

Pyostomatitis Vegetans: A Clue for Diagnosis of Silent Crohn's Disease

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ABSTRACT

Pyostomatitis vegetans is a very rare oral manifestation with unknown pathogenesis. Skin and other mucous membrane involvement may be seen. This lesion has strong association with Inflammatory Bowel Disease (IBD) and may be the first sign of it. The management of Pyostomatitis vegetans is usually based on the management of underlying bowel disease. We present a case of Pyostomatitis vegetans involving gingiva and oral mucosa with no skin lesion which led to the diagnosis of Crohn's disease to emphasize important role of dentists in diagnosis of rare oral lesions and management of patients' systemic disease.

Keywords: Exophytic pustules, Gingiva, Inflammatory bowel disease, Oral manifestation, Ulcers

CASE REPORT

A 39-year-old woman with chief complaint of oral ulcers, oral malodour, and soreness since one month, presented to the dental clinic. She had no significant medical and family history and neither did she took any medication. Extra-oral examination was normal. Intra-oral examination revealed multiple exophytic pustules with yellowish appearance on the tender, erythematous base that covered all of the gingiva and buccal mucosa and vestibular oral mucosa in some regions [Table/Fig-1]. Tongue and floor of the mouth were free from lesions. No skin lesion was seen. Because of "snail track" appearance of the lesions the provisional diagnosis of Pyostomatitis vegetans was considered. The differential diagnosis included pemphigus vulgaris, and pemphigus vegetans. Incisional biopsy was carried out from the affected area. Microscopic sections showed intraepithelial clefting and acantholysis [Table/Fig-2]. Accumulation of eosinophils within the spinous layer (intraepithelial abscesses) was also seen. The underlying connective tissue demonstrated infiltration of mixed inflammatory cells. Therefore, the diagnosis of Pyostomatitis vegetans was confirmed.

As a result of known association between Pyostomatitis vegetans and chronic IBD, patient was referred to gastroenterologist. After stool examination and colonoscopy, the Crohn's disease was diagnosed. Systemic medication with Mezalazin and Azeram was started and after one month the oral lesions disappeared. After one year the oral lesions recurred as a result of drug discontinuation [Table/Fig-3], following which the treatment of Crohn's disease was restarted.

DISCUSSION

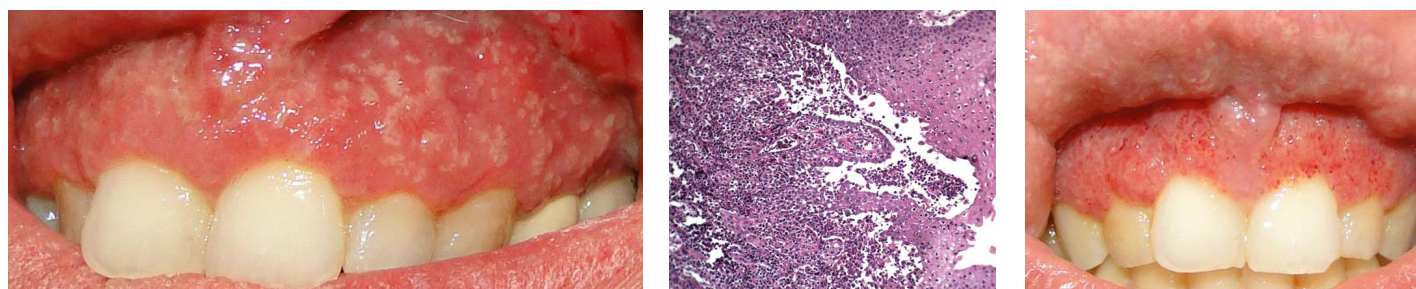
Pyostomatitis vegetans is a very rare oral disorder characterized by pustules that affect oral mucosa [1]. Multiple white or yellow pustules

on erythematous base may rupture and form folded, fissured appearance resembling a "snail-track" [2]. The labial gingiva, soft and hard palate, buccal and labial mucosa are most commonly affected [3]. Tongue and floor of the mouth was rarely involved [1]. Pyostomatitis vegetans is a rare oral lesion which may affect any age group but it is commonest in young and middle aged adults and has a predilection for males with male to female ratio of 2:1 to 3:1 [1,3,4].

Patients may have skin lesions, which was named pyodermitis vegetans, characterized by vesicular, pustular, exudative and vegetating plaques and usually involved the scalp, face, axillae and genital regions [5]. Cutaneous lesions can appear after or prior to the occurrence of Pyostomatitis vegetans lesions but Pyostomatitis vegetans can be seen without skin lesions [3]. Other mucosal membranes may be affected (vaginal, nasal, and rarely periocular mucosa) [3]. The present case was a 39-year-old female who presented only with oral lesions.

The diagnosis of Pyostomatitis vegetans is based on clinical features (snail tracks appearance), association with inflammatory bowel disease, peripheral eosinophilia, and characteristic histological features (intraepithelial abscesses with large amount of eosinophils). Direct and indirect immunofluorescence in Pyostomatitis vegetans lesions is usually negative or may be weakly positive in direct immunofluorescence and the results are inconclusive [3,4].

The differential diagnosis of Pyostomatitis vegetans include blistering mucocutaneous disorders such as pemphigus vulgaris and especially its rare variation pemphigus vegetans [3]. The main difference between Pyostomatitis vegetans and pemphigus vulgaris and pemphigus vegetans is the blistering nature of pemphigus. On the other hand, the association of Pyostomatitis vegetans with IBD is another important differentiating feature.



[Table/Fig-1]: Multiple exophytic pustules on the erythematous base that covered the gingiva and oral mucosa resembling "snail tracks". **[Table/Fig-2]:** Microscopic section revealed an intraepithelial clefting and inflammatory cells accumulation (eosinophilic microabscesses) $\times 100$. **[Table/Fig-3]:** The oral lesions recurred as a result of drug discontinuation.

Considering histopathology of the lesions, the characteristic feature of Pyostomatitis vegetans is intraepithelial abscesses filled with numerous eosinophils [3,6]. Histopathologic findings in pemphigus vulgaris is suprabasal clefting, row of tombstone, acantholysis, and presence of Tzank cells [6]. Pemphigus vegetans (Neumann type) showed intra-epidermal vesicles with suprabasal acantholysis without eosinophilic microabscesses. Hallopeau type pemphigus vegetans, showed eosinophilic spongiosis and microabscesses [3]. Direct and indirect immunofluorescence in pemphigus is positive [7]. Some authors believe that immunofluorescence is the only way to distinguish between Pyostomatitis vegetans and pemphigus [1,3]. But the others suggest that there is heterogeneity in the findings and the possibility of both false positive and false negative results must be considered [6-8].

One of the characteristic feature of pemphigus vegetans is tongue involvement with typical sulci and gyri pattern over the dorsum of the tongue called cerebriform tongue [9,10] and accepted as "Premalatha sign" [11]. Conversely, in Pyostomatitis vegetans tongue involvement is rare [1]. In the present case, the tongue and floor of the mouth were free from lesions.

Nico MM et al., [7], reported a case of Pyostomatitis vegetans with oral lesions and prolonged course of disease (several years) that never exacerbated into cutaneous lesions. This is unlike what usually occurs in non treated pemphigus. They suggested this finding as one of the issues while differentiating Pyostomatitis vegetans from pemphigus vegetans [7].

Pyostomatitis vegetans is a highly specific marker for IBD and may be the first sign of it [3]. IBDs, including Crohn's disease and ulcerative colitis, may have an extra-intestinal involvement such as oral manifestations [12,13]. Oral lesions are more prevalent in Crohn's disease, vice versa Pyostomatitis vegetans is the only oral lesion that occurs more commonly in ulcerative colitis [12,13]. IBD usually precede the onset of Pyostomatitis vegetans months or years, but may be asymptomatic or mild so the problem was not diagnosed [3,12]. Therefore, when Pyostomatitis vegetans is diagnosed in patient with no history of bowel disease, a thorough careful investigation of gastrointestinal tract must be taken into account. Treatment of any underlying bowel disease usually results in improvement of oral and skin lesions [3]. In this case, Pyostomatitis vegetans was associated with Crohn's disease which was diagnosed after appearance of oral lesion and referral to gastroenterologist. The treatment of Crohn's disease led to resolution of oral lesion.

As discussed above, management of Pyostomatitis vegetans is usually based on treatment of the underlying gastrointestinal disease. In the case of oral lesions without any gastrointestinal problem, oral lesions can be managed with local therapies such as

antiseptic mouthwashes (chlorhexidine), and topical corticosteroids (triamcinolone acetonide paste or betamethasone mouthwash). However, a limited success of topical steroid therapy was reported. Systemic steroid therapy (azathioprine and sulfamethoxypyridazine) led to resolving and controlling the lesions [2,3].

The pathogenesis of Pyostomatitis vegetans is unknown, but there is a theory that an abnormal immune response in the course of IBD led to cross reaction of antigens in the skin and small bowel, leading to this mucocutaneous manifestation [14].

CONCLUSION

Familiarity with rare oral lesions, especially those associated with systemic disorders, help general dentists and specialist in early diagnosis and appropriate management of the disease. Eventually, other important role of dentists in such condition is to prevent wandering the patients.

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