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Case Report

Penile verrucous lesion: not necessarily a genital wart

陰莖疣狀疹：不一定是性病疣

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A 46-year-old Chinese man presented with one year history of itchy verrucous lesions over penis and scrotum. Skin biopsy confirmed epidermolytic acanthoma. Epidermolytic acanthoma is a rare benign tumour. Before making such a diagnosis, exclusion of other diseases, especially genital warts and bowenoid papulosis is necessary. Treatment of multiple epidermolytic acanthoma remains unsatisfactory.

此病例之患者是一位 46 歲男性，近一年來於陰莖及陰囊出現癢性疣狀疹。皮膚組織病理標本檢查確診為表皮鬆解性棘皮症。表皮鬆解性棘皮症是一種罕有的良性腫瘤。要診斷表皮鬆解性棘皮症，必須先排除其他疾病，特別是性病疣及鮑温樣丘疹病。治療此病的效果並不理想。

Keywords: Epidermolytic acanthoma, epidermolytic hyperkeratosis

關鍵詞：表皮鬆解性棘皮症、表皮鬆解性角化過度症

Introduction

Epidermolytic acanthoma is a rare benign tumour that shows characteristic features of epidermolytic hyperkeratosis on histology. The aetiology of epidermolytic acanthoma is not defined yet though a mutation in K1 and K10 has been suggested in the pathogenesis of this rare tumour. It usually presents as solitary or multiple verrucous papules or plaques over the trunk, scrotum or the light-exposed skin of middle-aged people.

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Case report

A 46-year-old gentleman was seen at the Social Hygiene Clinic because of itchy verrucous papules on the penis and scrotum. The patient had been well until one year earlier, when he noticed a 2 mm itchy verrucous papule on his penis. Several months later, he noticed several new verrucous papules on his penis and

scrotum. He recalled venereal exposure involving a female sex worker one year before his first visit to the clinic. The lesions grew from between 2 and 4 mm in diameter to nearly 10 mm in about one year.

The patient had no fever. There was no history of ichthyosiform processes, genital warts or recent travel. No other relevant personal or family history was noted.

On examination, several verrucous papules were found on the penis (Figures 1 and 2). No other cutaneous abnormalities were noted on examination.

Laboratory investigations revealed that renal and liver function tests were normal and syphilis and HIV serology were negative.

The differential diagnoses were genital warts, bowenoid papulosis, penile malignant growth, epidermolytic acanthoma and lichen planus.

This gentleman underwent skin biopsy and the histopathology demonstrated an acanthotic epidermis, with reticular degeneration and occasional eosinophilic keratin inclusion in the upper epidermis (Figures 3 and 4). These features represent epidermolytic hyperkeratosis and are consistent with the diagnosis of epidermolytic acanthoma.

The diagnosis of epidermolytic acanthomas of the penis was made based on the histopathological findings. The primary lesion was excised and we offered treatment of the other lesions by cryotherapy, but the patient was then lost to follow-up.

Discussion

Epidermolytic acanthoma is a rare benign cutaneous tumour. Although usually solitary, a clinical variant known as disseminated epidermolytic acanthoma has been documented. The isolated form of epidermolytic acanthoma was first described by Shapiro and Baraf[1] in 1970 as a distinctive clinical entity showing epidermolytic hyperkeratosis histologically in the

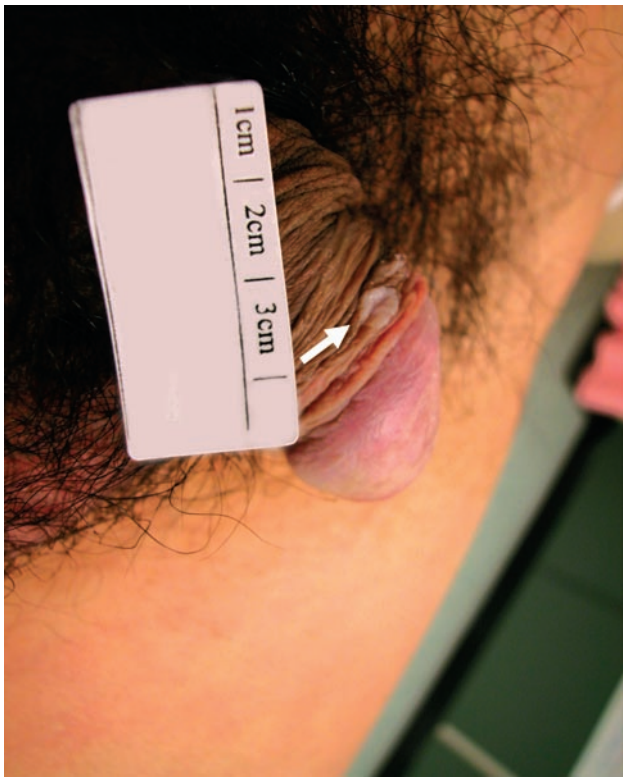

Figure 1. A 2 mm verrucous papule was found on the penis.

Figure 2. Another 1 mm verrucous papule on the penis.

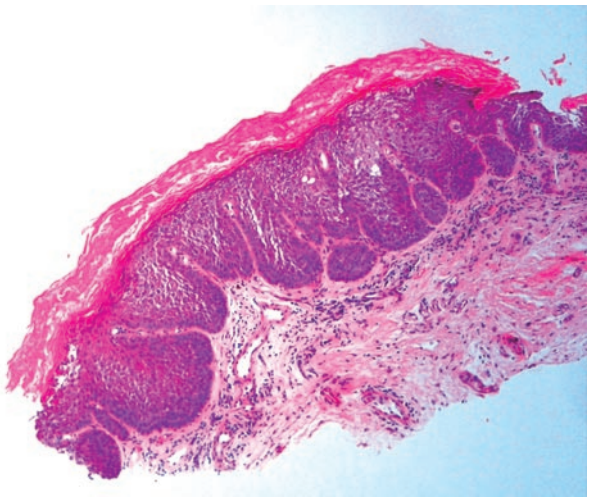

Figure 3. The epidermis shows a defined area of compact orthokeratosis and acanthosis. (H&E, Original magnification x 5)

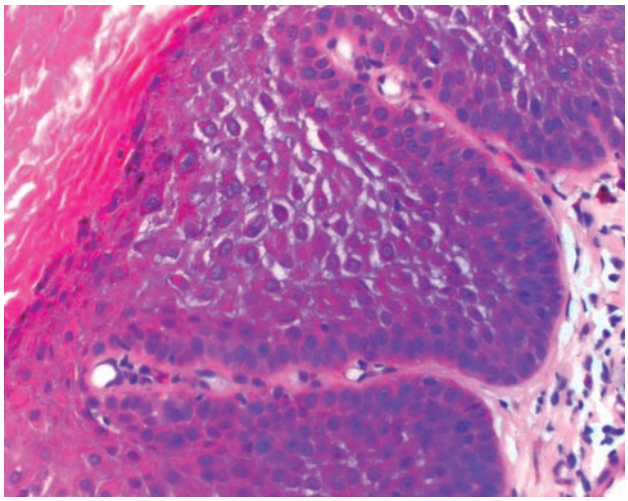

Figure 4. High power view shows vacuolar change of keratinocytes in the upper epidermis. (H&E, Original magnification x 20)

acquired solitary lesion. In 1973, the term disseminated epidermolytic acanthoma was described by Hirone and Fukushiro in a patient who had multiple lesions on the trunk, upper limbs and shoulders.[2]

In the isolated form, the trunk, scrotum, head and neck or the light-exposed skin of middle-aged people are the usually affected sites. The scrotum, trunk, scalp and lower extremities are commonly affected in the disseminated form.[3] Men were believed to be disproportionately more affected by epidermolytic acanthoma compared to women. However, there was no reported difference in frequency between race and sex. Epidermolytic acanthoma can occur at any age, but it most often occurs in middle-aged and older persons. The mean age of onset of disease is 40-60 years,[3] although the range is broad and disease arising in the elderly and children has been described.

In 1997, Cohen[3] reviewed 37 reported cases and classified epidermolytic acanthomas according to the number of lesions on the affected individual rather than by location. In 'solitary epidermolytic acanthoma', the lesion is "isolated" to a single site whereas in 'multiple epidermolytic acanthoma', the lesions may be either "localised" or "disseminated".

The clinical features of epidermolytic acanthoma are rather non-specific, and can resemble a variety of dermatoses, including seborrhoeic keratosis, viral warts and even Bowen's disease. In cases of disseminated variants which involve other parts of the body, the clinical appearance may sometimes be more confusing.

The cause and the mechanism involved in the pathogenesis of epidermolytic acanthoma are not yet known. Postulations include mechanisms involving immunosuppression, HPV infection, trauma and other genital disorders. The role of altered K1 and K10 genes in the pathogenesis of epidermolytic acanthoma has been revealed.[3] Cohen demonstrated that K1 and K10 expression in solitary epidermolytic acanthoma was less than that in normal skin.

It was once believed that there was an association between human papillomavirus (HPV) infection and epidermolytic acanthoma because of the clinical similarity of epidermolytic acanthoma and genital warts. Moreover, the histologic features of epidermolytic acanthoma mimic those of

human papillomavirus infection. Leonardi[4] examined eight cases of epidermolytic acanthoma for the presence of HPV types 6, 11, 16, 18, and 33 using polymerase chain reaction followed by dot-blot analysis. He demonstrated that human papillomavirus DNA was absent in epidermolytic acanthoma and concluded that epidermolytic acanthoma was not associated with the correspondent HPV infection.

Many other factors were also postulated as the aetiologies of epidermolytic acanthoma. Case reports showed that "multiple epidermolytic acanthoma" were seen in patients following severe sunburn,[5] long term immunosuppression[6] or during PUVA therapy.[7] Therefore, immunosuppression was suggested as a factor in the pathogenesis. Repeated trauma was also suggested as another causative factor.[8,9] However, the exact causation and mechanism remains unknown.

Because of the non-specific features of epidermolytic acanthoma, a biopsy is often required to establish the diagnosis. According to Ackerman,[10] characteristic histological features of epidermolytic hyperkeratosis include: (1) perinuclear vacuolisation, of variable size, of the keratinocytes in the stratum spinosum and stratum granulosum; (2) indistinct cellular boundaries; (3) a markedly thickened granular layer with increased numbers of keratohyalin granules; and (4) hyperkeratosis. In fact, because of its non-specific features, epidermolytic acanthoma is probably an under-reported benign cutaneous neoplasm.

Treatment options depend on the number of the lesions in the affected person. Treatment of isolated epidermolytic acanthoma is not essential as the lesions are benign and asymptomatic. Surgical excision, cryotherapy, curettage and laser ablation are options if treatment is contemplated.

Treatment of "multiple epidermolytic acanthoma" is challenging. Surgical and other destructive modalities may have poor cosmetic results. Topical 5% imiquimod cream has been shown to be effective in an anecdotal report.[11]

In summary, epidermolytic acanthoma is a rare benign cutaneous tumour of unknown aetiology that commonly affects middle-aged men. Treatment is not essential as it is benign and asymptomatic. Surgical excision and other destructive modalites can be adopted if treatment is contemplated.

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