White sponge nevus- A rare entity

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Introduction

White sponge nevus (WSN) is an autosomal-dominantly inherited form of mucosal leukokeratosis. It was first described by Hyde in 1909 and named by Cannon in 1935. Hence, it is also called Cannon's disease, hereditary leukokeratosis of mucosa, or white sponge nevus of Cannon. It is caused by a mutations in genes coding for keratin, which results in defective keratinisation. It is characterized by benign, painless, thick, white and spongy plaques of the oral mucosa. There is no gender predilection. Other extra oral sites include nasal, oesophageal, laryngeal, vaginal and anal mucosa.(1) The histopathologic features of WSN include epithelial thickening, hyperparakeratosis, and vacuolization of the keratinocytes in the suprabasal layers.(2) It is asymptomatic and therefore no treatment is required. Here is a case report of white sponge nevus in a 25 year old male affecting the right buccal mucosa.

Case Description

A 25 year old male reported to the department of oral medicine and radiology with a chief complaint of white patch on right side cheek mucosa since 14 years (Fig. 1). History of present illness revelled that the patch was noted by him while brushing and was smaller in size but has gradually attained its present size during the span of 5-6 years. There was no relevant dental history or medical history. In the family history, patient's brother also had similar type of patch in relation to his left buccal mucosa since 25 years. On general physical examination all the vital signs were in normal range. No abnormality was detected on systemic evaluation and extra-oral examination. On intra-oral examination a white translucent plaque with well defined margins, measuring approximately 2.5 x 2.0 cm was present on right buccal mucosa extending anteroposteriorly 0.5 cm away from retro commissural area to middle of buccal mucosa and supero-inferiorly 1cm above and 0.5cm below the occlusal line. On palpation it was smooth, soft, non-tender and non–scrappable. Based on history and clinical examination a provisional diagnosis of White sponge nevus was given with a differential diagnosis homogenous leukoplakia, pseudomembranous candidiasis and dyskeratosis congenita. On haematological investigations all the values were within normal limits and a punch biopsy was performed as an investigative procedure. The histopathological report under 4X magnification revealed hyperplastic epithelium with hyperparakeratinization and acanthosis, broad, and elongated rete ridges (Fig. 2). Whereas, 40X magnification showed the entire spinous layer was characterized by intracellular edema with basket-weave and "cell-within-a-cell" appearances. Characteristic perinuclear condensations were noted, along the presence of Nevus nests (Fig. 3). Hence, based on histopathological examination a final diagnosis of White Sponge Nevus was given. Though no treatment is required for WSN, patient was however advised surgical excision for cosmetic purposes.

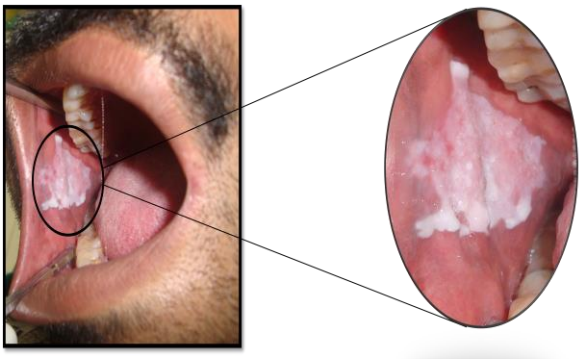

Fig. 1: Showing a white plaque with well defined margins in relation to right buccal mucosa

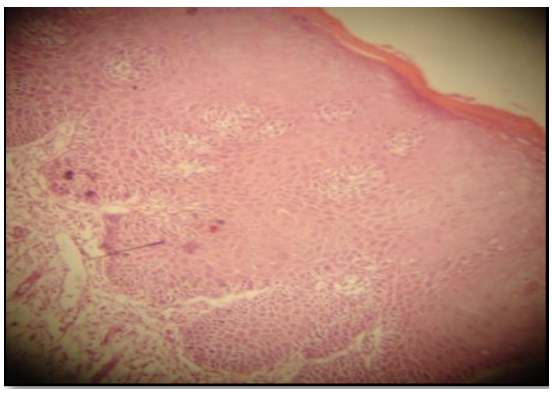

Fig. 2: Showing hyperplastic epithelium with hyperparakeratinization and acanthosis, broad, and elongated rete ridges under 4x magnification

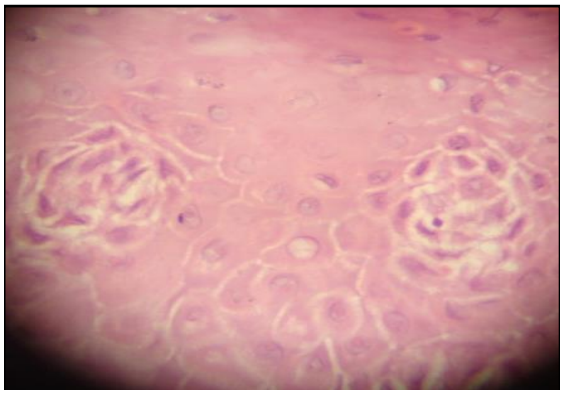

Fig. 3: Showing spinous layer characterized by intracellular edema with basket-weave and "cell-within-a-cell" appearances. Characteristic perinuclear condensations were noted, along the presence of Nevus nests under 40x magnification

Discussion

White sponge nevus has been listed as a rare disorder by the National Institutes of Health. Its prevalence is below 1 in 200,000. The clinical appearance usually commences during adolescence, and has equal gender prediliction.[3]

It should be noted that Keratins are a family of about 30 proteins. Non-keratinizing stratified epithelia i.e. buccal mucosa, express CK4 and CK13 with CK5 and CK14. Keratinizing stratified epithelia such as the tongue and the palate express CK1 and CK10 together with CK5 and CK14.[2,4,5] Mutation in the genes that code for epithelial keratin- K4 and K13 initiate White sponge nevus.[3,6]

The hallmark histopathologic feature is predominantly located within the stratus spinosum, showing pronounced intracellular edema with basket-weave and "cell-within-a-cell" appearances of the superficial epithelial cells, hyperplastic epithelium with hyperparakeratinization and acanthosis, broad, and elongated rete ridges. Deep fissures in the non-dysplastic epithelium may be seen with subepithelial tissue showing just mild infiltrations.[3]

As white sponge nevus may constitute a differential diagnostic problem so it must be differentiated from other white lesions.[7] If the lesions have been present since birth or at least since early life, this will largely eliminate Lichen Planus and leukoplakia because these lesions are quite unusual in patients under 30 years of age. Reticular variant of Lichen planus will have presence of Wickham's Stria, whereas WSN will not have any such characteristic appearance. Leukoplakia will be associated with habit history like consumption of tobacco products. Cheek chewing or Morsicatio buccrum can produce a rough-surfaced lesion. Once the habit is detected the diagnosis is made. Hereditary benign intraepithelial dyskeratosis may have similar whitish keratotic oral lesions, but it is differentially diagnosed because of its presence in a triracial population in North Carolina. Clinically, pachyonychia congenita might be easily confused with white sponge nevus because the keratotic white lesions may look quite similar and may be present from birth in both diseases. However, pachyonychia congenita shows the presence of nail anomalies and the skin lesions. Leukoedema is found to have only a milky opalescence, whereas white sponge nevus has a rough, granular, somewhat leathery surface. Furthermore, stretching the tissue makes leukoedema disappear but does not affect the appearance of white sponge nevus.[8,9,10]

Usually, proper identification is required, since the white sponge is benign. Occasionally a raw surface results from the desquamation of the thickened epithelium, and various palliative procedures are necessary to relieve the burning and tenderness. White sponge nevus does not show any symptoms, and no treatment is therefore required. Although intraoral skin grafts can be used as treatment modality and are easily recognized by most clinicians who have seen them. Systemic antibiotics have been used in an attempt to resolve the disorder, but with non-consistent results. When a positive effect is obtained, the recurrence rate is considerable.[3,8,9,10] To conclude White sponge nevus is a rare entity. Proper diagnosis can be made only after taking detailed case history, conducting efficient intra-oral and extra-oral examination and performing adequate investigations especially histopathologic investigations. Being a benign lesion it does not require any treatment. However, these days various methods are available for treating WSN, in case patient wants its removal for aesthetic reasons.

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