

Case Report

White Sponge Nevus: A Case Report

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Abstract

White sponge nevus (WSN) is a rare hereditary dyskeratotic hyperplasia of mucous membranes. It is an autosomal dominant disorder with variable penetrance. We report a case of WSN in a healthy 21-year-old male with no history of familial involvement. A white smooth plaque with no erythema or other structural abnormalities was observed, which confirmed the diagnosis of WSN histopathologically.

Key words: Dyskeratosis, white lesion, white sponge nevus.

Introduction

White sponge nevus (WSN) is a relatively rare cutaneous and mucosal lesion.^{1,2,3} Hyde reported the first case of WSN in 1909 and a detailed report was published in 1935 by Cannon.^{4,5} Etiologically, it is a rare developmental anomaly inherited as an autosomal dominant trait with variable expressivity and a high degree of penetrance. This condition is attributed to a defect in the normal keratinization (keratin 4 and keratin 13, which are specifically expressed in the spinous cell layer of the oral mucosa).⁶⁻¹¹ This keratotic mucosal alteration may be seen on vaginal and rectal mucosa but the great majority of cases involve the oral mucosa.¹

A search of dermatological and gynecological literature revealed very little about WSN in Iran. More information was available about this lesion in the oral cavity, which was retrieved from the dermatological and dental literature.¹²

Lesions of WSN usually appear at birth or in early childhood, but sometimes the condition develops during adolescence. The lesions consist of symmetric,

thickened, white, corrugated or velvety, diffuse plaques. Buccal mucosa is the most frequently affected, followed by the labial and gingival mucosa, and the floor of the mouth. Extra-oral mucosal sites, such as the nasal, esophageal, laryngeal, and anogenital mucosa, appear to be less commonly affected. Patients are usually asymptomatic. The white color does not diminish when the tissue is stretched in any mucosal site.^{1,2,6,13,14}

The recognition of this disorder is important in that it must be differentiated from other congenital or familial disorders of more widespread clinical significance. The clinical appearance is so distinctive that biopsy is usually unnecessary. The microscopic features of WSN are characteristic but not necessarily pathognomonic. Prominent hyperparakeratosis and marked acanthosis with clearing of the cytoplasm of the cells in the spinous layer are common features; however, similar microscopic findings may be associated with leukoedema and hereditary benign intraepithelial dyskeratosis. In some instances an eosinophilic condensation is noted in the perinuclear

region of the cells in the superficial layers of the epithelium, a feature that is unique to WSN.¹⁻⁵

In this paper, a case of WSN in a healthy white male with no history of familial involvement is described.

Case Report

The patient was a 21-year-old Iranian male referred to the Department of Oral Medicine at Tabriz University of Medical Sciences Faculty of Dentistry for diagnosis and management of a “white, itchy spot” on the buccal mucosa. White bilateral lesion in oral mucosa was the chief complaint of the patient. The patient complained of a white lesion which was present since birth.

The patient’s general health was reportedly good. The patient denied presence of a similar condition in immediate family members or any similar lesions elsewhere on his body.

In clinical examination, there were bilateral, symmetrical white plaques and patches on the buccal and labial mucosa, which could not be removed (Figure 1). The plaques were smooth with velvety texture and irregular, well-defined borders. There was no elevation or erythema. The margins were clear and no lymph nodes were noticeable. Oral hygiene was good and other oral structures were normal in appearance.

In histopathologic evaluation, oral mucosa covered by stratified squamous epithelium revealed prominent hyperparakeratosis and marked acanthosis with clearing of the cytoplasm of cells in the spinous layer. In addition, eosinophilic condensation was noted in the perinuclear region of the cells in superficial layers (Figure 2). Underlying connective tissue was normal in appearance with rare chronic inflammatory cell infiltration.

Based on clinical data and histopathologic findings, the lesion was consistent with white sponge nevus. Because of benign nature of this lesion, no treatment is necessary and only biopsy and correct diagnosis is necessary to rule out other similar lesions. Six-month follow-up was recommended.

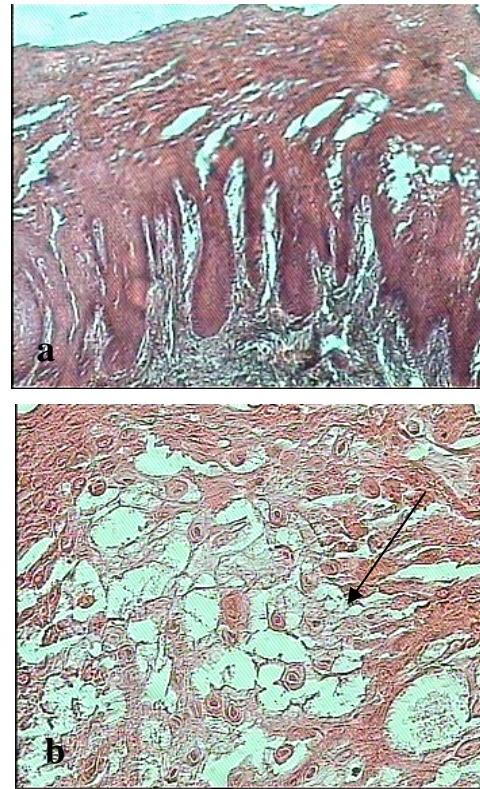


Figure 2. (a) Histopathologic view of the lesion (×10); (b) perinuclear condensation of keratin tonofilament (arrow) (×40) (H&E).

Discussion

WSN is a rare hereditary dyskeratotic hyperplasia of mucous membranes. This entity is also known by other names, such as Cannon's disease, familial white folded hypertrophy of the mucous membranes, hereditary leukokeratosis, white gingivostomatitis, and exfoliative leukoedema.¹⁻⁵ WSN is an autosomal dominant disorder with variable penetrance and hence familial reports are not very common, similar to the present case. WSN has been listed as a rare disorder, with a prevalence rate below 1 in 200,000.¹³ Most commonly, lesions appear at birth or in early childhood. Neither gender nor racial predilection exists.¹⁴ A case of WSN, in which human papilloma



Figure 1. Clinical views of the lesions.

virus type 16 was demonstrated, has been reported in the literature.⁴ Many different types of white lesions can occur in the oral mucosa and the appearance of WSN is not pathognomonic. There is a need for precise identification through prompt histopathologic examination to differentiate this condition from more serious, potentially premalignant lesions as well as other genodermatoses such as hereditary benign epithelial dyskeratosis, lichen planus, lichenoid drug reaction, lupus erythematosus, cheek chewing and possibly candidiasis. While some of these lesions are benign, others are pre-malignant or manifestations of some systemic diseases. Therefore, early diagnosis of this benign lesion is important,³ and often, these lesions need different treatment plans.¹⁵⁻²¹ In addition, these lesions reveal different epidemiological patterns and involve different societies and races. In Northwest Iran, this condition seems to be rare and no other similar documented cases are available. In this case, none of the family members had similar lesions. This lesion appeared early in life without any reported changes throughout the patient's life, but diffuse spreading of the lesion seems to be an alarming factor. Biopsy in such cases is necessary for treatment planning and ruling out of other lesions.

References

1. Neville BW, Damm DD, Allen CM, Bouquot JE. *Oral & Maxillofacial Pathology*, 3rd ed. St. Louis: WB Saunders; 2009:332-5.
2. Elder DE, Elenitsas R, Johnson BL, Murphy GF. *Lever's Histopathology of the Skin*, 9th ed. Philadelphia: Lippincott Williams & Wilkins; 2005: 715.
3. Regezi JA, Sciubba JJ, Jordan R CK. *Oral Pathology*, 5th ed. St. Louis: Saunders Elsevier; 2008: 74-77.
4. Cox MF, Eveson J, Porter SR, Maitland N, Scully C. Human papillomavirus type 16 DNA in oral white sponge nevus. *Oral Surg Oral Med Oral Pathol* 1992;73:476-8.
5. Cannon AB. White sponge nevus of the mucosa. *Arch Dermatol Syph* 1935; 31:365-70.
6. Jorgenson RJ, Levin LS. White sponge nevus. *Arch Dermatol* 1981;17:73-6.
7. Búchholz F, Schubert C, Lehmann-Willenbrock E. White sponge nevus of the vulva. *Int J Gynaecol Obstet* 1985;23:505-7.
8. Nichols G, Cooper P, Underwood P, Greer K. White sponge nevus. *Obstet Gyn* 1990;76:545-8.
9. Woo SB. Diseases of the oral mucosa. In: McKee P, Calonje E, Granter S, eds. *Pathology of the Skin, With Clinical Correlations*, 3rd ed. Philadelphia: Mosby; 2005: 387.
10. Dadlani C, Mengden S, Kerr AR. White sponge nevus. *Dermatol Online J* 2008;14:16.
11. Rugg EL, McLean WH, Allison WE, Lunny DP, Macleod RI, Felix DH, et al. A mutation in the mucosal keratin K4 is associated with oral white sponge nevus. *Nat Genet* 1995;11:450-2.
12. Jahanbani J, Sandvik L, Lyberg T, Ahlfors E. Evaluation of oral mucosal lesions in 598 referred Iranian patients. *Open Dent J* 2009;3:42-7.
13. Shibuya Y, Zhang J, Yokoo S, Umeda M, Komori T. Constitutional mutation of keratin 13 gene in familial white sponge nevus. *Oral Surg Oral Med Oral Pathol Oral Radiol Endod* 2003; 96:561-5.
14. Hernandez-Martin A, Fernandez-Lopez E, de Unamuno P, Armijo M. Diffuse whitening of the oral mucosa in a child. *Pediatr Dermatol* 1997; 44:316-20.
15. Lamey PJ, Bolas A, Napier SS, Darwazeh AM, Macdonald DG. Oral white sponge nevus: response to antibiotic therapy. *Clin Exp Dermatol* 1998; 23:59-63.
16. Everett FG, Noyes HJ. White folded gingivostomatitis. *J Periodontol* 1953; 24:32.
17. O'Leary PA, Montgomery H, Brunsting LA, Kierland RR. White sponge nevus: moniliasis? *Arch Dermatol Syphilol* 1950; 62:608.
18. Aloi FG, Moliner A. White sponge nevus with epidermolytic changes. *Dermatologica* 1988; 177:323-6.
19. Alinovi A, Benoldi D, Pezzarossa E. White sponge nevus: successful treatment with penicillin. *Acta Derm Venereol* 1983;63:83-5.
20. McDonagh AJ, Gawkrödger DJ, Walker AE. White sponge nevus successfully treated with topical tetracycline. *Clin Exp Dermatol* 1990;15:152-3.
21. Lim J, Ng SK. Oral tetracycline rinse improves symptoms of white sponge nevus. *J Am Acad Dermatol* 1992; 26:1003-5.

Erratum

The article entitled "Accuracy of digital subtraction radiography in combination with a contrast media in assessment of proximal caries depth" which appeared in *J Dent Res Dent Clin Dent Prospect* 2008; 2(3):77-81 incorrectly listed the second author's affiliation as Department of Oral and Maxillofacial Radiology, Shahid Beheshti University of Medical Sciences. The correct affiliation of Sara Ehsani is Dental Research Center of Shahid Beheshti University of Medical Sciences.