White Sponge Nevus: A Case Report and Clinical Update on the Diagnosis

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Abstract

Hereditary white lesions are a rare occurrence in the oral cavity. These lesions are usually harmless and benign. But these lesions resemble the other pathological oral white lesions having the risk of malignant transformation. The diagnosticians should be aware of the existence of these benign white lesions to avoid misdiagnosis and timely intervention for potentially malignant disorders. We have reported the occurrence of white sponge nevus in a young male patient and his mother. The patient's family history, complete clinical examination, and histological report validated the clinical diagnosis.

Keywords: Diagnosis, leukokeratosis, white sponge nevus

Introduction

The white lesions of the oral cavity vary from reactive, hereditary, or benign lesions to oral potentially malignant disorders. Diagnosing these lesions usually presents a challenge for dental practitioners because of their remarkable resemblance to each other.[1]

Detailed case history and good clinical examination can give a clue to the clinical diagnosis of the condition that can later be confirmed histopathologically. White sponge nevus (WSN) is a hereditary benign disorder that occurs because of the mutation in *Keratin 4* and *13* genes leading to a defect in the keratinization process.[2]

This disease is transmitted by the autosomal mode of inheritance, as seen in the present case.[1] This paper presents a case of familial transmission (four members) of WSN in the oral cavity and a clinical update on differentiating it from other potentially malignant disorders.

Case Report

Clinical findings

A 16-year-old boy accompanied by his mother visited the oral health center complaining of uneven, rough surfaces with whitish discoloration in the mouth since childhood. The patient was a nonsmoker with no significant medical or dental history. The patient's mother (41 years) and one sibling (22 years) also have similar lesions in their mouth. Further investigating, the patient's mother revealed that her father (67 years) also has similar patches in his mouth [Figure 1]. However, the lesions were not associated with a burning sensation or difficulty chewing or speech [Table 1].

Intraoral examination showed the presence of bilateral, leathery, thick white lesions. It is crumpled with ill-defined borders, covering the entire oral mucosa, excluding the gingiva, and floor of the mouth [Figures 2 and 3]. On palpation, the lesions were non-scrapable, leathery in consistency, rough and thick in texture, and did not disappear on stretching. The patient's mother had similar white, plaque-like lesions in her mouth with no skin lesions.

Diagnostic assessment

Because the lesions had been present from childhood, a provisional diagnosis of WSN was given. Furthermore,

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hereditary benign intraepithelial dyskeratosis, dyskeratosis congenita, and pachyonychia congenita were the clinical differential diagnosis. Incisional biopsy confirmed the diagnosis of WSN in the patient with cytoplasmic clearing and perinuclear condensation of cells in superficial layers.

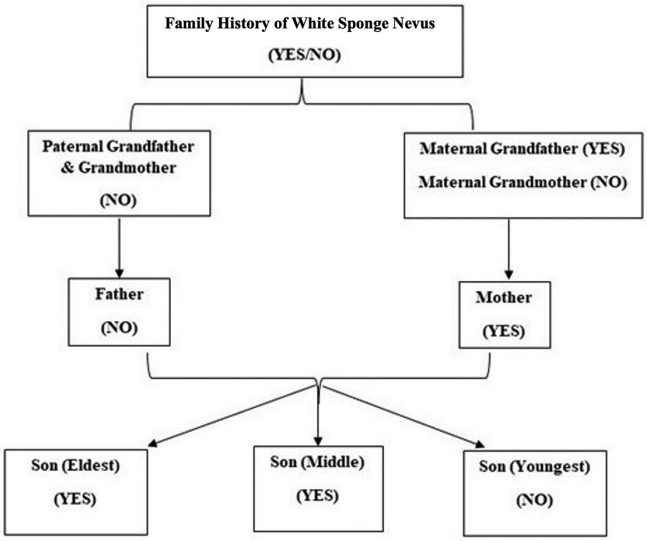

Figure 1: Family pedigree chart shows the presence or absence of the lesions

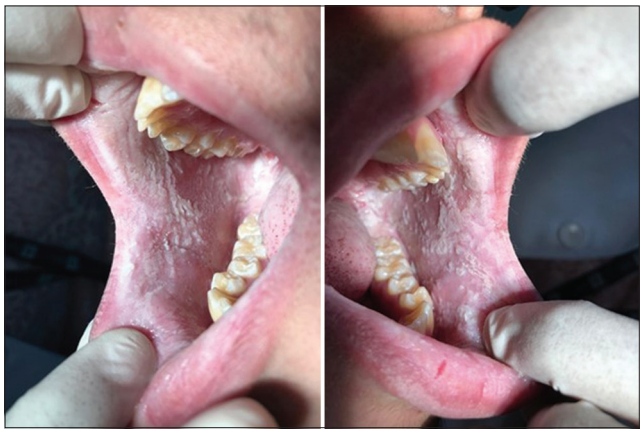

Figure 2: Intraoral examination shows white lesions on the right and left buccal mucosa

Therapeutic intervention

The patient was reassured that it is a hereditary lesion and will gradually regress over a period. No intervention was done for the patient.

Follow-up and outcome

The patient has been kept under regular follow-up for his oral conditions and was evaluated after 2 months and 6 months. No significant changes were reported in the patient's oral lesions.

Discussion

WSN is the rare, hereditary, non-scrapable white lesion affecting the mucosal surfaces. The detailed report of the first case of WSN was submitted in 1935 by Cannon.[3-5]

WSN is considered a rare pathology with an incidence of 1 in 200,000 and no gender predilection. Most cases have been reported either at birth or in childhood, although there are reports of the occurrence of WSN in adolescents.[6]

WSN presents as the whitish plaque-like lesions occurring bilaterally in the oral cavity involving the entire mucosa.[3] Sometimes, they represent a "cracked mud appearance," as seen in the cases of homogenous leukoplakia. The lesions are painless, non-scrapable, and do not disappear on stretching like leukoedema.[4,7] However, the clinical expression of this disease depends on the severity of the condition.

Candidiasis and lichen planus can be considered a differential diagnosis in the milder form of the disease. Proliferative verrucous leukoplakia can be excluded by the history of tobacco smoking

Table 1: Timeline of history

Age of patient	16 years
Uneven, rough surfaces with whitish discoloration in the mouth	Since childhood
Any associated symptoms such as difficulty in speech and burning sensation.	No
Any previous therapeutic interventions	No
The patient reported to OPD.	February 2020
Incisional biopsy	March 2020
Follow-up	May 2020 and September 2020

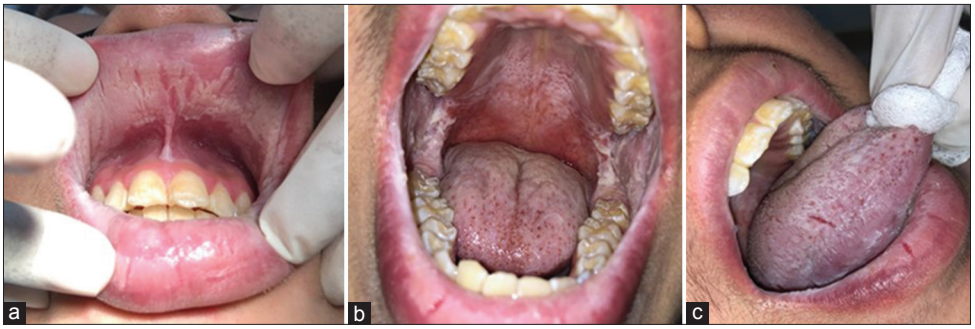

Figure 3: (a) Hyperkeratosis of upper labial mucosa (b) white plaques on the palate and retromolar pad area (c) diffused white lesions on the lateral border of the tongue

and more preference in older people. Cheek and lip biting can also be excluded from the patients' personal history. Dyskeratosis congenita is associated with other oral manifestations such as caries risk, short, blunted roots, hypodontia, and gingival inflammation, unlike WSN.[7,8] Pachyonychia congenita is associated with painful and dystrophic nails along with the leukokeratosis of oral mucosa and palmoplantar keratosis in the areas of friction. Hereditary benign intraepithelial dyskeratosis is associated with the white plaques in the bulbar conjunctiva along with the hyperkeratosis of the oral mucosa.[7]

The histopathological confirmation is not required in all cases of WSN because of its classical clinical appearance. However, cytoplasmic clearance and perinuclear condensation of the epithelial cells can be seen in WSN, which can help distinguish WSN from other lesions.

No specific treatment is needed for this condition, as it does not cause any symptoms in the patients. However, numerous therapeutic and surgical interventions have been tried to reduce the severity of this lesion.[8]

Further case studies are need to be done to reveal the different epidemiological patterns of this disease and various treatment interventions for WSN.

Conclusion

Early clinical diagnosis of these white lesions must differentiate them from other critical pathological lesions of the oral cavity.

Declaration of patient consent

The authors obtained the patient consent. The mother consented to process her son's and herself's data for research and publication without disclosing her identity.

Key message

White sponge nevus (WSN) is a rare white lesion in the oral cavity. Its significant resemblance to oral potentially malignant disorders usually leads to misdiagnosis. This report illustrates the rarity of WSN and its clinical differential diagnosis to avoid unnecessary interventions.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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