

White Sponge Nevus

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Summary

A 45-year old woman was referred to our Department with bilateral white lesions on the buccal mucosa. Detailed medical history, clinical appearance including a histopathologic finding revealed a diagnosis of white sponge nevus.

Key words: *white sponge nevus*.

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CASE REPORT

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Introduction

White sponge nevus is an uncommon autosomal dominant disease characterized by benign white spongy plaques (oral leukokeratoses). Lesions usually appear on the buccal mucosa, but also on other non-keratinized, moist mucosa (1). Histological finding suggests thickening and vacuolisation of the spinous layer, with extensive hyperparakeratosis and acanthosis that may produce thickness in excess of 40 cell layers. The cells of stratum granulosum and spinosum are noticeably swollen and acanthotic. An occasional abnormal mitotic change and minimal signs of dyskeratosis may be evident in the basal cell layer. The basement membrane is intact, and the rete pegs are long and thin. The connective tissue generally, has no, or very slight, inflammation (2). The lesions are frequently present in the oral

cavity, but may also occur on the penis, vaginal and anal mucosa. Gender predisposition does not exist and the majority of described cases refer to the white race. The disorder may be detected during childhood, but is not usually detected till adolescence or adult age. The lesions are asymptomatic and are accidentally discovered. The involved mucosa is white or greyish, thickened, folded and spongy. During mastication, the superficial keratotic layer can be destroyed, leaving the naked epithelium. There is no evidence to show that these lesions show dysplastic changes or that they predispose oral cancer development. Lesions are usually diagnosed as candidiasis in children and the true nature of the disease is discovered when they are resistant to antifungal therapy. In literature white sponge nevus is also known as Cannon's disease, white gingivostomatitis and exfoliative leukoedema (3,4).

Case report

A female patient MM (45) was referred to our Department with white lesions on the buccal mucosa bilaterally. Detailed medical history did not reveal any systemic disease, nor was the patient taking any medications. Complete blood count, iron, total iron binding capacity, unsaturated iron binding capacity, aspartate-aminotransferase, alanin-transferase, gamma-glutamyltransferase, triglycerides, cholesterol, urea, creatinin were within normal ranges given for the general population. Clinical finding in the oral cavity showed white hyperkeratotic areas, 3x3 cm, on both sides of the buccal mucosa, with partially exfoliated epithelium and mild erythematous areas underneath. *Candida* culture smear taken with cotton swab and placed on Sabourauds agarose gel was negative after 48 hours in an incubator at 37°C. Histopathologic finding showed thickened and parakeratotic epithelium, with marked swelling and vacuolisation in overlying surface areas.

The patient denied the parafunctional habit of cheek-biting. Medical history revealed an interesting fact that the patient's daughter has similar changes on the buccal mucosa. Other mucosa (vaginal and anal) were not involved.

Discussion

White sponge nevus is a hereditary disease, in most cases involving the oral cavity and is present after birth or appears during early childhood. It is characterized by white or grey thickening or folds of the mucosa which appear oedematous. When unfolded the lesions do not lose thickness as seen in leukoedema. All members of the family are usually affected which is in concordance with our finding. Differential diagnosis can also include cheek-

-biting, in this case denied by the patient. Candidiasis was also excluded when a *Candida* culture was found to be negative. In the absence of nail changes and palmoplantar hyperkeratosis diagnosis of pachyionichia congenita was rejected. Differential diagnosis should also include hereditary benign intraepithelial dyskeratoses, known as Witkop von Sallman syndrome. The disorder is histologically and cytologically distinguished from white sponge nevus, on the basis of specific intraepithelial dyskeratoses appearance as well as acantosis, and vacuolisation of the stratum spinosum. In this syndrome photophobia and blindness due to plaque and cicatrix formation can be seen. It should be emphasized that palmoplantar hyperkeratosis can be manifested as white sponge nevus and is also characterized with gingival and palmar hyperkeratosis and with the development of oesophageal carcinoma and oral leukoplakia.

Chemical and temperature burns, smoking, leukoplakia, lichen ruber planus may also have the appearance of white sponge nevus (7,8).

More recent studies have reported that two mutations on the gene K13 are found to be responsible for the development of white sponge nevus. T to C transition was found in one family and resulted in leucine 115 changing into proline. In another family very similar T to C transition was found in codon 108 which lead to methionine changing into threonine (1).

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 (9,10).

It should be emphasized that correct diagnosis of white sponge nevus should be established while many other possible "white" lesions could have malignant potential.