

## PYOSTOMATITIS VEGETANS

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**Abstract:** Pyostomatitis vegetans is a rare oral disorder with unclear pathogenesis, that may be associated with inflammatory gastro-intestinal diseases, cutaneous involvement or with other disorders of the joints, eyes or liver. The etiology of the extraintestinal manifestations of inflammatory bowel diseases is obscure, but it is believed that the injured intestinal mucosa may trigger immune responses related to inflammatory processes in extraintestinal areas. Oral manifestations include amicrobial pustules and yellow ulcerations with a typical aspect of snail tracts on the buccal and labial mucosa and on the dorsum of the tongue. Histopathological exam shows epithelial acanthosis and superficial ulcerations, as well as eosinophilic and neutrophilic infiltrate in the subjacent connective tissue. Intraepithelial and subepithelial milliary abscesses may also be present. The available therapeutical alternatives (systemic and topical corticosteroids, sulphones, retinoids) have a relatively poor and transitory effect. We present the case of a teenage girl with oral lesions and histopathological features suggestive for pyostomatitis vegetans, persistent for approximately one year, with no other cutaneous or systemic involvement.

**Key words:** Pyostomatitis vegetans, inflammatory bowel disease

### INTRODUCTION

Pyostomatitis vegetans is a rare oral disorder, considered to be a marker of an inflammatory bowel disease, as 70% of the cases begin concomitant with such a condition, most often represented by ulcerative colitis [1, 2]. The pathogenesis is still unclear, even though immunological and bacterial factors have been suggested as possible etiological factors [1]. It is believed that the disturbance of the enteric immune response leads to pathological alterations in various organs [3]. The most common extraintestinal disorders associated with an inflammatory bowel disease are dermatological, ophthalmological, musculoskeletal and hepato-biliary, but virtually any organ or system may be affected [4, 5].

### CASE REPORT

The adolescent female patient M.A.C., aged 16, without any notable family or personal history, describes the onset of the disorder at the end of the year 2006, with oral lesions accompanied by a relative discomfort and diagnosed as ulcerative

stomatitis. The histopathological features of a biopsy sample of the modified oral mucosa taken at the Oro-maxillo-facial Surgery Clinic in „Sf. Spiridon” Hospital, Iași revealed a non-specific inflammatory infiltrate: acanthosis, epithelial necrosis (Fig. 1), neutrophilic infiltrate forming intraepithelial and subepithelial microabscesses with perivascular disposition or inside the blood vessels' walls (Fig. 2, 3). She received topical and systemic antibiotics, oral topical nystatin and hydrated sodium borate in glicerine, with a partial and transitory effect.

In april 2007, when the patient presented to the Dermatology Clinic of the University Railways Hospital Iasi, the examination of the oral cavity revealed exofitic lesions with an erythematous border and a creamy-yellow surface presenting a crumbling membrane, easily desintegrating to generate small ulcerative lesions or superficial erosions on the dorsal aspect of her tongue, as well as pustules on the inferior labial mucosa and on the buccal mucosa (Fig. 4, 5 6), associated with a certain difficulty in eating and talking. The general physical

examination, as well as the hematological and biochemical paraclinical investigations

were within normal limits.

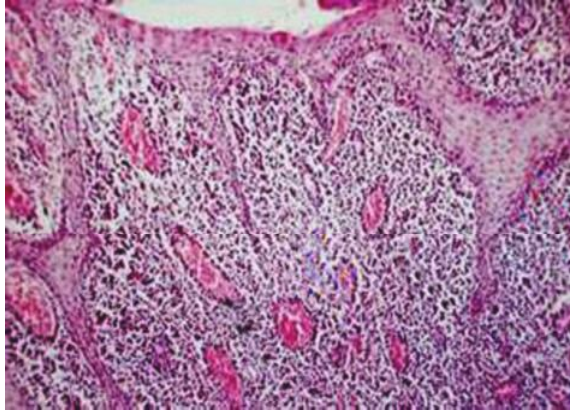


Figure 1. Acanthosis; dense perivascular inflammatory infiltrate with eosinophils, neutrophils and lymphocytes

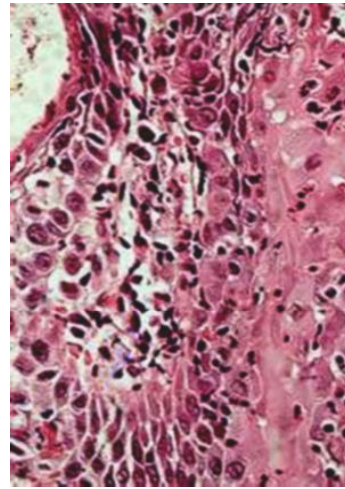


Figure 2. Intraepithelial neutrophilic microabscesses and intraepithelial oedema

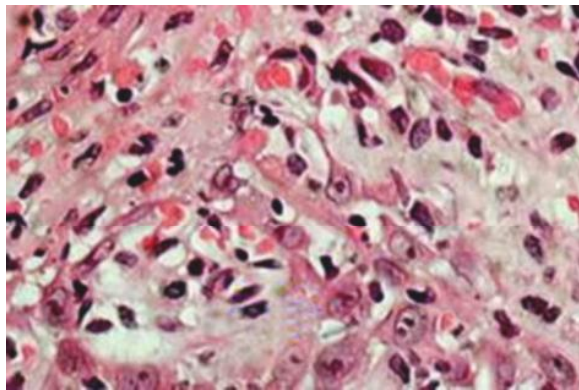


Figure 3. Ulceration: inflammatory granulation tissue



Figure 4. Exofitic creamy-yellow lesions with an erythematous border on the tongue



Figure 5. Pustules and erosive lesions covered with a crumbly membrane



Figure 6. Pustules on the inferior labial mucosa

The clinical aspect of the oral lesions, together with the histopathological findings leads to the diagnosis of pyostomatitis vegetans. The first line treatment consisted of systemic corticotherapy (Medrol 16 mg/day, 14 days, then 16 mg every other day, followed by Diprophos 1 ml every 30 days) associated with topical dexamethasone and ketoconazole in oleum helianthi. The therapeutic benefit was unsatisfactory after 2 months. Sulphonotherapy was then initiated, with Disulone 100 mg/day, 5 days a week, in association with vitamin C and Ferrofolgamma, with no therapeutic response after one month. A relatively favorable course was obtained in approximately 6 weeks, with Isotretinoin per os 10 mg/day and topical Tretinoin 0,025% in oleum helianthi (once every two days).

## DISCUSSIONS

Patients with inflammatory bowel disease may develop extraintestinal complications manifested in the skin, eyes, joints or liver [5]. The role of the gastrointestinal tract in the host's response to the foreign antigens present in the intestine is expressed through a high susceptibility of the enteric immune system to any external disturbances. Lymphocytic tissular infiltration seems to be the main feature of extraintestinal manifestations [5]. Recently the presence of a population of long life memory lymphocytes has been discovered, as a consequence of the intestinal

inflammation, that express homing receptors which direct their migration, not only in the intestine, but also in extraintestinal areas [4].

Cutaneous and mucous manifestations are usually correlated with the activity of the intestinal disease, but they may also have an independent course and are represented by pyoderma gangrenosum (severe, painful ulcerative disease) and erythema nodosum (in the skin) and by the recurrent aphthous stomatitis, cobblestoning mucous nodules and pyostomatitis vegetans (in the oral cavity) [6].

Oral lesions are invariably present, sometimes solitary, and they are represented by multiple yellowish pustules that form snail tracts and break easily [2]. The cutaneous lesions may precede, accompany or follow pyodermatitis-pyostomatitis vegetans that is usually extensive [7]. The cutaneous lesions begin as papulo-pustules with crusts that become coalescent to form big vegetant red-brown anular plaques especially in flexural areas, in genital area and on the scalp [8].

In the literature there are reports of cases of pyostomatitis vegetans with unusual clinical features and course. Thus, concomitant oral, labial and nasal manifestations were reported in a 28 year old patient in good general health and a 2 year history of an unpainful stomatitis, who presented pustules and crusts on the labial and narinar mucosa, with no intestinal manifestation and negative direct and indirect immunofluorescence, as well as an unusual case with relapses for 2

years, with no gastro-intestinal manifestation, dependent of corticotherapy, with no response to other therapies: disulone, isotretinoin, azathioprine [9].

Differential diagnosis must exclude the Neumann vegetant pemphigus, considering that the histopathological exam reveals similar important eosinophilic response, acanthosis and milliary intraepithelial microabscesses [10].

The treatment is unsatisfactory as a long term result and consists of corticosteroids, retinoids, sulphotherapy, azathioprine, antibiotherapy, multivitamines, nutritive supplements [10].

## CONCLUSIONS

Extraintestinal manifestations of inflammatory bowel disease may be diagnosed before, simultaneously or after the intestinal inflammatory disease in 20-

40% of the patients [11] and they significantly contribute to morbidity, as they negatively influence the life quality index in these patients. Even though pyostomatitis vegetans is considered a marker of an inflammatory intestinal malady (ulcerative colitis, Crohn's disease), cases with no intestinal involvement were reported [9]. The identification of common epitopes in the colon, eyes, joints and human biliary epithelium supports the autoimmune hypothesis of the extraintestinal manifestations [4]. Recent research suggests that biological anti-TNF $\alpha$  agents may have a beneficial effect on the course of these manifestations [11]. Further research will determine whether these extraintestinal disorders are manifestations of the inflammatory bowel disease, or if they represent associations of different autoimmune disorders.

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