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Case Report

Erosive Lichen Planus with Multisystem Involvement: A Multidisciplinary Diagnostic and Therapeutic Challenge

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Abstract:

Lichen planus (LP) is a rare autoimmune disorder that can affect multiple mucocutaneous sites simultaneously. Erosive lichen planus (ELP), a variant of LP, presents with chronic and painful ulcerations that may lead to significant morbidity if left untreated. We present a case of a 68-year-old woman with severe ELP involving the oral cavity, esophagus, vulva, and conjunctiva. The patient was initially diagnosed with separate dermatological conditions for each site involved, demonstrating the complexity of diagnosing ELP. Despite various treatment regimens, the patient's disease remained refractory and ultimately developed into squamous cell carcinoma of the vulva. This case highlights the need for a multidisciplinary team in managing complex autoimmune mucocutaneous diseases. It also illustrates patient-centered care, as the chosen treatment regimen was minimally invasive and broad enough to address differential diagnoses.

Key words: lichen planus; erosive variant; ocular; complex; refractory; pemphigoid; squamous cell carcinoma

Abbreviations

DIF: Direct Immunofluorescence

ELP: Erosive Lichen Planus

JAKi: Janus Kinase Inhibitor

LP: Lichen Planus

LPP: Lichen Planus Pemphigoides

MMP: Mucous Membrane Pemphigoid

SCC: Squamous Cell Carcinoma

Introduction

Lichen planus (LP) is a rare immune-mediated disorder with variable etiologies and mucocutaneous involvement. LP may occur solitarily but usually involves several surfaces, including the oral, laryngeal, ocular, and genital mucosae, skin, scalp, and nails. Not only does LP vary in its location, but also in morphology. Erosive lichen planus (ELP) is a variant of LP involving chronic and painful ulceration of mucocutaneous surfaces, most frequently of the oral cavity and genitals [1].

The pathological process of LP remains unclear but is proposed to be a T-cell-mediated autoimmune disease [1]. A virus, drug, or contact allergen alters epidermal self-antigens [1]. These altered self-antigens mimic normal self-antigens found on basal keratinocytes that CD8+ T cells then target for apoptosis [1]. The Hepatitis C virus is the most common virus associated with LP, yet the association between the hepatitis C virus and ELP has not been established [1,2]. Several medications are linked to LP and ELP, and few autoimmune diseases, specifically ulcerative colitis and alopecia areata, more commonly occur in patients with LP [2]. Usually, the cause of LP is unknown [2].

Erosive lichen planus is not only challenging to diagnose due to its variable presentation but also carries a risk of severe outcomes if left untreated, making timely management crucial. Ultimately, chronic ELP of the oral cavity and genital area holds a 1 to 5% risk of progressing to squamous cell carcinoma [2–4]. The painful and scarring nature of oral ELP often causes restrictive mouth opening and discomfort with eating [2,4]. ELP of the vulva may produce adhesions, which can cause urinary and defecation difficulties and interfere with a patient's sexual function, and chronic inflammation of conjunctival LP can progress to blindness [2]. Here, we present a case of severe ELP involving the gingivae, pharynx, esophagus, vulva, and conjunctiva that was initially diagnosed as a myriad of distinct diagnoses for each area involved. This case demonstrates the complexities of diagnosing ELP, particularly when distinguishing it from conditions with similar presentations like Lichen Planus Pemphigoides (LPP) and Mucous Membrane Pemphigoid (MMP). It also emphasizes the importance of a multidisciplinary approach to

multisystem mucocutaneous disease to ensure comprehensive patient care.

Case Presentation

A 68-year-old female presented with blurry vision, ocular burning, dysphagia, hoarseness, and vulvar erosions. She reported no history of blistering or pruritic skin lesions. A biopsy of the esophagus and mandibular area 7 years prior revealed erosive lichen planus, described as chronic lichenoid mucositis and hyperkeratosis. Direct immunofluorescence (DIF) was uninterpretable due to the lack of an intact epithelial-connective tissue interface. Five years before presentation, she was diagnosed with vulvar lichen sclerosis based on clinical presentation alone. Sixteen months before presentation, a biopsy of the oral cavity showed chronic lichenoid mucositis and hyperkeratosis with a negative DIF for fibrinogen, IgG, IgM, IgA, and C3. Direct immunofluorescence on the posterior pharyngeal wall yielded negative staining in the epithelium for IgG, IgM, IgA, and C3. Bullous Pemphigoid BP180 serum antibodies at that time were within a normal range (0-20 Units/mL). One year before presentation, the patient developed worsening ocular conjunctival injection and symblepharon, and she was referred to dermatology for suspected ocular cicatricial pemphigoid.

On physical exam, our patient was found to have erosions along the lower gingiva and more severe erosions on the upper gingiva and hard palate without Wickham's striae (Figure 1), a 2 cm erosion on the anterior labia minora and medial clitoral head with depigmented macules surrounding the introitus (Figure 2), and symblepharon and ankyloblepharon bilaterally on the ocular surfaces with conjunctival injection (Figure 3). Given severe oral ELP, a new biopsy of the vulva was obtained, revealing lichenoid dermatitis, favouring lichen planus, with superimposed features of lichen simplex chronicus. Specifically, the histopathology demonstrated a brisk lichenoid interface change characterized by dyskeratosis and vacuolization at the tips of the rete ridges in association with a dense bandlike lymphocytic infiltrate with numerous apoptotic keratinocytes (Figure 4). Neither hyalinized collagen bands nor loss of melanocytes were appreciated, making the previous diagnosis of lichen sclerosis less likely. A GMS stain, spirochete stain, and CD123 stain (Figure 5) were also performed on the vulvar biopsy and were negative for fungal infection, spirochete infection, and malignancy, respectively.

At the time of presentation, the patient was taking the following medications: cetirizine, paroxetine, and ocular lubricant, none of which are associated with drug-induced LP [2]. She had received all her childhood and influenza vaccines annually. Tests for complete blood count, liver, and renal function all showed results within normal limits. Additionally, the patient tested negative for hepatitis A, B, and C virus. A serum/urine immunofixation was performed to rule out hematologic abnormalities, showing no significant findings. Given biopsy-proven ELP of the vulva, oral cavity, and esophagus, a global diagnosis of ELP was reached. Our patient frequently followed up with a multidisciplinary team, including a dermatologist, dentist, otolaryngologist, ophthalmologist, and gynecologist. After several collaborative treatment plans (Figure 6), our patient continues to have severe refractory disease, with the unfortunate development of invasive squamous cell carcinoma (SCC) of the vulva.

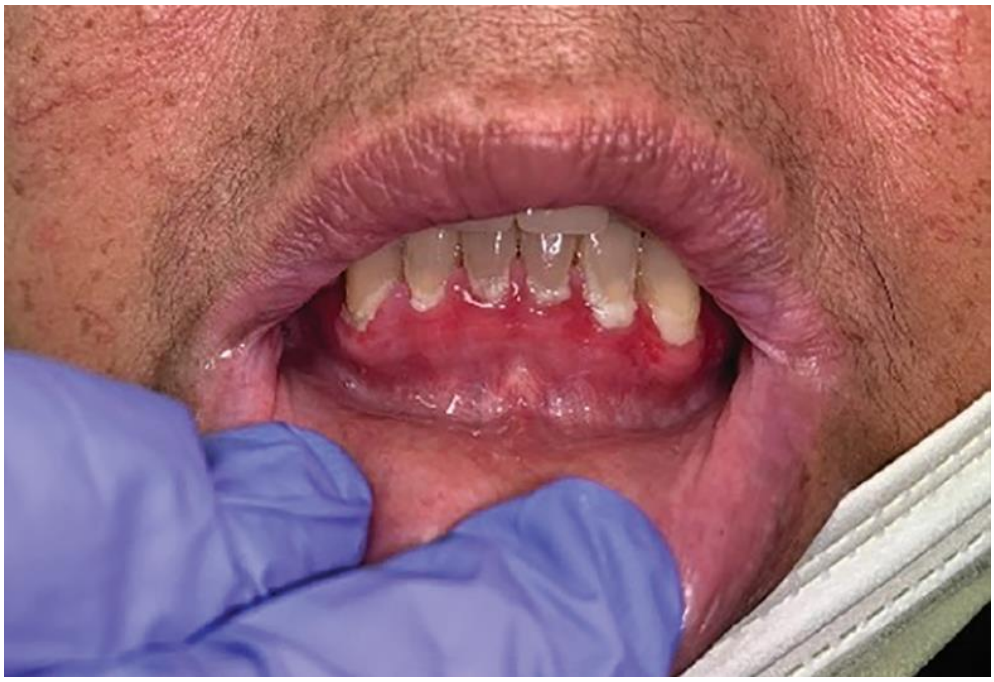

Figure 1: Oral erosive lichen planus: erosions along the lower gingiva

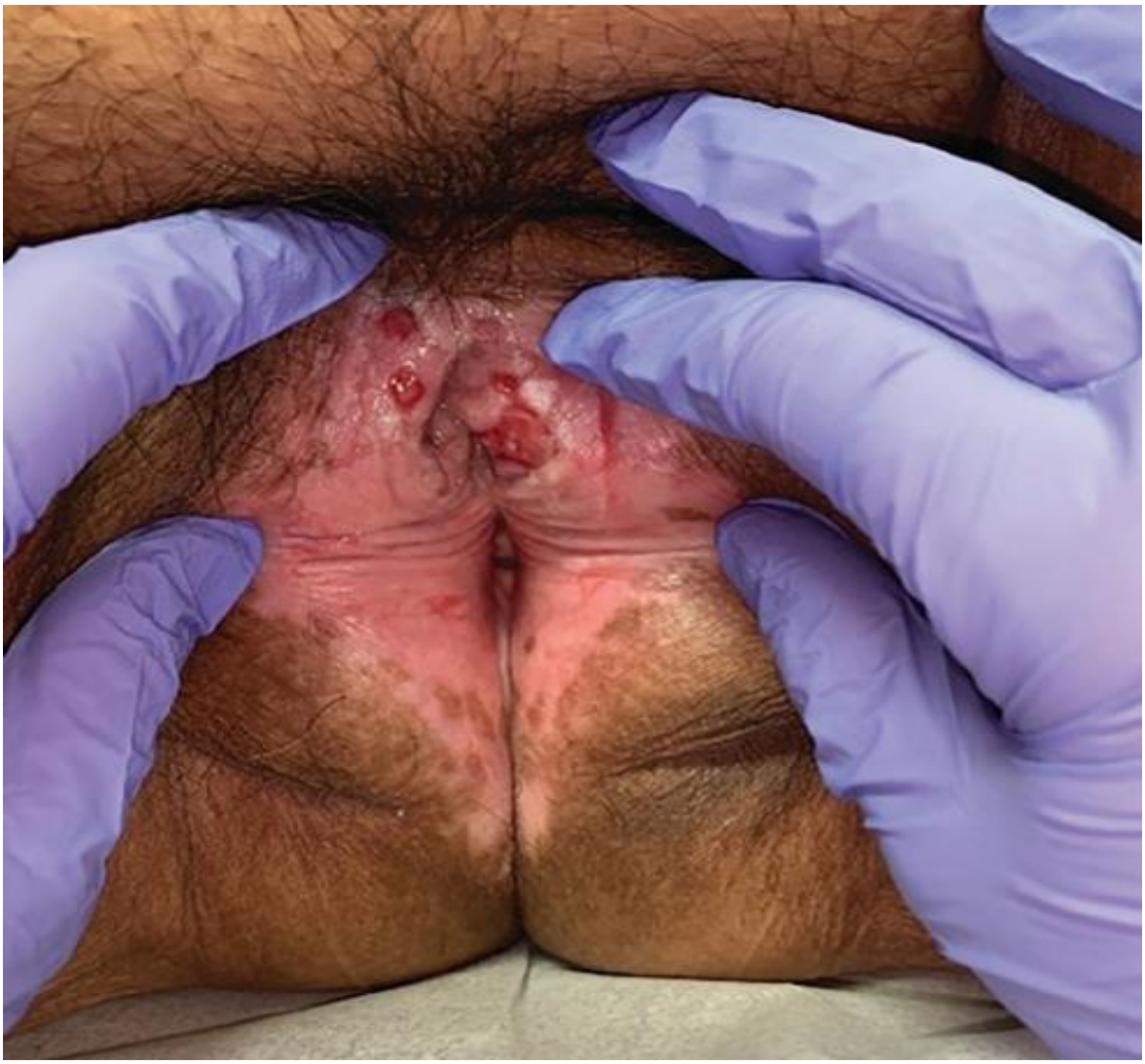

Figure 2: Vulvar erosive lichen planus: a 2 cm erosion on the anterior labia minora and medial clitoral head with depigmented macules surrounding the vaginal introitus.

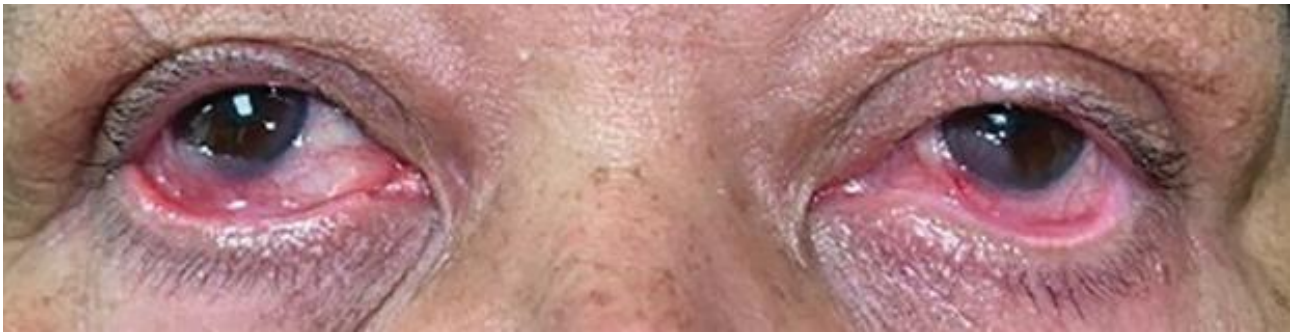

Figure 3: Ocular lichen planus: symblepharon and ankyloblepharon of the eyes with conjunctival injection

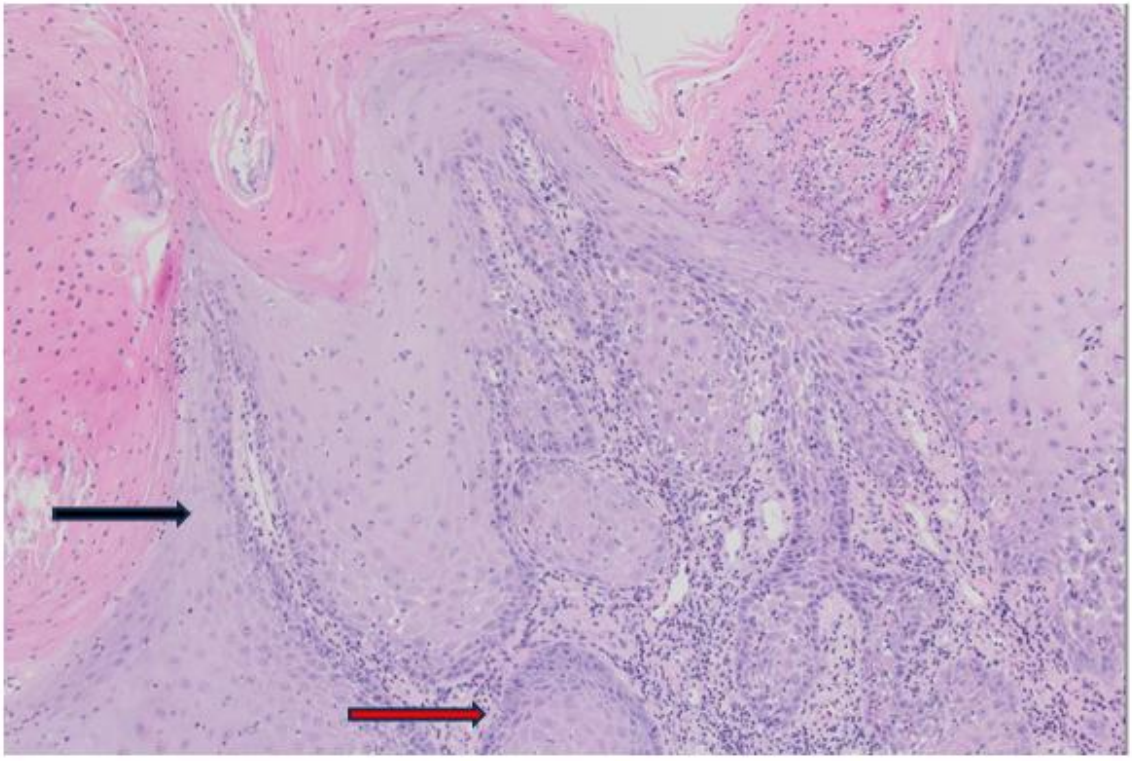

Figure 4: Histopathologic findings of the right vulva showing lichenoid dermatitis (red arrow) with superimposed features of lichen simplex chronicus (black arrow).

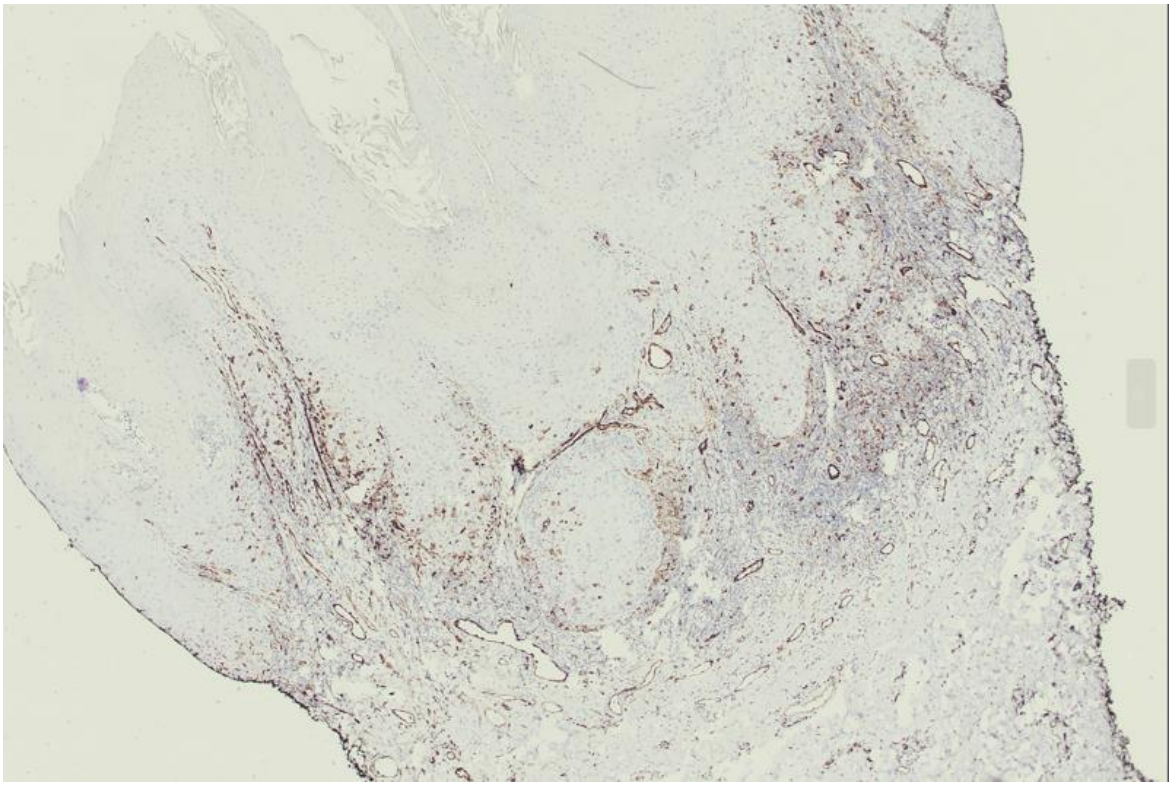

Figure 5: CD123 stain of a biopsy from the right vulva, negative for squamous cell carcinoma.

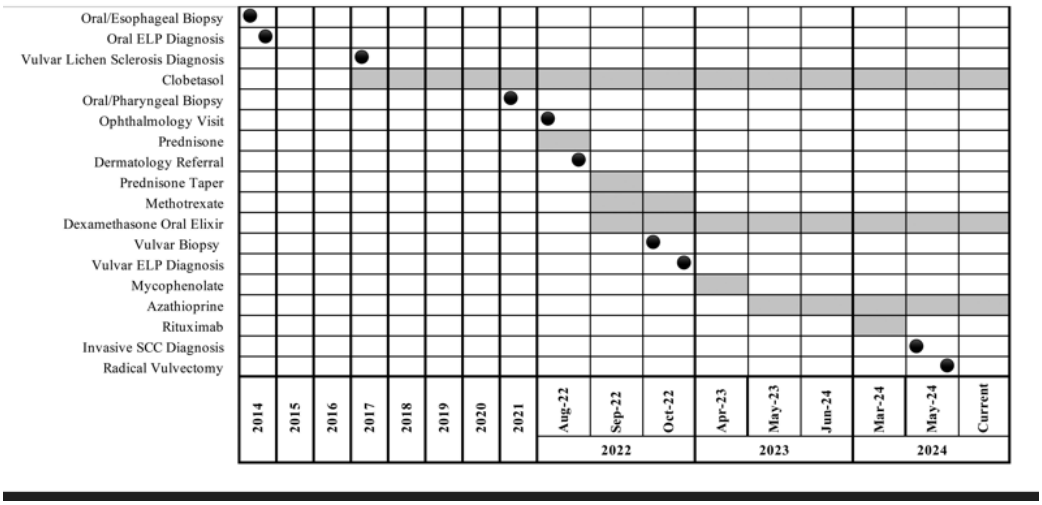

Figure 6: Biopsy/Diagnoses/Treatment timeline for our patient's case

Discussion

Erosive lichen planus often presents with significant diagnostic and therapeutic challenges, especially when it involves multiple mucocutaneous sites and overlaps with similar conditions such as LPP and MMP [1]. Our patient presented with features of ELP, including painful erosions in the oral cavity and vulva. These clinical findings, augmented by multi-site histopathological evidence of a dense band-like lymphocytic infiltrate and numerous apoptotic keratinocytes, strongly suggested ELP as the primary diagnosis [1]. The development of squamous cell carcinoma in the vulva further reinforced this diagnosis [2-4].

Direct immunofluorescence is a valuable diagnostic tool for identifying ELP and distinguishing it from other mucocutaneous diseases, such as MMP. However, negative DIF results can often occur in ELP, particularly in lesions without Wickham striae [5]. Buajeeb et al. found that patients with oral lesions without Wickham striae had negative DIF results 30% of the time, compared to an average rate of 17.1% across all lesions [5]. DIF results vary across studies, with favourable rates ranging from 37% to 97% [6]. Therefore, when DIF results are negative but clinical suspicion for ELP remains high, it is imperative to consider the complete clinical picture and a comprehensive histopathological analysis. In our patient's case, although the negative DIF did not exclude other diagnoses such as MMP or LPP entirely, the solid clinical and histopathological evidence supported the diagnosis of ELP.

The decision to forgo another DIF test was made with careful consideration. The multidisciplinary team, involving a dermatologist, ophthalmologist, otolaryngologist, and gynaecologist, agreed that performing an additional biopsy for DIF could increase the risk of further patient discomfort and potential complications. The care team also agreed that further testing would unlikely change the management plan [5-7]. This approach supports the notion that while DIF can help diagnose LP, particularly in ambiguous cases or differentiating from diseases like pemphigoid, its utility diminishes when clinical and histopathological evidence is clear [6].

When considering the ocular involvement in our case, we acknowledge that a conjunctival biopsy is the most definitive method for diagnosing ocular disease. However, previous studies have demonstrated that ocular LP can be diagnosed without a conjunctival biopsy [8,9]. In cases of ocular LP or cicatricial pemphigoid where extraocular skin lesions exist, obtaining the initial biopsy from a non-ocular site is recommended to minimize possible complications associated with ocular biopsies [9]. Therefore, our patient's extensive biopsy-proven ELP suggested that her conjunctival disease was most likely ELP as well, and we felt a conjunctival biopsy was not in the best interest of the patient.

We evaluated both LPP and MMP as potential diagnoses in this case due to their clinical similarities with ELP. The presence of lichenoid inflammation suggested the possibility of LPP [10]. However, our patient did not show subepidermal blistering on histopathology or cutaneous involvement, which is present in 82% of LPP cases [10]. Mucous membrane pemphigoid was also considered due to the extensive mucosal erosions, including the conjunctiva. However, the specific features of ELP—particularly the band-like lymphocytic infiltrate and lack of significant blistering—helped to differentiate it from MMP [11]. Additionally, BP180 antibodies were not elevated and ultimately did not bolster a case for LPP or MMP [10,12].

The primary objective in ELP management is to alleviate symptoms and improve quality of life, as complete resolution is often unattainable. Topical corticosteroids are the cornerstone of treatment for oral LP and have demonstrated significant efficacy in reducing inflammation and alleviating oral ELP pain [7]. Additionally, topical corticosteroids are the most cost-effective treatment for oral LP. This was carefully considered when selecting our patient's oral ELP therapy. Alternative treatments for erosive oral LP include topical calcineurin inhibitors, like tacrolimus and pimecrolimus, which are documented to be as effective as topical corticosteroids in treating oral LP [7]. However, calcineurin inhibitors carry a Food and Drug Administration warning due to a potential, though unconfirmed, association with cancer [7,13]. For anogenital ELP lesions, super-potent steroids like clobetasol are used to lessen symptoms and prevent scarring rather than achieve resolution [13]. Our patient continues to use topical clobetasol and reports symptomatic relief for vulvar lesion flares.

Patients with severe unresponsive ELP may require a 4-to-6-week course of oral prednisone while transitioning to steroid-sparing agents [1,13]. Due to the persistent nature of ELP, alternative therapies are often necessary. Recent studies have highlighted the potential of topical Janus kinase inhibitors (JAKi) in treating refractory ELP [14]. However, further controlled studies on alternative therapies are needed to establish definitive treatment options for patients with refractory ELP. In our patient's case, treatment with prednisone, hydroxychloroquine, methotrexate, mycophenolate mofetil, rituximab, and azathioprine was ineffective. Acitretin was considered but ultimately unaffordable for our patient. This comprehensive treatment regimen was carefully chosen to cover potential differential diagnoses, such as LPP or MPP [10,12]. Unfortunately, the patient required a radical vulvectomy with gynecology

oncology due to invasive SCC of the vulva; therefore, apremilast is being pursued as the next treatment option.

Conclusion

This case emphasizes the challenge of diagnosing and managing ELP. While ELP was strongly favored based on clinical and histopathological evidence, the comprehensive treatment regimen—including azathioprine, rituximab, and mycophenolate mofetil—was broad enough to address other potential diagnoses, including LPP or MMP. This tactic is patient-centered and ensures optimal care, regardless of the final diagnosis. Our case demonstrates that managing a patient with complex multisystem mucocutaneous diseases requires a multidisciplinary approach. Effective communication and collaboration among dermatologists, ophthalmologists, otolaryngologists, dentists, and gynaecologists ensure that treatment plans are timely and tailored to meet the unique needs of each patient.

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