White sponge nevus in the oral cavity: case report and a review of literature

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Abstract

Introduction
White sponge nevus (WSN) is an uncommon white lesion mainly affecting the oral mucosa. This is an autosomal dominant hereditary disease with mutations in Keratin 4 and 13 genes. Due to its variable penetrance sporadic cases are also reported rarely without any familial involvement. Usually WSN exhibits a benign course and can be managed conservatively.

Case report:
A 28-year-old boy presented with white spongy plaques on bilateral buccal mucosa, labial mucosa, floor of the mouth and tongue which was clinically suggestive of white spongy nevus. A histopathological examination confirmed the diagnosis and this case was managed conservatively.

Conclusion:
Our case report reinforces the suggestion that histopathological investigations are mandatory for the diagnosis and to exclude possible potentially malignant disorders. Further proper diagnosis will prevent over treatment of this condition. This is especially important when there is no familial involvement.

Key words: White Spongy nevus, Cannon disease, Oral Mucosa, White plaques, Case reports.

Introduction
White sponge nevus (WSN) is an inherited disease that produces a white, bilateral, keratinized appearance typically appearing on the buccal mucosa. This benign rare disorder was initially described by Hyde in 1909 and coined by Canon in 1935. Many synonyms are used in literature to identify this condition which include: Hyde-Cannon’s disease, familial white folded dysplasia, hereditary leukokeratosis, white gingivostomatitis, and exfoliative leukoedema.

WSN commonly affects the non keratinized mucosa of the oral cavity resulting in development of multiple nevi as an attribute to a defect in the normal keratinisation. These keratotic mucosal alterations may be seen on extra oral sites are reported which include mucosae of nose, oesophagus, genitals, and rectum. Classically these patients present with painless white spongy plaques on the bilateral buccal mucosa followed by tongue, floor of the mouth and alveolar...
mucosa\(^3\). The onset of the disease is in the early childhood, but is discovered incidentally at a later age due to lack of symptoms\(^4\). Although, this is known to be a genetic disorder, instances of de novo mutations also have been reported\(^5\).

Even though, clinical feature are not pathognomonic, diagnosis of this condition is important as it must be differentiated from other congenital or familial disorders of more widespread clinical significance. Histopathological investigations are necessary to confirm the diagnosis as similar microscopic findings may be associated with leukoedema and hereditary benign intraepithelial dyskeratosis however; microscopic features will aid to exclude other potentially malignant white lesions\(^6\). Further, correct diagnosis of this condition will prevent over treatment of this condition which is otherwise managed conservatively with very good prognosis.

A case of WSN in a healthy Asian male with no history of familial involvement is described.

**Case report**

An otherwise healthy male patient aged 28 year presented to the Oral Medicine Clinic, University Dental Hospital Peradeniya, Sri Lanka complaining of symptomatic soft white plaques on bilateral cheeks and tongue for 5 years duration. Patient denied the habits of betel chewing, alcoholism or smoking. On examination both upper and lower lips appeared depigmented and scaly. Intra orally white “spongy” plaque like lesions was noticed on bilateral buccal mucosa, dorsal and ventral surface of the tongue and on floor of the mouth which could not be removed by scraping (Figure 1). There were no extra oral lesions. Patient denied the presence of similar lesions among immediate family members.

![Figure 1](image1.png)

**Figure 1.** (A) Depigmented lips (B) white spongy plaques on right buccal mucosa (C) white spongy plaques on right lateral border of the tongue (D) white spongy plaques on ventral surface of the tongue
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Microscopic examination of the soft tissues taken from the buccal mucosa (Figure 2A & 2B) revealed marked hyperkeratosis and acanthosis of the stratified squamous epithelium together with perinuclear eosinophilic condensation and cytoplasmic clearing in keratinocytes. These histopathological features, taken in conjunction with the clinical findings, were consistent with a definitive diagnosis of white spongy nevus (WSN).

Patient was reassured and as he was asymptomatic only antiseptic mouth wash (0.2% chlorhexidine gluconate) was prescribed and was followed in six months intervals.

Discussion

WSN is a rare genetic disorder exhibiting autosomal dominant trait with wide variability of expression and penetrance and hence familial reports are not very common, as this case present here. This may also occur as a result of sporadic mutation. Lesions of WSN usually appear at birth or in early childhood, but sometimes the condition develops during adolescence. The reported prevalence rate is below 1 in 200,000.

Mutations in the KRT4 or KRT13 gene cause WSN. The keratin 4 protein (produced from the KRT4 gene) and the keratin 13 protein (produced from the KRT13 gene) partner together to form molecules known as intermediate filaments. The function of these filaments is to protect the mucosae from being damaged by friction or other everyday physical stresses by provide strength and resilience. Keratin 4 and 13 proteins are only produced in the oral, nasal, oesophageal and anogenital mucosae. Mutations in the KRT4 or KRT13 gene disrupt the structure of the keratin protein. This altered structure of keratin 4 and 13 results in easy damage of the mucosa with little friction or trauma. Damage to intermediate filaments leads to inflammation and promotes the abnormal growth and division (proliferation) of basal cells of epithelium, causing the mucosal hyperkeratosis and acanthosis resulting in white sponge nevus.

Diagnosis of WSN is challenging and many other different types of white lesions should be considered in the differential diagnosis. There is a need for precise identification through prompt histopathological examination to differentiate this condition from more serious, potentially premalignant lesions as well as other genodermatoses. The differential diagnosis should include hereditary benign epithelial dyskeratosis (Witkop’s disease), Oral lichen planus/ lichenoid drug reaction, lupus erythematosus, cheek biting and possibly candidiasis. Further, pachyonychia congenita can be excluded by lack of extra oral skin lesions (thickened nails and hyperkeratotic palms & soles). Histopathology of Witkop’s disease is identical to the WSN except in

Figure 2. (A) Hyperkeratosis and Spongiosis (B) nuclear clearing and perinuclear condensation of keratin (H&E100)
Witkop’s disease scattered spinous cells show premature keratinisation and pyknosis of nuclei. Differentiation of these two conditions is very important as Witkop’s disease may end up with blindness due to formation of gelatinous plaques on bulbar conjunctiva. Pseudomembranous candidiasis and chronic hyperplastic candidiasis can often be confused with WSN. But unresponsiveness to antifungal treatment, fungal examination and histopathological features help to differentiate these. Although cheek biting, chemical burns, smoking have similar clinical features they can be excluded by good history taking. Clinically, on examination the disappearance of the grey appearance upon stretching will differentiate Leukoedema from WSN. Lichen planus may look similar, but the age of onset and the presence of white reticular lesions will help the diagnosis clinically whereas histopathologically also these can be differentiated. Anyhow, biopsy is mandatory for diagnosis and to exclude the possible premalignant disorders such as leukoplakia and oral submucous fibrosis.

Usually WSN is asymptomatic, unless super infection is present. The corrugated plaques of WSN easily harbour these microorganisms and provide the best conditions for colonization. Maintenance of good oral hygiene, daily use of an antiseptic mouthwash is recommended for these patients to avoid such super infections. During acute severe exacerbations topical as well as systemic antibiotic treatment is reported in the literature. Topical tetracycline 0.25% or 1% has been used with good symptomatic control.

Failed attempts of surgical excision of the lesions using laser are also reported in the literature. This may be due to its genetic based pathogenesis. There is no definitive and standard treatment for WSN yet. Lesions may persist throughout the life but is completely benign in nature.

**Conclusion**

We report a case of WSN with no familial involvement. The clinical and histopathological findings of this case are similar to the literature. Correct diagnosis of the condition is important to exclude the other possible premalignant disorders. Although patients are asymptomatic good oral hygiene should be maintained to prevent bacterial and fungal super infections.

**References**


