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Case presentation

Acantholytic dermatosis of the vulva

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Abstract

Acantholytic dermatosis of the vulva is a rare condition, presenting with papular eruption in the genital area without history of Darier disease or Hailey-Hailey disease. We report a case with a papular pruritic eruption in the region of the vulva, coalescing into plaques. Biopsy specimen showed irregular acanthosis with an area of split-like bullous formation in the deeper part of the epidermis, as well as acantholytic cells, marked hypergranulosis and hyperkeratosis, compatible with the rare diagnosis of acantholytic dermatosis of the vulva. We review the clinical and histological characteristics of this uncommon disease.

Keywords: acantholytic dermatosis, vulva, genitalia

Introduction

Acantholytic dermatosis of the vulva is an extremely uncommon disease of the genital area. There are very few reports in the medical literature, and many of them are in gynecological journals [1,2]. Herein we describe a woman who presented with a papular eruption on the vulva, which was diagnosed following a skin biopsy as acantholytic dermatosis of the vulva.

Case synopsis

A 41-year-old woman presented with a one-year history of a papular pruritic eruption in the region of the vulva. On inspection, there were white papules merging into plaques over both sides of the labia majora (Figure 1). Other parts of the skin and mucous membranes were normal. The patient underwent a gynecological examination and

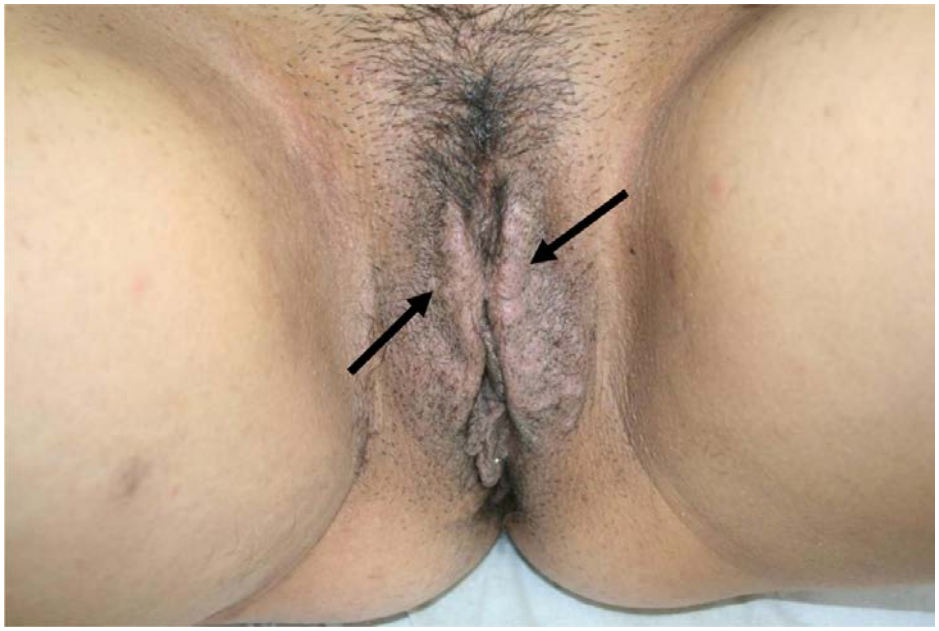
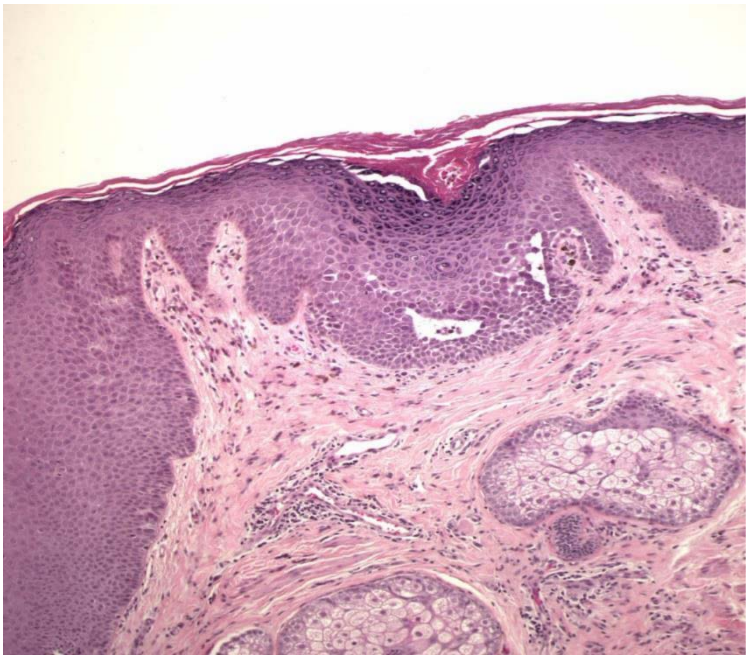


Figure 1. White papules merging into plaques on both labia majora (marked by arrows).

no vaginal lesions were found. Repeated cultures for candida were negative. No family history of skin disorders was reported. A biopsy specimen from the middle part of the right labia majora showed irregular acanthosis with an area of a split-like bullous formation in the deeper part of the epidermis, as well as acantholytic cells, marked hypergranulosis, and hyperkeratosis (Figures 2 and 3). These changes are compatible with the Darier-like Grover disease variant of acantholytic dermatosis of the vulva. Symptomatic treatment with betamethasone valerate cream was commenced, which led to partial improvement of her pruritus.



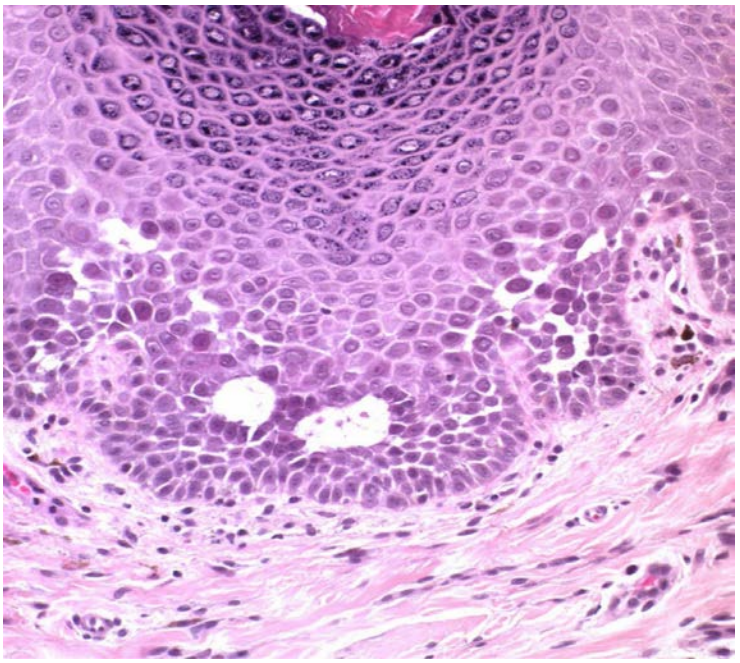


Figure 2. Lower magnification of histopathology of a biopsy taken from the right plaque, demonstrating irregular acanthosis with an area of a split-like bullous formation in the deeper part of the epidermis. **Figure 3.** Higher magnification, demonstrating acantholytic cells, marked hypergranulosis, and hyperkeratosis**.**

Discussion

Acantholytic dermatosis of the vulva is an uncommon entity that affects mainly women in their 3rd-7th decade [3]. Several case reports have also described genital lesions in both males [4] and children [5]. Clinically, diverse features including papules, vesicles, bullae, patches, and plaques have been documented [4]. The more common findings are papules, at times pruritic, that merge into plaques, and may resemble lichen planus, psoriasis, or lichen sclerosus. It is worth noting that a histological diagnosis of acantholytic dermatosis of the vulva was also made in a reported case of pruritus vulvae displaying no macroscopic lesions [6].

The histological features include dyskeratosis, a Hailey-Hailey pattern of acantholysis, parakeratotic plugging, and the absence of an inflammatory infiltrate containing eosinophils [4]. Direct immunofluorescence tends to be negative. However, one reported case of acantholytic dermatosis of the vulva showed a positive result with features of pemphigus vegetans [7].

In acantholytic dermatosis of the vulva, lesions tend to be localized to the genitalia with no involvement of other areas of the body. The clinical course is generally persistent and there is no reported family history. The absence of an eruption elsewhere on the body, the persistent course of the condition, and the absence of a positive family history, distinguishes acantholytic dermatosis of the vulva from Hailey-Hailey, Darier disease, or Grover'disease [4]. The differential diagnosis of a localized disease includes warty dyskeratoma and squamous cell carcinoma, which can be distinguished based on clinical and pathological features [8,9].

A range of different etiologies have been postulated including hormonal, viral, candidal [3], and physical (such as moisture and irritation) [4], yet none has been proven scientifically.

A variety of local treatments have been suggested, including topical steroids, nystatin, oxytetracycline cream, retinoid cream, and tar and sulphur-containing ointments [6]. Nevertheless, no treatment has achieved complete resolution of the rash, as seen in our case.

Ablation methods such as local excision, cryosurgery, or dermabrasion appear to be the only modalities to provide definitive treatment in this condition. An additional promising treatment is ablative laser therapy, such as with YAG or CO_2 lasers, which were utilized successfully in Hailey-Hailey and Darier disease [10,11]. Indeed, CO_2 laser was shown to be effective also

in the rare case of acantholytic dermatosis involving the vulva [1]. It should be remembered, though, that in many cases a definitive diagnosis and reassurance are all that is required.

Conclusion

Acantholytic dermatosis of the vulva is an uncommon condition, which can be encountered by dermatologists and gynecologists. This disorder is localized to the genital area and does not involve other areas of the body, distinguishing this condition from Hailey-Hailey or Darier disease. Knowledge of this disease is crucial, since it runs a more benign course, and oftentimes reassurance is sufficient.

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