Volume 02, Issue 03 (May-June 2019), PP 21-24 www.ijmsdr.org
ISSN: 2581-902X

International Journal of Medical Science and Dental Research

Cannon's Disease a Rare Genodermatosis - Case Report with Brief Review of Literature

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Abstract: *White sponge nevus (WSN) is a rare hereditary dyskeratotic hyperplasia of mucous membranes. It is an autosomal dominant disorder. We report a case of WSN in a 18-year-old male with a history of familial involvement. A white patch with no erythema or other structural abnormalities was observed, which confirmed the diagnosis of WSN histopathologically.*

Key words: *Dyskeratosis, white lesion, white sponge nevus.*

I. Introduction and brief review of literature

One of the rare autosomal dominant genodermatosis affecting the oral mucosa in the majority of the cases is White sponge nevus (Cannon's disease, hereditary leukokeratosis of mucosa, white sponge nevus of Cannon, familial white folded dysplasia or oral epithelial nevus). The nasal, esophageal, rectal and vaginal mucosas are less frequently affected. [1,2] Hyde in 1909, first reported WSN [3],Cannon coined the term "white sponge nevus" in 1935.[4,5]This disorder is caused by a defect in the normal keratinization process of the oral mucosal epithelium. Mutation of the genes responsible for production of cytokeratins (CKs) 4 and 13, which are specifically expressed in the spinous cell layer of oral mucosal epithelium is the etiopathogeneis behind WSN.[2,5,6] This genetic disorder affects about one in 200,000 people, and the condition often shows variable expressivity and irregular penetrance.[7]

Lesions of WSN usually appear at birth or in early childhood, and it consists of symmetric, thickened, white, corrugated or velvety, diffuse plaques. Buccal mucosa is the most frequently affected, followed by the labial and gingival mucosa, and the floor of the mouth. The other extra-oral mucosal sites include nasal, esophageal, laryngeal, and anogenital mucosa. Patients are usually asymptomatic. [8]
The diagnosis of this disorder is important in that it must be differentiated from other congenital or familial disorders of more widespread clinical significance. The clinical appearance is so distinctive that biopsy is usually not necessary. The histopathologic features of WSN are characteristic but not necessarily pathognomonic. Prominent hyperparakeratosis and marked acanthosis with clearing of the cytoplasm of the cells in the spinous layer are common features. In some instances an eosinophilic condensation is noted in the perinuclear region of the cells in the superficial layers of the epithelium, a feature that is unique to WSN. [9, 10]

II. Case report

Herein, we have aimed to present an 18-year-old male patient who reported to the Outpatient Department of the Dental College, with a chief complaint of white patches in his mouth since early childhood. Patient gives history that he himself noticed the white patches while brushing his teeth and has problem with esthetics while speaking and no other associated symptoms.

The patient's past history revealed that the lesions were present since early childhood. The family history revealed that similar lesions were present in the mother (aged 40 years) and the elder brother (aged 21 years) of the two siblings.

On clinical examination, there were bilateral, symmetrical white plaques and patches with a corrugated surface on the bilateral buccal and labial mucosa. The plaques were smooth with velvety texture and irregular, well-defined borders. There was no elevation or erythema. The margins were clear. Oral hygiene was good. The other oral structures were normal in appearance. The plaques extended from the retro-commissural area anteriorly to retromolar region posteriorly. The lesions could not be removed by scraping and were asymptomatic. The patient was referred to the Departments of Ophthalmology and Obstetrics/Gynecology to rule out the presence of any conjunctival and genital lesion, respectively. No other sites were identified. . No other contributing factors, such as tobacco use or chemical burn, could be elicited. Considering above clinical features and findings a provisional diagnosis of white sponge nevus was given, differential diagnosis of dyskeratosis congenital, pavhyonychia congenital, xeroderma pigmentosum, hereditary benign intraepithelial dyskeratosis was given.

A biopsy was advised to rule out other lesions. On histopathological examination of the H and E stained soft tissue section revealed hyperplastic parakeratinised stratified squamous epithelium showing braid and blunt reteridges, acanthosis, parakeratotic plugging extending into the prickle cell layer and there was evidence of intra cellular edema and pyknotic nuclei in parakeratotic layer giving basket weave appearence. the underlying connective tissue showed moderately dense collagen bundles, few endothelial lined blood vessels and muscle.

Based on clinical data and histopathologic findings, the lesion was consistent with white sponge nevus. Because of benign nature of this lesion, no treatment is necessary and only biopsy and correct diagnosis is necessary to rule out other similar lesions.

Figures

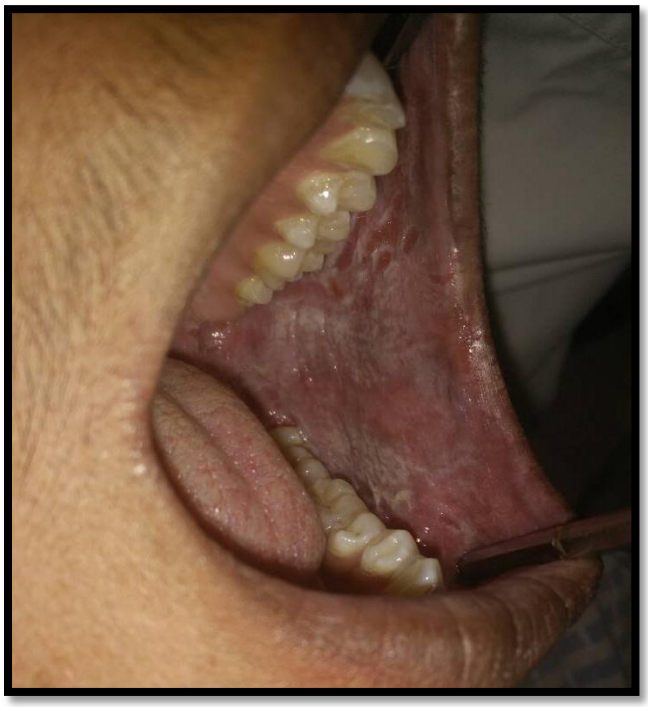

Figure 1: Left buccal mucosa

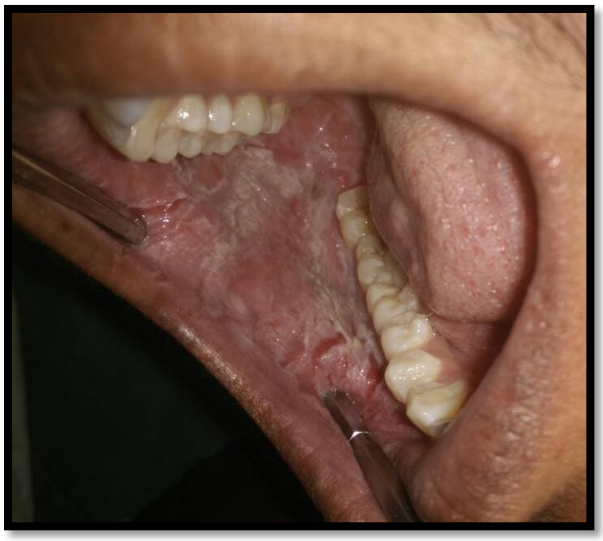

Figure 2: Right buccal mucosa

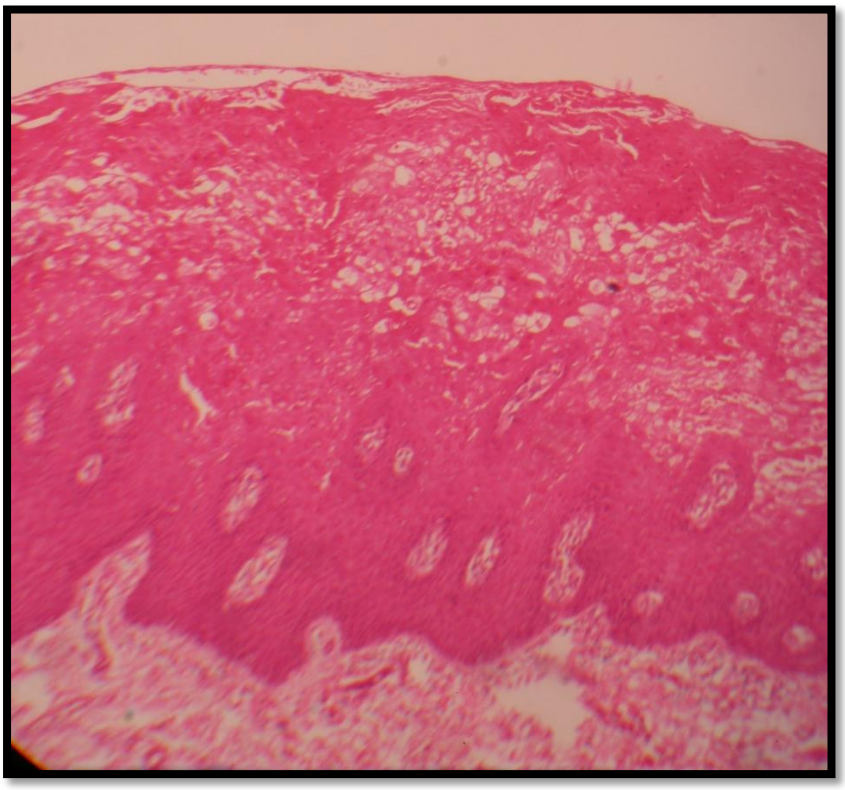

Figure 3: Histopathology

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