

***Pemphigus vulgaris* versus Mucous Membrane Pemphigoid: Case Series**

Pénfigo Vulgar versus Penfigoide de la Membrana Mucosa: Series de Casos

**Natalia Garcia Santaella¹; Brena Rodrigues Manzano¹;
Eduardo Stedile Fiamoncini¹ & Paulo Sérgio da Silva Santos¹**

GARCIA SANTAELLA, N.; RODRIGUES MANZANO, B.; STEDILE FIAMONCINI, E.; DA SILVA SANTOS, P.S. *Pemphigus vulgaris* versus mucous membrane. Pemphigoid: Case series. *Int. J. Odontostomat.*, 18(2):248-254, 2024.

ABSTRACT: *Pemphigus vulgaris* (PV) and mucous membrane pemphigoid (MMP) are bullous autoimmune diseases that reach the oral mucosa and have common clinical features. The objective of the study was to present and compare the clinical manifestations of PV and MMP and the results of applied treatments. A case series of a stomatology service from 1985 to 2018. Data collection included epidemiological data, comorbidities, medications in use, duration of symptoms before the first visit, previous treatment, symptomatology, clinical description of lesions, presumptive diagnosis, histopathological description, extraoral manifestations, final diagnosis, treatment and follow-up. The medical records of 25 patients were analysed, 19 of whom were diagnosed with MMP and 6 with PV. The female gender was prevalent in MMP (84 %) and the male gender in PV (67 %). More than 60 % of patients complained of pain at their first visit. Patients with MMP took on average 6 months to seek professional help and patients with PV, about 2 months. Desquamative gingivitis was the most common lesion (63 %) in MMP and non-gingival ulcers (67 %) in PV. Minimal therapy was effective in all cases of MMP, and in PV one individual required minimal adjuvant therapy due to worsening of the case. Patients with PV have more intense signs and oral symptoms and may need more intensive treatment than patients with MMP. The use of topical and/or systemic corticosteroids was sufficient for most cases in both diseases.

KEY WORDS: pemphigus vulgaris, mucous membrane pemphigoid, oral manifestations, treatment outcome.

INTRODUCTION

The terms *Pemphigus vulgaris* (PV) and mucous membrane pemphigoid (MMP) refer to autoimmune bullous dermatoses that affect the buccal mucosa and have distinct pathophysiological mechanisms. Their primary cutaneous manifestation consists of the formation of vesicles and blisters (Hammers & Stanley, 2017). They are differentiated microscopically according to the location of the blister on histopathological examination: in MMP separation between the surface epithelium and the underlying connective tissue of the basement membrane is observed, whereas in PV separation of the epithelium above the basal layer, which remains attached to the connective tissue, is seen (Schmidt & Zillikens, 2010).

PV often affects the mouth at an early stage, presenting isolated blisters, erosions or a combination of the two, leading rapidly to erosions and chronic

ulcers, seen mainly in the labial and jugal mucosa, palate and belly of the tongue (Ramos *et al.*, 2012; Sankar & Noujeim, 2017). Approximately 50 % of cases present exclusively oral clinical manifestation (Laskaris *et al.*, 1982; Mignogna *et al.*, 2001; Santoro *et al.*, 2003; Sultan *et al.*, 2017). Gingival lesions in *Pemphigus vulgaris* are less common (Scully & Mignogna, 2008; Said & Golitz, 2011). The aim of treatment is to minimize the burden of the disease and improve the patients' quality of life, which consists of eliminating lesions and, consequently, pain (Sultan *et al.*, 2017). Treatment includes topical or systemic corticosteroids, immunosuppressants and other biological therapies (McMillan, Taylor *et al.*, 2015).

MMP presents mainly with lesions of gingival affection in the form of desquamative gingivitis, presenting irregular, diffuse erythema and erosion of the

¹ Department of Surgery, Stomatology, Patology and Radiology, Bauru School of Dentistry, University of São Paulo, São Paulo, Brazil.

inserted gingiva (Scully & Lo Muzio, 2008; Mustafa *et al.*, 2015). When it starts by affecting the oral cavity exclusively, it presents a more benign evolution and may not manifest extra-oral lesions (Mobini *et al.*, 1998). However, most MMP patients have more than one site of manifestation (Chan *et al.*, 2002; Feller, Ballyram *et al.*, 2017), and the therapy consists mainly of topical and systemic corticosteroids, alone or in combination (Di Zenzo *et al.*, 2014; Taylor *et al.*, 2015; Feller *et al.*, 2017).

The clinical characteristics of PV and MMP have been described extensively, with several studies evaluating each disease separately (Feller *et al.*, 2017). However, the literature is scarce in studies comparing the clinical characteristics presented by patients affected by these 2 diseases in the same population (Sultan *et al.*, 2017), especially since they present very similar clinical characteristics. Moreover, there is not enough evidence in the literature to determine the efficacy of the available treatments. Therefore, the aim this study is to describe and compare the clinical manifestations of PV and MMP, as well as the results of the treatments used.

MATERIAL AND METHOD

This was a case series of individuals with PV and MMP diagnosed and treated in a graduate and post-graduate clinic in stomatology between the years of 1985 and 2018. The study was approved by the Committee of Ethics in Research (CAAE: 90561118.9.0000.5417).

The study was performed on an electronic medical record system, using the terms "pemphigus", "pemphigoid", "pemphigus vulgaris" and "benign pemphigoid", "benign mucosal pemphigoid" and "mucous membrane pemphigoid". At first, 131 medical records were found and distributed among 3 raters for data collection. The individuals included in the study were those with a confirmed diagnosis of PV or MMP, with a histopathological report.

Data were collected on age and gender, comorbidities, medications in use, duration of symptoms before the first visit, previous treatment or intervention, symptomatology, clinical description (desquamative gingivitis, non-gingival ulcers, erythema, erosion and blisters), oral region affected, diagnostic hypothesis, histopathological description of the lesions, presence of extraoral manifestations, previous treatment, number of consults, final diagnosis, treatment performed, follow-up and therapeutic response of the case.

The data collected were tabulated and presented in a descriptive way using graphs and tables, and descriptive statistics were used.

RESULTS

A total of 25 individuals were included in this study, 19 with MMP and 6 with PV. The majority of individuals with MMP were females (84 %), and the mean age was 56.94 years, whereas the group of PV had more males (67 %) and a mean age of 49.5 years. With respect to the comorbidities present, gastritis was the most frequent in both groups, present in 5 (26 %) individuals with MMP and 2 (33 %) with PV, and the other comorbidities found are described in Table I.

More than one comorbidity could be present in the same individual. The MMP group used more medication (21 drugs) than the PV group (18 drugs), and while the MMP group used more hormone therapy, individuals with PV used more antidepressants. In addition, several other drugs were used, and the same individual could have used more than one drug (Table I).

Over 60 % of subjects in both groups had pain complaints at the first visit (Table I), which presented a median duration of 6 months (range 1 to 180 months) in the MMP group and 2 months (range of 15 days to 3 months) in individuals with PV. We found that 47 % of individuals with MMP and 67 % of individuals with PV did not receive treatment prior to the first visit. Regarding the cases who had received previous treatment at the first consultation (3 cases with MMP and 1 with PV), 1/3 (33 %) of individuals from the MMP group had used a topic corticosteroid, and the only (33 %) individual with PV who had received prior treatment at the first visit used an antibiotic associated with an NSAID (non-steroidal anti-inflammatory) (Table I). The most common diagnostic hypotheses in the first evaluation in both groups were MMP, PV and Lichen planus (Table II).

Among individuals with PV, 3/3 (50 %) presented more than one type of lesion while 6/19 (32 %) individuals with PMM presented this characteristic. The desquamative gingivitis (DG) was the most common type of lesion found in individuals with MMP, present in 12 (67 %) individuals with MMP and only 1 (17 %) case of PV, and in 4/12 (33 %) individuals with MMP with DG, this was the only lesion present. In all cases of PV and 11/19 (58 %) with MMP, more than one region was affected, and individuals with MMP presented more lesions in the gingiva (79 %), whereas

Table I. Description of each case of Mucous Membrane Pemphigoid and Pemphigus vulgaris.

Diagnostic Patient	Gender /Age	Comorbidities	Medications	Symptomatology time	Previous treatment or intervention	Type of lesion and Regions affected	Extraoral manifestations	Diagnostic hypothesis	All treatments	Number of consults	Follow-up	Therapeutic response
1	F/45	Allergic (sun, dust, shrimp) Gastritis	Bromazepam®	+ 180 months	No related	Desquamative gingivitis, Left tonsil dunks, mouth floor, and alveolar mucosa * Throat blister history	No	MMP	Referral to dermatologist	1	-	-
2	F/76	Gastritis	No	3 months	No	Blister, the anterior region of the mandible	No	MMP	Referral to dermatologist	1	-	-
3	F/76	Gastritis Labyrinthitis	No	3 months	No related	Blister - gum and anterior region of the mandible	No	PV and MMP	Referral to dermatologist	1	-	-
4	F/45	Gastritis Hypothyroidism	Sodic levotyroxin	No related	No related	Desquamative gingivitis	No	MMP	Prednisone 5 mg and mouthwash with sodium bicarbonate for 19 days	3	2,5 months	Absence of signs and symptoms on minimal therapy
5	F/51	Labyrinthitis Hypothyroidism	Monoteam®, Tetrod®, Vimpoostina®	180 months	Yes, bippy	Desquamative gingivitis Erythema - alveolar mucosa Erosion - gum	No	MMP	Decadron® elixir and Droxaine® bicarbonate	8	84 months	Absence of symptoms on minimal therapy
6	F/8	No	No	No related	No related	Erosion - mucosa alveolar e gum	No	Herpetic stomatitis	Decadron® elixir and sodium bicarbonate	7	8 months	Partial remission off therapy
7	M/68	No	No	24 months	Yes, omelcolon-A orabase	Desquamative gingivitis Ulcer - buccal mucosa, alveolar mucosa, throat, tonsillar region Erythema - gum, and throat Blister - precorde ulcers	Pustules scalp	MMP and lichen planus	Droxaine®, milk of magnesia (paste), Hucomeidine®, and prednisone 5 mg	8	1 month	Partial remission on minimal therapy
8	F/59	Type II diabetes Hypothyroidism	Synthroid®, Repogen®	6 months	No related	Desquamative gingivitis Gum- erosion and Blister	No	Atrophic lichen planus	Cobetasol propionate, milk of magnesia, Phillips dentifrice®, Mucostatin®, sodium bicarbonate, Carax®, and prednisone	8	97 months	Partial remission on minimal therapy
9	F/55	No	No	12 months	No related	Desquamative gingivitis Gum- Erosion and Blister - precorde ulcers	No	MMP and PV	Omcilon orabase®	3	8 months	Absence symptoms on minimal therapy
10	F/66	Osteoporosis Cysticercosis	Alexofedim®, Omeprazole®, Gardinal®, Thalidomide®, Raloxifene®	No related	No related	Desquamative gingivitis Ulcer - lower alveolar rebordo, buccal mucosa, anterior vestibule mandible and floor mouth Blister - precorde ulcer	No	MMP and PV	Periogaro®, propionate cobetasol, and Carax®	13	66 months	Permanency of lesions on minimal therapy
11	M/82	Hypertension	Venac®, Mocarelic®, Actonil®	1 month	No	Ulcer - Buccal mucosa	Skin peeling and erythema	MMP, PV and lichen planus	0.05% cobetasol propionate	8	21,5 months	Absence signs and symptoms on minimal therapy
12	F/74	Diabetes Gastritis	Omeprazole®, Glyphage®, Diamicron®	4 months	No	Desquamative gingivitis	No	MMP and lichen planus	Dexamethasone elixir®	2	4 months	Absence signs and symptoms on minimal therapy
13	F/83	No	No	1 months	No	Ulcer - Buccal mucosa and alveolar mucosa	No	Candidiasis, MMP and lichen planus	No related	1	No related	No related
14	F/52	Contrast allergy	No	8 months	Yes, surgical removal	Desquamative gingivitis Ulcer -alveolar mucosa	No	MMP and PV	Endovenous corticoid	2	1,5 months	Partial remission on minimal therapy
15	M/51	Gastritis	Levoid®	60 months	No	Desquamative gingivitis Blister - anterior and posterior inserted gingiva	No	MMP and PV	Desamethasone elixir®	8	26 months	Absence signs and symptoms on minimal therapy
16	F/18	Allergic (turkey balm, cobalt chloride, lanolin, and thimerosal)	B complex	12 months	No	Desquamative gingivitis Erythema - gingiva	Peeling on the skin of the face	PV and hypersensitiv	Cobetasol propionate	3	7 months	Absence signs and symptoms of therapy
17	F/72	No	Losartan®	48 months	No	Erythema - Buccal mucosa, hard palate, alveolar mucosa, and gingivae	No	PV	Cobetasol propionate	7	12 months	Relapse
18	F/56	No	Iodine	3 months	No	Ulcer - Buccal mucosa, labial mucosa, and tongue side	No	PV and lichen planus	Cobetasol propionate, 1 week after systemic corticoids introduced later. 2 months cobetasol propionate; 2 months systemic corticoids (5 mg prednisone)	2	2 month	Permanence of lesions
19	F/45	No	Alexofedim®	2 month	Yes, mystalin 7 days	Desquamative gingivitis Erythema - Alveolar mucosa and upper labial mucosa	No	PV and lichen planus	Pasalix®, cobetasol propionate (after 3 months)	3	3 months	Permanence of lesions

F: female; M: male; MMP: mucous membrane pemphigoid; PV: Pemphigus vulgaris

in the individuals with PV, the jugal mucosa (83 %) was the most affected region. Information on the type and prevalence of lesions and regions affected is shown in Table II.

The prevalence of histopathological features found in common with mucous membrane pemphigoid (MMP) was: subepithelial cleft 53 % and chronic infiltration 47 %).

The prevalence of histopathological features found in common with *Pemphigus vulgaris* was: intraepithelial cleft 36 %, chronic infiltration 36 % and acantholysis 28 %.

At the initial consultation, topical therapy was prescribed exclusively for most individuals (12 with MMP and 4 with PV) and only 1 (5 %), who was in the MMP group, required an associated systemic corticosteroid (Table III). Regarding systemic therapy, 3 (16 %) cases of MMP and 2 (33 %) of PV had systemic corticosteroids at some point during follow-up, and in 1 (1/2) of these PV cases (Patient 23-Table I), Azathioprine was administered concomitantly after worsening of the general condition.

Diagnostic	Patient	Gender/ Age	Comorbidities	Medications	Symptomatology time	Previous treatment or intervention	Type of lesion and Regions affected	Extraoral manifestations	Diagnostic hypothesis	All treatments	Number of consults	Follow- up	Therapeutic response
Pemphigus Vulgaris	20	F/44	Gastritis Depression	Paracetamol®, Sertraline®, Vitamin B12®, Risperidone®, Oxcarbazepine®, Bromazepam®, Escitalopram®	No related	No	Erosion - Buccal mucosa, gum, alveolar mucosa, hard and soft palate, and tonsillar pillar Erythema - soft palate Dysphagia - throat, alveolar mucosa Ulcers - Tongue soft palate, right tonsillar pillar, upper lip brake, and lingual brake Erythema - Soft palate and throat Erosion - Buccal mucosa	Scalp	PV	Made by Dermatologist Referral to dermatologist	3	2 months	Absence of signs and symptoms
	21	M/57	Smoking alcoholism	No	3 months	-		No	Ulcerative stomatitis and erythema multiforme	Hexomedine®, clobetasol propionate, prednisone	1	-	Exacerbation- Appearance of new larger lesions under minimal therapy
	22	M/49	Gastritis	Acetylsalicylic acid Cloxacillin®	15 days	No		No	MMP and PV	Hexomedine®, Lactimel®, prednisone, Azathioprine®, and calcium carbonate + vitamin D3	6	8 months	Partial remission
	23	F/48	No	Amoxicillin Diclofenac®	2 months	Yes, Amoxicillin Diclofenac		Edema on the face, the abdomen on the crusts on the skin, inability to eat		Hexamedine®, Retigard®, Clobetasol propionate base®, Lactimel®, prednisone, Azathioprine®, and calcium carbonate + vitamin D3	5	15 months	Partial remission
	24	F/40	Allergic Sulfonamide and iodine	Melantapus®	2 months	No		No	PV erosive lichen planus	Dexamethasone elixir®	2	1 month	Partial remission on minimal therapy
25	M/59	No	Nimesulid®	No related	No		No	MMP and lichen planus	Clobetasol propionate	4	8 months	Absence of signs and symptoms on therapy	

Table II. Prevalence of the type and oral regions with lesions in the initial consultation.

Type of lesion	MMP (n=19) N (%)
Erosion and / or gum ulcer (desquamative gingivitis)	12 (63 %)
Non-gingival ulcers	7 (37 %)
Bulla	7 (37 %)
Erythema	4 (21 %)
Erosion	1 (5,2 %)
Oral regions affected	
Gum	15 (79 %)
Alveolar mucosa	8 (42 %)
Mucosa jugal	6 (32 %)
Throat	2 (11 %)
Floor of the mouth	2 (11 %)
Tonsillar region	2 (11 %)
Mucous labial	2 (11 %)
Palate	1 (5 %)
Labial fold	1 (5 %)
Alveolar ridge	1 (5 %)
Anterior mandible	1 (5 %)
Tongue	1 (5 %)
Labial frenum	0 (0 %)
Retromolar	0 (0 %)

MMP: Mucous membrane pemphigoid; PV: *Pemphigus vulgaris*.

Table III. Treatments introduced in the initial consultation.

	MMP (n=19) N (%)	PV (n=6) N (%)
Topic	12 (63 %)	4 (67 %)
+ Systemic topic	1 (5 %)	0 (0 %)
Referred to Dermatologist	3 (16 %)	2 (33 %)
Not reported	2 (11 %)	0 (0 %)
None	1 (5 %)	0 (0 %)
Prescribed Medications		
Topic		
Dexamatomasone elixir	5 (26 %)	1 (17 %)
Clobetasol Propionate	7 (37 %)	1 (17 %)
Sodium bicarbonate	3 (16 %)	0 (0 %)
Droxaine ¹	2 (11 %)	0 (0 %)
Toothpaste without Sodium Lauryl Sulfate	2 (11 %)	0 (0 %)
Milk of magnesia	2 (11 %)	1 (17 %)
Chlorhexidine 0.12%	1 (5 %)	1 (17 %)
Triamcinolone acetoneide	1 (5 %)	1 (17 %)
Hexomedine ²	1 (5 %)	2 (33 %)
Antifungal	1 (5 %)	0 (0 %)
Systemic		
Prednisone	1 (5 %)	0 (0 %)

¹Aluminum hydroxide, Magnesium hydroxide, Oxetacaine ²Hexamidine isethionate, tetracaine hydrochloride

The first follow-up visit occurred on average 1.4 months after the initial assessment in the MMP groups and 1.1 months later in the PV group. In this consultation, lesions were controlled in 10/19 (53 %) individuals with MMP; of these cases, in 4/10 (40 %) the lesion completely regressed. In 8/10 (80 %) cases of MMP with controlled lesions, they were treated with exclusive topical therapy, 1/10 (10 %) was treated with associated topical and systemic therapy, and 1/10 (10 %) reported intravenous corticosteroid use due to upper airway infection.

In the individuals with PV, the lesions were controlled at the first follow-up visit, in 3 (50 %) individuals, and there was a total regression of the lesions in 1/3 (33 %) of which there was no description of the type of medical treatment and the other subjects (2/3) were treated with exclusive topical corticosteroids.

The lesions were not controlled at the first follow-up visit in 5/19 (26 %) individuals with MMP, and 3/5 (60 %) of these cases were using topical corticosteroids associated with other topical therapies, 1/5 (20 %) was receiving another type of topical therapy without corticosteroid and 1/5 (20 %) individuals was not receiving any therapy. The only individual with PV in whom the lesions were exacerbated was using topical corticosteroid therapy with an anaesthetic.

Regarding clinical follow-up, the group of individuals with MMP underwent more consultations (mean of 4.6 queries in MMP and 3.5 queries in PV) and were followed up for longer than individuals in the PV group (mean of 22.9 months and 6.8 months, respectively).

The therapeutic response of the diseases was performed in 17 individuals with MMP and in 5 individuals with PV. Thus, most individuals in both groups had absence of signs and oral symptoms (41 % and 40 %) and partial remission of the oral lesions (24 % and 40 %), respectively.

DISCUSSION

In the present study, a descriptive analysis revealed that the majority (84 %) of cases of MMP occurred in women (Taylor *et al.*, 2015; Broussard *et al.*, 2016; Bagan *et al.*, 2018), whereas most cases of PV were in men (67 %), contrary to what we see in the literature (Laskaris *et al.*, 1982; Svecova, 2015; Bai *et al.*, 2016; Sultan *et al.*, 2017), a fact that can be attributed to the small number of patients included in

this study. On the other hand, a very positive fact was that a large number of individuals had signs and symptoms regression only with minimal therapy, both in PV and MMP.

The presence of symptomatology at the initial consultation was slightly higher in individuals with PV (67 % in PV and 63 % in MMP) and, although this difference did not seem relevant, we observed that patients with a PV diagnosis had sought the dental surgeon previously (2 months and 6 months prior, respectively), suggesting that, although both diseases present symptoms, those of PV are more intense, leading the individual to seek professional help in a shorter period of time after the onset of symptoms (Sultan *et al.*, 2017).

Desquamative gingivitis is a common clinical manifestation in individuals with MMP and less common in individuals with PV (Patel *et al.*, 2016; Sultan *et al.*, 2017; Bagan *et al.*, 2018; Maderal *et al.*, 2018), being present in 63 % and 17 % of patients diagnosed with MMP and PV in this study, respectively. The prevalence of desquamative gingivitis between MMP and PV is diverse, but it is always more prevalent in individuals with MMP (Laskaris *et al.*, 1982; Lo Russo *et al.*, 2009; Sultan *et al.*, 2017).

Other intraoral manifestations include non-gingival ulcers, blisters, erythema and erosion, which vary in location and can affect both diseases (Broussard *et al.*, 2016). In the present study, non-gingival ulcers, erythema and erosion were more prevalent in individuals with PV, and blisters affected more individuals with MMP, which is to be expected because, in this disease, the clefts are subepithelial (Petruzzi, 2012; Srikumaran & Akpek, 2012; Arduino *et al.*, 2017) presenting tissue; therefore, they remain longer in the mouth and can be visualized during a physical examination.

The extraoral manifestations described were more prevalent in individuals with PV (33 %) than MMP (16 %), according to a previous study (Laskaris *et al.*, 1982; Sultan *et al.*, 2017). In PV, the site most affected by extraoral manifestations is the skin; less commonly, there is ocular involvement. In MMP, ocular involvement is more common, followed by cutaneous involvement, but it can affect any other mucosa (Broussard *et al.*, 2016). Although ocular lesions are common in MMP, they were not present in this study (Sultan *et al.*, 2017), possibly because it was a retrospective cohort study, and the presence of extra-oral manifestations may not have been included in the medical records.

There is a wide variety of options for the treatment of PV and MMP, which include the use of topical and systemic corticosteroids, alone or in combination, biological therapies and immunosuppressants (Chan *et al.*, 2002; McMillan *et al.*, 2015; Taylor *et al.*, 2015; Broussard *et al.* 2016). Treatment for MMP is generally effective only with minimal therapy (Murrel Marinovic *et al.*, 2015; Sultan *et al.*, 2017); such was the case in this study, and in those with PV, only one individual required adjuvant minimal therapy due to worsening of the clinical condition (Murrel *et al.*, 2008; Sultan *et al.*, 2017).

Also, the cases that most presented good therapeutic response, presented absence of the signs and symptoms, and partial remission of oral lesions, principally on minimal therapy. The minimal therapy, in turn, when used in patients with MMP, included: corticosteroids, antibiotics from the tetracycline group, conchicine, sulazopyrine, sulfapyridine, sulfamethoxypridazin and nicotinamide (Murrel *et al.*, 2008), while in patients with PV, it included only the use of corticosteroid prednisone or equivalent, in low doses (Murrel *et al.*, 2008).

Regarding the mean follow-up time, individuals with MMP in this study were followed up more often by the dental surgeon than those with PV (22.9 months and 6.8 months, respectively). Knowing that more PV patients had cutaneous manifestations and were referred to the dermatologist, the authors of this study believe that said individuals, when referred to the dermatologist, remained with this professional for treatment and did not return for dental consultations, resulting in a smaller number being followed up by dental professionals. This fact reinforces the importance of the close relationship between the dentist and dermatologist, as communication between these professionals will reduce the non-continuity of dental treatment in these patients.

In conclusion, the patients with PV have more intense signs and symptoms and may require more intensive treatment than patients with MMP, despite the fact that most patients, both with PV and MMP, show complete remission of signs and symptoms with only minimal therapy.

FUNDING: This study was financed by the Coordination for the improvement of Higher Education Personnel – Brasil (CAPES) - Finance Code 001.

GARCIA SANTAELLA, N.; RODRIGUES MANZANO, B.; STEDILE FIAMONCINI, E.; DA SILVA SANTOS, P. S. Pénfigo vulgar *versus* penfigoide de la membrana mucosa: Serie de casos. *Int. J. Odontostomat., 18(2):248-254, 2024.*

RESUMEN: Pénfigo vulgar (PV) y Penfigoide de la Membrana Mucosa (PMM) son enfermedades autoinmunes ampollas que llegan a la mucosa oral y tienen características clínicas comunes. El objetivo de este estudio fue presentar y comparar las manifestaciones clínicas de PV y PMM y los resultados de los tratamientos aplicados. En el análisis se incluyó una serie de casos de un servicio de estomatología de 1985 a 2018. La recolección de información incluyó datos epidemiológicos, comorbilidades, medicamentos en uso, duración de los síntomas antes de la primera visita, tratamientos previos, sintomatología, descripción clínica de las lesiones, diagnóstico presuntivo, descripción histopatológica, manifestaciones extraorales, diagnóstico final, tratamiento y seguimiento. Se analizaron las historias clínicas de 25 pacientes, 19 de los cuales fueron diagnosticados de PMM y 6 de PV. El sexo femenino fue prevalente en PMM (84 %) y el sexo masculino en PV (67 %). Más del 60 % de los pacientes se quejaron de dolor durante la primera consulta. Los pacientes con PMM tardaron en promedio 6 meses en buscar ayuda profesional y los pacientes con PV, alrededor de 2 meses. La gingivitis descamativa fue la lesión más común (63 %) en PMM y las úlceras no gengivales (67 %) en PV. La terapia mínima fue efectiva en todos los casos de PMM, y en PV un individuo requirió terapia adyuvante mínima debido al empeoramiento del caso. Los pacientes con PV tienen signos y síntomas orales más intensos y pueden necesitar un tratamiento más intensivo que los pacientes con PMM. El uso de corticosteroides tópicos y/o sistémicos fue suficiente para la mayoría de los casos en ambas enfermedades.

PALABRAS CLAVE: pénfigo vulgar, penfigoide de la membrana mucosa, manifestaciones orales, resultado del tratamiento.

REFERENCES

- Arduino, P.; Broccoletti, R.; Carbone, M.; Conrotto, D.; Pettigiani, E.; Giacometti, S.; Gambino, A.; Elia, A. & Carrozzo M. Describing the gingival involvement in a sample of 182 Italian predominantly oral mucous membrane pemphigoid patients: A retrospective series. *Med. Oral Patol. Oral Cir. Bucal*, 22(2):e149-e152, 2017.
- Bagan, J.; Jiménez, Y.; Murillo, J. & Bagan, L. Oral mucous membrane pemphigoid: a clinical study of 100 low-risk cases. *Oral Dis.*, 24(1-2):132-4, 2018.
- Bai, Y. X.; Zhang, L. M.; Xiao, T. & Chen, H. D. A 6-year treatment experience for pemphigus: retrospective study of 69 Chinese patients. *Dermatol. Ther.*, 29(2):84-7, 2016.
- Broussard, K. C.; Leung, T. G.; Moradi, A.; Thorne, J. E. & Fine, J. D. Autoimmune bullous diseases with skin and eye involvement: Cicatricial pemphigoid, pemphigus vulgaris, and pemphigus paraneoplastica. *Clin. Dermatol.*, 34(2):205-13, 2016.
- Chan, L. S.; Ahmed, A. R.; Anhalt, G. J.; Bernauer, W.; Cooper, K. D.; Elder, M. J.; Fine, J. D.; Foster, C. S.; Ghohestani, R.; Hashimoto, T.; *et al.* The first international consensus on mucous

- membrane pemphigoid: definition, diagnostic criteria, pathogenic factors, medical treatment, and prognostic indicators. *Arch. Dermatol.*, 138(3):370-9, 2002.
- Di Zenzo, G.; Carrozzo, M. & Chan, L. S. Urban legend series: mucous membrane pemphigoid. *Oral Dis.*, 20(1):35-54, 2014.
- Feller, L.; Ballyram, R.; Khammissa, R. A.; Altini, M. & Lemmer, J. Immunopathogenic oral diseases: an overview focusing on *Pemphigus vulgaris* and mucous membrane pemphigoid. *Oral Health Prev. Dent.*, 15(2):177-82, 2017.
- Hammers, C. M. & Stanley, J. R. Mechanisms of disease: pemphigus and bullous pemphigoid. *Annu. Rev. Pathol.*, 11:175-97, 2017.
- Laskaris, G.; Sklavounou, A. & Stratigos, J. Bullous pemphigoid, cicatricial pemphigoid, and pemphigus vulgaris. A comparative clinical survey of 278 cases. *Oral Surg. Oral Med. Oral Pathol.*, 54(6):656-62, 1982.
- Lo Russo, L.; Fierro, G.; Guiglia, R.; Compilato, D.; Testa, N.; Lo Muzio, L. & Campisi, G. Epidemiology of desquamative gingivitis: evaluation of 125 patients and review of the literature. *Int. J. Dermatol.*, 48(10):1049-52, 2009.
- Maderal, A. D.; Salisbury 3rd, P. L. & Jorizzo, J. L. Desquamative gingivitis: Clinical findings and diseases. *J. Am. Acad. Dermatol.*, 78(5):839-48, 2018.
- McMillan, R.; Taylor, J.; Shephard, M.; Ahmed, R.; Carrozzo, M.; Setterfield, J.; Grando, S.; Mignogna, M.; Kuten-Shorrer, M.; Musbah, T.; *et al.* World Workshop on Oral Medicine VI: a systematic review of the treatment of mucocutaneous pemphigus vulgaris. *Oral Surg. Oral Med. Oral Pathol. Oral Radiol.*, 120(2):132-42.e61, 2015.
- Mignogna, M. D.; Lo Muzio, L. & Bucci, E. Clinical features of gingival pemphigus vulgaris. *J. Clin. Periodontol.*, 28(5):489-93, 2001.
- Mobini, N.; Nagarwalla, N. & Ahmed, A. R. Oral pemphigoid. Subset of cicatricial pemphigoid? *Oral Surg. Oral Med. Oral Pathol. Oral Radiol. Endod.*, 85(1):37-43, 1998.
- Murrel, D. F.; Dick, S.; Ahmed, A. R.; Amagai, M.; Barnadas, M. A.; Borradori, L.; Bystry, J. C.; Cianchini, G.; Diaz, L.; Fivenson, D.; *et al.* Consensus statement on definitions of disease, end points, and therapeutic response for pemphigus. *J. Am. Acad. Dermatol.*, 58(6):1043-6, 2008.
- Murrell, D. F.; Marinovic, B.; Caux, F.; Prost, C.; Ahmed, R.; Wozniak, K.; Amagai, M.; Bauer, J.; Beissert, S.; Borradori, L.; *et al.* Definitions and outcome measures for mucous membrane pemphigoid: recommendations of an international panel of experts. *J. Am. Acad. Dermatol.*, 72(1):168-74, 2015.
- Mustafa, M. B.; Porter, S. R.; Smoller, B. R. & Sitaru, C. Oral mucosal manifestations of autoimmune skin diseases. *Autoimmun Rev.*, 14(10):930-51, 2015.
- Patel, S.; Kumar, S.; Laudenbach, J. M. & Teruel, A. Mucocutaneous diseases: oral lichen planus, mucous membrane pemphigoid and pemphigus vulgaris. *J. Calif. Dent. Assoc.*, 44(9):561-70, 2016.
- Petruzzi, M. Mucous membrane pemphigoid affecting the oral cavity: short review on etiopathogenesis, diagnosis and treatment. *Immunopharmacol. Immunotoxicol.*, 34(3):363-7, 2012.
- Ramos, W.; Chacon, G. R.; Galarza, C.; Gutierrez, E. L.; Smith, M. E. & Ortega-Loayza, A. G. Endemic pemphigus in the Peruvian Amazon: epidemiology and risk factors for the development of complications during treatment. *An. Bras. Dermatol.*, 87(6):838-45, 2012.
- Said, S. & Golitz, L. Vesiculobullous eruptions of the oral cavity. *Otolaryngol. Clin. North Am.*, 44(1):133-60, 2011.
- Sankar, V. & Noujeim, M. Oral manifestations of autoimmune and connective tissue disorders. *Atlas Oral Maxillofac. Surg. Clin. North Am.*, 25(2):113-26, 2017.
- Santoro, F. A.; Stoopler, E. T. & Werth, V. P. *Pemphigus*. *Dent. Clin. North Am.*, 57(4):597-610, 2013.
- Schmidt, E. & Zillikens, D. Modern diagnosis of autoimmune blistering skin diseases. *Autoimmun. Rev.*, 10(2):84-9, 2010.
- Scully, C. & Lo Muzio, L. Oral mucosal diseases: mucous membrane pemphigoid. *Br. J. Oral Maxillofac. Surg.*, 46(5):358-66, 2008.
- Scully, C. & Mignogna, M. Oral mucosal disease: pemphigus. *Br. J. Oral Maxillofac. Surg.*, 46(4):272-7, 2008.
- Srikumaran, D. & Akpek, E. K. Mucous membrane pemphigoid: recente advances. *Curr. Opin. Ophthalmol.*, 23(6):523-7, 2012.
- Sultan, A. S.; Villa, A.; Saavedra, A. P.; Treister, N. S. & Woo, S. B. Oral mucous membrane pemphigoid and pemphigus vulgaris-a retrospective two-center cohort study. *Oral Dis.*, 23(4):498-504, 2017.
- Svecova, D. Pemphigus vulgaris: clinical study of 44 cases over a 20-year period. *Int. J. Dermatol.*, 54(10):1138-44, 2015.
- Taylor, J.; McMillan, R.; Shephard, M.; Setterfield, J.; Ahmed, R.; Carrozzo, M.; Grando, S.; Mignogna, M.; Kuten-Shorrer, M.; Musbah, T.; *et al.* World Workshop on Oral Medicine VI: a systematic review of the treatment of mucous membrane pemphigoid. *Oral Surg. Oral Med. Oral Pathol. Oral Radiol.*, 120(2):161-71.e20, 2015.

Corresponding author:

Paulo Sérgio da Silva Santos
Alameda Doutor Octavio Pinheiro Brisolla, número 9-75
Vila Universitária
Bauru SP
BRASIL

E-mail: paulosss@fob.usp.br