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CASE REPORT

Depigmentation of oral mucosa as the earliest possible manifestation of oral submucous fibrosis in Sri Lankan preschool children

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Abstract

This article reports the presentation of oral submucous fibrosis in five Sri Lankan preschool children, aged 2–3 years, with loss of pigmentation of the lips as the sole clinical feature. Oral submucous fibrosis has not been reported in this age group of children. The five 2–3 year olds did not display any classical features of oral submucous fibrosis, due to the disease having been detected at a very early stage. The present study attempts to establish that depigmentation of the lips and oral mucosa is perhaps the earliest feature to develop in the natural history of oral submucous fibrosis. The differential diagnosis of oral mucosal depigmentation relevant to these cases is also discussed. Previously-reported cases of oral submucous fibrosis in children are reviewed.

Introduction

Oral submucous fibrosis (OSF) is a chronic, insidious, generalized, and debilitating condition of the oral mucosa that causes burning sensation and progressive limitation in the mouth opening, including a reduced ability to protrude the tongue.[1] OSF is a well-known potentially-malignant disorder, and is predominantly encountered in South Asian and South–East Asian countries, and among South Asian immigrants in Western countries.[2–4] It has been established that OSF is etiologically linked to the consumption of the areca nut in flavored formulations or as an ingredient in the betel quid chewed by the communities in these countries.[5]

Apart from the burning sensation and limitation in the mouth opening, including a reduced ability to protrude the tongue, the other important clinical feature of OSF is the loss of pigmentation of the vermilion border of the lips and the oral mucosa, followed by the development of a leathery texture and blanching of the oral mucosa.[6,7] Depigmentation of the lips can easily be recognized in many of these patients, even without close examination. Although OSF is generally considered an adult disease, it has been reported among school children from communities with betel and areca nut chewing habits.[2,3,8]

The aim of the present article was to report the presentation of OSF in five 2- to 3-year-old Sri Lankan preschool children. To the best of our knowledge, this is the first study to report on OSF in such a young age group. We also aimed to highlight that loss of pigmentation appears to be the earliest manifestation of the condition. The reported childhood cases of OSF to date are also reviewed in this paper.

Case reports

Table 1 summarizes the salient information regarding the five children who presented to the oral medicine clinic of the University Dental Hospital, Peradeniya, Sri Lanka, between 2000 and 2009, with loss of pigmentation of the lips as the sole complaint. Although case 1, HM, was initially seen in 2000, she was periodically reviewed until

Table 1. Case summaries of five Sri Lankan preschool children diagnosed with OSF

Case no./ reference	Age (years)/ sex	Year of presentation	Presenting complaint/ duration‡	Associated chewing habit	Other characteristic features of OSF	Biopsy findings§	Details of chewing habit
01 HM	3/Female	2000	White color of lip/ 4 weeks	Betel with areca nut	None	Mildly-atrophic, stratified squamous epithelium with increased amount of collagen in the upper corium Fibrosis extends into muscle in a few foci. No associated inflammatory infiltrate	2–3 times/day Copied from grandmother
02 NCK	3/Male	2008	Loss of lip pigmentation/ >1 month	Areca nut only	None	Mild, atrophic changes in the surface epithelium and features suggestive of OSF seen, although no significant increase in fibrosis found in the underlying corium. Histopathological features in this patient appeared to be of a lesser degree	2–3 times/day Copied from grandmother
03 RMN	3/Male	2009	Whitish shade of lip/ >2 weeks	Betel with areca nut	Mild burning sensation later, but occasional	Biopsy not performed	1–2 times/day
04 SDR	3/Male	2009	Loss of lip color/ 3–4 weeks	Betel with areca nut	None	Biopsy not performed	2–3 times/day Copied from grandmother
05 DLH	2†/Female	2009	Loss of lip pigmentation/ >3 weeks	Areca nut only	None	Biopsy not performed	No reliable information

†2 years and 3 months; ‡duration since noticed by family; §in hematoxylin–eosin-stained sections. OSF, oral submucous fibrosis.

2004. During the review period, the child claimed to have abstained from betel chewing, but there was neither significant improvement nor worsening in the depigmentation of the lips. In 2004, she underwent biopsy. Case 2, NCK, also underwent biopsy, but without periodic review. The histological findings from these two biopsies are presented in Table 1, and the clinical features of cases 2 and 3 are shown in Figures 1 and 2. Figure 3 shows the histological features of the biopsy in case 1.

No biopsy was performed on the last three cases because of the striking similarity with the two previous cases and the existence of a very clear association with a betel/areca nut chewing habit.

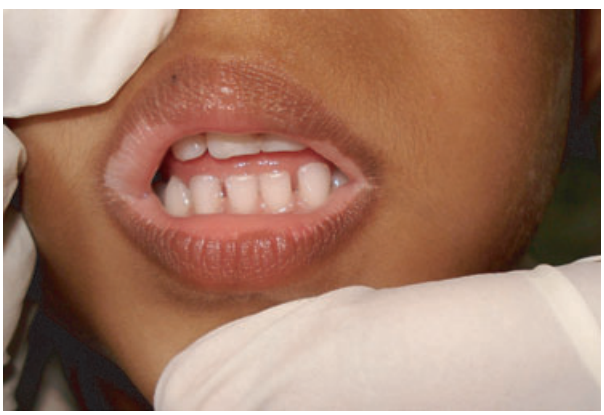

Figure 1. Clinical photograph of case 2.

Discussion

OSF has not been reported previously in 2- to 3-year-old children in the English-published literature. Sri Lanka has a moderately-high prevalence of OSF.[9] The oral medicine clinic of the University Dental Hospital possesses a large databank of patients with OSF and has no previous record of 2- to 3-year-old children with the disease. It is very rare for children of such young age in Sri Lanka to indulge in betel/areca nut chewing. It is important to note that depigmentation of the lips was the only complaint in these children, and this would appear to be the earliest feature of OSF. Most reported cases of OSF in the literature concern young adults or older individuals with

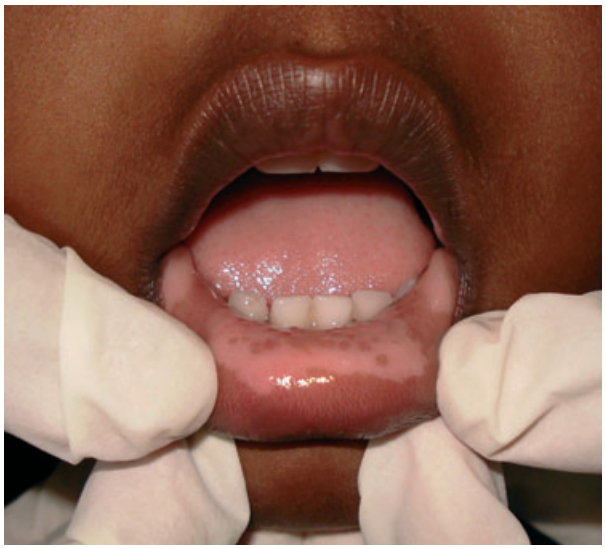

Figure 2. Clinical photograph of case 3.

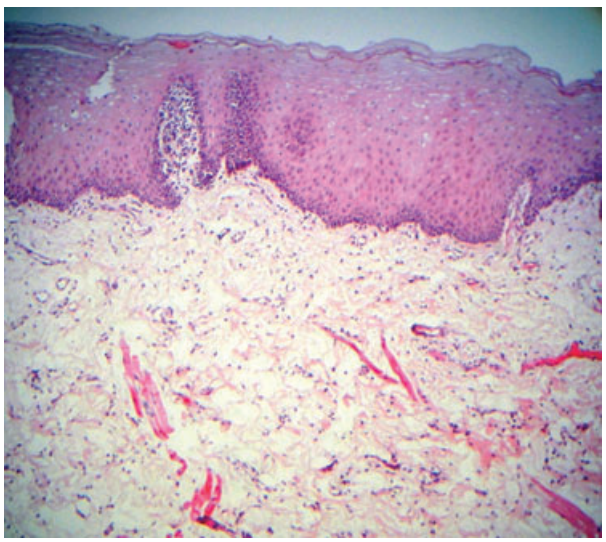

Figure 3. Photomicrograph of hematoxylin–eosin-stained section of biopsy from case 1. Magnification ×200.

moderately-advanced or late-stage disease. OSF presenting in such stages in the natural history of the disease would exhibit many classical symptoms and signs. None of the five 2–3 year olds in the present study displayed any such advanced clinical features of OSF. This is evidently due to the disease having been detected at a very early stage.

The paucity of information on lip depigmentation presenting as an early symptom might be due to the fact that OSF patients generally present after the disease has progressed and causes burning sensation and/or significant limitation in the mouth opening. Concurrently features, such as a leathery texture and blanching of the mucosa, would also be present. Although depigmentation and blanching are described in the literature as overlapping entities, our clinical experience shows that these are two distinct stages. A depigmented oral mucosa still retains its glossy appearance, whereas blanched mucosa exhibits a matt appearance. These differences are conceivable when the probable underlying factors are considered. Depigmentation might occur as a result of diminishing melanocytes, due to an early reduction of vascularity of the mucosa, whereas blanching probably results from the disappearance of the capillary bed in the corium, which is associated with increasing fibrosis.

According to the histological reports on the biopsies of the first two cases, the degree of fibrosis seen had been very minimal, and was expected to be so in a very early stage of OSF. The diagnosis of early cases of OSF, especially in a child patient, should be achieved through a clinicopathological correlation.

Differential diagnosis

The only other conditions that need to be considered in the differential diagnosis of oral mucosal depigmentation are vitiligo and lichen sclerosus et atrophicus. Among the different subtypes of vitiligo, vitiligo vulgaris, the generalized type, could be easily excluded, as there was no depigmentation in other parts of the body in any of the patients. The segmental type can also be excluded because the condition is usually distributed within a trigeminal dermatome, and is thus likely to extend beyond the mucocutaneous junction onto the facial skin, which was not the case in any of our patients. Thus, the only type that needs to be differentiated is focal-type vitiligo. The definite association with the consumption of areca nuts in each of these five cases can be considered sufficient evidence for the diagnosis of OSF, as it is highly improbable that all five children with vitiligo were areca nut consumers. The use of a Wood's lamp test was not considered suitable to exclude vitiligo in these cases, as the test is not specific to vitiligo.

Lichen sclerosus et atrophicus of the oral mucosa is very rare and has never been reported in children. Histologically, the absence of vascular lumen obliteration or reduction distinguishes lichen sclerosus et atrophicus from submucous fibrosis,[10] and this was true of the histology of the two cases biopsied.

Review of previous reports of OSF in children

Table 2 summarizes previous studies[2–4,8,11] of OSF in children. All of these children had a habit of chewing areca nuts, with or without betel quid, and almost all of them had been diagnosed with moderately-advanced OSF.

Table 2. Summary of previously-reported cases of OSF in children

Author(s)	Year of report	Country	Age (years)/ sex of children	Ethnicity†	No. cases
Hayes[2]	1985	Canada	4/Female	Indian	1
Anil and Beena[11]	1993	India	12/Female	Indian (Kerala)	1
Shah *et al.*[3]	2001	UK	11/Female	Bangladeshi	1
Yusuf and Yong[4]	2002	UK	12/Male	Bangladeshi	1
Oakley *et al.*[8]	2005	Commonwealth of the Northern Mariana Islands	16.3 ± 1.5; both sexes	Polynesian	27

†of reported cases; OSF, oral submucous fibrosis.

None of these reports highlighted the loss of pigmentation of lips or oral mucosa as an early finding.

Several other studies reported areca nut addiction among school children in various countries, such as Taiwan[12,] and Pakistan,[13] and among South Asian immigrants to the UK,[14] although these studies have not documented the prevalence of OSF in the population of school children studied.

In conclusion, although OSF has been previously reported in children, its occurrence in 2- to 3-year-old preschool children has not been reported in the English-published literature, and to our knowledge, the present report is the first to do so. It is noteworthy that the loss of pigmentation had been the sole clinical feature of OSF in these children, and it would appear to be the earliest feature to develop in the natural history of this condition. Thus, it would be prudent to ascertain the consumption of areca nuts when a patient presents with depigmentation of the lips in the absence of other clinical features of OSF.

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