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Case Report

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VARIATION IN THE CLINICO-PATHOLOGIC BEHAVIOR OF ORAL CROHNS DISEASE: A CASE REPORT

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ABSTRACT

Crohns disease is poorly understood inflammatory bowel disease. It is characterized by accumulation of noncaseating granuloma formed as an inflammatory response to intestinal microbes. Clinical features include mucosal ulceration or erythemas associated with abdominal pain and diarrhea that often follow periods of relapse and remission. It occurs usually in 3rd to 5th decade of life with 60% individual presents early manifestation in oral cavity. The exact etiology is unclear but studies suggest that inflammatory pathway plays an intricate role in maintaining high levels of cytokine IFN-γ in disease progression. The author aims to highlight the case of oral crohns disease which has preceded before the emergence of gastrointestinal features in an adult patient.

KEYWORDS: Crohns Disease, Orofacial granulomatosis, cobblestone.

INTRODUCTION

Crohn's disease (CD) and ulcerative colitis (UC) are multisystem granulomatous disease primarily affecting the alimentary canal.[1] It was first reported by Burrill Crohn in 1932 as chronic entity involving intestine while association of oral symptoms was first outlined by Dudeney in 1969.[2] It has a bimodal age of distribution with first peak affecting early adulthood and second during 5th to 6th decade of life. It has been observed that the patient presented with CD may manifest initial features in the oral cavity which precede or occur concomitantly with lesions associated with gastric mucosa.[3] The common oral finding include swelling, linear ulcerations and cobblestone appearance of buccal mucosa while systemic features presents as abdominal pain, diarrhea, anemia, arthralgia and weight loss.[1,4] Histopathological features include non caseating granulomas, epitheliod shaped macrophages, multinucleated giant cells intermixed with lymphocytes and fibroblast.[5] Other oral and systemic conditions characterized by granulomatous inflammation must be excluded by appropriate investigations. Medical treatment focuses on the use of corticosteroid and immunosuppression to alleviate the symptoms while surgery is considered as a last resort to treatment.[6,7] The article presents a distinctive case of oral CD with widespread involvement of oral cavity in an adult.

CASE REPORT

A male patient aged 24 years reported to the outpatient department with the chief complaint of slow growing painless swelling of left side of the face since 6-7 months. Extraoral examination revealed diffuse swelling of left lower one third of face and gross enlargement of the lower lip with marked vertical fissures. Submandibular lymph node of ipsilateral side was slightly enlarged. (Fig.1A) Intraoral examination revealed multiple areas of soft, non-tendered, exophytic lesion involving marginal, interpapillary and attached gingiva extending to the mucogingival junction in relation to mandibular incisors. The overall appearance of gingiva was erythematous extending from the lingual surfaces of the mandibular teeth up to the distal surface of left mandibular 3rd molar region. Bleeding on probing was positive with grade I mobility in 31 and 41. The buccal mucosa of the involved side showed mucosal tagging surrounded by greyish white linear indentations forming cobblestone appearance. The overall oral hygiene status was poor with missing 46 (Fig.1B,C). On palpation, the lesion was soft in consistency without any

signs of surface ulceration. Intra oral periapical view revealed bone resorption in relation to alveolar crest of 31, 32, 41and 42. The patient's personal, family and medical histories were non-contributory. Considering the clinical features in view, the provisional diagnosis of periodontitis with orofacial granulomatosis was made.

Further, Hematological investigation was advised which included complete blood count, angiotensin converting enzyme and C-reactive protein levels in order to rule out the possibility of sarcoidosis. Chest X ray was also advised with Mantaux test to exclude tuberculosis.

The incisional biopsy was planned after improving oral hygiene status. Hematoxyllin and eosin (H/E) staining of tissue sections showed the presence of non caseating granuloma with epithelioid like cells interspersed with multiple giant cells. The perivascular lymphocytic infiltrate was also present (Fig.2A). This confirmed the granulomatous nature of lesion and the patient was referred to general physician for thorough gastrointestinal tract (GIT) checkup. Endoscopy was advised which revealed erythematous multiple polyps in GIT highlighting the inference of crohn's disease. (Fig.2B) With the above mentioned tests and results, the confirmative diagnosis of Oral crohn's disease was made and patient was prescribed triamcinolone acetonide 10mg intralesional injection in a tapering dose followed by topical application supplemented with micronutrients. Partial resolution of intraoral lesion was observed after 3 months of continuous treatment.

Table 1: Common Pathologies presenting in Oral cavity.[14,15,16]

	Diseases with granulomatous Histopathology	Oral Presentation	Diagnostic Methodology
1.	Oral Crohn's Disease	Diffuse lip and buccal swelling, cobblestoning of buccal mucosa, deep linear ulceration.	Gastroscopic evaluation, anti-Saccharomyces cerevisiae antibodies, Calprotectin, anti-neutrophil cytoplasmic antibodies (ANCA)
2.	Orofacial granulomatosis	Facial and lip swelling.	Nonspecific- Diagnosis based on exclusion
3.	Foreign body granulomatosis	Erythema, localized or generalized edema, pain and/or ulceration.	Nonspecific- Diagnosis based on etiology
4.	Sarcoidosis	Painless ulcers or nodules, generalized gingival enlargement, Salivary glands are affected, xerostomia, bilateral parotid swelling.	Angiotensin converting enzyme in blood, Urine calcium
5.	Tuberculosis	Painless ulcers in soft mucosa, gingival inflammation, hyperaemic, nodular or papillary proliferation of gingival tissues.	Chest X ray, Manotux test, PCR
6.	Mycotic infections	Ulcerative or vegetative lesion of oral mucosa, gingiva, tongue and palate.	Fungal Culture, Grocott-Gomori methenamine silver stain
7.	Syphilis-Tertiary	Gumma (palate and tongue) Mucopapular eruptions, Atrophic and interstitial glossitis.	Treponemal antibody testing, Rapid Plasma Reagin (RPR) or Venereal Disease Reference Laboratory (VDRL) tests.

DISCUSSION

CD is an intractable, immune mediated, multisystem disease manifesting initial signs and symptoms as orofacial granulomatosis. The first published case of CD was reported in 1970 in India showing peak age of occurrence in 3rd to 4th decade of life while the incidence rate in Indian subcontinent is still unknown due to lower number of reported cases.[8] In addition to this, it has been observed that more cases of CD are being diagnosed as compared to previous years with oral symptoms occurring in approximately in 60% of patients while 5-10% patients manifest the initial signs in oral cavity.[9]

The pathogenesis of CD is multifactorial in nature associated with complex interplay between genetic, immunologic and environmental factors. Epithelial barrier function, cell specific microbial responses, innate and adaptive immunity are targeted areas of research to understand the complexity of disease. It has been mentioned that commensal bacteria of intestine normally maintain gut homeostasis to repair the damage epithelial cells while they may also dysregulate immune pathways leading to inflammatory bowel diseases. The major success in understanding the pathogenesis was nailed with the association of defect in NOD2 gene which is expressed intracellularly in monocytes and paneth cells that trigger innate immunity through toll like receptor to activate NF-Kβ.[10]

The meta-analysis of genome wide association (GWA) studies traced more than 30 loci altered in inflammatory bowel disease (IBD) though few loci are common to both ulcerative colitis (UC) and CD. GWA studies revealed single nucleotide polymorphisms (SNPs) that affect ATG16L1 variant rs 2241880 to be closely linked to pathogenesis of intestinal CD in European study group. The alteration in Adenine to Guanine polymorphism

(T300A) further interacts with caspase 3 pathway which triggers TNF α resulting in autophagy of intestinal epithelial cells.[11] It is now known that abnormalities in ATG16L1 is not usually seen in studies conducted on Indian subgroups, though increased predisposition to disease is common on migration to other western countries.[8]

The clinical presentation is nonspecific characterized by nodular granulomatous swelling of buccal mucosa, oral cobblestoning, vertical fissures in lip, linear ulceration, angular cheilitis, lymphadenopathy, perioral erythema and rarely serpentine pustules coalescing in snail track pattern.[12]
Histologically, the oral sections show close resemblance to intestinal sections chiefly consisting of deep fissures in superficial mucosa and formation of noncaseating granuloma in connective tissue intermixed with multiple Langhan type giant cells and perivascular lymphocytic infiltrate. Presence of granuloma formation is a hallmark of OCD and other orofacial granulomatosis (OFG) diseases includes sarcoidosis, tuberculosis, foreign body reactions, pyostomatitis vegetans and Melkersson-Rosenthal syndrome. The diagnosis of OCD is based on exclusion criteria since terminology of OFG lacks specificity to any particular disease and poses a challenge to be differentiated from other clinical entities.[13] (Table-1).

Though the disease follow the phases of relapse and remission, localized surgical excision and medication like anti-inflammatory drugs like corticosteroids, immunosuppressants and immunomodulators are preferred choice of treatment to alleviate signs and symptoms.[3]

CONCLUSION

Conspicuous nature of granulomatous inflammation poses a great challenge to pathologist to diagnose and differentiate from other similar lesions. The caution should be exercised to carefully determine the underlying prevailing pathology involving other organs to conclude final diagnosis. These lesions demands thorough evaluation adjunct to routine histopathology for optimal management considering the frequent risk of relapse following treatment.

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