

Acantholytic Dermatitis of the Vulvocruural Area

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GOAL

To discuss a case of acantholytic dermatosis of the vulvocruural area

OBJECTIVES

Upon completion of this activity, dermatologists and general practitioners should be able to:

1. Describe the clinical presentation of acantholytic dermatosis of the vulvocruural area.
2. Outline etiological possibilities and the differential diagnosis of acantholytic dermatosis.
3. Delineate treatment for acantholytic dermatosis.

CME Test on page 220.

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Acantholytic dermatosis of the vulvocruural area is a rare skin disorder characterized by solitary or multiple skin-colored to white, smooth papules or plaques. Histopathological features of both Hailey-Hailey disease and Darier's disease are present. There is acantholysis, which may involve the full thickness of the epidermis, and dyskeratosis with corps ronds and grains. There may be marked hy-

perkeratosis and focal parakeratosis. We report a case of this rare disease and discuss its differential diagnosis and treatment.

Acantholytic dermatosis of the vulvocruural area is an uncommon clinical and histopathological entity that belongs to the spectrum of focal acantholytic dyskeratoses described by Ackerman.¹ In 1984, Chorzelski et al² proposed the term *papular acantholytic dyskeratosis of the vulva* to describe the disorder and suggested that this dermatosis may represent a unique entity. Since the initial description by Chorzelski et al, there have been sporadic reports of acantholytic dermatoses of the vulvocruural area,^{3,7} as well as a report of clinically and histologically similar lesions in males.⁸

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Figure 1. Confluent skin-colored papules on the medial thighs, inguinal folds, and labia majora.

Case Report

A 27-year-old Hispanic woman presented with a 9-month history of a slightly pruritic eruption in her groin. There was no family history of similar lesions. On physical examination, the patient showed confluent skin-colored, smooth, 2- to 4-mm papules on her medial thighs, inguinal folds, and labia majora that coalesced into plaques (Figure 1).

A 4-mm punch biopsy revealed acantholysis involving the suprabasal layers of the epidermis and dyskeratosis with some cells resembling corps ronds and grains (Figure 2). Direct immunofluorescence was negative, and treatment with topical tretinoin 0.05% cream produced no significant improvement.

Comment

Acantholytic dermatosis of the vulvocrural area presents as solitary or multiple skin-colored to white, smooth papules or plaques.^{2,6} The lesions are most commonly located on the labia majora but also may be located elsewhere in the perineum, in the inguinal folds, or on the superior medial aspects of the thighs.⁴ Two cases have been reported in which the submammary region also was involved.^{5,6} The papules may be painful or pruritic.^{3,5} Concurrent *Candida* vaginitis was observed in a small minority of cases.⁴

Acantholytic dermatosis of the vulvocrural area displays histological features of both Hailey-Hailey

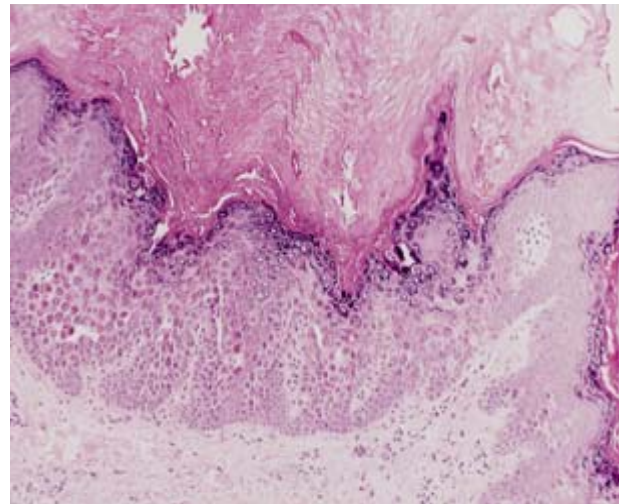


Figure 2. Acantholysis involving the suprabasal layers of the epidermis and dyskeratosis with some cells resembling corps ronds and grains (H&E, original magnification $\times 250$).

disease and Darier's disease. There is prominent acantholysis, which may involve the full thickness of the epidermis, and dyskeratosis with corps ronds and grains.⁵ There may be pronounced hyperkeratosis and focal parakeratosis.^{4,6} Direct immunofluorescence studies are consistently negative, with no deposition of immunoglobulin or complement in the skin.^{2,6} Indirect immunofluorescence studies are also negative.^{2,3,6}

The differential diagnosis of acantholytic dermatosis of the vulvocrural area consists of acantholytic disorders that can produce lesions localized to the genital region,^{7,9-11} including Darier's disease, Hailey-Hailey disease, pemphigus vegetans, warty dyskeratoma, and squamous cell carcinoma. These disorders are usually distinguished from each other on the basis of clinical and histopathological features. When performing a diagnostic evaluation, it is crucial to establish a negative family history. This helps to distinguish acantholytic dermatosis of the vulvocrural area from Darier's disease and Hailey-Hailey disease,^{9,10} which are autosomal dominant acantholytic disorders. Immunofluorescence studies also are significant because negative results will rule out pemphigus vegetans.⁷

Grover's disease (transient acantholytic dermatosis) also produces histologically similar lesions. However, it is excluded from the differential diagnosis because, unlike acantholytic dermatosis of the vulvocrural area, it consistently involves the trunk and often resolves within weeks to months.¹²

The etiology and pathogenesis of acantholytic dermatosis of the vulvocrural area are not fully elucidated. Electron microscopy shows a disappearance

of intercellular connections and a reduced number of desmosomes.⁵ In addition, there is detachment of tonofilaments from the desmosomal plate and perinuclear aggregations of tonofilaments.⁵ The underlying cause of these changes is unknown. It has been hypothesized that the warm, moist environment of the genital region triggers this disorder and accounts for the localization of lesions to the vulvocrural area.^{4,8} Furthermore, Coppola et al³ suggest a possible connection between ovarian hormones and the pathogenesis of this disease, based on their patient's premenstrual exacerbations of itching and burning.

Effective treatment of acantholytic dermatosis of the vulvocrural area is difficult. The lesions tend to persist for years. With the exception of topical tretinoin cream, which has been reported to give only a transient improvement in a single patient,⁴ there is no evidence that any topical or systemic medication can ameliorate or cure this dermatosis. Chorzelski et al² described the failure of corticosteroids (local and systemic) and sulfones to improve the eruption. Coppola et al³ also reported the failure of multiple treatments.

Surgical treatments, although impractical for multiple lesions, have proven more effective than medications. Cooper⁴ reports that excision of localized lesions is curative, based on follow-up periods ranging from 3 to 30 months. Furthermore, some papules have been successfully removed by electrocautery.²

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