WHITE SPONGE NAEVUS

(Report on 9 cases)

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Summary

Occurrence of white sponge naevus among Indians appears to be rare. In this paper 9 cases in a kindred of 42 from Kerala, India, have been reported. The lesions were located on the buccal mucosa and the patients were unaware of their presence. Of the 9 individuals with the lesions, 2 were from the 2nd generation, 4 from the 3rd generation and 3 from the 4th generation. The findings in our series supports the autosomal dominant mode of inheritance of white sponge naevus.

White sponge naevus is an asymptomatic mucosal disorder, first described by Cannon in 1935[1]. The oral mucosa is the most often reported site[2-4], although the involvement of other mucosal sites of nasal cavity, vagina and rectum also has been reported[3,5,6]. The oral mucosa in this condition appears white or grey, thickened, folded and spongy. The lesions may be widespread, involving almost all areas of the oral mucosa. Shafer et al[7] indicated that the disease might follow a hereditary pattern as an autosomal dominant trait.

The lesion does not seem to be very common. Banoczy et al[8] indicated that they were able to trace 120 cases of white sponge naevus in the literature, including those reported under other terms. Their own series consisted of 45 cases, the largest reported so far. Though the family members of most of the cases could not be examined, the authors were able to ascertain that there was familial occurrence in 25% (12) of their cases.

Most of the reports of the white sponge naevus have come from the Western countries. The only report from India was published by Kamalamma et al[4], from Manipal (South Kanara), India. Including the proband, 9 cases were detected in a kindred spanning three generations in their series, which strongly supported the autosomal dominant mode of inheritance.

In this paper we report on 9 cases in a kindred in Kerala, India. The proband was diagnosed during the epidemiologic study of oral cancer and precancerous lesions in Indian rural populations[9]

Case Report

The proband was a 28 years old Hindu female. She did not practise

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any tobacco habits. On both sides of the buccal mucosa, a white, thickened, folded lesion with spongy texture was observed (Fig. 1). A tentative diagnosis of white sponge naevus was made and a punch biopsy was obtained from the left buccal mucosa. The patient was the mother of 2 boys aged 8 and 4 years. Both her children were examined and the elder one also had white spongy lesions on the right and left buccal mucosa. The proband was unaware of the lesions.

Histology

The formalin fixed tissue was processed by the routine laboratory methods and the section stained by hematoxylin-eosin method.

The section showed hyperplastic epithelium with uneven thickness of para-keratin on the superficial surface. The parakeratin surface also exhibited vacuolated cells, at places with faintly stained nuclei. Intercellular oedema was observed in the lower spinous cell layer. Areas of darkly stained bluish-masses were seen on the surface indicative of clusters of bacteria. The stratification of the cellular layer was normal. A slight degree of mononuclear inflammatory cell infiltrate was evident more in the sub-epithelial region rather than in the deeper part of the submucosa (Figs. 2 and 3).

Histologic findings were compatible with that of white sponge naevus.

Pedigree findings

Information could be obtained for 42 individuals spanning 4 generations. Of the 42 people, 5 had died (4 unrelated spouses) and one was not available for examination. Among the 36 people examined, 6 were unrelated spouses (3 males and 3 females). The lesion was detected in 9 members of the kindred (Fig. 4).

Of the 9 persons with the lesion, 2 were from the second generation, 4 from the third generation and 3 from the fourth generation. 4 among the affected members were males and 5 females. Their ages ranged from 7 years to 57 years (Table 1).

TABLE 1.

Distribution of the lesions according to age, sex, generation and tobacco habits

Case No.	Age	Sex	Generation	Tobacco Habits	
				Smoking	Chewing
1.	57	F	2	—	—
2.	43	M	2	+	+
3.	34	F	3	—	—
*4.	28	F	3	—	—
5.	25	M	3	—	—
6.	16	F	3	—	—
7.	16	M	4	—	—
8.	8	M	4	—	—
9.	7	F	4	—	—

* Proband

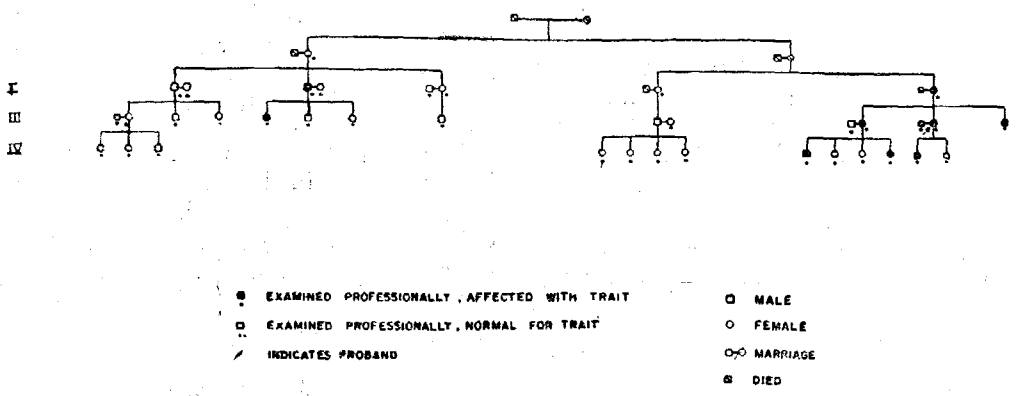

Fig. 4 Pedigree

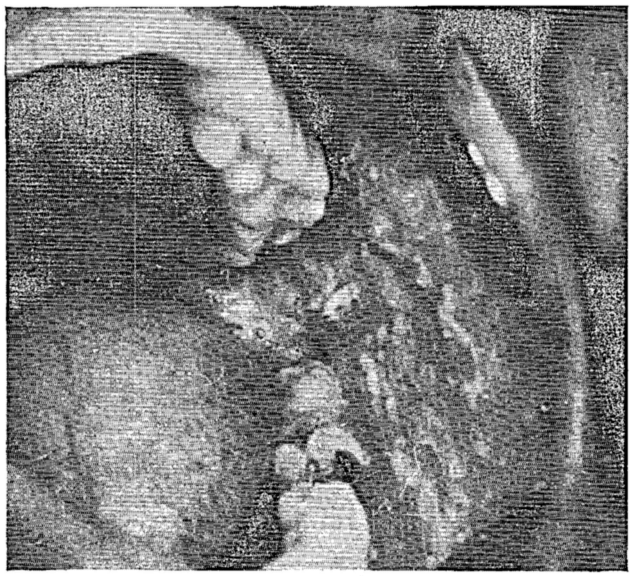

Fig. 1 White sponge naevus on the left buccal mucosa of the proband

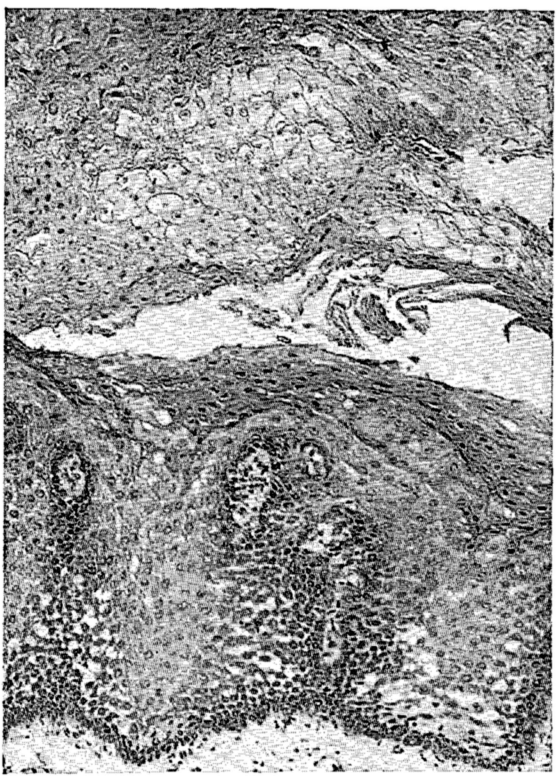

Fig. 2 Thick uneven para-keratin layer with vacuolated cells and hyperplasia of the epithelium H & E × 40.

Fig. 3 Intercellular Oedema and vacuolated cells in the lower spinous cell layer. H & E X 200

Comments

To our knowledge this is the second report of this condition among Indians. The buccal mucosa was exclusively involved in all and the lesions were bilateral in 4 patients. Examination of the other mucosal sites was not feasible in this series. None of the patients revealed any cutaneous, occular or nail anomalies suggestive of other hereditary anomalies like Darier-White's disease, hereditary benign intra-epithelial dyskeratosis or pachyonychia congenita.

Our findings strongly support the autosomal dominant mode of inheritance of white sponge naevus.

Acknowledgements

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