WHITE SPONGY NEVUS (FAMILIAL CONGENITAL LEUKO KERATOSIS)

NOOROLIAH TAID KASHANI

Summary

A 35 years old white male, smoker with a whitish lesion involving both sides of oral mucosa and tongue is presented. Similar lesions were discovered on the oral mucosa of patient's father and daughter. Clinically and histologically the diagnosis of congenital familial leukokeratosis was made. No treatment was given other than advice on oral hygiene.

To the best of our knowledge, this is the first case report of "White Spongy Nevus" from Iran.

White Spongy Nevus was first described by Cannon in 1935[1,2]. The condition represents a congenital ectodermal dysplasia affecting the ectodermal tissue of oral mucosa.

Case Report

35 years old white male, a mechanical engineer was referred to the dental department of Razi Hospital for extraction of the right third upper molar tooth. On examination whitish lesion covering oral mucosa and tongue was noticed. The lesion was hard, folded and hyperkeratotic. Removal of superficial layers did not produce any bleeding.

Patient was a thin, tall nervous individual. He gave the history that his mouth lesion was present as long as he could remember. His 70 year old father had a similar but thicker lesion involving the right side of the oral mucosa from the age of 20 years. Patient's 8 year old eldest daughter also had a similar lesion. He had 2 other children who were unaffected. His mother had died 10 years ago due to heart disease.

Patient was a smoker, smoking 2 packets of cigarettes per day. His father was a nonsmoker.

Histopathology

The covering squamous epithelium was hyperplastic and there was marked parakeratosis. Rete pegs which had intermingled and fused together appeared broadened. Marked intra and extracellular edema was evident in prickle cell layer. Many of the prickle cells were keratinized. These appeared discrete, circular and red. Submucosa was fibrotic and hyalinized. On the basis of the clinical and pathological features of this lesion, congenital leukokeratosis was diagnosed. (Fig. Page No. 50).

Discussion

White spongy nevus is a familial and congenital ectodermal defect which has been given different names[2,3]. White folded gingivostomatitis, white folded familial dysplasia of mucosa, intraoral leukonevus[4].

Assistant Professor of Oral Medicine,
Section Dignosis of Oral Diseases,
University of Tehran, Iran.

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More than 30 cases have been reported in literature. All patients were of caucasian origin[5-7]. Any ectodermal tissue can be involved. The oral mucosa, and mucosae of vulva, vagina and anus [5, 8, 9] can be involved at birth and reach its full development by adolescence[5]. Both sexes are equally affected in this autosomal dominant disease[5,7]. Sporadic cases have also been reported.

White lesions in the mouth are often reactive in origin The clinical appearance of these chronic white lesions are seldom indicative of their true nature, [5, 10]. Several etiologic factors may be responsible for these lesions. They vary from simple traumatic irritation and burns of oral mucosa to fungus infection and specific infectious diseases involving oral mucosa[10]. They can simulate leukoplakia, a premalignant condition. The white spongy nevus occurs without symptoms and are detected accidentally[5]. Mucosa becomes thick and folded. Superficial Keratin layer desquamates and on scraping yields no bleeding[2]. Free borders of the gum look thin and probably are not involved[6].

The lesion is almost always symmetrical but occasionally unilateral[11]. Besides oral and genital mucosa, nasal and pharyngeal mucosa can also be involved[6]. In histology epithelium is thickened and parakeratotic. Superficial cells show pyknotic nuclei and spongiosis. Prickle cells are usually vacuolated[5].

Differential Diagnosis

Lesions produced following bite of patient's own mucosa, pachyonychia congenita, leukoplakia, Lichen planus, moniliasis, familial and beningn intraepithelial dydkeratosis should all be considered in the differential diagnosis[5, 7]. Extensive leukokeratosis in a child specially when associated with a positive family history, should alert the physician to make the diagnosis of familial white folded gingivostomatitis.

Treatment

In most reports no special treatment is suggested other than good oral hygiene. One report recommends surgical removal of the lesion[4]. X-ray treatment is not recommended.

REFERENCES

1. Cannon AB : White nevus of the mucosa (nevus spongious albus mucosae), Arch-Derm. (Chicago) 31 : 365, 1935.
2. Thoma KH and Goldman HM : Oral pathology, 5th Ed, St Louis, CV Mosby Company, 1960, p 94.
3. Cohen L and Stream IP : Clinical benodermatology, Baltimore, Williams Wilkins Company, 1962, p 141.
4. Csila A and Egervary G : Dental digest Dec. 1970 p 518.
5. Gorlin RJ : Genetic disorders affecting mucous membranes, Oral Pathology, 1968 p 512.
6. Cohen L and Young AH : The white sponge nevus, Bright J Oral Surg, 5 : 206, 1968.
7. Kamalammes MK, Prabhu SR, Shetty JN, et al ; The white sponge nevus, Oral Surg Med Path, 30 : 51, 1970.
8. Sempson HE : White sponge nevus; report of three cases, J Oral surg 24 : 493, 1966.
9. Withip CI Jr and Gorlin RJ : Four hereditary mucosal syndromes, Arch Dermat, 84 : 762, 1961.
10. Rushton MA and Cooke BED : Oral Histopathology, ED 5, St Louis CV Vasby Company, 1960 p 941.
11. Cooke BED : Keratinizing lesions affecting the oral mucosa, Proc-Rey-Soc-Med, 60 : 819, 1967.