

Pyodermatitis-pyostomatitis vegetans: case report and review of medical literature*

Piodermatite-pioestomatite vegetante: relato de caso e revisão de literatura

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Abstract: Pyodermatitis-pyostomatitis vegetans is a rare inflammatory dermatosis of unknown etiology, with a typical mucocutaneous involvement. We report the case of a woman with pustular and vesicular lesions in the axillae, evolving with vegetating plaques and pustules with annular grouping. The disease progressed with vulvar and inguinal involvement as well as involvement of the oral, nasal and ocular mucous membranes. She started the treatment with prednisone (40 mg/day), with remission of the lesions after one month of use of such medication. Association with inflammatory bowel disease occurs in 70% of the cases. Immunofluorescence, which is typically negative, helps to characterize the disease. A rapid response to systemic steroids is expected.

Keywords: Corticosteroids; Vesiculobullous skin diseases, eosinophilia; Adrenal cortex hormones

Resumo: A piodermatite-pioestomatite vegetante é uma rara dermatose inflamatória de etiologia desconhecida, com típico comprometimento mucocutâneo. Relatamos caso de paciente feminina com lesões pustulosas e vesiculosas em axilas, evoluindo com placas vegetantes e pústulas com agrupamento anular. Houve progressão com comprometimento vulvar, inguinal e mucosas oral, nasal e ocular. Proposto o diagnóstico, optou-se por iniciar prednisona 40mg ao dia, com remissão das lesões após um mês de uso da medicação. A associação com doença inflamatória intestinal ocorre em 70% dos casos. A imunofluorescência é um fator que ajuda a caracterizar a doença, sendo tipicamente negativa. A rápida resposta à terapêutica com corticosteroides sistêmicos é esperada.

Palavras-chave: Corticosteroides; Dermatopatias vesiculobolhosas; Eosinofilia

INTRODUCTION

Pyodermatitis-pyostomatitis vegetans (PD-PSV) is a rare inflammatory dermatosis characterized by pustular and vesicular lesions and vegetating plaques, with typical mucocutaneous involvement. Cutaneous lesions can affect the genital and axillary areas as well as the scalp. Less frequently it affects the face, chest and extremities.^{1,2} Mucosal involvement is characterized by multiple pustules and vesicles on erythematous base that, in general, disrupt themselves resulting in exulcerations. Oral mucosa is the most affected and there might occur lesions in the nasal and conjunctival mucosae.^{1,2,3,4} PD-PSV has strong asso-

ciation with gastrointestinal diseases, being considered a marker of inflammatory bowel disease (IBD).^{1,2,5}

Hallopeu⁶ described in 1889 a new clinical entity characterized by vegetating pustular lesions with benign outcome, not associated with constitutional symptoms. Nine years later, five similar cases were, once more, described by him and the entity was named pyodermatitis-vegetans. Two of these five cases presented the vegetating cutaneous lesions associated with pustular lesions in the oral mucosa. McCarthy,⁷ in 1949, introduced the term pyostomatitis vegetans, that he supposed to be part of Hallopeu's

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Pyodermatitis Vegetans, after reporting the case of three patients who presented isolated lesions in the oral mucosa with two of these cases evolving with skin lesions.

There are disagreements about the real origin of this entity. Some believe that it is a form of pemphigus, others, an idiosyncratic reaction to skin infections, a variant of dermatitis herpetiformis or a distinct entity.^{6,7, 8-10}

CASE REPORT

Forty-seven year old female patient, previously healthy, presented in november 2006, insidious onset of pustular and vesicular lesions, pruritic, and painful in axillae, evolving with vegetating plaques coated with pustules, papules and vesicles with annular grouping in intensively erythematous (Figure 1A). There was progression with involvement of the vulva, skin of the inguinal and hypogastric areas with annular vegetating plaques coated by pustules besides exulcerated lesions in lower left periorbital region and lower perioral region (Figure 1B). Absence of lesions in the oral mucosa at the early stage of the disease. The patient denied gastrointestinal or constitutional symptoms with the condition.

The patient had to be hospitalized due to the exuberance of the clinical condition and systemic antibiotic therapy (cephalothin 2g every six hours) was carried out during seven days without success. During hospitalization it was performed biopsy of the axillary vegetative lesion and histopathological study which showed: ulceration and adjacent epithelial hyperplasia, focal subepidermal cleft, mixed inflammatory infiltrate predominantly eosinophilic with perivascular

distribution, adnexa and interstitial, associated with papillary eosinophilic abscesses and eosinophilic spongiosis (Figure 2). Presence of neutrophilic papillitis in rare areas. Direct and indirect immunofluorescence were carried out and were negative.

Laboratory investigation revealed peripheral eosinophilia (11% - 979/mm³) and increase in the speed of ESR (erythrocyte sedimentation rate) (55 mm in the 1st hour). Antineutrophil cytoplasmic antibodies and antinuclear antibody were negative. Enzymes and liver function tests were normal. Colonoscopy and barium enema unchanged.

Proposed the diagnosis of PD-PSV, it was decided to start prednisone 40mg/day, with remission of the lesions after a month of use of the medication, presenting only residual hyperchromic macules (Figure 3). The medication was gradually withdrawn over 6 months. Before the use of prednisone 5mg in alternate days the patient presented lesion recurrence being necessary to return to the use of the dose of 40 mg/day.

In an attempt to adequately control the condition and reduce the dose of prednisone, dapsone was started with partial remission of the clinical condition (dapsone 100mg/day associated with prednisone 5mg/day). Intermittent recurrences with the appearance of lesions in the oral and nasal mucosae and in the left upper eyelid characterized by vesicles and pustules on the erythematous base besides the impairment of the left conjunctival mucosa by intense enanthema (Figure 4).

Associated with clinical improvement there was normalization of eosinophil count in peripheral blood.



FIGURE 1: A. Erythematous papules, pustules, vesicles and lesions coated by crusts that form vegetating plaques in axillary region; B. Involvement of the vulvar, inguinal and hypogastric regions

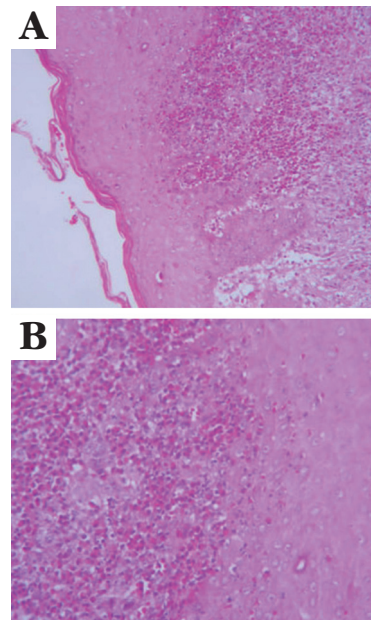


FIGURE 2: A. Epithelial hyperplasia, focal subepidermal cleft, mixed inflammatory infiltrate predominantly eosinophilic with perivascular distribution, adnexa and interstitial, associated with papillary eosinophilic abscesses and eosinophilic spongiosis. B. Detail of the interstitial inflammatory infiltrate predominantly eosinophilic



FIGURE 3: Residual hyperchromic macules in axillary and vulvar regions after introduction of corticosteroid treatment



FIGURE 4: A. Exulcerated lesions on palate; B. Vesico-pustular lesion on the left upper eyelid on erythematous base besides involvement of the left conjunctival mucosa by intense enanthema

DISCUSSION

Pyodermatitis-pyostomatitis vegetans (PD-PSV) is a rare eosinophilic inflammatory dermatosis of unknown cause and difficult diagnosis characterized by vesiculous and pustulous lesions and vegetating plaques with mucocutaneous involvement.² It generally occurs in individuals at the end of their third decade of life being the proportion of incidence between the female and male sex of 3:1.¹

In most cases, the oral mucosa is affected, presenting pustulous exophytic lesions with erythematous halo, friable membrane that easily disintegrates leading to exulcerated lesions. Lips, gums, soft and hard palate, vestibule and tonsils are the most frequently affected areas.^{1,5,11} Vaginal, nasal and ocular mucosae can also be affected.^{2,3,4} Mucosal lesions may precede, may appear simultaneously or succeed skin lesions.^{5,11,12} The patient presented lesions in the oral, ocular and nasal mucosae during the evolution of the condition with clinical characteristics similar to the ones described in the medical literature.

Cutaneous involvement is frequent, occurring in 58% of the cases.² It is characterized by erythematous pustulous papules, vesicles and lesions coated by crusts which coalesce into vegetating plaques. Lesions are asymmetric and affect mainly axillae, sculp, vulva and groin. Less frequently, other regions can also be affected like face, chest and extremities.^{1,2,13} In the reported case, the patient presented the characteristic cutaneous manifestations in typical areas as first manifestations of the disease. The association of PD-PSV with IBD is well established, occurring in 70% of the cases.¹ More commonly associated with ulcerative colitis (53%), being Crohn's disease seen in 11% of the patients.² Although PD-PSV is considered a

marker of IBD this connexion is not absolute and it is not found in all described cases, suggesting the existence of a variable exclusively mucocutaneous.¹¹

However, skin lesions may precede gastrointestinal symptoms in approximately 15% of the cases indicating the need of regular and careful gastroenterological monitoring of these patients.^{2,5,12} Approximately 26% of the cases of PD-PSV present some kind of liver dysfunction such as sclerosing cholangitis, chronic hepatitis and pericolangitis.²

In the present case, gastroenterological investigation including laboratory tests, barium enema and colonoscopy were performed and showed no changes. However, regular gastroenterological monitoring is kept aiming at detecting IBD or liver dysfunctions at an early stage in case they occur.

There are reports of association with zinc deficiency, possibly caused by malabsorption resulting from the inflammatory bowel disease and improvement of the skin condition with the supplementation of this mineral.¹³

Etiology of PD-PSV remains unknown. Some authors suggest a spectrum of neutrophilic dermatoses and others propose that it represents a clinical manifestation of pyoderma gangrenosum. Others classify it as a hypersensitivity reaction such as the erythema nodosum or pyoderma gangrenosum, associated with IBD, and there are still some that associate it with other mucosal diseases that present intraepithelial abscesses and acantholysis such as pemphigus vegetans and vulgaris.^{2,5,12} Unlike PD-PSV, pemphigus vulgaris is potentially fatal and both, the vegetans and vulgaris forms, are well characterized as autoimmune bullous diseases revealing intercellular

deposits of IgG and C3 by direct immunofluorescence and positivity of circulating antibodies by indirect immunofluorescence.¹⁴ Besides that, many times they are refractory to therapy in opposition to the excellent response to steroids presented by PD-PSV.

In PD-PSV, histopathology reveals pseudoepitheliomatous hyperplasia with intraepithelial microabscesses and eosinophilic spongiosis. Acantholysis and suprabasal clefts can be seen. A dense perivascular inflammatory infiltrate formed by neutrophils, eosinophils, lymphocytes and plasma cells can be seen on the dermis. As for the present case, the histopathological findings were similar to those found in the medical literature. Although very similar to the findings observed for pemphigus vegetans there are also some characteristics of dermatitis herpetiformis such as the presence of neutrophilic papillitis in scarce areas. These morphological aspects possibly contribute for understanding the existing disagreements reported by some authors concerning the real nature of this disease.^{6,7,10} Concerning the predominance of eosinophils in the inflammatory infiltrate further studies, retrospective and prospective, could be valuable in trying to enframe the entity, from a morphological point of view, within the pattern of eosinophilic dermatoses and not in the pattern of neutrophilic dermatoses.

Immunofluorescence constitutes another fac-

tor that helps to characterize the disease. It facilitates the differentiation between pemphigus vegetans and PD-PSV, as this has both direct and indirect immunofluorescences negative. However, a direct immunofluorescence slightly positive does not exclude PD-PSV. As for the case presented here, both direct and indirect immunofluorescence were negative confirming the findings of medical literature.

In the reported case, the hematological and biochemical assessments are within the normal limits in most cases except for an increase in the erythrocyte sedimentation rate and for the peripheral eosinophilia that can be found in 90% of the cases, having diagnostic value.² Eosinophilia can be 20% bigger than the differential leukocyte count. Cultures for fungi, bacteria and viruses are generally negative.

The quick response to therapy with systemic corticosteroids is common in PD-PSV as observed in the reported case. Topical corticosteroids, dapsone, sulfasalazine, azathioprine and cyclosporins have already been used with success.^{1,2,4,5,11,15} There is report of the effectiveness of infliximab associated with methotrexate to control PD-PSV.¹⁵ Total remission has already been observed after total colectomy for treatment of subjacent IBD.¹⁶

Prognosis for PD-PSV is excellent. When associated with IBD its development and severity tend to follow the progression of the basic disease. □

REFERENCES

- Ko HC, Jung DS, Jwa SW, Cho HH, Kim BS, Kwon KS, et al. Two cases of pyodermitis-pyostomatitis vegetans. *J Dermatol*. 2009;36:293-7.
- Nigen S, Poulin Y, Rochette L, Lévesque MH, Gagné E. Pyodermitis-pyostomatitis vegetans: two cases and a review of the literature. *J Cutan Med Surg*. 2003;7:250-5.
- Leibovitch I, Ooi C, Huilgol SC, Reid C, James CL, Selva D. Pyodermitis-pyostomatitis vegetans of the eyelids case report and review of the literature. *Ophthalmology*. 2005;112:1809-13.
- Peuvrel L, Barbarot S, Gagey-Caron V, Tessier MH, Cassagnau E, Stalder JF. Pyodermitis-pyostomatitis vegetans with nasal involvement. *Ann Dermatol Venerol*. 2008;135:753-6.
- Storwick GS, Prihoda MB, Fulton RJ, Wood WS. Pyodermitis-pyostomatitis vegetans: a specific marker for inflammatory bowel disease. *J AM Dermatol*. 1994;31:336-41.
- Hallopeau H. "Pyodermitis vegetante". ihre Beziehungen zur dermatitis herpetiformis und dem pemphigus vegetans. *Arch Dermatol Syphilol*. 1898;43:289-306.
- McCarthy P. Pyostomatitis vegetans: report of three cases. *Arch Dermatol Syphilol*. 1949;60:750-64.
- Elder DE, Elenitsas R, Johnson BL, Murphy GF, Xu X. *Lever's Histopathology of the skin*. 10th ed. Philadelphia: Lippincott Williams & Wilkins, 2009. p. 546.
- Degos R, Carreaud A. Pyodermitis végétante de Hallopeau. *Ann Dermatol Syph*. 1953;80:254-62.
- McCarthy P, Shklar G. A syndrome of pyostomatitis vegetans and ulcerative colitis. *Arch Dermatol*. 1963; 88:913-9.
- O'Hagan AH, Irvine AD, Allen GE, Walsh M. Pyodermitis-pyostomatitis vegetans: evidence for an entirely mucocutaneous variant. *Br J Dermatol*. 1998;139: 552-3.
- Soriano ML, Martinez N, Grilli R, Farina MC, Martin L, Requena L. Pyodermitis-pyostomatitis vegetans: report of a case and review of the literature. *Oral Surg Oral Med Oral Pathol Oral Radiol Endod*. 1999;87:322-6.
- Ficarra G, Cicchi P, Amorosi A, Piluso S. Oral Crohn's disease and pyostomatitis vegetans. *Oral Surg Oral Med Oral Pathol*. 1993;75:220-4.
- Cunha PR, Barraviera SRCS. Dermatoses bolhosas autoimunes. *An Bras Dermatol*. 2009;84:111-24.
- Bens G, Laharie D, Beylot-Barry M, Vergier B, Noblesse I, Beylot C, et al. Successful treatment with infliximab and methotrexate of pyostomatitis vegetans associated with Crohn's disease. *Br J Dermatol*. 2003;149:181-4.
- Yasuda M, Amano H, Nagai Y, Tamura A, Ishikawa O, Yamaguchi S. Pyodermitis-pyostomatitis vegetans associated with ulcerative colitis: successful treatment with total colectomy and topical tacrolimus. *Dermatology*. 2008;217:146-8.

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