International Journal of Women's Dermatology 3 (2017) 225–227

Contents lists available at ScienceDirect

International Journal of Women's Dermatology

Case Report

Vulvar lichen planus pemphigoides☆,☆☆

J. Loyal [a,*], S. Rashtak [b]

[a] University of Vermont College of Medicine, Burlington, VT
[b] Department of Dermatology, University of Cincinnati College of Medicine, Cincinnati, OH

ARTICLE INFO

Article history:
Received 21 June 2017
Accepted 3 July 2017

ABSTRACT

Lichen planus pemphigoides (LPP) is a rare blistering disease with features of both lichen planus and bullous pemphigoid. LPP typically appears on the extremities and occasionally involves the oral mucosa. Herein, we describe a case of LPP of the vulva of an 80-year-old woman, an uncommon location for this disease process. This clinical scenario can be confused with a number of similarly appearing entities such as erosive vulvar lichen planus, mucous membrane pemphigoid, and erosive lichen sclerosus et atrophicus. In fact, our patient carried a diagnosis of lichen sclerosus by an outside physician for 2 years prior to being properly diagnosed and treated. A detailed discussion of the epidemiology, clinical, and pathogenesis as well as the histologic and immunofluorescence characteristics of this uncommon diagnosis is presented. Our case emphasizes the necessity of microscopic analysis to differentiate lookalike disease states when making a diagnosis and choosing the correct therapeutics.

Introduction

Lichen planus pemphigoides (LPP), a rare blistering disease with features of both lichen planus (LP) and bullous pemphigoid (BP), frequently appears on the extremities and occasionally orally (Zaraa et al., 2013). We report a case of LPP of the vulva, which is an atypical location for this disease process.

Case report

An 80-year-old female patient presented with a 2-year history of itching and painful erosions of the vulva. She carried a diagnosis of lichen sclerosus by an outside physician. The patient also reported blisters and sloughing of the gums, resulting in significant pain to the point that she could only eat soft solid foods. A pertinent review of systems was otherwise negative, and a family history of blistering or autoimmune diseases was denied.

An examination of the genitalia revealed adhesions of the labia minora and white sclerotic patches admixed with erythematous smooth patches on the labia majora and minora. Small areas of white lacy reticular streaks and erosions on the labia and vaginal introitus were noted (Fig. 1). An oral examination showed recession and areas of sloughing of the gingival mucosa (Fig. 2). A histopathologic examination of the labia showed subepidermal clefts and band-like lymphocytic infiltrates with melanophages in the papillary dermis (Fig. 3). An analysis of the maxillary gingiva showed focal subepithelial clefts and superficial lymphocytic infiltrates (Fig. 4). Direct immunofluorescence (DIF) of labial tissue revealed linear deposition of immunoglobulin (Ig) G, IgA, and fibrin along the basement membrane zone (BMZ; Fig. 5). A salt-split skin technique (using 2M NaCl) revealed depositions of IgG and IgA that were limited to the epidermal side confirming mucosal pemphigoid (Figure 6).

A diagnosis of vulvar and oral LPP was rendered. Treatment consisted of topical clobetasol 0.05% ointment twice daily to the genitalia and dexamethasone 0.5mg/5mL oral solution four times daily. At follow-up, there was improvement in symptoms, but genital and oral erosions persisted. Initial blood tests were within normal limits and initiation of dapsone 50mg daily was planned.

Discussion

LPP is an uncommon acquired autoimmune blistering disease with features of both LP and BP (Zaraa et al., 2013). LPP has a peak incidence in the fifth and sixth decades of life and is most common in women (Zaraa et al., 2013). LPP is typically idiopathic; however, some report an association with medications, phototherapy, and malignancy (Zaraa et al., 2013). LPP is characterized by the development

☆ Conflicts of interest: None.
☆☆ Funding sources: None.
* Corresponding author.
E-mail address: Jameson.Loyal@med.uvm.edu (J. Loyal).

http://dx.doi.org/10.1016/j.ijwd.2017.07.002

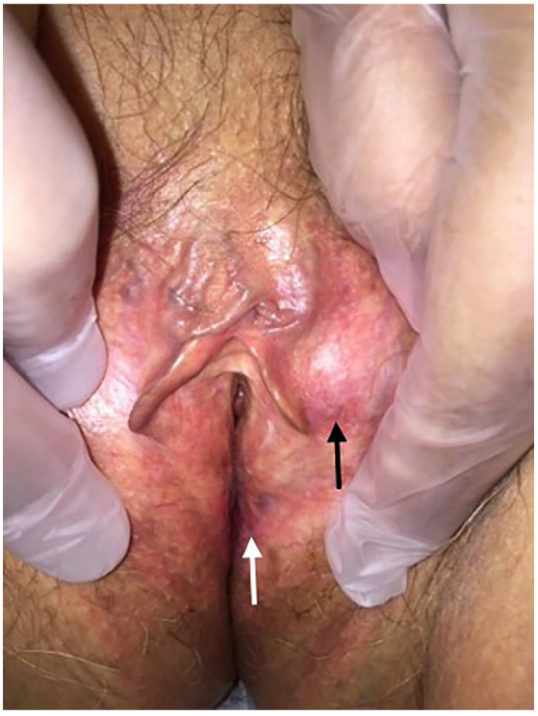

Fig. 1. Genital examination: Vulva shows white and erythematous patches on bilateral labia with a small area of white lacy striae (black arrow) and erosion (white arrow).

of bullae on LP lesions and on normal-appearing skin. However, LPP can be observed in patients with bullae on only LP-inflicted areas (Archer et al., 1992; Zaraa et al., 2013). Linear depositions of IgG and/or C3 on the BMZ is characteristic (Zillikens, 1999). LPP occurs predominately on the upper and lower extremities with occasional

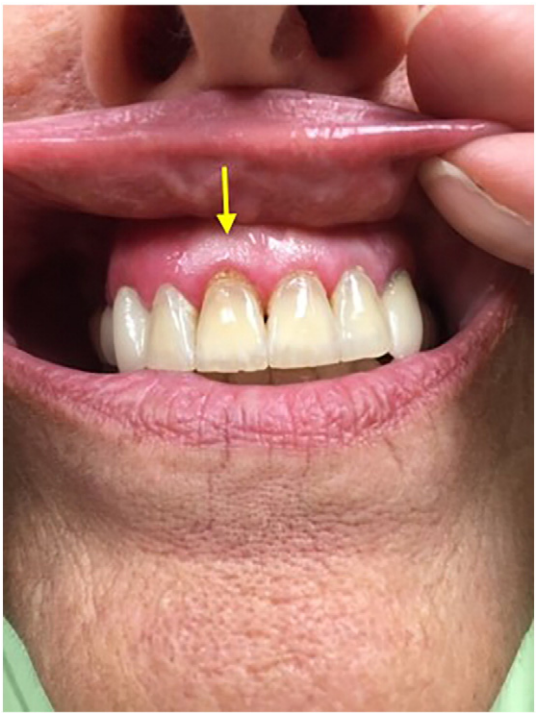

Fig. 2. Oral examination: Recession of maxillary and mandibular gingiva with mild white sloughing (yellow arrow).

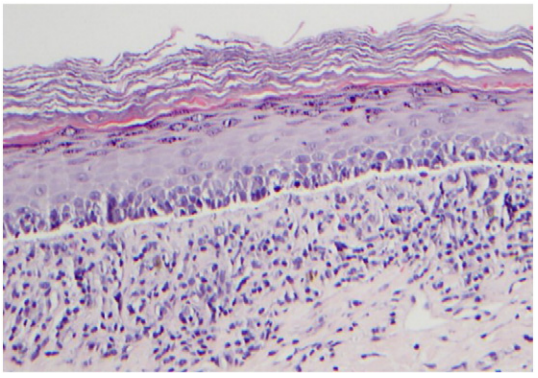

Fig. 3. Subepidermal clefts and band-like lymphocytic infiltrates with melanophages of vulva (hematoxylin–eosin; ×200 original magnification).

oral involvement (Zaraa et al., 2013). To our knowledge, this is the first reported case of vulvar LPP.

The pathogenesis of LPP results from inflammation of LP, which causes damage to the BMZ and subsequent exposure of BP 230-kDa and 180-kDa antigens (Zaraa et al., 2013; Zillikens, 1999). A unique 200-kDa antigen has also been demonstrated in LPP (Zillikens, 1999). Exposure of these antigens results in the production of IgG autoantibodies against the BMZ that contribute to the bullae development (Zaraa et al., 2013; Zillikens, 1999). Autoantibodies in LPP react with the C-terminus of the NC16A reactivity domain of the 180-kDa antigen (Zillikens, 1999).

Vulvar LPP should be distinguished from similar-appearing conditions such as erosive vulvar lichen planus, mucous membrane pemphigoid, and erosive lichen sclerosus et atrophicus. Erosive vulvar lichen planus, an inflammatory scarring disorder, results from T-cell-mediated damage to basal keratinocytes and is distinguished by its lack of linear IgG and/or C3 staining in DIF (Cooper et al., 2005). Mucous membrane pemphigoid, another scarring inflammatory disorder, is characterized by positive DIF staining of IgG, C3, and/or IgA but lack of fibrin staining at the BMZ (Chan et al., 2002). Erosive lichen sclerosus et atrophicus results in epidermal atrophy and dermal sclerosis with reports of preferentially staining for only fibrin on DIF (Keith et al., 2015).

Conclusion

Thorough history and clinical examination along with histopathology and DIF examinations can lead to the correct diagnosis and treatment. Potential treatment options include topical and systemic corticosteroids, dapsone, cyclosporine, azathioprine, methotrexate,

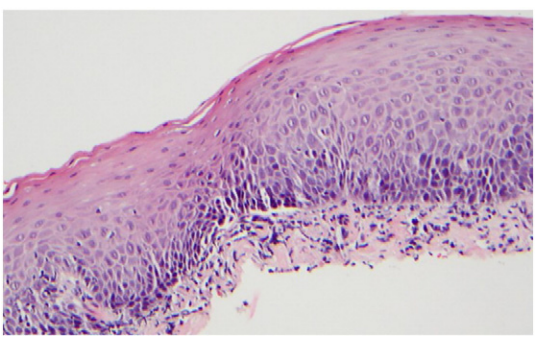

Fig. 4. Subepithelial clefts and lymphocytic infiltrates of maxillary gingiva (hematoxylin–eosin; ×200 original magnification).

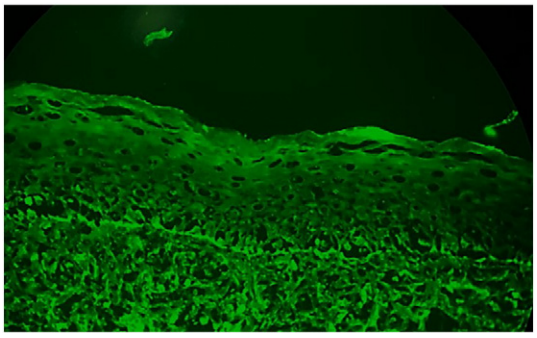

Fig. 5. Direct immunofluorescence shows linear immuglobulin G positivity on the basement membrane zone of the vulva.

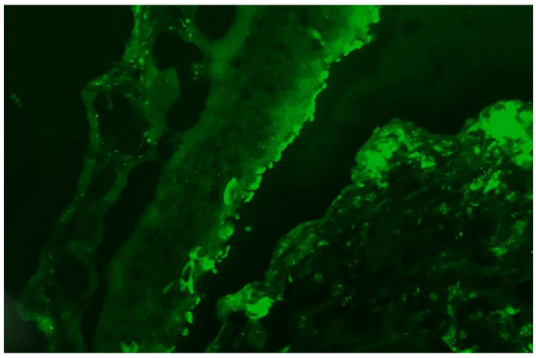

Fig. 6. Salt-split direct immunofluorescence is immuglobulin G-positive on the epidermis of the vulva.

and retinoid medications because they have been previously used to treat cutaneous LPP and have been shown to be effective (Allen et al., 1987; Zaraa et al., 2013).

Acknowledgements

The authors are indebted to Diya Mutasim, MD, and Kerith Spicknall, MD, both from the Department of Dermatology at the University of Cincinnati College of Medicine, for their invaluable assistance with the dermatopathology analysis and photo preparation for this report. No compensation was received for their role.

References

Allen CM, Camisa C, Grimwood R. Lichen planus pemphigoides: Report of a case with oral lesions. Oral Surg Oral Med Oral Pathol 1987;63:184–8.
Archer CB, Cronin E, Smith NP. Diagnosis of lichen planus pemphigoides in the absence of bullae on normal-appearing skin. Clin Exp Dermatol 1992;17:433–6.
Chan LS, Ahmed AR, Anhalt GJ, Bernauer W, Cooper KD, Elder MJ, et al. The first international consensus of mucous membrane pemphigoid. Arch Dermatol 2002;138:370–9.
Cooper SM, Dean D, Allen J, Kirtschig G, Wojnarowska F. Erosive lichen planus of the vulva: Weak circulating basement membrane zone antibodies are present. Clin Exp Dermatol 2005;30:551–6.
Keith PJ, Wolz MM, Peters MS. Eosinophils in lichen sclerosus et atrophicus. J Cutan Pathol 2015;42:693–8.
Zaraa I, Mahfoudh A, Sellami MK, Chelly I, El Euch D, Zitouna M, et al. Lichen planus pemphigoides: Four new cases and a review of the literature. Int J Dermatol 2013;52:406–12.
Zillikens D. Acquired skin disease of hemidesmosomes. J Dermatol Sci 1999;20:134–54.