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BRIEF COMMUNICATION

Labial Adhesion Secondary to Lichen Sclerosus Masked by Vulvar Hailey–Hailey Disease

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Labial adhesions in postmenopausal women are caused by various inflammatory processes in the context of estrogen deficiency. Lichen sclerosus (LS) is a chronic, progressive condition characterized by idiopathic epithelial thinning of the anogenital region with symptoms of significant pruritus, discomfort, and dyspareunia. Early diagnosis and treatment of LS can prevent disease progression, leading to labial adhesions and malignancy. We present an 84-year-old woman with long-standing vulvar Hailey–Hailey disease who developed labial adhesions. Clinical examination with vulvar biopsy revealed histopathological findings consistent with LS. Surgical separation of the labia by blunt dissection was then performed, and clobetasol ointment and vaginal dilatators were postoperatively prescribed. This resulted in a significant improvement in her symptoms. This is the first reported case of LS in a patient with Hailey–Hailey disease.

Key Words: Genital diseases, Female, Vulvar diseases, Vulvar lichen sclerosus

INTRODUCTION

Labial adhesion is defined as the partial or complete fusion of the labia minora or majora that has an incidence of 2% among prepuberal females [1,2]. Predisposing factors include chronic inflammation secondary to poor hygiene, underlying skin conditions, recurrent urinary infections, and local trauma and estrogen deficiency [2,3], which ultimately causes denudation of the surface epithelium and fusion of the labia during the healing process [4,5]. Among dermatologic conditions associated with labial adhesion are lichen sclerosus (LS) and lichen planus [6].

Hailey–Hailey disease (HHD), also called benign familial pemphigus or benign chronic pemphigus, was first described in 1939 [7]. It is a rare autosomal dominant genodermatoses caused by a loss-of- function mutation in the *ATP2C1* gene, which encodes the hSPCA protein [8]. Prevalence is unknown and there is no link between particular racial groups nor sex predilection. The age of onset is commonly during the second to fourth decade of life. Clinical presentation is characterized by involvement of flexural areas with symmetrical distribution, such as axillary folds, groin, inframammary region and neck folds. Skin lesions initially appear as flaccid bullae over an erythematous background or normal appearing skin, which give rise to erosions, maceration, and crusts with a circinated border, leading to foul smelling moist vegetations and fissures [9]. Vulvar involvement presents in approximately 50 percent of females with HHD [10] which can be misdiagnosed for intertrigo or superficial mycoses. Diagnosis is made on clinical basis and confirmed by skin biopsy, which shows widespread loss of cohesion among suprabasal keratinocytes, causing a 'dilapidated brick wall' appearance [9].

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CASE REPORT

An 84-year-old woman (gravida 3 para 3) with a 40-year history of axillary and inframammary HHD presented to Hospital Zambrano Hellion, TecSalud gynecology clinic for evaluation of vulvar pain. She associated the appearance of symptoms to the development of vulvar lesions of HHD 3 years prior, for which she occasionally applied low and high potency topical corticosteroid (TCS) without improvement. She denied irritative symptoms of the lower urinary tract such or discharge and denied previous history of pelvic or genital trauma.

Clinical examination (Fig. 1) revealed well-circumscribed erythematous plaques with hyperpigmented borders, overlying crusts and lineal fissures affecting the vulva, perineum, perianal area and medial aspect of inner groin area, consistent with HHD. External genitalia presented total fusion of the labia minora in the midline, causing obstruction of the urethral meatus, vestibule and vagina, with an introitus opening of 1 cm. Bilateral fusion of the labia majora was also observed as well as absence of the clitoris and clitoral hood. The patient was referred to the dermatology department for evaluation. Since HHD is a non-cicatricial dermatosis, dermatology in conjunction with our gynecology team (E.S. and L.F.G.) decided to perform a vaginoscopy under epidural anesthesia, separating the labia by blunt dissection (Fig. 2). Vaginoscopy by hysteroscope revealed mucosal atrophy without erosions or other lesions and two punch biopsies were performed from the mucosal lining of the labia majora and minora. The histopathology examination was consistent with LS. Treatment with vaginal dilators 2 times per week together with topical clobetasol ointment once daily was initiated with symptom improvement after 1 month. The written informed consent for the publication of this article was obtained from the patient.

DISCUSSION

The above case shows a striking clinical presentation of vulvar HHD that masked the diagnosis of LS and led to development of labial adhesion. HHD generates superficial vesicles, thus a clinical presentation with scars and atrophy is unusual, which is why other causes of labial adhesion had to be thoroughly investigated in this case. Clues for the diagnosis of LS in this case, where the lesions of both conditions where overlapping, includes dyspareunia, pruritus and fusion of the labia, which are indicative of a scarring condition and should prompt to perform a biopsy.

Treatment for mild cases of labial adhesion includes treating the underlying condition together with TCS and topical estrogen creams [3]. If there is no response to topical therapy, surgical separation under anesthesia can be performed [5]. Recurrence of adhesions has been reported in 14%–20% of patients who have undergone surgical or manual separation; thus it is important to emphasize topical estrogen application and regular digital separation of the vulva, especially in patients who are not sexually active [6].

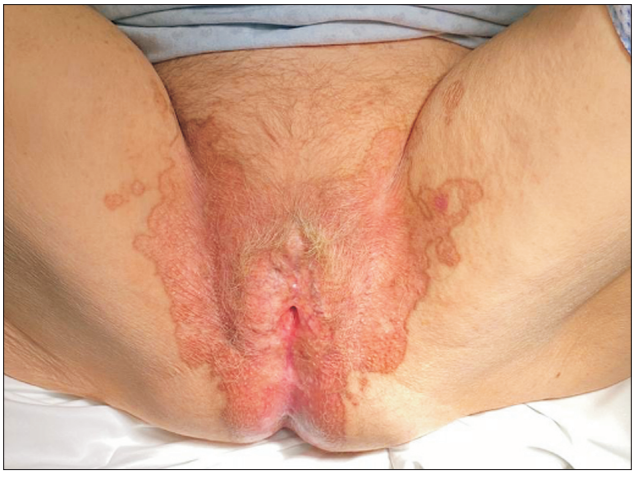

Fig. 1. Complete labial fusion with flat appearance of the vulva and a 1-cm opening in the vaginal introitus.

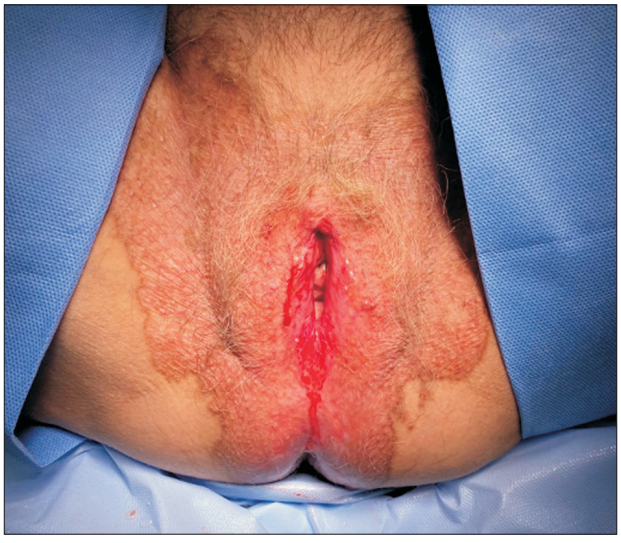

Fig. 2. Vulvar appearance after surgical labial separation.

Obtaining a complete history, in addition to careful clinical examination and skin biopsy confirmation may facilitate early diagnosis of LS. Prevention of chronic complications of LS such as labial adhesion may improve the quality of life of patients affected by this condition.

To our knowledge, this is the first case of LS in the context of vulvar HHD. Further research could determine if these two entities are associated.

CONFLICT OF INTEREST

No potential conflict of interest relevant to this article was reported.

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