

PYOSTOMATITIS VEGETANS: REPORT OF A RARE CASE

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Pyostomatitis vegetans is a rare chronic disorder of the oral mucosa. It is a highly specific marker for inflammatory bowel disease.

Although immunological and microbial factors have been suggested as predisposing factors, the exact aetiology is unknown.

Here, a case of pyostomatitis vegetans in a 30-year-old woman is reported. No bowel involvement was observed. Report of such cases emphasizes the importance of early diagnosis of oral findings in control and management of disease.

Key words: pyostomatitis vegetans, oral mucosa, inflammatory bowel disease.

Introduction

Pyostomatitis vegetans (PV) is a rare, benign condition characterized by pustular and vegetative lesions on an erythematous base that affects the oral mucosa [1]. It is frequently associated with gastrointestinal and/or cutaneous involvement. The association of pyostomatitis vegetans with inflammatory bowel disease (IBD), especially Crohn's disease, is well known in the literature [2]. The aetiology of PV is unknown but immunological, microbial and psychogenic factors may play a role [2, 3].

Hallopeau reported the first cases of PV in 1898. He described 2 patients with pustular dermatosis and oral lesions. He named them "pyodermite végétant".

The name "pyostomatitis vegetans" was proposed for the first time in 1949 by McCarthy when he observed 3 patients with isolated oral lesions similar to those reported by Hallopeau [3-5].

Since the initial conception, approximately 37 cases have been reported in the literature and most of them supported the theory that pyodermatitis vegetans and pyostomatitis vegetans are the same disease [1, 3].

Although pyostomatitis vegetans may be seen at any age, it is more prevalent between 20 and 59 years, with an average of 34 years. This disease predominantly affects men [3].

Clinical presentation and distribution of the disease may vary among patients. Pyostomatitis vegetans is associated with inflammatory bowel disease (IBD) in 75% of cases [6]. Inflammatory bowel disease (IBD) often appears months or years before oral and skin findings, although symptoms may not be sufficient to make a diagnosis [2].

Oral mucosa is usually thickened and vegetating lesions on erythematous mucosa are seen. Multiple white to yellow pustules that often easily rupture and convert to erosive-ulcerative chronic lesions are usually observed. Granular appearance of oral mucosa, degeneration of vegetative pustules and erosive lesions always lead to a folded, fissured appearance named "snail tracks" [1-4].

All parts of the oral mucosa may be involved, but labial mucosa, attached gingiva, soft and hard palate and buccal mucosa are more commonly affected [7].

Cutaneous lesions are usually seen in the axillaries, groin and scalp areas in an asymmetrical pattern.

They present as crusted, erythematous papulo-pustules that extend peripherally and coalesce to form large vegetating plaques [8].

Controversy exists about the pathogenesis of PV. Many researchers believe that PV is an immunologi-



Fig. 1. Prominent fissured and velvety patient's lower lip



Fig. 2. Multiple yellow pustules on erythematous base on the labial and gingival mucosa. "Snail tracks"

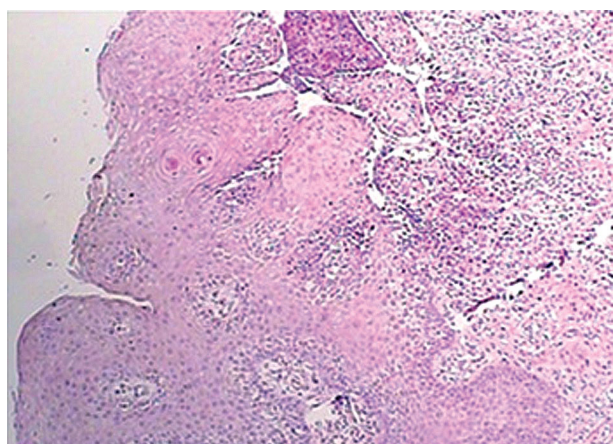


Fig. 3. Low magnification demonstrating the presence of intraepithelial clefting and an admixture of acute and chronic inflammatory cells with prominent eosinophils. HE, magnification 400×

cal disorder. Deposits of proteins in skin vessels in PG lesions are suggestive of an Arthus-like reaction [9, 10]. Although some authors have considered microbial pathogenesis for this disease, no bacteria, fungi or viruses have been found in research [11].

Histopathological findings are similar in the skin and oral lesions. Epidermal hyperplasia is a constant feature in skin specimens. Focal acanthosis and hyperkeratosis may be seen in oral lesions. Intraepithelial and subepithelial microabscesses with abundant eosinophils and neutrophils are another finding in biopsy specimens [3]. The inflammatory infiltrate shows a predominance of cytotoxic (CD3+/CD8+) lymphocytes [12]. As the lesion matures there are fewer eosinophils [13]. Viral, bacterial and fungal cultures of oral lesions of PV are usually negative. Both direct and indirect immunofluorescence studies are usually negative [3].

Case description

A 30-year-old woman was referred to the Oral Medicine Department of Mashhad Dental Faculty with a chief complaint of pain and a burning sensation in the lower and upper lips from one year ago.

On clinical examination prominent thickening of vermillion of both lips with spongy fissures was observed (Fig. 1).

Intraoral examination showed multiple yellow pustules on an erythematous base on the buccal mucosa and labial mucosa of the upper and lower lips, and also on the gingival and soft palate (Fig. 2). Some pustules ruptured, resulting in ulceration and erosions. The typical linear "snail track" appearance was also seen on the buccal mucosa and soft palate. Her familial history was unremarkable, and she had not received any treatment for her disease. She was in good overall health and denied any drug allergies.

Incisional biopsy from the labial mucosal lesions was performed. In histopathological examination, acanthosis, hyperparakeratosis and intraepithelial clefting with acantholysis were found. Intra and subepithelial abscesses containing numerous eosinophils and neutrophils were also observed. The lamina propria displays a chronic inflammatory cell infiltrate containing eosinophils, lymphocytes and plasma cells which aggregate to form small abscesses (Figs. 3-5).

The patient's gastro-intestinal status was evaluated by an internist and no abnormality was reported.

The patient was treated with oral methylprednisolone 20 mg/day. After 4 weeks oral lesions showed marked improvement and after 2 months they disappeared completely. The patient was followed for recurrence of oral lesions and IBD.

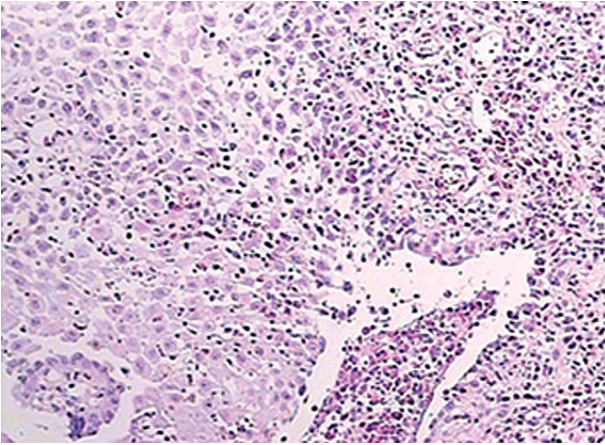


Fig. 4. Intraepithelial vesiculation with formation of eosinophilic abscesses. HE, magnification 400×

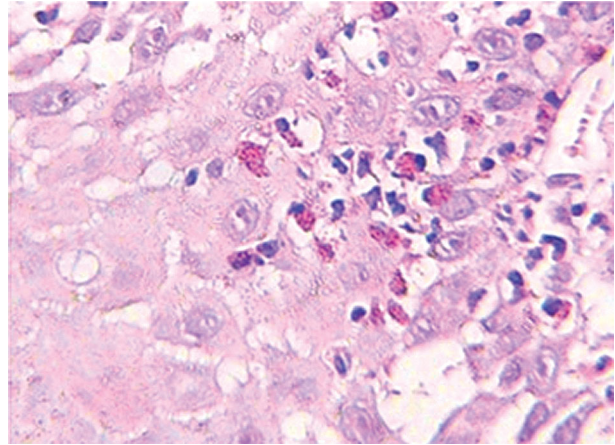


Fig. 5. High magnification of intraepithelial eosinophils. HE, magnification 400×

Discussion

The diagnosis of PV is based on clinical features, presence of inflammatory bowel disease, peripheral eosinophilia, negative culture of pus from lesions and histological features [13].

The differential diagnosis of PV includes blistering disease affecting the oral cavity such as pemphigus vegetans, pemphigus vulgaris, bullous pemphigoid, erythema multiforme, herpes simplex and bullous drug eruption. However, clinically the presence of pustules with characteristic “snail track” ulcers and histopathological features of intraepithelial eosinophilic abscesses suggests PV rather than other blistering diseases [3, 4]. The differential diagnosis should be established with Neumann type pemphigus vegetans since in both cases lesions are clinically similar and histology reveals an important eosinophil response, acanthosis and the formation of intraepithelial miliary microabscesses [14].

Although IBD may precede the onset of oral or cutaneous lesions by months or years, sometimes the symptoms may be minimal and not sufficient to make an early diagnosis. In these cases the identification of PV could represent a reason to encourage diagnostic investigations intended to reveal subclinical intestinal diseases [2].

Nigen in 2003 reported two patients with pyodermitis-pyostomatitis vegetans. Both patients had lesions on the skin and mouth [7].

Leibovitch also reported the first case of PDPSV in 2005 that involved the eyelid in addition to the skin and mouth [15].

If PV is associated with pyodermitis vegetans, cutaneous lesions are included of vesicopustular, vegetating plaques that are observed in 58% of patients [4].

Nigen reported pseudoepitheliomatous hyperplasia in two cases with pyodermitis-pyostomatitis

vegetans [7]. Deleporte suggested that pyodermitis-pyostomatitis vegetans belongs to the spectrum of neutrophilic dermatoses [6].

Management of PV is often based on treating the underlying gastrointestinal disease.

The oral lesions can be managed with local therapies utilizing antiseptic mouthwashes such as chlorhexidine, and topical corticosteroids such as triamcinolone acetonide paste or betamethasone mouthwash. However, topical steroid therapy has limited success. Strategic treatment initially consists of systemic steroid therapy aimed at resolving and controlling the lesions [16, 17]. As there were no signs or symptoms of IBD in our patient, we only used topical corticosteroid for oral lesions.

Conclusion

Report of such cases emphasizes the importance of early detection and diagnosis of oral lesions in control and management of the disease.

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