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A white beauty: A case report of white sponge nevus of oral cavity

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Abstract
White sponge nevus is a developmental and a rare cutaneous and mucosal lesion. It is an autosomal dominant disorder with variable penetrance. We report a case where a white smooth plaque with no erythema or other structural abnormalities was observed. White sponge nevus was also known with names like cannon's disease etc.

Keywords: white sponge nevus, smooth plaques, cannons disease, autosomal dominant, oral mucosa

Introduction
White sponge nevus (WSN) is a rare oral mucosal lesion. Hyde first described in 1909, but Cannon coined the term in 1935 [1]. This entity is also known as Cannon's disease, familial white folded dysplasia, hereditary leukokeratosis, white gingivostomatitis, and exfoliative Leukoedema [2]. It is characterized as benign leukokeratotic lesion of early onset with periods of remission and exacerbation. It is usually asymptomatic, although pruritus, burning, and pain have been reported following irritating stimuli [3].

WSN represents as a gray-white spongy plaque, which in full expression may be thick and deeply folded. It most frequently affects the oral mucosa, although the nasal cavity, esophagus, vagina and rectum also may be involved [3]. Instances of de novo mutations also have been reported [4]. Here we report a case of WSN in a seventeen year old female who suffers from type 1 diabetes & with no familial involvement.

Case Report
A female patient aged 17yrs reported to our dental outpatient department with a chief complaint of pain in the upper left back tooth region since 1 month. Patient gave a medical history of having type 1 diabetes since twelve years and on medication, On examination, bilateral, soft, white and "spongy" plaques was noticed on the buccal & labial mucosa, dorsum of the tongue and floor of the mouth. These could not be removed on scraping. The surface appeared smooth, thickened, folded & corrugated with velvety texture having a peculiar opalescent hue & were irregular in shape. No lymph nodes were palpable. Multiple carious teeth were present. The patient denied the presence of a similar condition in her immediate family members.

Biopsy could not be performed due to patients parents and patients unwillingness. Also keeping in mind the outcome of the diagnosis does not overrule the trauma caused as a consequence of a biopsy as it was asymptomatic. Therefore, diagnosis was restricted to purely clinical data, the lesion was diagnosed with white sponge nevus. Because of the benign nature of the lesion, no treatment was necessary.

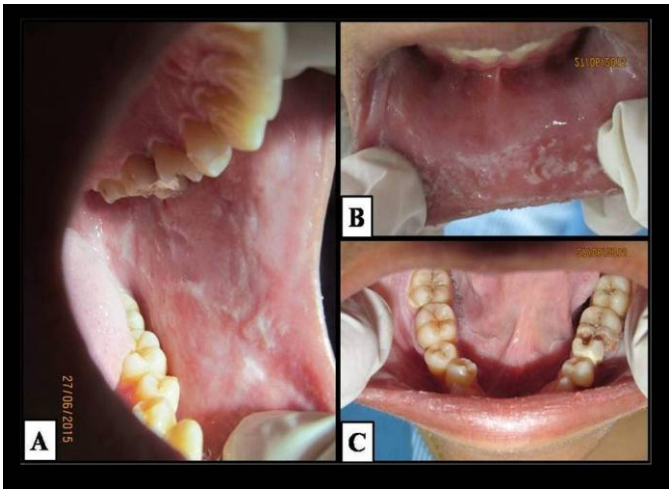

Fig 1: Presence of white spongy plaques seen in Left buccal mucosa (A) Labial mucosa (B) Floor of the mouth (C)

Discussion
Women are more affected than men by a ratio of 3:1. WSN is inherited as an autosomal dominant trait with wide variability of expression and high penetrance. Histologically, the lesion is confined to the epithelial layer, which appears parakeratotic and acanthotic with pyknotic cell nuclei. Cellular vacuolization may affect all layers of the epithelium or be distributed irregularly [5]. The basement membrane remains intact, although a mild inflammatory reaction may be present in the submucosa. This finding, aids in differentiating this lesion from leukoplakia or lichen planus.

Differential diagnosis should include pachyonychia congenita, dyskeratosis congenita, hereditary benign intraepithelial dyskeratosis, hypertrophic lichen planus, leukoplakia, and traumatic keratoses. A white smooth plaque with no erythema

or other structural abnormalities was observed, thus we arrived at the diagnosis of WSN.

WSN is considered a rare disorder, affecting one in 200,000 people. Some authors claim that the condition is related to mutations in *K4* and *K13* genes, characterized by defects in the maturation and desquamation of epithelial cells [6]. It is characterized by white or grey thickening or folds of the mucosa which appear occasionally oedematous. When unfolded the lesions do not loose thickness as seen in Leukoedema [6]. All members of the family are usually affected which is not in concordance with our finding. Cases without familial background have been reported [7]. In this case, none of the family members had similar lesions suggestive of not having a family predilection. Onset is in early childhood with about 50% cases being diagnosed before the age of 20 which in this case was noticed at the age of 17. This lesion appeared early in life without any reported changes throughout the patient's life.

This lesion appears at birth or in early childhood which was reported in our case. Neither gender nor racial predilection exists [1 & 7]. WSN is highly variable, as well as the size of the plaques and their distribution in the oral mucosa.

Differential diagnosis can also include cheek-biting, which in this case is denied by the patient, in addition to it, there was absence of any sharp cusps [7]. In the absence of nail changes and palmoplantar hyperkeratosis diagnosis of pachyionichia congenita was rejected. Chemical and temperature burns, leukoplakia, may also have the appearence of white sponge nevus. But these are denied as the patient does not give a history which can favour these.

Although benign in nature and not needing treatment, recent studies have proposed the use of topical tetracyclines for reduction of the lesions, pointing out that the oral microflora could stimulate the appearance of the hyperkeratosis [8, 9]. It should be emphasized that correct diagnosis of white sponge nevus should be established while many other possible "white" lesions could have malignant potential. WSN is usually present since childhood & without any signs and symptoms.

Conclusion

In our experience, we do not recommend any medication as the patient had not mentioned any kind of discomfort or issues regarding it. Moreover the patient was not even aware of the condition making it easier for us to understand that her condition was completely asymptomatic. Proper clinical examination and history helps in providing a better treatment. Oral hygiene maintaining methods are to be explained to the patient as well.

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