subjected to mucosal biopsy for histopathological examination. Also, all the biopsy specimen were subjected to direct & indirect immunofluorescence examination.

Statistical analysis

Data was compiled using Ms Excel and analysed using IBM SPSS software version 20. Categorical data was expressed as frequency and percentage whereas numerical data was expressed as mean and standard deviation.

Results

The present study included a total of 30 patients with MMP with mean age of 62.3 ± 22.2 years.

Sociodemographic variables		Frequency (n = 30)	Percentage
Age (years)	21 - 40	2	6.7
	41 - 60	3	10
	61 - 80	25	83.3
Gender	Male	8	26.7
	Female	22	73.3

Table 1: Distribution of patients according to sociodemographic variables.

Majority of patients with MMP belonged to 61 to 80 years of age (83.3%). Male predominance was observed for cases of MMP with male: female ratio of 1:2.75.

Site of involvement	Frequency (n = 30)	Percentage
Oral mucosa	29	96.7
Ocular conjunctiva	19	63.3
Larynx	14	46.7
Nasal Mucosa	12	40
Skin	12	40
Pharynx	7	23.3
Oesophageal	3	10

Table 2: Distribution of patients according to site of involvement.

Out of 30 patients, the most common site of involvement was oral mucosa (96.7%), followed by ocular conjunctiva in 63.3% and laryngeal mucosa (46.7%). Pharyngeal mucosa and oesophagus were the least commonly involved site in 23.3% and 10% cases respectively.

Out of 29 cases with oral mucosal lesions, 100% cases presented with complaint of pain and inability to eat food. Among them, 55.8% patients presented with oral erosions/ulceration and remaining 44.2% patients had Desquamative gingivitis. Pseudo membrane formation was noted in 3 (10.3%) cases whereas vesicles formation and mild scarring with atrophy was seen in 1 (3.4%) cases.

Similarly, among 19 cases with ocular MMP, 47.4% cases presented with non-specific conjunctivitis associated with photophobia and photosensitivity. Other manifestations were corneal erosion and damage, symblepharon formation, ankyloblepharon, trichiasis and entropion in 10.5% cases each. However, none of the patients with ocular pemphigoid were diagnosed to be suffering from blindness.

Out of 14 cases with laryngeal involvement, mode of presentation was Sore throat and hoarseness in 100% cases. On examination, scarring with stenosis of airway was found only in 28.6% cases which was associated with breathing difficulty too.

Patients with nasal mucosal involvement (n = 12) came with complaints of discharge (33.3%) and excessive crust (8.3%). About 51.3% cases had nasal mucosal erosions.

Among Skin lesions (n=12), 66.7% presented with localized erythematous plaque near affected mucosal surfaces followed by generalized bullous eruption on the head and upper body in 33.3%. No cases were found of Scarring or cicatricial alopecia.

All the cases with pharyngeal and oesophageal MMP had dysphagia and odynophagia respectively. However stricture formation was observed in none of the cases.

Discussions

Mucous membrane pemphigoid is a rare disorder which may involve multiple system including oral mucosa, conjunctiva, nasopharynx, larynx and occasionally skin. In present study, a total of 30 cases with MMP with mean age of 62.3 ± 22.2 years were enrolled over the period of 3 years. Majority of cases with mucous membrane pemphigoid belonged to age range of 61 to 80 years (> 80%) and about 73.3% cases were females. These findings were supported by findings of Dharman., *et al*. in which the authors concluded that the disease has predilection for female and is reported after the fifth decade of life [9]. Chiou., *et al*. documented that mucous membrane pemphigoid occurs primarily in older women. About 68% cases were females and more than 80 percent patients were over 50 years at onset [10].

In present study, oral mucosa was the most common site involved in 29 (96.7%) cases. All these cases presented with difficulty in eating food. Predominant manifestation among these patients was oral erosions/ulceration and desquamative gingivitis. Our study findings were concordant with the findings of Chiou., *et al*. in which oral mucosal lesions were documented in > 90% cases and about 63% cases exhibited erosive or ulcerative lesions [10]. Dharman., *et al*. also reported desquamative gingivitis to be the main clinical sign in MMP which is characterised by erythematous gingiva and loss of stippling that extend apically from gingival margins to alveolar mucosae [9]. Silverman *et al*. [11] and Agbo-Godeau., *et al* [12]. Also reported a higher prevalence of gingival involvement in cases with MMP.

Another common site of involvement was conjunctiva in our study observed in 19 cases. Among them, 47.4% cases presented with non-specific conjunctivitis with photophobia and photosensitivity. Higgins., *et al*. assessed the presence of ocular lesions in patients with oral mucus membrane pemphigoid. They observed ocular lesions in 30% cases at the time of presentation and about 37% cases developed ocular manifestations over the course of time [13]. Georgoudis., *et al*. documented that MMP is the leading cause of cicatricial conjunctivitis which can affect both young and adult patients. Though disease commonly affect older patients, but the disease tend to be severe in case of younger patients [14]. Previous studies have reported cicatrizing conjunctivitis as the most common mode of presentation. However, our study observed that patients with MMP may also present with corneal erosion and damage, symblepharon formation, ankyloblepharon, trichiasis and entropion apart from characteristic conjunctivitis.

Patients with laryngeal involvement predominantly presented with sore throat and hoarseness whereas patients with nasal involvement presented with erosions, discharge and crust formation. MMP lesions involving skin appeared as localized erythematous plaque near affected mucosal surfaces in majority of cases and few cases presented as generalized bullous eruption on the head and upper body. However, patients with pharyngeal and oesophageal MMP presented with dysphagia and odynophagia respectively. Literature regarding clinical manifestations of MMP other than ocular and oral manifestations is scarce. However, Madgar., *et al*. documented that the patients with oral MMP may present with certain complications such as blindness in case of ocular involvement and airway obstruction in case of laryngeal involvement [15]. These findings were supported by previous studies in which the mode of presentation varied depending upon site of involvement. The patients with MMP may present with burning mouth; ocular dry-

ness and irritation; nasal obstruction; rhinorrhoea; odynophagia; dysuria; sexual dysfunction; dysphagia; hoarseness; and breathing difficulty. However, active lesions may present with erosions, ulcerations, vesicles, blisters, crusts and pseudomembranes [16].

Conclusion

Mucous membrane pemphigoid is a rare disorder which most commonly involve oral mucosa. However, ocular, nasopharynx, larynx, oesophagus involvement can also be observed. Depending upon site of involvement, patients may present with varied manifestations such as oral erosions and ulcers, desquamative gingivitis in case of oral lesions, cicatrizing conjunctivitis in case of ocular involvement. The involvement may also range from mild erosion to airway obstruction and dysphagia in severe cases depending upon site of involvement.

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