Focal epithelial hyperplasia (Heck’s disease): a case report from Iran

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INTRODUCTION

Focal epithelial hyperplasia, or Heck’s disease, is a relatively rare benign disease which is characterized by multiple circumscribed soft elevated papules. The papules usually have the color of the normal mucosa but may be white or pale. Heck’s disease is common in native South Americans, native Greenlanders, and in South African communities and is rare in Caucasians. Heck’s disease is especially associated with HPV-13 or -32. HPV 1 and 18 were demonstrated in rare cases. This disease was first described by Archand et al in 1965. It is also known as hyperplasia multilocularis mucosae oris.

Based on the study population and geographic region, the prevalence of this disease varies from 0.002 to 35%. This disease often regresses spontaneously in most cases although it can persist through old age. It is not difficult to make a diagnosis through accurate clinical assessment and review of the patient’s medical history.

Pathological examination shows acanthosis, bullous extension of rete ridges, and focal or diffuse ballooning degeneration of the epithelium. There are viral particles in lesional keratinocytes, and viral antigens have been detected by polymerase chain reaction (PCR). Treatment is not necessary for all patients.

CASE REPORT

A 47-years-old woman referred to Farshchian Hospital, Hamedan, Iran, with oral cavity lesions. They developed spontaneously when the patient was 10 years old. The patient received several treatments with no healing during these years. Her lesions were on both bucal mucosa, lower lip and tongue. Other parts of the mouth were not involved. Except for aesthetic problems, she had no complaint and was symptom free. On clinical assessment, papules and papillary form lesions with a color similar to the oral mucosa were seen. The size of lesions was about 2 to 7 mm in diameter (Figure 1). On physical examination, other parts of the skin and mucosal membrane were normal. Familial history
and past medical history were negative and she had no history consistent with immunodeficiency.

After a clinical diagnosis of focal epithelial hyperplasia, an incisional biopsy was taken from the buccal mucosa. Histopathology reported prominent acanthosis, hyperkeratosis, and parakeratosis with elongated and broad rete ridges. The epidermis was composed of mature stratified squamous epithelium. Focal pallor of epithelial cells in some areas was noted. Few mitotic activities and a mild lymphocytic infiltration were also observed in the underlying stroma (Figure 2). PCR showed HPV 13 DNA. All these clinical and histopathological data confirmed the diagnosis of focal epithelial hyperplasia.

**DISCUSSION**

Heck’s disease has been reported particularly in Indians and Eskimos but few Caucasian cases have also been reported. Most patients are children but adults may also be at risk.\(^9\)\(^-\)\(^11\) This disease may
continue for years with a significant reduction in the quality of life, but spontaneous remission of FEH is probable, too. Affected areas in order of frequency are the labial mucosa, buccal mucosa, tongue and the lingual mucosa. Gingival and tonsillar lesions have also been reported. The palatal mucosa and oropharynx are usually intact. There are reports of the involvement of several members of one family and genetic predisposition is suspected. Malignant transformation does not occur in FEH. Clinical presentation of the disease is multiple, soft, elevated papules with a whitish color or color similar to other mucosa. The size of the papules varies from 1 to 5 mm. The lesions are usually painless.

Differential diagnoses include verruciform xanthoma, verrucous carcinoma, Cowden’s disease, Crohn’s disease, Cannon’s disease (White Sponge Nevus), condyloma acuminatum, florid oral papillomatosis, and inflammatory papillary hyperplasia. Due to the similarity with condyloma acuminatum, it is important to differentiate between them. PCR is useful for differentiation. Histopathology shows acanthosis with broadening and elongation of the rete ridges. The rete ridges are widened and are often connected together. Epithelial cells show vacuolization changes which is most obvious in the upper portion. Histopathological analysis is necessary to establish a definitive diagnosis.

If the patient is symptomatic or if cosmetic problems are significant, treatment is necessary. Surgical techniques are excision, electrocauterization, cryotherapy and curettage, especially for localized lesions. Systemic treatments include acitretin, etretinate, interferon-α2, and methotrexate. Carbon dioxide laser is useful, too. It seems that CO2 laser surgery is one of the best treatment modalities. Because of the side effects and risks of other treatments, topical agents such as interferon-β and imiquimod are preferred. Imiquimod has been reported to be a good treatment in children.

REFERENCES