

Case 2: Epidermolytic Acanthoma

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Date:	10 December 1997
Venue:	Yaumatei Skin Center
Organizer:	HKSDV, Clinico-pathological Meeting

CASE SUMMARY

History

Mr. Chen, a 61-year old teacher, noted multiple brownish papules over his scrotum for many years. They gradually increased in size and number, and were asymptomatic. There was no history of venereal exposure. There was no family history of similar disease or ichthyosiform disease. He was treated with caustic solution by private doctor with no improvement. His past health was unremarkable.

Examination

There were multiple skin-coloured keratotic papules of irregular size and shape over his scrotum (Figure 1).

Differential diagnoses

The diagnoses considered include viral wart, seborrhoeic keratosis, skin fibroepithelioma, Bowenoid papulosis, and epidermolytic acanthoma.

Investigations

Skin biopsy showed focal areas of hyperkeratosis, papillomatosis, and epidermolytic hyperkeratosis. The latter was limited to the upper stratum malpighii, with intracellular and intercellular epidermal cellular edema, coarse keratohyaline granules and dyskeratosis.

Diagnosis

The diagnosis of Epidermolytic Acanthoma (EA) was made histologically.

Discussion

It was raised whether Epidermolytic Acanthoma was a distinct disease entity or just an reactive pattern and CO₂ laser might be helpful in our patient.



Figure 1: Multiple hyperkeratotic papules over the scrotum

The pathologist thought that Epidermolytic Acanthoma was probably a distinct disease due to a defect of keratinization, and the presence of E.A. in various skin disease could well be due to spontaneous mutation of the keratinization gene.

REVIEW ON EPIDERMOLYTIC ACANTHOMA

It is an uncommon benign skin tumor firstly described as an entity by Shapiro and Baraf in 1970, clinically appearing as a solitary acquired verrucoid papule. It shows light-microscopic features of epidermolytic hyperkeratosis.

Aetiology

The cause is unknown but no human papillomaviral DNA (types 6,11,16,18,33) can be isolated by the polymerase chain reaction¹. In case of disseminated epidermolytic acanthoma, immunosuppressive status² and PUVA therapy³ may be the possible etiology.

Clinical features

It can be solitary, usually reported to occur over the genital area (but can occur anywhere), or disseminated. In disseminated cases, immunosuppression and PUVA therapy may be associated. Clinically it may look like viral wart⁴, molluscum contagiosum, skin fibroepithelioma, or Bowenoid papulosis. Usually they are asymptomatic.

Histological features

Epidermolytic hyperkeratosis (EH) with granular degeneration is the main feature. Other characteristics included epidermal acanthosis, compact papillomatous hyperkeratosis, and dissolution of the suprabasilar epithelium, resulting in perinuclear clear zones. Granular keratohyaline clumping, hypergranulosis, and dyskeratosis resulting in intracellular eosinophilic globules, hence the "cells within a cell" appearance. The findings with hematoxylin-eosin stain and Fontana-Masson stain indicated that the tumor is originated from the hair follicle. However, EH can occur in 3 hereditary conditions: bullous congenital ichthyosiform

erythroderma, systematized nevus verrucosus and hereditary palmoplantar keratoderma of Verner. EH can also be seen in some acquired skin disorders such as pilar cyst, seborrheic keratosis, cutaneous horn, actinic keratosis, basal and squamous cell carcinoma, leukokeratosis, intradermal nevus, malignant melanoma, granuloma annulare and lichen amyloidosis⁵.

Diagnosis is made in the context of clinical and histologic features. It is important to differentiate Epidermolytic Acanthoma from other more serious conditions associated with epidermolytic hyperkeratosis.

Management

No treatment is required. In case of solitary lesion, excisional biopsy may be both diagnostic and therapeutic.

Learning points:
Epidermolytic acanthoma is frequently misdiagnosed as genital wart.

References

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