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Acantholytic Dermatosis of the Vulva: Case Series and Review of Literature

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Abstract

Papular Acantholytic dyskeratosis; also known as acantholytic dermatosis of the vulvocrural area, is a rare inflammatory disorder of the genital location. This review discusses the clinical and pathologic correlation of this rare entity, its differential diagnosis and a recent case series with analysis of all previous reported cases in the literature.

Keywords: Acantholytic Dermatosis; Dyskeratosis; Acantholysis Papular Acantholytic Dyskeratosis

Introduction

Papular Acantholytic dyskeratosis; also known as acantholytic dermatosis of the vulvocrural area, is a rare but distinct histopathologic cutaneous disorder of the genitalia which was first described as a subset of focal acantholytic dyskeratosis in 1972 and can present with a variety of clinical manifestations. This review discusses the clinical and pathologic presentation of this entity and its differential diagnosis as well asreview of literature.

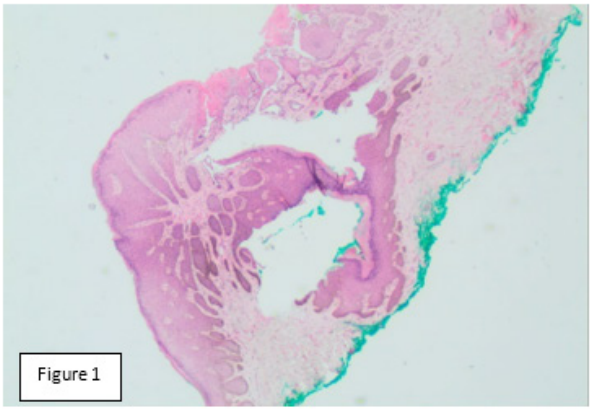
Case Series

The first case is a 72 year old African American woman; post-hysterectomy, came with the complaint of pruritus on the medial aspect of left labia majora. The patient's most recent pap smear was normal and she denied any associated discharge or post-menopausal vaginal bleeding. Physical examination showed a slightly raised, grayish lesion, measuring 5 mm on the medial aspect of the left labia majora with normal external genitalia, no adnexal masses or tenderness, no bladder tenderness, and an unremarkable rectovaginal septum. Clinical considerations included vulvar dystrophy and condyloma. Excisional biopsy was obtained. Histologic examination found acantholysis with dyskeratotic keratinocytes and associated chronic inflammation (Figures 1 & 2). Periodic acid–Schiff (PAS) stainwas negative for fungi. HSV1 immunostain was negative. There was no personal or family history of Hailey-Hailey or Darier's disease. There was no associated skin rash or dermatologic papules and vesicles in the axilla, central chest or trunk. Microscopy did not show any dysplasia or atypical mitoses. The clinical features personal and family history and microscopic findings were diagnostic of acantholytic dermatosis. A second case is a 44 year old Caucasian woman who presented with a single asymptomatic, skin colored, solitary papule, measuring 4mm on the vulva. The patient reported no pruritic or papulovesicular lesions elsewhere on the body with no associated personal or family history of Hailey-Hailey or Darier's disease. The lesion was surgically excised and submitted for routine pathologic evaluation. On microscopic examination, the sections showed hypergranulosis, focal acantholysis and dyskeratotic keratinocytes. HSV1 immunostain was negative and PAS stain did not demonstrate fungal organisms. The clinical features, personal and family history and microscopic findings were diagnostic of acantholytic dermatosis (Figures 3 & 4).

Discussion

Acantholytic dermatosis of the vulva has been regarded as a subset of focal acantholytic dyskeratosis; a term first proposed by Ackerman in 1972 [1]. Papular acantholytic dyskeratosis has been reported as a distinct histopathologic entity which can present with varying clinical manifestations. The common sites of occurrence are vulva, penis, scrotum, perianal area, and inguinal folds. A rare case involving vaginal epithelium has also been reported [2]. Clinical presentation ranges from solitary, discrete lesion to multiple grouped or confluent, flesh-colored papules, vesicles and bullae as well as white macerated patches and plaques [3]. The involved area is generally asymptomatic, but pruritis, erythema

and burning has also been noted [4]. It has a chronic course and shows variable response to different treatment options including; topical retinoids, steroids, cryotherapy, electrocoagulation, laser therapy and surgery [5]. Histologically, intraepidermal acantholysis is seen, along with dyskeratosis including corps ronds and grains. Other histologic findings include parakeratosis, hyperkeratosis, suprabasal cleft formation and corp ronds (dyskeratotic cells composed of basophilic nucleus with a perinuclear halo and eosinophilic cytoplasm) and corp grains (small cells with elongated, hyperchromatic nuclei in stratum corneum and granulosum) in the acantholytic layer of the epidermis2 3. Direct Immunofluorescence consistently shows negative results.



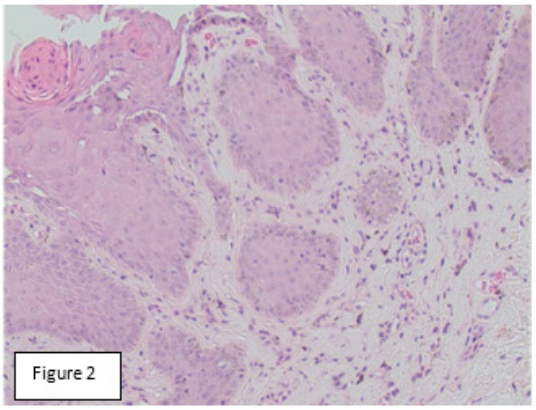
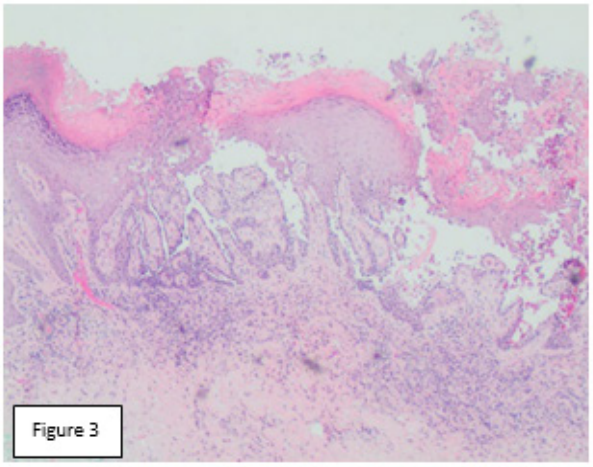


Figure 1 & 2: (10X) and (40X) Hematoxylin-eosin stain showing focal acantholysis, and dyskeratotic keranitnocytes.



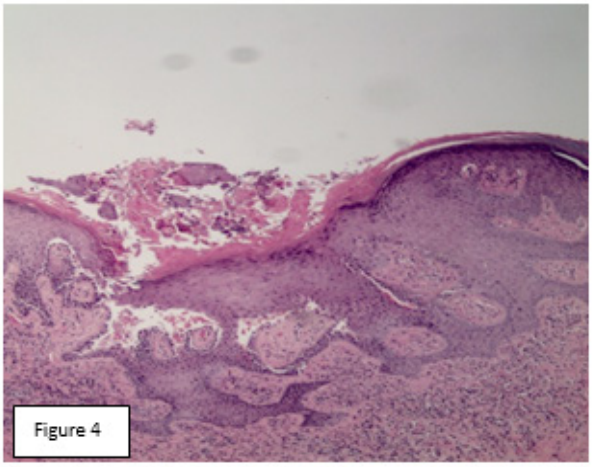


Figure 3 & 4: Hematoxylin-eosin stain slides at 10X, showing acantholysis with dyskeratotic keratinocytes and background chronic inflammation.

The histologic findings of acantholysis and dyskeratosis can be seen in other cutaneous disorders include including Darier's disease, Hailey-Hailey disease and transient acantholytic dermatosis (Grover's disease) and Pemphigus Vulgaris. Hailey-Hailey disease demonstrates epidermal acantholysis, resembling a 'dilapidated brick wall appearance' and infrequent dyskeratosis. Darier disease has similar histopathologic findings of acantholytic dyskeratosis but usually presents clinically as verrucous papules and plaques in seborrheic regions, such as the flexures of the groin and axilla with characteristic nail changes. Both Hailey-Hailey disease and Darier's disease exhibit an autosomal dominant mutation in the calcium pump genes; ATP2C1 and ATP2A2; respectively, with a distinct personal and family history of recurrent lesions [4]. Transiennt acntholytic dermatosis (Grover's disease) shows suprabasal acantholysis with or without dyskeratosis and commonly occurring in middle aged to elderly patients, with multiple papulovesicular eruptions in a photo distributive pattern. Pemphigus vulgaris also shows acantholysis, typically in the spinous layer of the epidermis and is a well established autoimmune disorder presenting with flaccid blisters and positive direct immunofluorescence stain

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findings, including positive IgG and C3 immunoreactivity in the spinous epidermal layer. A PubMed search of "Acantholytic dermatosis of vulva" found 25 reported cases of this entity; which have been summarized in the table below (Table 1). In conclusion, careful assessment of the clinical presentation, course of disease, personal and family history play an important role alongside characteristic histopathological findings to make a definitive diagnosis of acantholytic dyskeratosis. The pathogenesis still remains uncertain but a recent study implicated somatic mosaicism of the ATP2A2 mutation in multiple biopsies of acantholytic dyskeratosis lesions [6]. Dermatopathologists should be aware of this entity, as it can mimic other cutaneous disorders histologically with careful clinicopathologic correlation essential to correct diagnosis and appropriate treatment and management.

Table 1: Acantholytic dermatosis of vulva found 25 reported cases of this entity.

Published Cases of Acantholytic Dermatosis of Vulva	Presentation	Age	Family History
Mansura A, Maly A, Ramot Y, Zlotogorski A 2015 [7]	Papular pruritic eruption	-	Negative
Elena Hadjicharralambous, Stephanie Diamond, 2017 [8]	-	-	Questionable
García-Morales I, Requena-Caballero L,2018 [9]	Ulcerated lesion	infant	Negative
Yu WY, Ng E, Hale C,2016 [10]	-	-	Familial hailey-hailey disease
Wong KT, Wong KK 1994 [11]	Warty lesion	22	Negative
Cooper PH 1989 (6 cases reported) [12]	Multiple papules, solitary papules, and plaques	-	-
Baliu-Piqué C, Iranzo P 2017 [13]	-	-	-
Haddadeen C, Theaker J, Rowen D, Lotery H 2020 [14]	Erosive, scarring lesion; chronic	-	Negative
Dittmer CJ, Hornemann A, Rose C, Diedrich K, Thill M 2010 [15]	Multiple papules, chronic	45	Negative
Flores-Terry MA, Zamberk Majlis P 2017 [16]	Whitish papules and erosion	30	Negative
Bell HK. 2001 (2 patients) [17]	Asymptomatic, popular lesion	-	Negative
Sáenz AM, Cirocco A 2005 [18]	Pruritic popular eruption; chronic	11	Negative
Lee S, 1989 [19]	Multiple papules	32	Negative
Roh MR, Choi YJ, Lee KG 2009 [20]	Asymptomatic popular eruption	63	Negative
Wang L, Yang XC, Hao F, Mei Y, Ye QY 2009 [21]	-	-	-
Peştereli HE 2000 [22]	Pruritic papule, chronic	44	Negative
Van Joost T 1991 [23]	Pruritic, persistent eruption on chest and vulva	35	Negative
Weedon D 1986 [24]	-	-	-
Chorzelski TP 1984[25]	Multiple, whitish-grouped papules	23	Negative

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