Focal Epithelial Hyperplasia (Heck's disease) – Case report in a Nigerian child

and short review of the literature.

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ABSTRACT

Focal epithelial hyperplasia or Heck's disease is an oral mucous membrane disease caused by the human papillomavirus. It presents with multiple papules or nodules in the oral cavity. It is a rare disorder commoner in children but can occur in any age. The clinical presentation is however somewhat different in children and in adults. Despite the fierce appearance and the viral etiology of this condition, it is a relatively benign condition that resolves spontaneously over time.

INTRODUCTION

Focal epithelial hyperplasia or Heck's disease is a rare viral infection of the oral mucosa caused by human papillomavirus. It was first reported by Archard et al[1] in 1965 in Inuit and Indian children from North and South America but since then it has been reported from other parts of the world[2,3]. The frequency of this disease varies widely from one geographic region to another[4]. Sporadic reports have been reported all over the globe including a few cases from Nigeria[5-6]. It is a condition that can present either to the dermatologist or to the oral physicians (dentists and orthorhinolaryngologists) and these specialists must be aware of it despite its rarity.

The patient presented below is one of these rare cases from south-south Nigeria and this will hopefully contribute to the literature on this rare entity that a dermatologist might sometimes come across.

CASE REPORT

A four years old girl presented to our clinic with multiple warty growths on the mucosal of both the upper and lower lips starting a year ago - Figures 1 & 2. The growth has been spreading inwardly despite being completely asymptomatic. There were no constitutional or any other systemic symptoms. No similar symptoms or lesions in the parents. She is the first of the two children of the parents and the other sibling does not have a similar illness.

HIV testing was negative in the child and parents.

Histology of biopsy specimen was in keeping with a diagnosis of Heck's disease. Other sophisticated investigations such as a polymerase chain reaction (PCR) and sequencing could not be done due to limited resources and non-availability. The parents were counseled on the generally benign nature of the pathology and the child is being closely followed up for any development of unusual symptoms.

DISCUSSION

Focal epithelial hyperplasia or Heck's disease is a rare viral infection of the oral mucosa caused by human papillomavirus. It is a benign condition of the oral mucosa produced by the subtypes 13 or 32 of human papillomavirus (HPV) [7]. It primarily occurs in children with no gender predilection[8]. The subtype 32 of HPV tends to cause the disease in the older age groups while the subtype 13 of HPV seems to be equally involved in the development of the disease in both young and old patients[9]. It was first described by Archard et al[1] in Native Americans in 1965 but since then several cases has been reported from other parts of the world including those from African countries[10]. Sawyer et al[5] and Akinwande et[6] al had reported similar cases from the western part of Nigeria however this is the first case reported from the southern part of this country. The frequency of this disease varies widely from one geographic region

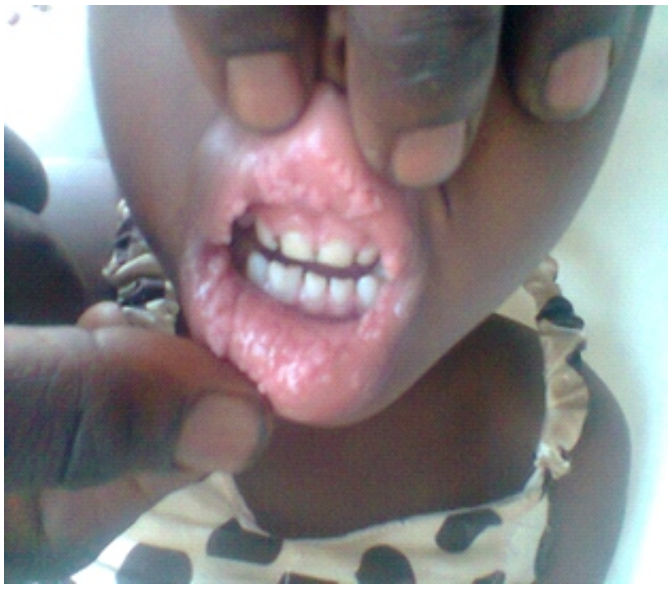

Figure 1: Multiple warty growth

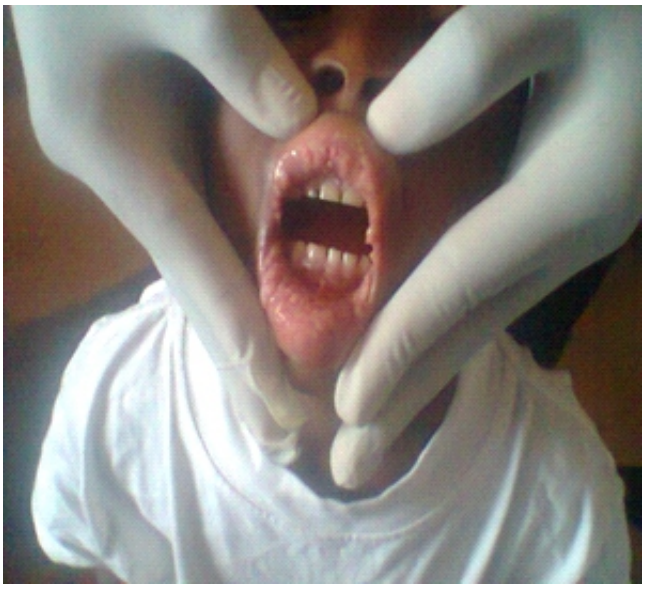

Figure 2: Multiple mucosal growth

to another[4] and in numerous populations and ethnic groups. A higher incidence in close communities and among family members is suggestive of an infective pathogenesis[11]. Communal living[12] and eating and drinking together from the same plate is postulated to be the main mode of transmitting the disease in earlier publications however this has not been confirmed by other reports of the condition.

FEH is characterized by the occurrence of multiple or unique whitish or normal coloured papules or nodules in the oral cavity (as seen in the case above), mostly on the labial and buccal mucosa, the lower lip and tongue, and less often on the upper lip, gingiva and palate[13]. Younger patients with FEH often have multiple lesions (as seen in our patients above), whereas older patients may have few or even single lesions, which tend to be flat and papular. A site-specific predilection for keratinized and non-keratinized surfaces has been observed in these HPV infections[3].

The diagnosis of FEH can usually be made on the basis of clinical observations but histological examination may show characteristics of viral infection (parakeratosis, epithelial hyperplasia, focal acanthosis, fusion, and horizontal outgrowth of epithelial ridges and the cells named mitozoids) [13]. However polymerase chain reaction (PCR) is a more sensitive and useful investigation to identify the viral etiology of FEH lesions where available[14].

The main differential diagnosis in tropical dermatologic practice is condyloma acuminatum because of the high prevalence of HIV infection. This is because suppression of the immune system leaves the patient vulnerable to opportunistic infections, particularly HPV infections[15-16]. The early detection of some oral diseases including FEH sometimes results in earlier diagnosis of HIV infection. Other rare conditions to be excluded during evaluation include inflammatory fibrous hyperplasia, inflammatory papillary hyperplasia, verruciforme xanthoma, verrucous carcinoma, Cowden's disease and focal dermal hypoplasia syndrome (Goltz–Gorlin syndrome)[17].

Treatment ranges from doing nothing to laser surgery to topical or injectable chemotherapy[18]. This condition tends to be more persistent in patients with HIV infection and can be resistant to treatment[16]. However in otherwise healthy children, some regress spontaneously and others respond to liquid nitrogen or laser treatment. Other forms of therapy include intra-lesional injections or topical chemotherapy, cryotherapy, electrocoagulation, treatment with carbon dioxide laser or systemic treatment with interferon-α or topical treatment with interferon-β and retinoic acid.

Despite its viral etiology, the disease is a relatively benign condition and spontaneous resolution is the usual course though it may last for several months, or years, before running its course[9]. This may be related to the less developed immunologic system in children and the disappearance of the lesions as the child's immune system matures. This may also explain why these lesions are rarely found in adults.

Current research has shown not shown any malignant potential complicating cases of FEH lesions associated with HPV 13 and 32 subtypes[19].

REFERENCES

1. Archard HO, Heck JW, Stanley HR. Focal epithelial hyperplasia: an unusual mucosal lesion found in Indian children. Oral Surg. 1965;20:201–212.
2. Axéll T, Hammarström L, Larsson A. Focal epithelial hyperplasia in Sweden. Acta Odontol Scand. 1986;39:201–205.
3. Bassioukas K, Danielides V, Georgiou I, Photos E, Zagorianakou P, Skevas A. Oral focal epithelial hyperplasia. Eur J Dermatol. 2000;10:395–397. [PubMed]
4. Praetorius-Clausen F. Geographical aspects of oral focal epithelial hyperplasia. Pathol Microbiol. 1973;39:204–213. [PubMed]
5. D.R Sawyer, G.Arole, A. Mosadomi. Focal epithelial hyperplasia. Report of three cases from Nigeria, West Africa. Nigeria Oral Surgery, Oral Medicine, Oral Pathology. 09/1983;56(2)185-189.
6. Akinwande J.A, Arain A, Taiwo E.O. Focal epithelial hyperplasia (Heck`s disease).Report of case. Nigerian Dental Journal.1986;7:53-56.
7. Bassioukas K, Danielides V, Georgiou I, Photos E, Zagorianakou P, Skevas A.Oral focal epithelial hyperplasia. Eur J Dermatol 2000; 10(5): 395–397.
8. Silverberg N. Human papillomavirus infections in children. Curr Opin Pediatr. 2004 Aug;16(4):402-409.
9. Summersgill KF, Smith EM, Levy BT, et al. Human papillomavirus in the oral cavities of children and adolescents. Oral Surg Oral Med Oral Pathol Oral Radiol Endod. Jan 2001;91(1):62-69.
10. Chindia ML, Awange DO, Guthua SW, Mwaniki DL. Focal epithelial hyperplasia (Heck's disease) in three Kenyan girls: case reports. East Afr Med J. 1993 Sep;70(9):595-596.
11. Harris AMP, van Wyk CW. Heck's disease (focal epithelial hyperplasia): a longitudinal study. Community Dent Oral Epidemiol 1993; 21(2):82–85.
12. Dos Santos PJ, Bessa CF, de Aguiar MC, do Carmo MA. Cross-sectional study of oral mucosal conditions among a central Amazonian Indian community, Brazil. J Oral Pathol Med 2004; 33(1):7–12.
13. González LV, Gaviria AM, Sanclemente G, Rady P, Tyring SK, Carlos R, Correa LA, Sanchez GI. Clinical, histopathological and virological findings in patients with focal epithelial hyperplasia from Colombia. Int J Dermatol. 2005 Apr;44(4):274-279.
14. Jayasooriya PR, Abeyratne S, Ranasinghe AW, Tilakaratne WM. Focal epithelial hyperplasia (Heck's disease): report of two cases with PCR detection of human papillomavirus DNA. Oral Dis 2004; 10(4):240–243.
15. Marvan E, Firth N. Focal epitelial hyperplasia in an HIV positive man. An illustrated case and review of the literature. Aust Dent J 1998; 43(5):305–310.
16. Moerman M, Danielides VG, Nousia CS, Van Wanzeele F, Forsyth R, Vermeersch H. Recurrent focal epithelial hyperplasia due to HPV13 in an HIV-positive patient. Dermatology 2001; 203(4):339–341.
17. Gorlin R, Cohen MM et al. Syndromes of the Head and Neck. 1990 Oxford University Press. Pages 357-360.
18. Steinhoff M, Metze D, Stodefleth E, Luger TA. Successful topical treatment of focal epithelial hyperplasia (Heck's disease) with interferon-beta. Br J Dermatol2001; 144(5):1067–1069.
19. Bennett LK, Hinshaw M. Heck's disease: diagnosis and susceptibility. Pediatr Dermatol. 2009 Jan-Feb;26(1):87-89.