Bullous lichen sclerosus: isolated vulvar involvement

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Abstract

We present a patient with the bullous form of lichen sclerosus of the vulva. She had no lesions in other cutaneous and mucosal areas. We used topical tacrolimus and topical clobetasol propionate. The patient was lesion free at the first-year follow-up.

Keywords: bullous, lichen sclerosus, vulva

Introduction

The bullous type is a rare variant of lichen sclerosus (LS). Although vulvar involvement has been discussed as a component of generalized bullous LS (BLS), [1], our patient has isolated BLS in the vulva.

Case Synopsis

A 44-year-old woman presented with a 2-month history of pruritic white and bullous lesions in the vulva. Family and previous medical histories were noncontributory.

Dermatologic examination revealed depigmented sclerotic plaques, as well as vesicles and eroded lesions in the vulva. A large hemorrhagic blister was observed in the lower part of labium majus (**Figure 1**). Nikolsky sign was negative. Other cutaneous and mucosal areas including oral mucosa, vagina, and cervix were not involved.

Anti TPO, antinuclear antibody, HIV, hepatitis B and C, VDRL, and *Borrelia burgdorferi* IgG and IgM serologies were all within normal limits or negative.

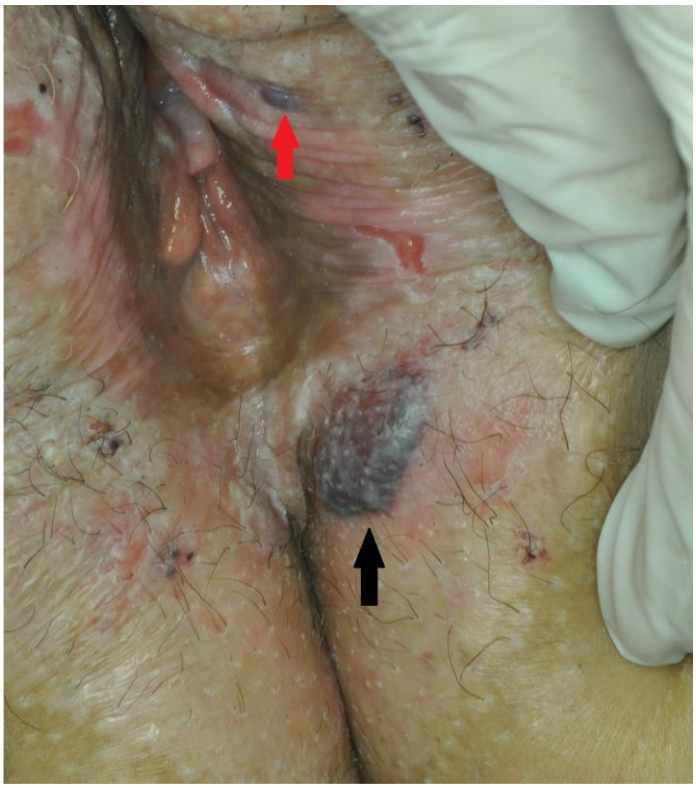

Figure 1. *Hemorrhagic bulla (⬆), vesicle (⬆), erosions and ivory-white plaques.*

TSH was 5.94 µIU/mL (normal range 0.27-4.2). Anti TG was 382.5 IU/ml (normal range <115). Bacterial culture of bulla was negative.

Punch biopsies from white plaque, bulla, and eroded lesions were obtained. Histopathological examinations revealed atrophy and vacuolar degeneration of the basal cell layer in the epidermis, homogenization of the collagen fibers, and pronounced edema in the papillary dermis. Biopsy of the lower part of the labium majus also showed

subepidermal vesiculation (**Figure 2**). A PAS-positive basal layer was seen on the dermal side. Direct immunofluorescence showed scattered C3 deposition on the basement membrane. Serologies for anti-BP 180 and anti-BP 230 were negative.

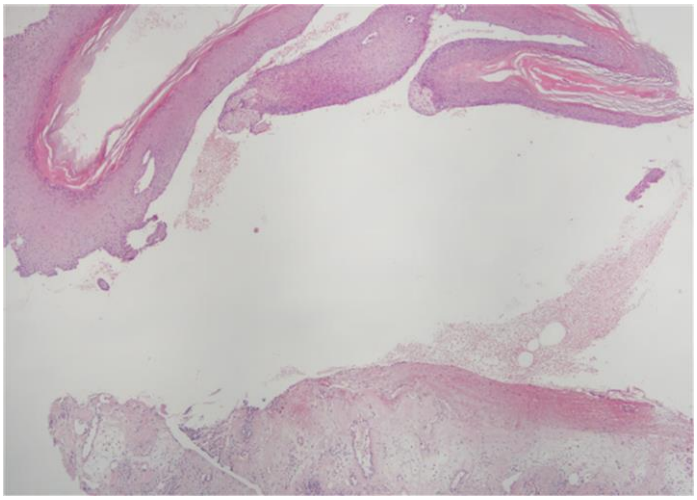

Figure 2. *Subepidermal vesiculation with blister formation,. H&E, 40×.*

Clobetasol propionate 0.05% ointment was started and all lesions and pruritus were improved except for depigmented plaques in the first month of follow-up. Treatment was maintained with tacrolimus 0.03% ointment on weekdays and clobetasol 0.05% ointment on weekends. At the end of one year, the patient had no remaining skin lesions except for mild hypopigmentation.

Case Discussion

In 2010, Kimura et al. [2] mentioned at least 39 BLS cases reported in the Japanese literature with only 4 of those having genital involvement. Unfortunately, our research in PubMed revealed no detailed information about these cases. In a case with generalized bullous and erosive LS involving the vulva, a possible association was found with *Borrelia afzelii* [1].

Vacuolar degeneration, epidermal atrophy, edema of the papillary dermis, and decreased support of collagen fibers may lead to blister formation [3]. Hemorrhagic character might be related to trauma.

The rate of BLS in the genital area is expected to occur more than other areas owing to the possibility of more frictional trauma and occlusion. However, BLS has been reported more frequently in extragenital than in genital LS [2, 3]. The reason for this discrepancy might relate to underreporting or referral hesitation.

Direct immunofluorescence is helpful to differentiate BLS from autoimmune bullous disorders. Furthermore, pemphigoid and pemphigus might develop in patients with long-standing LS [4]. BP180 and BP230 antibodies have been found in some LS patients as well [3]. The DIF finding was interpreted as nonspecific in our patient.

Extragenital BLS does not clearly have a negative prognostic risk [5]. Although vulvar LS has the potential to be premalignant or malignant, the possible risk of the bullous form is uncertain.

Treatment of extragenital BLS is similar to the classical form [3]. There is no previous report about using topical calcineurin inhibitors in genital BLS. To prevent new blisters, we preferred to use topical tacrolimus to maintain clearing. However, to decrease the possible irritative effect of the medication, we used it after vesiculobullous lesions had been completely epithelized.

Conclusion

In conclusion, this presented case is unique in terms of having isolated vulvar involvement of BLS. As Sauder et al. [3] claimed previously, BLS might be more common than dermatologists realize. Future reports may clarify the mechanism, the risk of malignant transformation, treatment response, and the relapse rate of vulvar involvement in BLS.

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