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Original Research Paper

Oral Medicine

HECK'S DISEASE – A RARE CASE REPORT

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ABSTRACT Heck's disease is a familial benign lesion affecting the oral mucosa and skin caused by Human papilloma viruses 13 and 32. This condition is quite rare in the Indian subcontinent and till date less than 10 case reports have been published. Females are predominantly affected. It is important for an oral physician to be aware and possess sufficient knowledge about these lesions as these lesions are highly contagious. Further, the presence of these lesions may indicate an underlying immunodeficient state of the patient. This paper reports an unusual case of Heck's disease in a 30 year old Indian male.

KEYWORDS : Focal epithelial hyperplasia, Heck's disease, Human Papilloma viruses-13 and 32

INTRODUCTION

Heck's disease was first described by Heck, Archard and Stanley in 1965 [1]. Several terminologies have been used to describe this disease namely focal epithelial hyperplasia, multifocal epithelial hyperplasia and multifocal papilloma virus epithelial hyperplasia. It is caused by Human papilloma viruses- 13 and 32 [2]. The virus induces localized proliferation of oral squamous epithelium. It is more common among Native Americans and Inuits. It is uncommon in Asian, European and African populations [3].

Very few cases of Heck's disease have been reported in Indian literature till date [4-7]. Heck's disease usually affects children and young adults and occasionally affects middle-aged adults. Females are more commonly affected than males. It is characterized by numerous soft, well-circumscribed papules or nodules commonly involving the labial, buccal and lingual mucosa. Histologically, acanthosis of the oral epithelium and koilocytes are observed. The present article reports one such case of Heck's disease in an Indian male with unusual oral findings.

CASE REPORT

A 30- year old male patient reported to the Department of Oral Medicine and Radiology, D. A. P. M. R. V Dental College, Bangalore, with a complaint of stains on his teeth. Incidentally, clusters of papules and nodules were noted on the lower labial mucosa and dorsal surface of the tongue. Patient was aware of these lesions since 10 years which gradually progressed in size without undergoing any secondary changes. The lesions were asymptomatic. No history of similar lesions elsewhere on the body. Patient reported no relevant medical or familial history. The lesions over the lower labial mucosa alone were surgically excised for cosmetic purpose a year ago. After which he developed new lesions in the same region within a span of 6 months which gradually progressed in size.

On general examination, patient was moderately built with normal gait and posture. All vital signs were within normal limits. No extra-oral abnormalities were detected.

On intra-oral examination, multiple pinkish-white, smooth surfaced papules measuring approximately 3x3mm were noted on lower labial mucosa adjacent to 31, 32 (Fig: 1) and labial and lingual attached gingiva with respect to 31, 32 (Fig: 2, 3). Numerous closely clustered papules each measuring approximately 5x4mm were noted on the left anterior dorsal surface of the tongue (Fig: 4). Two pales nodules with a papillary surface measuring 6x4mm and 4x2 mm were noted on the left dorsum (Fig: 4) and lateral border of tongue (Fig: 5) respectively. On palpation, the papules were non-tender and firm in consistency. The nodules were sessile, non-tender, firm, non-reducible, non-compressible, and non-pulsatile and with negative diascopy. Based on the patient's history and clinical presentation a provisional diagnosis of Heck's disease was arrived at.

The following differential diagnoses were considered- Squamous papilloma, Verruciform xanthoma, Verruca vulgaris, Condyloma acuminatum, Cowden's syndrome, Neurofibroma, Multiple endocrine neoplasia (MEN'S syndrome), Molluscum contagiosum as all these conditions present with asymptomatic, smooth surfaced papules or nodules on labial and buccal mucosa, tongue, gingiva in children and young adults.

Excisional biopsy of two papules on lower labial mucosa was performed under local anesthesia and sutures were placed. The postoperative healing was uneventful. The biopsy specimens (Fig: 6) were sent for histopathological evaluation. Histopathological report revealed stratified squamous epithelium with hyperparakeratosis and hyperplasia. The rete ridges were long, broad and confluent (Fig: 7). Mitoses was observed in the basal and parabasal layers. The spinous cell

layer showed epithelial clear cells with dark pyknotic nuclei suggesting koilocytes (Fig: 8). The underlying connective tissue contained collagen fibres, many blood vessels and few inflammatory cells.

Based on the patient's history, clinical features and histopathological report, a final diagnosis of Heck's disease was arrived at.

FIGURES

Fig:1 Multiple, pinkish-white, smooth surfaced papules measuring about 3x3mm on lower labial mucosa adjacent to 31, 32.

Fig: 2 Multiple, pinkish-white, smooth surfaced papules measuring about 3x3mm on labial attached gingiva.

Fig: 3 Multiple, pinkish-white, smooth surfaced papules measuring about 3x3mm on lingual gingiva with respect to 31, 32.

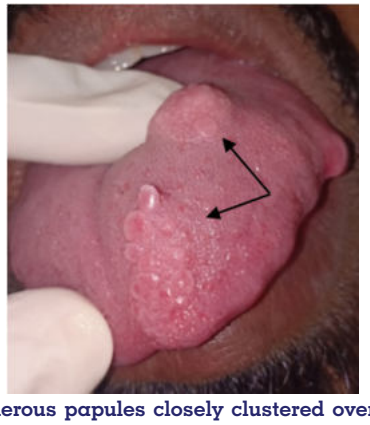

Fig: 4 Numerous papules closely clustered over tip of the tongue towards the left dorsal surface that measured 5x4 mm each and a pale nodule with papillary surface measuring 6x4mm on the left dorsum of tongue.

Fig: 5 A pale nodule with papillary surface measuring 4x2 mm on the left lateral border of tongue.

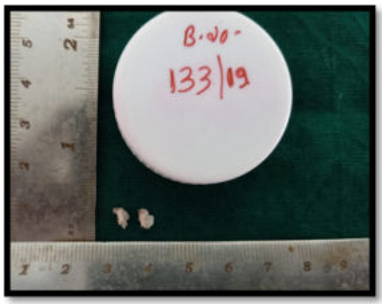

Fig: 6 Biopsy specimens sent for histopathological evaluation.

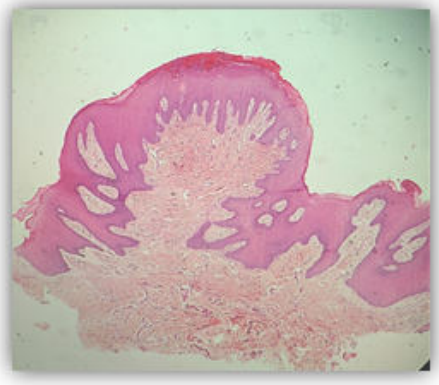

Fig: 7 Histopathological section under 4x reveals epithelium with hyperparakeratosis and broad confluent rete ridges.

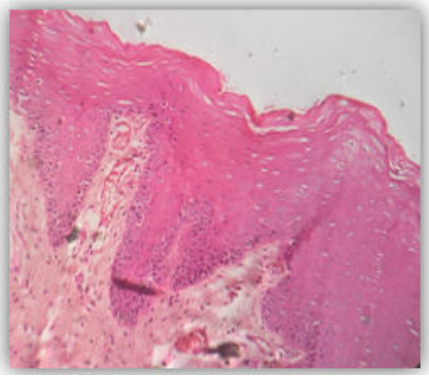

Fig: 8 Histopathological section under 10x reveals koilocytes and mitoses in basal and parabasal layers.

DISCUSSION

Heck's disease is a rare familial benign condition affecting the oral mucosa. The prevalence rate greatly varies from one geographical region to another (0.002%–35%) [9]. It is commonly found in Native Indian residents in North, Central and South America (40%), Eskimos (7-36%), and descendants of Khoi-San in South Africa (7-13%) [8]. It is less common in

Europe, Africa, and Asia [2]. Recent findings among Puerto Ricans and blacks suggest that this lesion is quiet wide spread.

In Indians, its occurrence appears to be sporadic and so far less than 10 cases have been reported [4-7].

It is caused by Human papilloma virus types 13 and 32. It is proposed that the HLADR4 allele is linked to the increased susceptibility to HPV infection particularly HPV-13 [10]. The possible risk factors include lower socio-economic status, close living conditions, poor hygiene, dietary insufficiency and immunodeficient states such as HIV infections [8, 11]. Family history may be positive in most of the patients. In the present case, there was no familial history, absence of an immunodeficient state and the possibility of other risk factors contributing to the lesion was less known.

Heck's disease occurs predominantly in females with F: M ratio of 5:1 [8]. It mainly affects children with lower labial mucosa being the most common involved site [11]. Other sites include buccal and lingual mucosa. In the present case, the lesions presented in a male at an age of 20 years. Multiple sites in the oral cavity were involved such as lower labial mucosa, labial and lingual gingiva and dorsal surface and left lateral border of the tongue.

Clinically, the condition presents as asymptomatic multiple soft, flattened or round papules which are usually clustered although may be scattered. The papules are usually pale pink and rarely white. Individual lesions vary in size from 3 to 10mm [12]. According to Said AK et al 2013, there are two distinct clinical forms of the condition-(1) papulonodular commonly involving buccal and/or labial mucosa and commissures; (2) papillomatous commonly involving the tongue and gingiva. The papulonodular variant is more common than the papillomatous type [8]. In the present case, both the clinical variants of the lesion were evident. The lesions may increase in size, remain unchanged or may undergo spontaneous regression within months to years. In the present case, the lesions persisted and progressed over a period of 10 years. In the present case, provisional diagnosis of Heck's disease was made on the basis of patient's history and clinical features.

A number of differential diagnoses were considered. Squamous papilloma and Verruciform xanthoma were ruled out as they usually present as solitary reactive lesions and, in most cases, an irritant can be identified. Verruca vulgaris commonly affects the skin rather than oral mucosa; lesions of Condyloma acuminatum are usually larger in size (1-3 cm), thus were excluded. Conditions such as Cowden's syndrome, Neurofibroma, Multiple endocrine neoplasia (MEN'S syndrome), Molluscum contagiosum were also excluded as they are associated with several systemic and skin manifestations which were absent in the present case.

The diagnosis is often made based on patient's history, clinical examination followed by histopathological examination for confirmation [8]. Histological findings such as abrupt acanthosis of oral epithelium, koilocytic changes are the characteristic features of this lesion. The other methods of detection of HPV within the lesional tissue include PCR, immunohistochemical analysis, in situ hybridization [8, 11]. In the present case, the histopathological evaluation confirmed the diagnosis of Heck's disease.

As the lesions are asymptomatic, often regress spontaneously and have no malignant potential, the lesions are excised primarily for aesthetic or functional reasons. The first line of therapy includes conservative surgical excision, cryotherapy, CO_2 laser ablation or electrocoagulation/ electrodessication [8, 11]. The other modalities of treatment described in literature [8, 11] include topical Imiquimod (5% cream), Interferon-beta [13], trichloroacetic acid (80%concentration) [14], intralesional interferon alpha, systemic retinoids. However, in the present case, the patient being asymptomatic refused treatment.

Recurrence may occur despite effective initial treatment or spontaneous regression. Recurrence and the site of new lesions are unpredictable thus; periodic review of the patient is often necessary [8]. A few reasons such as latent infection, altered immune response or new infections may be attributed for the recurrence of lesions. In the present case, the patient revealed the recurrence of the lesions over the lower labial mucosa within 6 months after conservative excision of the previous lesion for cosmetic purposes. Hence, long-term follow up is advised for monitoring the status of the lesions.

The table no.1 summarizes the cases of Heck's disease reported in Indian literature.

CASE REPORTS	AGE& GENDER	CLINICAL FEATURES		DURATION	OTHERS	TREATMENT
		EXTRAORAL	INTRAORAL			
M.P.V Prabhat et al 2013[4]	65yrs/ Female	Multiple warts involving face, scalp and trunk	Predominant papillary lesion involving multiple sites like gingiva, palate, tongue & commissural areas of both right and left buccal mucosa	6-7 months	PCR revealed presence of HPV type 16	No treatment Periodic follow up
D. Asha et al 2015[5]	8yrs/ Male	-	Single, white, plaque, of 1 × 1.5 cm in size on the anterior one third of the tongue	1 year	-	No treatment
Nallanchakravarthy S et al 2018[6]	5yr/ Male	-	Soft, sessile papules varying 2–10cm in dimension, two papules present on the right and left the side of the lower lip region, and one on the left ventral aspect of the tongue	3 months	PCR revealed the presence of HPV type 32	Diode soft tissue laser excision (810 nm of 3–3.5W power for 3–60 seconds intermittently)
Karthikeya Patil et al 2019[7]	65yrs/ Male	-	Multiple grayish white papules over the buccal mucosa bilaterally and on the upper and lower labial mucosa measuring 1 to 4 mm in size	-	H/o smoking 20beedis/ day for past 30 yrs (possible attribute to the disease)	No treatment Long term follow up advised

Present case report	30yrs/ Male	-	Multiple pinkish-white, smooth surfaced papules 3x3mm approx. noted on lower labial mucosa, labial and lingual attached gingiva with respect to 31, 32. Numerous closely clustered papules each 5x4mm approx. on the left anterior dorsal surface of the tongue. Two nodules measuring 6x4mm and 4x2 mm with a papillary surface on the left dorsum and lateral border of tongue respectively.	10 years	H/o cosmetic excision on the lesions over labial mucosa a year before and new lesions developed in the same region in a span of 6 months.	No treatment Long term follow up advised

CONCLUSIONS

Heck's disease is a rare condition encountered in the Asian populations including Indians. Although it has no malignant potential and carries no risk of morbidity, it is caused by HPV which is highly contagious. Furthermore, the presence of this disease may indicate an underlying immunodeficient state of the patient. This emphasizes the need for oral physicians to possess adequate knowledge about the condition which would help in appropriate intervention and timely management.

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