CASE REPORT

Ackerman’s tumour of buccal mucosa in a leprosy patient

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Summary  Leprosy (Hansen’s disease) is a chronic granulomatous disease caused by Mycobacterium leprae (Hansen’s bacillus). Oral manifestations occur in 20–60% of cases, usually in lepromatous leprosy, and are well documented. They may involve both the oral hard and soft tissues. Incidence of verrucous carcinoma/Ackerman’s tumour developing in anogenital region and plantar surfaces of feet in lepromatous leprosy has been sufficiently documented in the literature. However, association of oral verrucous carcinoma with lepromatous leprosy has not been established. We report for the first time a case of verrucous carcinoma of the buccal mucosa occurring in a leprotic patient, with brief review of literature on orofacial manifestations of leprosy.

Introduction

Leprosy (Hansen’s disease) is a chronic, contagious granulomatous disease caused by Mycobacterium leprae (Hansen’s bacillus). The disease presents polar clinical forms (the ‘multibacillary’ or lepromatous leprosy, and ‘paucibacillary’ or tuberculoid leprosy), as well as other intermediate forms with hybrid characteristics.1,2 Oral manifestations usually appear in lepromatous leprosy and occur in 20–60% of cases. They may take the form of multiple nodules (lepromata) that progress to necrosis and ulceration.1 These lesions are usually located on hard and soft palate, uvula, tongue, lips and gingiva.1,2
Verrucous carcinoma is a low grade squamous carcinoma occurring most commonly in the oral cavity. It appears as a well demarcated exophytic growth showing fine papillary architecture. Its occurrence has been described as developing in chronic ulcers of skin in leprosy and diabetes mellitus with insufficient documentation.

Here we report probably the first documented case of verrucous carcinoma in the buccal mucosa in a patient with Hansen’s disease.

Case Report

A 60 year old male patient presented to the Oral Medicine and Radiology out-patient department with chief complaint of a growth on the inner surface of the cheek for 1 year. A detailed history revealed that it started as a painless ulcer a few years earlier. He was diagnosed as a case of lepromatous leprosy 5 years earlier, but had not received any treatment. He was a non-smoker, had no chewing habits (tobacco, areca nut, betel quid) and was a teetotaller, with unremarkable dental and family history. On general physical examination, the patient was conscious and orientated but evidently malnourished. His skin was dry, pale and sclerotic with several hypopigmented areas all over his body. Bilateral submandibular lymph nodes were palpable, partially fixed and tender. He had no motor nerve defect but sensory loss of the extremities with associated gross deformity of several digits on both feet.

Facial examination revealed numerous hypopigmented, polymorphous, poorly-defined skin lesions. The upper eyelids were swollen creating a sleepy appearance with sparse eyelashes. An opthalmic consultation revealed that there was damage to iris and sclera with complete blindness of the left eye. The ear showed multiple lepromatous nodules along the helix and lobule.

Intraoral examination demonstrated an exophytic, fungating mass on the buccal mucosa, on the right hand side, extending antero-posteriorly from the right commissure to approximately 3 cm anterior to the pterygomandibular raphe. The mucosa over the lesion was whitish, hyperplastic with papillomatous surface (Figure 1). On palpation, it was indurated and non-tender with mild sensory loss over the lesion. Examination of tongue revealed presence of a firm nodule, approximately 1 × 1 cm, on the dorsal surface of posterior one third of tongue along with atrophic changes of dorsal surface giving a pavement stone appearance (Figure 2). A scrapable white slough was present on the palate, which after scraping left an erythematous base. His dental health was very poor and revealed a few root remains in the lower jaw and edentulous maxilla.

On the basis of history and clinical examination a provisional diagnosis of verrucous carcinoma of buccal mucosa, leproma of the tongue and pseudo-membranous candidal infection of the palate was made. Orthopantomogram revealed marked destruction of the maxilla, more in the anterior region with respect to the residual alveolar ridges (Figure 3).

Punch biopsy was performed for the oral lesions on the buccal mucosa and the tongue under local anaesthesia after routine blood investigations. Histopathological report of the H&E stained sections of the growth on buccal mucosa showed hyperplastic keratinized epithelium with bulbous rete pegs and areas extending deep into the connective tissue. Other dysplastic changes along with keratin pearls were also evident (Figure 4).

Tongue sections showed hyperplastic epithelium with chronic inflammatory cells in the connective tissue along with degenerative changes (Figure 5).
Hence, a final diagnosis of verrucous carcinoma of buccal mucosa and leproma of the tongue was confirmed. The treatment plan consisted of advice for a balanced diet with a prescription for nutritional supplements, and a consultation with a leprologist for a comprehensive treatment of his general condition. Dental management, which included extractions of root remains, topical antifungal, excision of the verrucous carcinoma of buccal mucosa with placement of graft and prosthetic rehabilitation, was planned in our institute. The patient was, however, lost to follow up.

**Discussion**

Leprosy is a chronic granulomatous disease primarily affecting the skin and peripheral nervous system. The principal means of transmission of the *Mycobacterium leprae* is
by aerosol/droplet from infected nasal secretions to exposed nasal and oral mucosa.\textsuperscript{1,2} The worldwide prevalence of leprosy is reported to be just less than 1 case per 10,000 population. India, with an annual case detection rate of 53 per 100,000, accounts for 80\% of detected cases worldwide.\textsuperscript{4} The distribution of leprosy cases by clinical type varies among

\begin{figure}[h]
\centering
\includegraphics[width=\textwidth]{figure2.png}
\caption{‘Cobblestone’ appearance of the dorsum of the tongue due to a well demarcated depapillated and atrophic region along with the biopsy site.}
\end{figure}

\begin{figure}[h]
\centering
\includegraphics[width=\textwidth]{figure3.png}
\caption{Panoramic radiograph demonstrating marked destruction of the anterior maxilla in an otherwise edentulous upper arch.}
\end{figure}
countries. In India pure neuritic cases are more common, unlike in Central America where it is predominantly the lepromatous variety. The areas most commonly affected are the superficial peripheral nerves, skin, mucous membranes of the upper respiratory tract, anterior chamber of the eyes, and the testes which are the cooler parts of the body. Tissue damage depends on the degree to which cell-mediated immunity is expressed, the type and extent of bacillary spread and multiplication, the appearance of tissue-damaging immunologic complications (i.e. lepra reactions), and the development of nerve damage and its sequelae.5

Lepromatous leprosy usually begins in the form of chronic rhinitis leading to ulceration of the nasal mucosa. The facial skin shows areas of localised swelling, with deep furrows between them. The resultant nodules may become ulcerated. The swollen upper eyelids create a sleepy appearance. There may be damage to sclera and iris producing blindness. The nodular dermal manifestation often referred to as “leonine facies” is a facial deformity characteristic of leprosy.1 Multiple skin macules are present in various cooler regions of body appearing hypopigmented in dark skinned people and erythematous in those with light skin.

Leprotic oral lesions (Table 1),1,2,4–7 which are more common in the lepromatous form, indicate a late manifestation and have great epidemiological importance as a source of infection. These lesions develop insidiously and are generally asymptomatic. Development of ulcers on various sites in the body is an important complication in leprosy. The loss of sensation as a consequence of neural involvement of the disease and subsequent exposure to

Figure 4. H&E stained photomicrograph of verrucous carcinoma showing hyperplastic keratinized epithelium with bulbous rete pegs, areas extending deep into the connective tissue, dysplastic epithelial changes along with parakeratin plugging.
trauma are important predisposing factors. Therefore chronic irritation may be incorporated into multistep concept of cancer development as a promoting factor. Incidence of cancer is more common in lepromatous leprosy and it’s occurrence in chronic ulcers is well documented in literature but verrucous carcinoma of buccal mucosa in the oral cavity is a rare finding. Our case report is unique as there was a verrucous carcinoma on buccal mucosa inspite of the absence of any local causative agent.

Table 1. Orofacial manifestations of Hansen’s disease

<table>
<thead>
<tr>
<th>Region affected</th>
<th>Manifestations</th>
</tr>
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<tbody>
<tr>
<td>Tongue</td>
<td>Superficial erosions with loss of papillae and longitudinal fissures to nodular infiltration, ‘Paving stone appearance’ on the dorsal surface of tongue.</td>
</tr>
<tr>
<td>Lips</td>
<td>Macrocheilia, microstomia.</td>
</tr>
<tr>
<td>Palate</td>
<td>Enanthomas, erosions, ulcers, perforations and scars of hard palate.</td>
</tr>
<tr>
<td></td>
<td>Nodules (lepromata).</td>
</tr>
<tr>
<td>Teeth</td>
<td>Fibrosis or complete destruction of uvula.</td>
</tr>
<tr>
<td>Bones</td>
<td>Pink tooth due to infection of the pulp by infiltration of granulomatous tissue.</td>
</tr>
<tr>
<td></td>
<td>Loosening of teeth.</td>
</tr>
<tr>
<td>Regional nerve</td>
<td>Osteolysis of the premaxilla and alveolar process of the jaws.</td>
</tr>
<tr>
<td></td>
<td>Destruction of nasal bone and cartilage leading to nasal collapse.</td>
</tr>
<tr>
<td>Skin and adnexae</td>
<td>Hypopigmentation, loss of eye lashes and eyebrows, ‘sleepy eyes’, ‘leonine facies’.</td>
</tr>
<tr>
<td>Ophthalmic complications</td>
<td>Lagophthalmous, keratitis, conjunctivitis, uveitis.</td>
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Verrucous carcinoma is a low grade squamous cell carcinoma mainly recognized in three forms, all of them occurring in areas of maceration. Most common form occurs in oral cavity (oral florid papillomatosis). Second most common form is seen in anogenital region (called as Bushke Lowenstein tumor). Third, planter verrucous carcinoma called carcinoma cuniculatum, at first shows a striking resemblance to an intractable plantar wart. Classical verrucous carcinoma is an exophytic mass and microscopically it shows an exophytic and endophytic proliferation of well-differentiated squamous epithelium. In the oral cavity, proliferative verrucous leukoplakia represents its precursor although many cases are closely associated with use of smokeless tobacco or split tobacco. Our patient did not have any tobacco-related habits although he did suffer poor nutrition and poor dental health which may be the predisposing factors for the Ackerman’s tumour. In English literature adequate documentation exists for the development of this tumour in chronic ulcers on skin in leprosy and diabetes mellitus. However, a thorough PUBMED search revealed no prior report of oral verrucous carcinoma in leprosy patients.

To the best of our knowledge this is the first reported case of oral verrucous carcinoma in a leprosy patient. Further reports and documentation would be required to confirm this interesting association.

References