棘層鬆解性棘皮瘤

－二例病例報告－

余文欽　廖怡華*　蔡呈芳*　蕭光祥*　蕭正祥**
台北市立和平醫院 皮膚科 國立台灣大學附設醫院 皮膚科* 病理科**

Acantholytic Acanthoma

－Two Case Reports－

Wen-Chin Yu　Yi-Hua Liao*　Tsen-Fang Tsai　Guang-Hsian Hsiao*
Cheng-Hsian Hsiao**

Acantholytic acanthoma is a benign acanthoma unrecognized previously, which was first described by Brownstein in 1985. Here we report two cases of acantholytic acanthoma, the first case was a 62-year-old male patient presented with one erythematous, hyperkeratotic, rice grain-sized papule with elastic, firm consistency over his left axilla for several months. Histopathological examination of excisional biopsy revealed focal acanthosis with suprabasal clefts and intraspinous acantholysis of epidermis. The second case was a 68-year-old female patient presented with one brownish, bean-sized, hyperkeratotic plaque on her right upper arm for several months without any symptom. Microscopic study showed hypergranulosis and acanthosis with prominent suprabasal clefts and intraspinous acantholysis of epidermis. Both of these two cases were typical acantholytic acanthoma with characteristic histopathologic features and differentiations from other disease with acantholytic dyskeratosis were also made. (Dermatol Sinica 19 : 345-350, 2001)

Key words: Acantholytic acanthoma, Benign acanthoma, Acantholysis

棘層鬆解性棘皮瘤首先於1985年由Brownstein提出，是一種先前尚未被分類的良性棘皮瘤。本篇文章報告二例具有典型病理變化之棘層鬆解性棘皮瘤。第一例是一位62歲男性病人，在其左側腋下發現一個米粒大小，紅色具有彈性，硬的圓形小丘疹。組織病理學檢查發現表皮層有局部棘皮層增生，以及基底層裂隙和棘層鬆解的現象。第二例爲一位68歲女性病人，在其右上臂發現一個咖啡色，蠶豆大小的斑塊，其組織病理學檢查可以發現在表皮層有顆粒層及棘皮層的增生，並伴隨有明顯的基底層裂隙和棘層鬆解的現象。本文並討論了其他需要與棘層鬆解性棘皮瘤鑑別診斷的疾病。(中華皮誌19 : 345-350, 2001)

From the Department of Dermatology, Taipei Municipal Hoping Hospital, Departments of Dermatology and Pathology,** National Taiwan University Hospital*
Accepted for publication: July 9, 2001
Reprint requests : Wen-Chin Yu, M.D., Department of Dermatology, Taipei Municipal Hoping Hospital, No.33, 2nd Section, Chung-Hua Road, Taipei 100, Taiwan　TEL: 02-23889595 ext. 2226

INTRODUCTION

Benign acanthomas are benign tumors of epidermal keratinocytes, which may show a wide range of aberrant proliferation and keratinization with a variety of histopathologic patterns. These include epidermoid keratinization (seborreic keratosis), epidermolytic hyperkeratosis (epidermolytic acanthoma), dyskeratosis (warty dyskeratoma), cornoid lamellation (porokeratosis) and absence of keratinization (clear cell acanthoma).[1] In 1988, Brownstein reported 31 cases of benign acanthomas,[1] in which acantholysis was an outstanding histopathologic feature as first described by himself in 1985,[2] and he termed these tumors as acantholytic acanthoma. Since the original description of Brownstein, only few other cases has been reported.[3,4,5,6] Moreover, acantholytic acanthoma has never been reported in Taiwan. In this article, we report two cases of acantholytic acanthoma with the characteristic histopathologic features.

CASE REPORTS

CASE 1

A 62-year-old male patient had suffered from a reddish, rice-grain-sized papule over his left axilla (Fig.1) for several months. On physical examination, this skin lesion was a solitary, 2mmx2mm-sized, erythematous, hyperkeratotic, round papule with elastic, firm consistency. No tenderness, pruritus or other symptoms had been complained. According to the clinical presentation, skin appendage tumor was suspected and total excisional biopsy of this papule was performed. The histopathological examination showed hyperkeratosis with focal parakeratosis of stratum corneum, irregular hyperplasia of epidermis with prominent acantholytic suprabasal clefts and intraspinous acantholysis (Fig. 2 & 3). There were many acantholytic keratinocytes with few dyskeratosis in the suprabasal clefts. No lymphocytes, neutrophils, or eosinophils were found in the epidermis. Band-like infiltration of epithelioid histiocytes and lymphocytes in upper dermis and

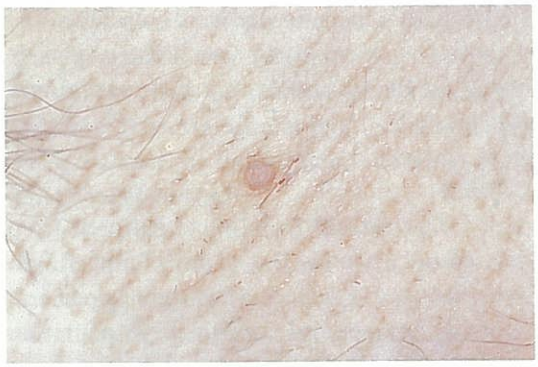

Fig. 1
The lesion is solitary, 2mmx2mm-sized, erythematous, hyperkeratotic, round papule with elastic, firm consistency on left axilla

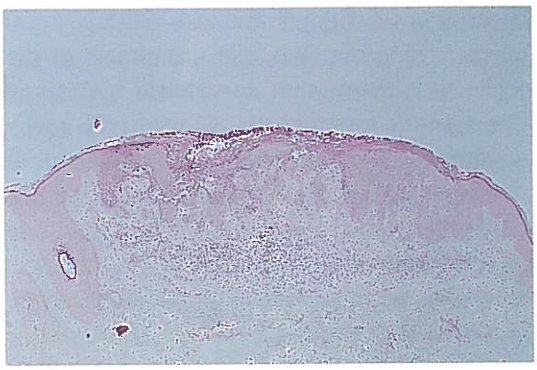

Fig. 2
Histopathologically, the lesion is symmetrical and well-circumscribed with hyperkeratosis, focal parakeratosis and irregular hyperplasia of epidermis under scanning view. (H & E 40x)

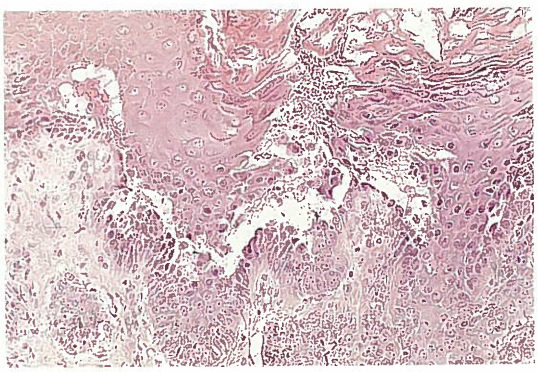

Fig. 3
In higher power, there are acantholytic suprabasal clefts and intraspinous acantholysis with acantholytic keratinocytes and few dyskeratosis. (H & E 200x)

slight melanin incontinence were noted, too. The diagnosis of acantholytic acanthoma was made based on the characteristic histopathologic features.

CASE 2

A 68-year-old female had suffered from one round, brownish, 15mmx8mm-sized, hyperkeratotic plaque on her right upper arm (Fig. 4) for several months. No pruritus, tenderness or other symptoms had been complained. According to its clinical picture, the diagnosis of seborrheic keratosis was considered and large cell acanthoma must to be ruled out, so a total excisional biopsy was performed. Microscopically, there were moderate hyperkeratosis with parakeratosis on the corneal layer, hypergranulosis and acanthosis of the epidermis. Prominent suprabasal clefts and intraspinous acantholysis (Fig. 5) with many acantholytic keratinocytes and occasional dyskeratosis (Fig. 6) were noted and band-like infiltration with lymphocytes was also found in upper dermis. The diagnosis of acantholytic acanthoma was made based on the characteristic histopathologic features.

DISCUSSION

Acantholytic acanthoma is a benign cutaneous tumor of epidermal keratinocytes, which is probably seen much more commonly than is reported. The most common clinical presentation is a solitary, well-circumscribed, hyperkeratotic papule or nodule in the size of 5mm to 15mm in diameter, and occasional with crusting on the surface.[1] Molluscum contagiosum-like lesion due to central umbilication had also been reported.[4] It is usually asymptomatic, but pruritus is sometimes present. The tumor is predominantly found in men (male:female ratio 2:1) and most patients are over 50 years of age.[1] There is a truncal predilection and the palms, soles, face, and mucous membrane are usually spared. The most frequent clinical diagnosis is seborrheic keratosis or actinic keratosis.[6] The pathogenesis

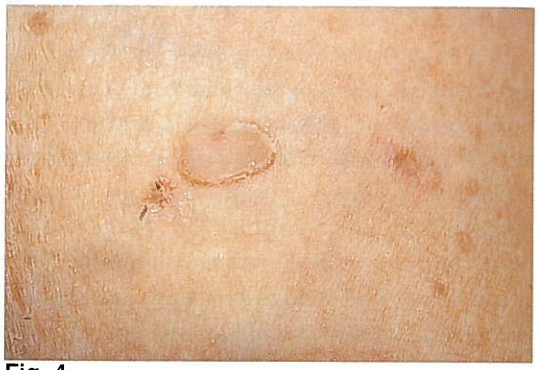

Fig. 4
One round, brownish, 15mmx8mm-sized, hyperkeratotic plaque on right upper arm.

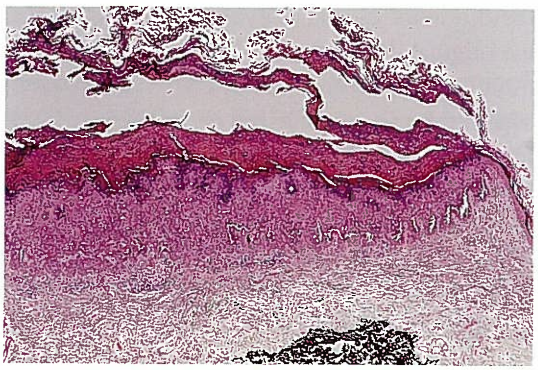

Fig. 5
Histopathologically, the lesion is well-circumscribed, moderate hyperkeratosis with parakeratosis, hypergranulosis and acanthosis of the epidermis under scanning view. (H & E 40x)

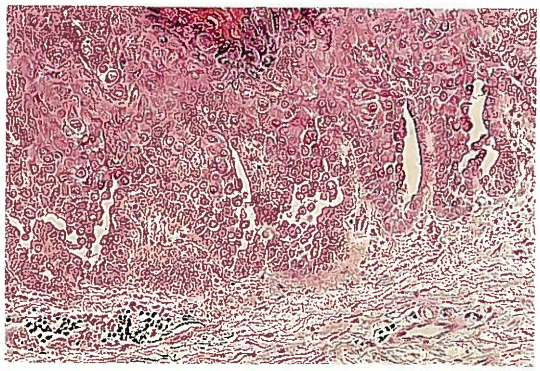

Fig. 6
In higher power, there are prominent suprabasal clefts and intraspinous acantholysis with many acantholytic keratinocytes and occasional dyskeratosis. (H & E 200x)

of acantholytic acanthoma and the relationship to other disease is still unknown. One case of acantholytic acanthoma in an immunosuppressed patient, a receipient of renal transplantation, has been reported,[5] but the role of decreased immune surveillance is uncertain.

Histopathologically, the lesion is symmetrical and well-circumscribed. The epidermis demonstrates hyperkeratosis, parakeratosis, papillomatosis and the most characteristic feature, prominent acantholysis. The level of acantholysis varies. It commonly involves all layers of epidermis, but it may be suprabasal, intraspinous, or at the level of the granular layer.[3] Sometimes, mild dyskeratosis with occasional grains can be seen.[4] A nonspecific perivascular lymphohistiocytic infiltrate with occasional eosinophils is present in the underlying dermis.

On histopathology, a list of cutaneous disorders must be differentiated from acantholytic acanthoma (Table I). Other acantholytic disorders such as pemphigus vulgaris, pemphigus foliaceus, pemphigus vegetans, Grover's disease, Hailey-Hailey disease and Darier's disease may mimic an acantholytic acanthoma. There are prominent eosinophils in most pemphigus group and the immunofluorescent studies are useful to differentiate pemphigus from acantholytic acanthoma. The acantholysis in Hailey-Hailey disease is full-thickness and very extensive with the characteristic appearance of a dilapidated brick wall. In Darier's disease, there is prominent dyskeratosis with corps ronds and grains, which is seldom seen in acantholytic acanthoma. Only small foci of acantholysis are seen in Grover's disease, and the acantholysis may occur in four histologic patterns: small suprabasal clefts as seen in pemphigus vulgaris, acantholytic dyskeratosis like Darier's disease, full-thickness acantholysis resembling Hailey-Hailey disease or a spongiotic dermatitis pattern. Besides, these disorders are eruptions with more widespread papular, vesicular, or papulovescular lesions, so they can easily differentiated clinically from the solitary, usually hyperkeratotic acantholytic acanthoma. It is therefore important to correlate the histopathologic features with the clinical pictures. Because acantholytic acanthoma is a benign cutaneous tumor, simple excision is the treatment of choice.

Differential diagnosis should also be made on certain solitary lesions with features of acantholysis, which include warty dyskeratoma, acantholytic seborrheic keratosis, acantholytic actinic keratosis and papular acantholytic dyskeratosis of the genitalia. Warty dyskeratoma displays a typical flask-shaped cystic invagination with papillary projections connected with the surface by a channel filled with keratinous material (parakeratotic plug). Acantholysis with suprabasal lacunae and villi are seen in the lower portion of invagination, and numerous acantholytic, dyskeratotic cells with corps ronds and grains are seen above the velli. But few dyskeratosis is found in acantholytic acanthoma. In acantholytic seborrheic keratosis, the acantholysis is seen exclusively in the squamous cell nests in the upper portion of the basic histologic architecture of a general configuration of seborrheic keratosis, which shows squamous cell proliferation, dyskeratotic cells and disorderly arrangements of the squamous cells. There is no suprabasal acantholysis, villi or corps ronds.[7,8] In the acantholytic type of actinic keratosis, the acanthosis with clefts or lacunae similar to those seen in Darier's disease occurs immediately above the basal cell layer of the actinic keratosis,[8] which is composed by atypical cells. Papular acantholytic dyskeratosis of the genitalia, is a papular eruption on genital or perineal area with itching and burning sensation. Histopathologically, there are prominent hyperkeratosis, superficial acantholysis, and dyskeratosis with corps ronds and grains, and the immunofluorescent studies of skin and serum are negative.[9] In addition, epidermolytic acanthoma, which are small brownish papillomatous papules resembling seborrheic keratosis on trunk, has also to be differentiated

Table I. Differential Diagnosis of Acantholytic Acanthoma

Disease	Clinical picture	Histopathologic feature
Acantholytic acanthoma	Solitary, hyperkeratotic, asymptomatic papule or nodule on the trunk	Hyperkeratosis, parakeratosis and acanthosis with prominent acantholysis
Pemphigus group	Multiple flaccid bullae of skin on face, chest, scalp and intertriginous areas Oral mucosa involved	Suprabasal acantholysis with blister formation Variable eosinophil infiltration DIF : intercellular deposition of IgG and passible C3
Grover's disease	Pruritc papulovesicles on chest, back, and thighs	Four histological pattern: Darier's disease pattern Hailey-Hailey disease pattern Pemphigus vulgaris pattern Spongiotic dermatitis pattern
Hailey-Hailey disease	Fragile flaccid blisters over intertriginous areas Autosomal dominant	Full-thickness acantholysis in The appearance of "dilapidated brick wall"
Darier's disease	Scaly papules and plaques of head, back, chest, and groin Autosomal dominant	Acanthosis and parakeratosis Suprabasal acantholysis Dyskeratosis with corps ronds and grains
Warty dyskeratoma	Single, slow-growing papulonodule with central pore or umbilication	Flask-shaped cystic invagination Suprabasal acantholysis and dyskeratosis with corps ronds and grains
Acantholytic seborrheic keratosis	Fresh-colored to black papules with smooth to warty surface	Focal acantholysis in the upper portion of a general architecture of ordinary seborrheic keratosis No suprabasal acantholysis
Acantholytic actinic keratosis	Small, erythematous, keratotic lesions with sharply margin on the sun-damaged skin	Focal acantholysis above the atypical cells of the basal cell layer of the underlying actinic keratosis
Papular acantholytic dyskeratosis of the genitalia.	Discrete dome-shaped, smooth-surfaced papules on external genital and perineal area	Prominent hyperkeratosis, superficial acantholysis, and dyskeratosis with corps ronds and grains
Epidermolytic acanthoma	Small brownish papules with verrucoid features on trunk	Epidermolytic degeneration with coarse, enlarged keratohyaline granules over granular and spinous layers, basal layer is sparing

from acantholytic acanthoma.

Histopathologically, there is pronounced epidermolytic degeneration, also referred to as granular degeneration, with prominent vacuolar changes andmanycoarse,enlarged keratohyaline granules over granular layer and spinous layer, but basal layer is usually sparing.

To the best of our knowledge, this is the first report of acantholytic acanthoma in Taiwan, which shows typical pathological features. Although only a simple papule is found clinically, acantholytic acanthoma shows histopathologic interest which must be differentiated from a lot of cutaneous disorders with features of acantholytic dyskeratosis. Therefore, a correct diagnosis can be made according the clinical pictures and the outstanding pathohistologic features, and to avoid unnecessarily extensive treatment due to misdiagnosis.

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