White Sponge Nevus: Report of a Case and Review of the Literature

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Abstract- White sponge nevus is a rare benign autosomal dominant disorder with variable penetrance. It is characterized by asymptomatic white plaques affecting mainly the oral mucosa. Careful clinical and histopathological examination is indicated to exclude other more serious conditions presenting as oral white lesions. Herein, we present a new case of oral white sponge nevus in a 17-year-old Iranian male with no familial background.

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Keywords: Oral mucosa; White sponge nevus; White lesions

Introduction

White sponge nevus (WSN) is a rare autosomal dominant hereditary disorder characterized by white thickened spongy plaques of the mucous membranes. It most commonly affects the oral mucosa, but extraoral mucosal involvement (mostly anogenital, esophageal and nasal) has also been reported. The disorder is usually congenital with no gender predilection but occasionally does not appear until adolescence. Typical lesions of WSN are asymptomatic thickened white plaques that bilaterally affect buccal, labial, and gingival mucosa as well as the floor of the mouth. The surface of the plaques may peel away from the underlying tissues (1). Histopathological examination of the lesions usually reveals hyperproliferation of the affected epithelium, intracellular edema of the spinous layer, and keratin tonofilaments aggregation in the form of perinuclear eosinophilic condensation (2). Mutations of keratin 4 and/or keratin 13 genes have been claimed to be linked to the development of WSN (3-8). These two keratins are specifically expressed in the spinous layer of the oral, esophageal, anogenital, and other non-cornified stratified squamous epithelia affected by the disorder (5). It is important to diagnose WSN correctly because there are several more serious pathologic conditions, some with malignant potential, which may resemble this benign condition (9). There seems to be paucity of literature on WSN from Iran. We herein present a case of WSN in a male patient which is just the second case of this rare disorder in the Iranian literature (10).

Case Report

A 17-year-old non-smoker Iranian male presented to the Department of Dermatology at Hamadan University of Medical Sciences with the chief complaint of lifelong widespread white lesions in the oral cavity. There was no immediate family member affected by the same lesions. His general health was good, and he did not have similar lesions elsewhere on the body. He was almost symptom-free except for episodic burning sensations when eating spicy food.

On physical examination, there were white velvety plaques with symmetric distribution on the buccal and labial mucosa, as well as the floor of the mouth and palate (Figure 1a,b).

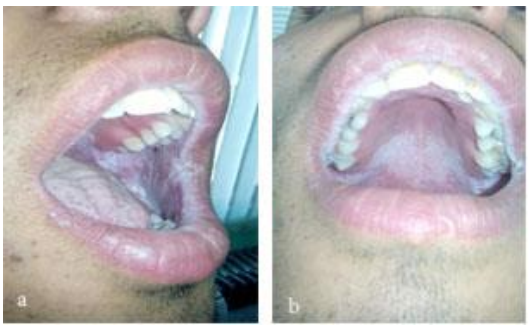

Figure 1. White velvety plaques on the buccal and labial mucosa (a) and palate (b)

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The plaques had irregular, but well-defined borders and no associated erythema were present. No lymphadenopathy was detected. Based on history and physical examination, his oral lesions were diagnosed as WSN, and histopathological examination was performed to confirm the diagnosis. Superficial parakeratosis, acanthosis, intracellular and intercellular edema, as well as eosinophilic condensation in the perinuclear space of some superficial spinous layer cells were revealed. Basal layer was intact, and a minimal lymphocytic infiltration was present in the stroma (Figure 2a,b).

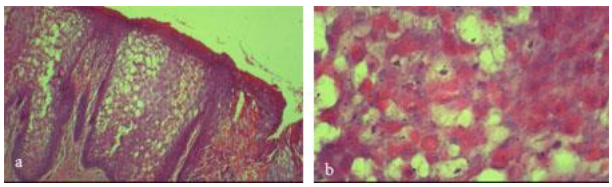

Figure 2. Parakeratosis, acanthosis, intracellular and intercellular edema (a) and eosinophilic condensation in the perinuclear space of superficial spinous layer cells (b)

Histopathological findings along with the clinical data supported the diagnosis of WSN, and because of the benign and almost asymptomatic nature of the disease, no medication was prescribed.

Discussion

WSN is considered a rare hereditary disorder which affects one in 200 000 people (11). Since the mode of transmission is autosomal dominant with variable penetrance, several non-familial cases have also been reported so far (12-14). Although mutations of keratin 4 and keratin 13 genes have been attributed to the development of WSN, Liu *et al*., have recently compared sporadic and familial cases with WSN and observed that only one of the five sporadic cases had keratin mutation (15). Since the characteristic clinical presentation of WSN is that of white plaques in the oral cavity, it should be differentiated from other white lesions such as oral leukoplakia, focal epithelial hyperplasia (Heck's disease), proliferative verrucous leukoplakia, and even squamous cell carcinoma. Oral lichen planus is especially difficult to be differentiated from WSN. However development of this disorder is quite uncommon in young people, in contrast to WSN. Oral candidiasis could also be excluded by fungal examination and unresponsiveness to antifungal agents. Oral lesions of Pachyonychia congenita, Darier disease, hereditary benign intraepithelial dyskeratosis, and Dyskeratosis congenita may also be confused with WSN, but other associated clinical manifestations of these disorders help to differentiate them from WSN.

A biopsy is usually indicated to distinguish lesions of WNS from the above mentioned conditions. Histopathological examination of WSN lesions typically reveals a hyperparakeratotic and thickened epithelium as well as vacuolization of the spinous layer cells. Perinuclear eosinophilic condensation of high Malpighian cells is a feature that has been claimed to be characteristic to WSN, although not pathognomonic (16).

Some investigators have hypothesized that viral, bacterial, and even fungal infections may contribute to the expression of WSN lesions (17-19). This idea is supported by the improvement of these lesions in some patients treated with antimicrobial medications such as tetracycline and chlorhexidine mouthwashes (20-22). Nonetheless, the disease runs a benign course, and no treatment is usually required. Development of squamous cell carcinoma within a white sponge nevus has been reported in the literature, but this malignant transformation took place in the setting of chronic prednisone use and no other similar reports exist (23).

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