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White Sponge Nevus: A Non-hereditery Presentation

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Abstract

White sponge nevus is a rare, autosomal-dominant disorder that affects the uncornified stratified squamous epithelia. Clinically, the presence of white, spongy plaques mostly in the buccal, labial, and gingival mucosa and the floor of the mouth characterize the lesions. The differential diagnosis of the lesion may be difficult and it is best diagnosed by biopsy. No standard treatment for the condition exists although numerous treatments have been tried. We report a case of white sponge nevus in the oral cavity of a 28-year-old man and review of the literature.

Keywords: Dyskeratosis, white lesion, white sponge nevus

Introduction

White sponge nevus (WSN) is an uncommon disease that Hyde first described in 1909, but Cannon coined the term in 1935. This entity is also known by other names: Cannon's disease, familial white folded dysplasia, hereditary leukokeratosis, white gingivostomatitis, and exfoliative leukoedema (1-2).

WSN is a rare pathology with a genetic based pathogenesis, a benign course and a localization affecting the mucosal membranes. The disorder may be detected in early childhood.

Lesions are easily recognized and clinically valuable: they appear as bilateral white spongy plaques, typically found on the cheek mucosa, and the patients dont complain about pain (3). Mucosal alteration usually affects oral soft tissues, but it sometime involves vaginal and rectal mucosa (2).

Correct diagnosis of WSN, which is a benign condition, should be established because other possible "white" lesions could have malignant potential. The histological findings are characteristic but not pathognomonic. Histopathologic features of WSN include epithelial thickening, hyperparakeratosis, and vacuolization of the keratinocytes in the suprabasal layers. In addition, compact cytokeratin (CK) aggregates are visible in the upper epithelial layers, such as those found in epidermal disorders associated with CK defects (4-5).

WSN typically affects several individuals in a same family, further confirming its autosomal domi- nant transmission (6-7).

We herein present a case of WSN of the oral cavity in a patient with no history of familial involvement.

Case report

A 28-year-old man presented with a 20 year history of asymptomatic, white, folded, soft, diffuse plaques bilaterally on his cheek, labial

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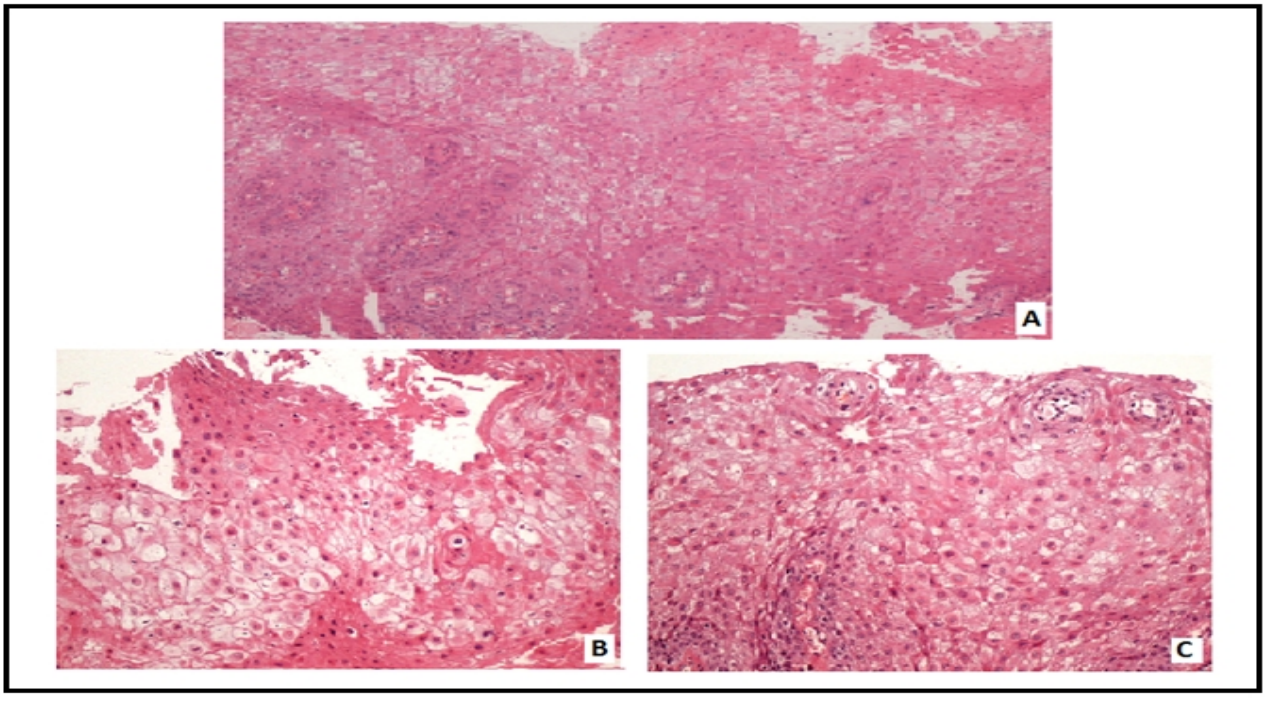
mucosa and lateral surfaces of his tongue. Lesions could not be removed. The margins were well defined, and no lymph nodes were palpable. Oral hygiene was adequate and oral examination was normal. Lesions never changed despite numerous interventions such as vitamin A and antibiotic therapy. The patient doesn't smoke and rarely consumed alcohol. He had seen a dentist who referred him to our Oral Medicine and Surgery Clinic. He was told he had leukoplakia or oral cancer. There was no smilar oral lesions in any other family members. No lesions in other body sites were reported. A punch biopsy was obtained from his buccal mucosa. Histopathologic evaluation revealed an, oral mucosa covered by stratified squamous epithelium with prominent hyperparakeratosis and marked acanthosis. Cytoplasmic clearing of the keratinocytes was detected. Underlying connective tissue was normal in appearance with rare chronic inflammatory cells. The lesions were painless. Un-esthetic appearance of the mucosa was the only complaint of the patient. Blood analysis and salivary flow rate showed no anomalies. The saliva analysis didn't show the presence of Candida albicans or other fungal infectious agents. Based on clinical and histopathologic findings, the lesion was consistent with WSN. Patient's saliva was examined in diagnostic oral microbiology laboratory: the analysis revealed the presence of Staphylococus aureus.

In the following days 2 daily rinses with mouthwash containing chlorhexidine digluconate at 0,2% was prescribed in order to decrease the bacteria. After one month, mild improvement was observed in the lesions. Six-month follow-up was recommended.

Figure 1. A: shows the histopathologic wiew of the lesion: marked epithelial thickening with spongiosis (HEx100). B, C: show the clear cell change and characteristic perinuclear condensation of keratin (HEX400). Figure 2. A,B,C,D,E show the clinical pictures of the patient.



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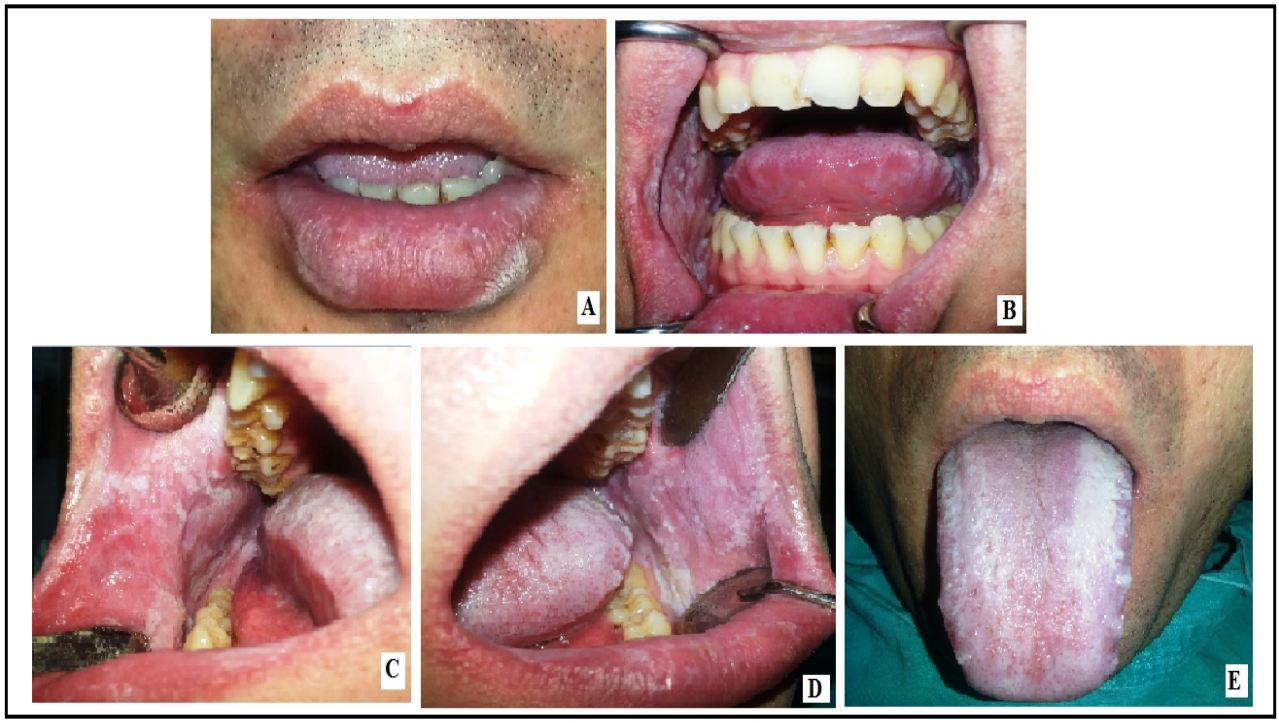

Figure 2. A,B,C,D,E show the clinical pictures of the patient.

Discussion

WSN is considered a rare disorder, affecting one in 200,000 people (8). The onset is usually during early infancy, often before 20 years, and there is no gender predilection (2). 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 (9-10). Lesions usually occur with significant predilection for the cheek mucosa, followed by the ventral surface of the tongue, labial mucosa, the alveolar ridge and floor of the mouth (11). As seen in our patient the absence of pain is an important clinical feature in these patients (12).

Other conditions presenting as white lesions on the oral mucosa was taken into account in the differantial diagnosis. These include genodermatoses and acquired conditions such as leukoedema, linea alba, bitten mucosa, dyskeratosis congenita (DKC), pachyonychia congenita focal epithelial hyperplasia (Heck disease), systemic lupus erythematosus (SLE), vegetative pioestomatitis, proliferative verrucous leukoplakia (PVL), oral florid papillomatosis, mucosal syphilis (mucous plaques), candidiasis, leukoplakia, frictional keratosis, and even squamous cell carcinoma (13). However, the most challenging differantial diagnosis of WSN is oral lichen planus (especially the reticular and plaque variants), since both diseases show predilection for the cheek mucosa and usually present bilaterally (14). In our case, lesions general view was the primary factor for approaching the diagnosis as well as the patients age. It is known that lichen planus lesions are observed later in life between the 4th and 6th decades (15).

The lesions on mucous membranes persist through life, but the condition is benign. There is no evidence that these lesions show dysplastic changes or predispose to oral cancer development (16).

The clinical condition is painless. Patients complain only from unaesthetic appearance or symptomatic condition deriving from the altered texture of the mucosa. In most of the cases, WSN requires no treatment because of its benign and asymptomatic behaviour: up to now, no protocol of treatment for this condition was standardized (17).

When pain is present, some authors reported reduction of symptoms by penicillin (18**)** or oral tetracycline rinses (19**)**, suggesting that a bacterial overgrowht could be the reason of painful symptoms. However, almost all cases reported in the literature describe WSN as a benign condition that does not require any treatment.

Furthermore, in the study of Marrel et al., successful results were obtained in two patients chlorhexidine digluconate mouthwash, and they reported that treatment of the staphylococcus aureus infection had also been achieved (20). Moreover in the study of Satriano et al. (21) two patients with WSN were recommended to take morning and evening rinses with chlorhexidine mouthwashes 0.12% (5 ml for 45 s), that induced a significant regression of plaques after only 8 days. Lamey et al. reported on six patients with WSN who were prescribed long-term low dose systemic antibiotic therapy. They stated that the efficacy of antibiotics in this disease may in some way be related to their possible effects on epithelial maturation (22). In our experience, we do not recommend any medication but we recommend to use mouthwash for oral hygiene.

Conclusions

Our case demonstrate that the clinical and histopathologic findings of WSN were similar to previous reports. Proper evaluation for a correct diagnosis is of utmost importance. Detailed history and biopsy are necessary to establish the diagnosis. There is no treatment protocol for this condition. However we suggest to control the oral hygiene and daily use of a mouthwash containing chlorhexidine digluconate if a *Staphylococcus aureus* superinfection is accompanying documented.

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