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Short Communication

The Purulent Estuary-Pyostomatitis Vegetans

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Preface Pyostomatitis vegetans is an exceptional, benign, chronic inflammatory disease of mucous membranes demonstrating an insidious onset. Characteristically, the condition delineates an ulcerated oral mucosa exhibiting numerous pustules or exudative plaques with an elevated perimeter along with intraepithelial abscesses imbued with innumerable eosinophils. Pyostomatitis vegetans, an uncommon manifestation of inflammatory bowel disease as ulcerative colitis or Crohn's disease, may simultaneously be associated with a cutaneous condition designated as pyodermatitis vegetans. Pyostomatitis vegetans is contemplated as an oral counterpart of pyodermatitis vegetans which appears to incriminate various cutaneous surfaces. Cogent disease discernment is possible upon clinical evaluation, characteristic morphology, exclusion of diverse infective diseases or concordant malignant conditions. Disease Characteristics Pyostomatitis vegetans is intensely concurrent with inflammatory bowel disease (IBD), commonly ulcerative colitis or Crohn's disease. Nevertheless, an inflammatory bowel disease may not precede the emergence of cutaneous or oral lesions associated with pyostomatitis vegetans and systemic examination may be unremarkable [1,2]. Oral lesions with extra-intestinal manifestation of inflammatory bowel disease are prevalent in ulcerative colitis or Crohn's disease. Asymptomatic or mild inflammatory bowel disease may precede the onset of pyostomatitis vegetans by an extended duration. Therefore, pyostomatitis vegetans discerned in subjects with an absence of bowel disease mandates a thorough evaluation of the gastrointestinal tract in order to exclude various inflammatory bowel conditions [1,2]. Besides, the condition may be associated with autoimmune hepatitis, severe psoriatic arthritis, myelodysplastic syndrome, infection with organisms such as nocardia vinacea, staphylococcus aureus or human immune deficiency virus (HIV), malignant lymphoma, alcoholism, nutritional deficiencies or diabetes mellitus [1,2]. Pyostomatitis vegetans commonly emerges within young to middle aged adults between third decade to fourth decade although no age of disease emergence is exempt. Lesions demonstrate a male predominance with a male to female proportion of around ~ 3:1. Generally, miniature, pustular, vesicular, painful, ulcers or plaques are enunciated upon diverse sites of oral mucosa or the hard palate. Subsequently, cutaneous lesions may arise as oedematous papules upon the trunk, arm, axilla, nipple, groin or inguinal area [1,2]. Upon disease regression, cutaneous lesions characteristically demonstrate a brownish pigmentation whereas oral lesions appear decimated [3,4]. Of obscure aetiology, the condition represents as a neutrophil-mediated, altered muco-cutaneous reaction to a concomitant systemic disease process or may emerge as a cutaneous infection engendered by diverse pyogenic agents [3,4]. Also, concurrent immune-mediated mechanisms may engender the condition with the integration of identical pathogenic pathways. An aberrant immune response emerging during the clinical course of inflammatory bowel disease may initiate a cross reactivity of diverse cutaneous and gastrointestinal antigens with consequent muco-cutaneous manifestations [3,4]. Precise disease determination is contingent to manifestation of pathognomonic clinical pattern, peripheral eosinophilia, aseptic lesion exudate, typical histological features and an association with inflammatory bowel disease [3,4]. Clinical Elucidation Pyostomatitis vegetans manifests as circinate, mildly elevated, yellow-white mucosal lesions which predominantly incriminate diverse oral mucosal sites as the buccal mucosa, labial mucosa, gingivae or palatal mucosae. Commonly, the labial gingiva, soft palate, hard palate, buccal mucosa and labial mu-

cosa are implicated. The tongue, floor of the mouth, vagina, nasal mucosa or periocular mucosa is exceptionally implicated [3,4]. Oral malodorous ulcers are comprised of multiple, yellowish, exophytic pustules with a tender, erythematous inferior surface. Lesions may be disseminated within the gingiva, buccal mucosa or vestibular oral mucosa [3,4]. Lesions may be asymptomatic or associated with pain and tenderness. Incriminated subjects depict painful oral ulcers or accompanying nodules with an erosive superficial surface. Upon clinical examination, the lesions demonstrate a characteristic "snail-track" appearance [3,4]. Well defined, oral ulcers and erythematous erosions may appear upon the hard palate, buccal mucosa or lips wherein extraneous cutaneous lesions demonstrate crusting [3,4]. Multiple, yellow or whitish pustules appear situated upon an erythematous base and may rupture to configure a folded, fissured mucosal surface simulating a "snail-track" [3,4]. Generally, oral lesions are composed of multiple, miniature, yellowish pustules with a tender, erythematous inferior surface and a superimposed ulcerated buccal mucosa [5,6]. Upon clinical and morphological grounds, lesions may simulate pemphigus vegetans. Nevertheless, upon direct immunofluorescence or indirect immunofluorescence lesions are devoid of autoantibodies [5,6]. Cutaneous lesions of pyodermatitis vegetans, characterized by vesicular, pustular, exudative or vegetative plaques, appear concordant to oral lesions and generally involve the scalp, face, axillae or genital region [5,6]. Cutaneous lesions may precede or emerge subsequent to lesions of pyostomatitis vegetans. Nevertheless, pyostomatitis vegetans may appear as an indolent disease with an extensive clinical course, oral lesions and a lack of accompanying cutaneous lesions [5,6]. Histological Elucidation Upon microscopy, hyperplasia and moderate spongiosis of superimposed stratified squamous epithelium may be observed along with supra-basal clefts and multiple, intraepithelial abscesses imbued with eosinophils and neutrophils [5,6]. Morphological alterations are comprised of intraepithelial or subepithelial abscesses incorporating innumerable eosinophils and neutrophils. The dermis may demonstrate a mixed, chronic inflammatory infiltrate composed of lymphocytes and macrophages [5,6]. Intraepithelial clefts, acantholysis, accumulated eosinophils within spinous epithelial layer with the configuration of intraepithelial abscesses is exemplified. Subjacent connective tissue is infiltrated with a mixed, chronic inflammatory cell exudate [5,6]. The unique micro-abscesses are predominantly composed of eosinophils confined to the spinous layer of superimposed stratified squamous epithelium. Intraepithelial oedema may be variable and simulates an intraepithelial cleft [5,6]. The superimposed stratified squamous epithelial layer is associated with hyperkeratosis, acanthosis and demonstrates several reactive, multinucleated keratinocytes [5,6].

Figure 1: Pyostomatitis vegetans demonstrating yellowish-white pustules and ulcerated plaques with an erythematous base and a "snail track" appearance [9].

Figure 2: Pyostomatitis vegetans exhibiting an acanthotic stratified squmoue epithelium with sub-epithelial abscesses imbued with neutrophils and eosinophils [9].

Figure 3: Pyostomatitis vegetans enunciating a hyperkeratotic stratified squamous epithelium with acanthosis and sub-epidermal clefts packed with neutrophils and eosinophils and a mixed, chronic inflammatory cell infiltrate [10].

Figure 4: Pyostomatitis vegetans exemplifying a stratified squamous epithelial layer overlying abscesses incorporated with neutrophils and eosinophils and a chronic, dermal inflammatory infiltrate [10].

Figure 5: Pyostomatitis vegetans delineating a stratified squamous epithelial layer with acanthosis, hyperkeratosis and sub-epidermal abscesses incorporated with eosinophils and neutrophils and a chronic inflammatory dermal infiltrate [11].

Figure 6: Pyostomatitis vegetans depicting a stratified squamous epithelial layer with acanthosis, hyperkeratosis and intra-epidermal abscesses imbued with eosinophils and neutrophils along with a mixed, dermal, chronic inflammatory cell exudate [12].

Figure 7: Pyostomatitis vegetans displaying an acanthotic stratified squamous epithelial layer with intra-epidermal neutrophils and a subjacent, dermal chronic inflammatory infiltrate [13].

Differential Diagnosis Pyostomatitis vegetans requires a segregation from muco-cutaneous disorders associated with blister formation as pemphigus vulgaris which demonstrates a definitive cutaneous blister and is devoid of association with inflammatory bowel disease. Upon histology, pemphigus vulgaris demonstrates supra-basal clefts, row of tombstone with adherent basal layer, intra-epidermal vesicles with acantholytic keratinocytes, intra-epidermal eosinophils and Tzanck cells [2,4]. Pemphigus vegetans, especially the Neumann variant, is constituted of intra-epidermal vesicles with supra-basal acantholysis and a lack of eosinophilic micro-abscesses. Classically, Hallopeau variant of pemphigus vegetans enunciates eosinophilic spongiosis and micro-abscesses. The superimposed, hyperplastic stratified squamous epithelium demonstrates pseudo-epitheliomatous hyperplasia. Pemphigus may depict autoantibodies with direct immunofluorescence and indirect immunofluorescence [2,4]. Investigative Assay Pyostomatitis vegetans can be accurately determined by typical clinical manifestations as “snail-track” appearance, association with inflammatory bowel disease, peripheral eosinophilia and characteristic histological features as an intraepithelial abscess with significant eosinophils [7,8]. Lesions of pyostomatitis vegetans appear devoid of autoantibodies upon direct immunofluorescence and indirect immunofluorescence [7,8]. Therapeutic Options Pertinent, efficacious, standardized therapy of pyostomatitis vegetans remains obscure. Nevertheless, corticosteroids are contemplated to be an optimal, first line treatment strategy [7,8]. Appropriate management of pyostomatitis vegetans is contingent to precise assessment and alleviation of associated gastrointestinal disorder [7,8]. Oral lesions in the absence of gastrointestinal disease can be accurately treated with localized therapies as antiseptic mouthwashes

or topical corticosteroids. Systemic corticosteroids are beneficial in curtailing and resolving the lesions [7,8]. Systemic corticosteroids or topical corticosteroids appear advantageous in alleviating lesions which may manifest as verrucous, pigmented plaques [7,8]. Additionally, agents such as dapsone, azathioprine, mycophenolate mofetil, cyclosporine or infliximab may be efficaciously employed to treat the disorder [7,8]. Generally, pyostomatitis vegetans may resolve with the abatement of concordant inflammatory bowel disease. Adequate therapy of associated bowel disease ameliorates oral and cutaneous lesions of pyostomatitis vegetans. Occasionally, lesions may arise prior to detection of inflammatory bowel disease and appear responsive to topical corticosteroids. However, oral lesions may reappear following cessation of therapy [7,8].

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9. Image 1 and 2 Courtesy: JOMS.org

10. Image 3 and 4 Courtesy: Dermatology advisor

11. Image 5 Courtesy: JAAD.org

12. Image 6 Courtesy: ijd.org

13. Image 7 Courtesy: Wiley online library.