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Epidermolytic hyperkeratosis of the vulva

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Abstract

Vulvar epidermolytic hyperkeratosis is a benign entity that mimics other malignant and inflammatory vulvar dermatoses clinically and histologically requiring careful clinical pathologic correlation for diagnosis.

Keywords: vulvar epidermolytic hyperkeratosis, epidermolytic hyperkeratosis

Introduction

Epidermolytic hyperkeratosis (EHK) of the vulva is an uncommon condition [1]. Although benign, vulvar EHK is often confused with other more commonly encountered inflammatory and malignant vulvar disorders, both clinically and histologically. Proper recognition is necessary to avoid overtreatment and unnecessary gynecologic surveillance.

Case Synopsis

A 68-year-old woman presented with a chronic pruritic unilateral vulvar eruption of several years duration that had been refractory to topical estrogen and high potency topical corticosteroids. Evaluation at a tertiary women's health center raised concern for possible vulvar malignancy. Physical examination revealed multiple, discrete, whitish, well-defined, sessile papules with variable confluence into small plaques, localized to the right labia majora. There was focal involvement of the inner mucosal surface (**Figure 1**). A 6mm punch biopsy showed prominent hyperkeratosis and papillomatosis (**Figure 2A)** with coarse basophilic keratohyalin granules and pallor (**Figure 2B)**. Correlation of the clinical findings with the histopathologic features led to the diagnosis of EHK of the vulva. The patient was reassured that the eruption was benign.

Case Discussion

Epidermolytic hyperkeratosis is a histologic reaction pattern characterized by a distinctive combination of hyperkeratosis, papillomatosis, acanthosis, and hypergranulosis with variable epidermal reticular degeneration [1]. It was originally described and used synonymously with bullous congenital ichthyosiform erythroderma, an inherited autosomal dominant skin disorder, but has subsequently been seen in other hyperplastic processes such epidermal nevi, seborrheic keratoses, and verrucae [2,3].

Vulvar EHK is a distinct clinical entity presenting with multiple, often unilateral, well-defined papules with variable confluence into plaques [4]. Larger exophytic hyperkeratotic nodules have also been described [2,4]. Complaints of itching are common and superimposed findings of chronic dermatitis may be seen [5]. The condition is rare and in one large case series represented 0.5% of 183 non-infectious non-neoplastic vulvar biopsies [6]. As the histopathology of EHK is shared with a number of conditions, careful clinical-pathologic correlation is required for proper diagnosis.

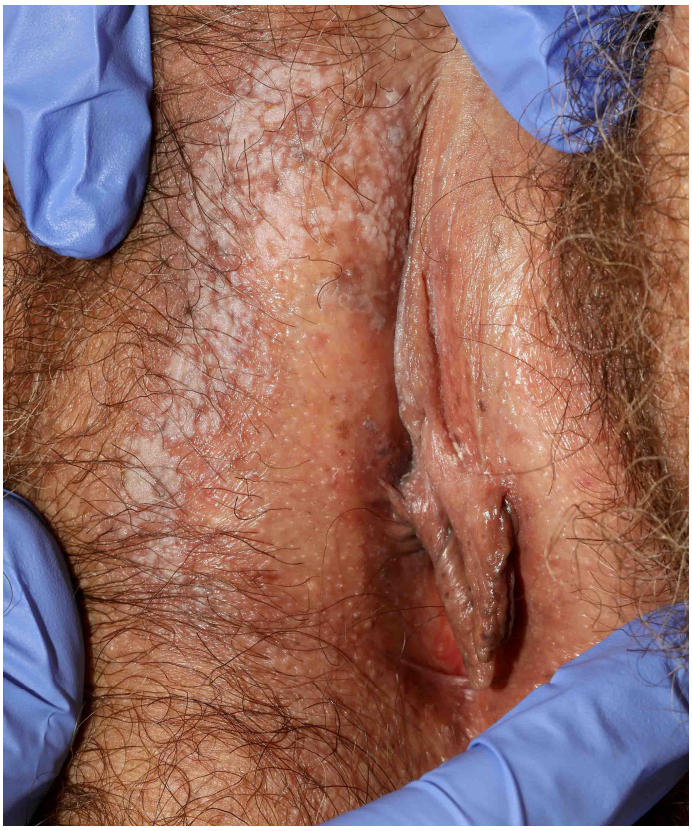

Figure 1. *Well-defined, greyish sessile papules with variable confluence into small plaques.*

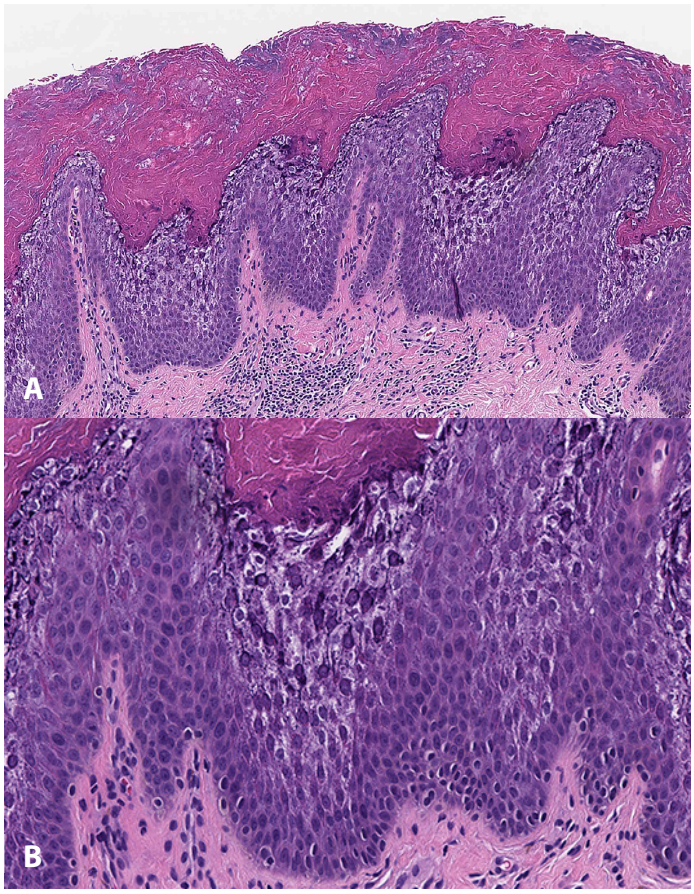

Figure 2. ***A)*** *Epidermal pallor with papillomatosis, hyperkeratosis, and hypergranulosis. H&E, 8×.* ***B)*** *Variable vacuolar degeneration of the spinous and granular layer with course basophilic keratohyalin granules. H&E, 20×.*

Retrospective reviews of vulvar biopsies have established that EHK was frequently misdiagnosed at initial clinical presentation [6]. Clinically, vulvar EHK may resemble inflammatory disorders such as chronic dermatitis as well as benign and neoplastic conditions including white sponge nevi, condyloma, and squamous cell carcinoma. Another major diagnostic pitfall is in the histopathology, which shows both hypergranulosis and cytolysis and can be easily confused with viral cytopathic effect and/or cytological atypia.

Studies have found no relationship between the human papilloma virus and vulvar EHK [4]. The etiology of vulvar EHK remains unknown but may represent a unique form of Koebernization related to trauma or chronic rubbing [1]. Mutations in keratin 1 and 10 have also been suggested to play a role in the pathogenesis [7].

Hypoallergenic bland emollients, low-potency topical corticosteroids, and topical calcineurin inhibitors may aid in attenuating the itch scratch cycle. Rarely, locally destructive methods such as cryotherapy or excision may also be utilized in more nodular presentations, but are not considered a first line treatment [1].

Conclusion

The natural history of vulvar EHK is benign with no potential for malignant transformation. Treatment focuses on symptomatic improvement of pruritus rather than full clearance of the lesions, which tend to be persistent.

Potential conflicts of interest

The authors declare no conflicts of interests.

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