SYSTEMIC CORTICOSTEROID IN THE TREATMENT OF A 9-YEAR-OLD BOY WITH ORAL MUCOUS MEMBRANE PEMPHIGOID: A CASE REPORT

UTILIZAÇÃO DE CORTICÓIDE SISTÊMICO NO TRATAMENTO DO PENFIGÓIDE BENIGNO DAS MEMBRANAS MUCOSAS EM UMA CRIANÇA DE 9 ANOS DE IDADE: UM RELATO DE CASO

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ABSTRACT

Objective: The aim of this report is present a rare case of oral Mucous Membrane Pemphigoid (MMP) in a 9-year-old-boy successfully treated with low doses of systemic corticosteroid. Case description: A 9-year-old boy was referred to our service with the complaint of painful gingiva and generalized burning mouth sensation over 1 year. Intraoral examination revealed desquamative lesions affecting the entire attached gingiva of both the maxilla and the mandible. After the incisional biopsy the MMP diagnosis was confirmed, and the patient was initially treated with dexamethasone 0,1mg/Ml mouth rinse twice daily for 24 weeks. Due the difficulty in eating and the presence of persistent large oral lesions, systemic prednisone 20 mg (1 mg/Kg) was prescribed for 4 weeks with a 5 mg gradual reduction per week over 3 weeks. After initiating the systemic corticosteroid therapy, the patient showed total regression of the lesions, and no indication of recurrence has been observed in the past 6 months. Conclusion: There is no cure for MMP; however, treatment can provide complete and long-lasting remission. Based on this premise, it is plausible to consider not only topical but also low doses of systemic corticosteroids in resistant cases of oral MMP, even in children. These measures could improve the quality of life of these patients by reducing pain and, consequently, improving the child's eating behavior.

KEYWORDS: Oral Mucous Membrane Pemphigoid; Benign Mucosal Pemphigoid; Child.

INTRODUCTION

Mucous membrane pemphigoid (MMP) constitutes a group of chronic inflammatory autoimmune diseases that affect the oral, ocular, genital, nasopharyngeal, esophageal and laryngeal mucosa and, occasionally, the skin. It is characterized by the linear deposition of IgG, IgA, or C3 along the epithelial basement membrane zone (BMZ)[1], resulting from the formation of antibodies against components of the BMZ, and causing subepithelial blistering and consequent erosion of the mucosa[2].

The oral cavity is the most common site for MMP, and, in some cases, oral MMP is the single manifestation of this disease[1]. Clinical presentation includes erosions, erythematous patches, blisters, and desquamative gingivitis[3]. Although, scarring is common on skin lesions, it is unusual in the oral cavity. MMP is a diagnosis that should be considered in cases with chronic (>6 months) and multiple oral ulcers, predominantly affecting women and elderly patients, between the ages of 60-80 years[4]. Oral manifestation of MMP is extremely rare in children[5], with just a few reports in the literature.

Topical corticosteroids are indicated for the treatment of localized cases, but in some cases, the control of the lesions is not achieved by these agents[5]. Systemic corticosteroids are usually avoided in treating young patients having MMP with oral lesions, because of their serious side effects. However, the rational use of these drugs could be helpful and safe in the treatment of juvenile patients with MMP[6].

The aim of this article was to report a rare case of oral MMP in a 9-year-old boy treated successfully with low doses of systemic corticosteroid.

CASE DESCRIPTION

A 9-year-old boy presented to our service with a chief complaint of painful gingiva and generalized burning mouth sensation. The patient's clinical history revealed that he had these

lesions over 1 year; however, the symptoms and difficulty eating arose in the past 2 months. There was not any medical history of skin or pulmonary disease or drug intake, or of any kind of previous infection. His family history was not contributory.

Apparently, the patient was presenting normal height and weight for his age. On extraoral examination, no skin or other mucosal involvements were observed. Intraoral examination revealed generalized erythema, erosions and desquamative lesions affecting the entire attached gingiva of both the maxilla and the mandible (Figure 1). Gentle pressure application on his gingiva caused gum peeling, indicating a positive Nikolsky's sign. Based on this clinical presentation, our clinical hypothesis was mucous membrane pemphigoid, pemphigus or erosive lichen planus.

Biopsy of the lesional gingival tissue, including intact mucosa, was performed for histopathological study. Microscopically, the specimen showed subepithelial detachment at the level of the basement membrane, with consequent blister formation and a chronic inflammatory infiltrate in the lamina propria (Figure 2). The clinical and microscopic findings confirmed the diagnosis of MMP, and the patient was initially treated with dexamethasone 0,1mg/Ml mouth rinse twice daily for 12 weeks, associated with rigorous homecare and professional oral hygiene regimens. However, only a minor clinical improvement was seen. The topical corticosteroid treatment was continued for another 12 weeks, but, again, no significant improvement was reported.

Since the difficulty in eating and the presence of large oral lesions persisted, systemic prednisone 20 mg (1 mg/Kg) was prescribed for 4 weeks with a 5 mg gradual reduction per week over 3 weeks. During the two years of follow-up, there was no sign of improvement in the first 6 months. After initiating the systemic corticosteroid therapy, the patient showed total regression of the lesions, and no indication of recurrence has been observed in the past 6 months (Figure 3).

DISCUSSION

MMP is an autoimmune subepithelial blistering disease that involves the mucous membranes and skin. Although skin involvement is unusual, occurring in less than 25% of the cases[7], the consequences of this disease can be severe, resulting in blindness or critical laryngeal stenosis[1]. The oral cavity is the most usual site for MMP[1], and clinically presents erosions, erythematous patches, blisters, and desquamative gingivitis[3].

MMP affects more often women than men, and is commonly observed in patients over 60 years of age. Currently, approximately 20 cases of MMP in childhood have been reported[5]. The occurrence of oral MMP in children is extremely rare. About 8 cases in patients under the age of 18 years have been reported so far in the English literature[6, 8-15]. This case report presents

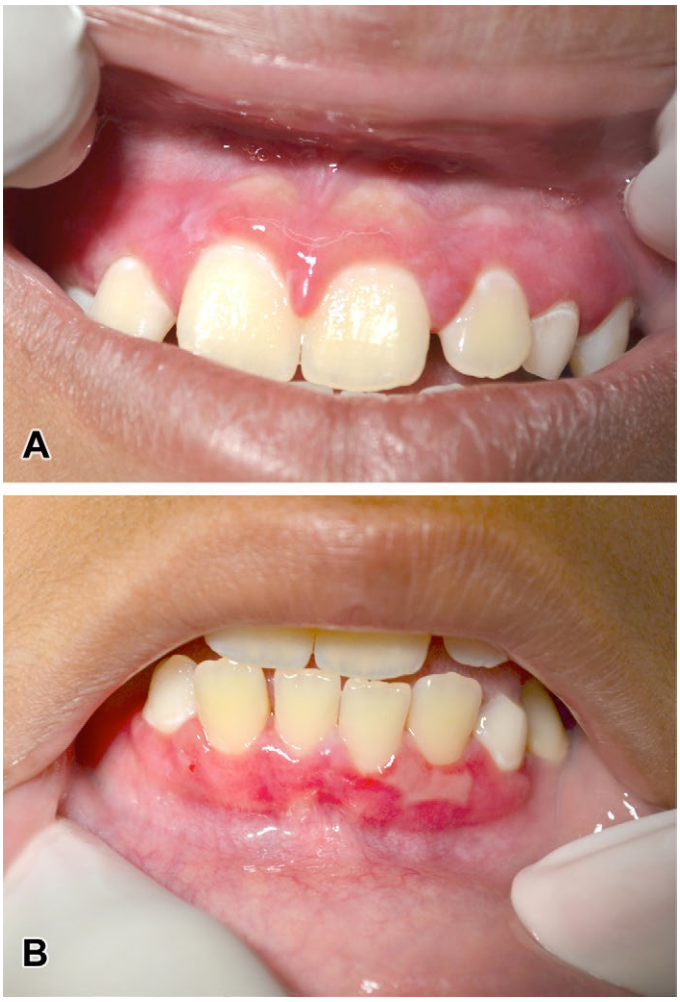

Figure 1 - Generalized erythema and desquamative gingivitis affecting the attached gingiva of (A) the maxilla and (B) the mandible.

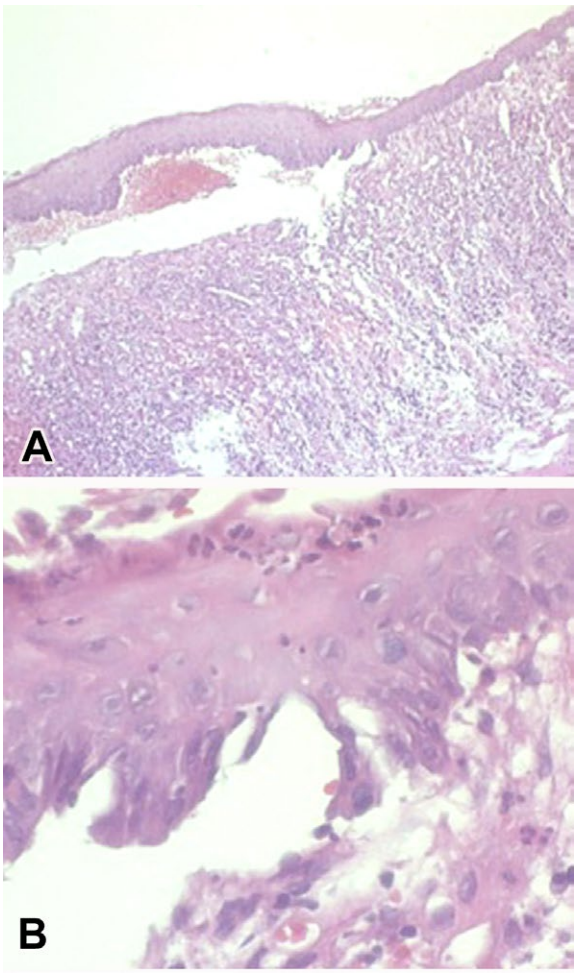

Figure 2 - Histopathology showing (A) subepithelial detachment and blister formation, and (B) chronic inflammatory infiltrate observed in the lamina propria (H-E, 200X – 400X).

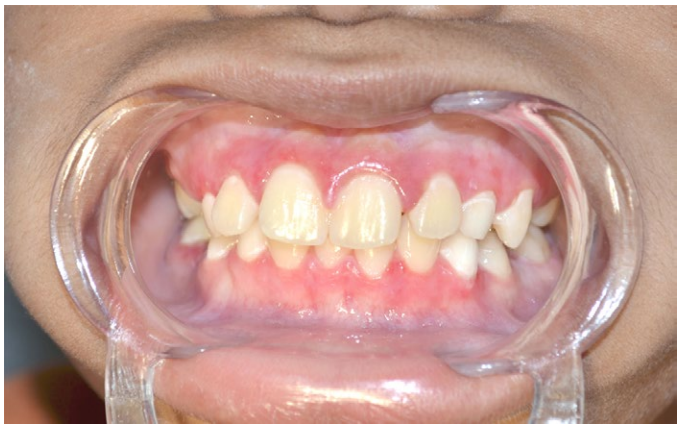

Figure 3 - Complete regression of the lesions and no indication of recurrence in the past 6 months.

an additional case of oral MMP in a 9-year-old boy with erosions and desquamative gingivitis on attached gingiva of both the maxilla and the mandible; the main clinical concern was the presence of dysphasia and a possible developmental delay.

Laryngeal and esophageal lesions of MMP are probably the most frequent manifestations related to dysphagia[16,17]. On the other hand, oral erosions or other forms of painful oral lesions could cause some eating problems in pediatric patients[18], demanding effective therapeutic actions to prevent disturbances in the physical development of the child.

The diagnosis of MMP, as defined in a consensus statement[1], is based on a combination of clinical, histological and immunopathological studies, which have concluded that clinical and direct immunopathology are essential for its diagnosis. However, the immunopathological exam can fail, and the diagnosis, in some cases, is based on clinical and histological aspects[8], as was done in the present case report.

The treatment for pemphigoid is based on clinical presentation, severity, site of involvement and age of the patient. Immunosuppressive medication is the main treatment modality; topical and systemic corticosteroids have also been used. In mild and moderate cases, topical agents are the first choice of treatment. However, in severe cases, systemic corticosteroid is used alone or combined with topical immunosuppressive agents[6]. When MMP is localized only in the oral mucosa, topical corticosteroid or dapsone should be the first choice. If either is not enough for clinical improvement, or if there is ocular, nasopharyngeal, esophageal or laryngeal involvement, then a systemic corticosteroid or immunosuppressive agents (azathioprine or mycophenolate mofetil) can be indicated[5, 19]. In this case, we chose systemic prednisone after an unsatisfactory response of the patient to a topical corticosteroid agent, and because it was a simpler therapeutic regimen than dapsone.

As with adults, the first choice of treatment for children should be topical corticosteroid; systemic corticosteroid can be prescribed for advanced cases or in resistant cases that do not respond to topical therapy[8]. It is important to state that long-term use of systemic corticosteroids can produce a range of side effects, including secondary infections, hypertension, hyperglycemia, gastric ulcers and osteoporosis. However, meticulous monitoring and use of preventive strategies, like low duration and dose of the corticosteroid, can reduce these side effects[5].

The response of MMP patients to treatment can be quite distinctive and the prognosis may be unpredictable. Based on this presupposition, the treatment should be individualized, according to disease severity and response to prior treatment[5].

Since 1977, different treatment modalities have been published for childhood MMP. Mostafa et al. (2010)[11] described a case of oral MMP in a 6-year-old boy who presented desquamative gingivitis, causing pain and eating difficulty. In their report, they described the treatment of oral MMP with topical betamethasone for 12 weeks, showing a great improvement in the gingival mucosa, marked only by persistent redness.

In 1986, Moy et al.[9] described a case of MMP in a 9-year-old girl, involving the oral mucosa and the eyes. According to this report, the patient was treated with 40 mg of systemic prednisone for about 8 weeks, and after lesion remission, prednisone was gradually decreased.

More recently, Lourenço et al.[8] (2006) described the case of oral MMP in a 4-year-old girl, initially treated with 50 mg of dapsone daily for 12 months. After 20 months, and regression of the lesions, dapsone was slowly discontinued and the patient was free from the disease after a 4-month follow-up.

In the present report, we showed a challenging case with a 2-year follow-up. Initially, the boy was treated with topical corticosteroids, according to the consensus in the literature. Unfortunately, no significant improvement was observed after a total of 6 months of treatment. A low dose of systemic prednisone (20 mg) was then given for 4 weeks, with a gradual 5 mg dose reduction per week over 3 weeks. With this systemic corticosteroid therapy regimen, the patient showed total regression of the lesions and has presented no indication of recurrence in the past 6 months.

According to a recent study, gingival MMP sites have greater plaque accumulation compared with non-affected sites, and seem to be more susceptible to gingival recession and clinical attachment loss[20]. This presumably presents an increased risk and severity of periodontal disease. In view of this scenario, we proposed a rigorous home and professional oral hygiene regimen to our patient, to control periodontal inflammation associated with bacterial biofilm. We believe that these measures were also important in controlling oral comorbidities associated with treating topical or systemic corticosteroids.

There is no cure for MMP; however, treatment can provide complete and long-lasting remission[21]. Based on this premise, it is plausible to consider not only topical but also low doses of systemic corticosteroids in resistant cases of oral MMP, even in children. These measures could improve the quality of life of these patients by reducing pain and, consequently, improving the child's eating behavior.

The present report adds an extra case of oral MMP in a 9-year-old boy to the literature, and demonstrates treatment results with a low dose of a systemic corticosteroid, associated with a rigorous oral hygiene regimen, as a therapeutic option for children with oral MMP.

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RESUMO

Objetivo: O objetivo deste relato é apresentar um caso raro de Penfigóide Benigno das Membranas Mucosas (PBMM) em um menino de 9 anos de idade tratado com sucesso com baixas doses de corticosteróide sistémico. Relato de caso: Um menino de 9 anos de idade foi encaminhado ao nosso serviço com a queixa de gengiva dolorida e sensação de queimação na boca há aproximadamente 1 ano. Ao exame físico intraoral constatou-se a presença de lesões descamativas que afetavam toda a gengiva inserida, tanto na maxila quanto na mandíbula. Após a realização de uma biópsia incisional o diagnóstico de PBMM foi confirmado, e o paciente foi inicialmente tratado com elixir de dexametasona 0,1mg / ml duas vezes por dia durante 24 semanas. Devido a dificuldade em comer e a presença de grandes lesões orais persistentes, foi prescrito prednisona sistêmica 20 mg (1 mg / Kg) por 4 semanas com uma redução gradual de 5 mg por semana durante 3 semanas. Depois de iniciar a corticoterapia sistêmica, o paciente apresentou regressão total das lesões, e nenhum sinal de recorrência tem sido observado nos últimos 6 meses. Conclusão: Não há cura para o PBMM; no entanto, o tratamento pode proporcionar a remissão completa e de longa duração. Com base nesta premissa, é plausível considerar a terapia tópica, mas também a utilização de corticosteroides sistêmicos em casos resistentes de PBMM oral, mesmo em crianças. Estas medidas poderiam melhorar a qualidade de vida desses pacientes, reduzindo a dor e, consequentemente, melhorando a alimentação da criança.

PALAVRAS-CHAVE: Penfigóide benigno das membranas mucosas; Penfigóide mucoso benigno; Criança.

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