

# Erosive Lichen Planus-Vulvovaginal Gingival Syndrome: A Case Report

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## ABSTRACT

**Objective:** Lichen planus is a chronic mucocutaneous disease characterized by oral ulcerations with or without cutaneous lesions. Erosive lichen planus is a rare form of lichen planus characterized by painful oral ulcers, and often associated with ulcers in the genital area. Erosive lichen planus is considered a pre-malignant condition. The objective of this paper is to report a case of Erosive lichen planus (vulvovaginal gingival syndrome) in a 67-year-old female with oral and genital lesions; to highlight the nature of a severe variant of lichen planus, its clinical presentation and management.

**Case report:** A 67-year-old female presented with a one-year history of recurrent mouth ulcers. This was associated with itchy cutaneous lesions around the navel and genital region. Patient had been placed on steroid therapy alongside antibiotics by general practitioners. This gave her temporary relief. Intra-oral examination revealed wide spread erosions and erythema, worse on both buccal mucosae and the tongue. Some areas were covered with pseudomembrane which could not be scrapped off. There was also marked desquamative gingivitis. An impression of Erosive lichen planus-vulvovaginal gingival syndrome, was made based on a history and clinical assessment and patient was commenced on prednisolone 50mg daily, dapsone 50mg 12hourly, and dexamethasone oral rinse 12hourly. Subsequent reviews showed healing oral and cutaneous lesions. Patient is currently on maintenance dose of prednisolone 10mg, dapsone 100mg daily, and oral steroid rinse.

**Conclusion:** This study reports a case of Erosive lichen planus 'vulvovaginal- gingival syndrome' in an elderly female, with refractory oral and genital ulcers. Early and accurate diagnosis with appropriate treatment is very important for alleviation of symptoms and improvement of patient's wellbeing.

**Keywords:** Lichen planus, oral ulcers, genital ulcers

## INTRODUCTION

Lichen planus is a chronic inflammatory mucocutaneous disease affecting the mucous membranes, skin, hair and nails (Gorouhi et al., 2014). In majority of patients, there may be only oral lichen planus, with no cutaneous involvement or lichen planus at other mucosal sites. This may be called isolated oral lichen planus (Gorouhi et al., 2014).

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Erosive lichen planus (ELP) is a variant of lichen planus, characterized by painful persistent ulcerations affecting mainly the mouth (oral lichen planus) and genital (vulva and penile planus) (Gall & Navarro-Fernandez, 2022). Other mucosae rarely involved

include the oesophagus, eyelid, larynx, anus, bladder, and external auditory canal (Katzka et al., 2010; Webber et al., 2012).

The aetiology of ELP is unknown (Balaji et al., 2011). It is, however, considered to be an auto-immune condition with T-cells destroying the epithelial cells (Balaji et al., 2011; Simpson et al., 2013). Precipitating factors include psychological stress, dental materials, viruses, or drugs, nutritional deficiency, diabetes, hypertension, and genetic predisposition (Elenbaas et al., 2022).

The condition affects women more (Lewis & Bogliatto, 2013), occurs commonly between the 5<sup>th</sup> and 8<sup>th</sup> decades of life and rarely affects children (Eisen, 2003). ELP may occur in conjunction with other clinical types of lichen planus (Gall & Navarro-Fernandez, 2022) and patient usually present with painful irregular erosions and ulcerations in the oral cavity, occurring on the buccal mucosa, labial mucosa, tongue, vermillion border, and gingiva (desquamative gingivitis). The lesions are often symmetrical and bilateral (Gaurav et al., 2016). ELP involving the genitals typically presents with dysuria, pain, burning sensation, dyspareunia with possible post-coital bleeding (Cooper et al., 2008). Female patients may have blood-tinged vaginal discharge (Gall & Navarro-Fernandez, 2022). In women, ELP presenting with oral ulcerations, desquamative gingivitis, as well as ulcers on the vulva and in the vagina is termed vulvovaginal gingiva syndrome, in men the equivalent condition is termed penogingival syndrome (Danesh-Sani et al., 2012). Diagnosis of ELP is often made via history and clinical assessment. A biopsy may however be done when diagnosis is in doubt or to rule out dysplastic changes (Gall & Navarro-Fernandez, 2022). Direct immunofluorescence is helpful in reaching a conclusive diagnosis (Schlosse, 2010). It is important to note that the histology often appear non-specific because the presence of ulcerations leads to the absence of the epidermis, thus preventing the typical histological findings of lichen planus (Eisen, 2002). The main stay of treatment is steroid therapy, which may be combined with other immunomodulators (Dunaway et al., 2020). Treatment is often challenging and relapses are common (Gall & Navarro-Fernandez, 2022). Being a chronic condition, treatment is often long term, given intermittently, or continuously (Amanda, 2015). ELP is considered a premalignant condition as it has been associated with malignant transformation, hence the need for long term follow-up (Balaji et al., 2011; Gaurav et al., 2016). The aim of this article is to report a case of vulvovaginal gingival syndrome in an elderly woman, in order to emphasize the need for thorough clinical assessment to aid correct diagnosis and appropriate treatment.

## CASE REPORT

A 65 year-old female was referred to the Oral medicine clinic of the University of Benin Teaching Hospital (UBTH), on account of multiple painful ulcers in the mouth of 1 year duration. Lesions (ulcerations) also appeared in the navel, genital and anal region around the onset of the oral lesions. Patient had been treated by general practitioners, but had little relief. General examination revealed a distressed patient, lymph nodes not palpable and blood pressure was 130/80mmHg. Intra-oral examination revealed poor oral hygiene with marked halitosis. There was wide spread erosions and erythema, worse on both buccal mucosae and lateral surfaces of the tongue. This was associated with peppery painful sensation which affected her feeding. Some areas were covered with pseudomembrane which could not be scraped off. There were also features of desquamative gingivitis. An impression of Erosive lichen planus-vulvovaginal gingival syndrome was made based on clinical assessment. Fasting blood sugar (FBS), full blood count (FBC) and retroviral screening (RVS) were requested for.

Patient was immediately commenced on 40mg prednisolone daily, dapsons 50mg 12hourly, dexamethasone oral rinse (2mg in 10mls water after meals), chlorhexidine oral rinse 8 hourly (before meals); all for one week. Kenalog (skin) cream for the cutaneous lesions with a plan for possible referral to Dermatology clinic. On a one-week review, the oral condition had improved. (Figure 4)

The genital lesions had improved too but still present. Results of the investigations were as follows: FBC showed low packed cell volume (PCV) of 28.2% suggestive of anaemia, RVS negative, FBS normal 87mg/dl. Prednisolone was tapered to 30mg daily, dapsons and dexamethasone regimen continued for 1 week. Patient was placed on haematinics to improve the PCV and chlorhexidine oral rinse discontinued. On a second review a week later, oral lesions had improved significantly. The marginal gingiva erythema and pseudomembrane had cleared remarkably. Patient's oral hygiene and feeding had improved. The drugs were continued for 1 week.

Patient was monitored on a weekly basis with improving condition noticed. Prednisolone was tapered weekly till patient was placed on a maintenance dose of 10mg daily, with the dapsons and dexamethasone regimen maintained. On the 5<sup>th</sup> review, oral lesions had cleared remarkably with little areas of erythema. The genital lesions also showed marked improvement; hence patient was told to continue the kenalog skin cream till the lesion clears completely.



Figure 1 (a): Wide spread intra-oral Erosions with Pseudomembrane on the tongue



Figure 1(b): Desquamative gingivitis with slough on the gingiva



Figure 2: Ulceration in the navel



Figure 3: Genital Ulceration



Figure 4: one week review showing improved oral condition





Figure 6: 5<sup>th</sup> week review showing resolving mucocutaneous lesions A-C (Oral lesion), D (Navel), E (Genital)

#### DISCUSSION

The bilateral and symmetrical presentation of the oral lesions of ELP, occurring on the buccal mucosae, sides of the tongue, labial mucosa, gingiva (desquamative gingivitis) are almost pathognomonic of oral ELP (Amanda, 2015). This patient had oral lesions in the sites stated. A consensus on Diagnostic criteria for Erosive lichen planus with vulva involvement (ELPV) has been reached to aid the diagnosis of the condition (Simpson et al., 2013). They are: (1) well demarcated erosions/erythematous area at the vaginal introitus (2) presence of hyperkeratotic border to lesions and/or Wickham striae in the surrounding skin (3) symptoms of pain and burning (4) scarring/ loss of normal architecture (5) presence of vaginal inflammation (6) involvement of other mucosal surfaces (7) presence of well defined inflammatory band involving the dermoepidermal junction (8) presence of inflammatory band consisting predominantly of lymphocytes (9) signs of basal cell inflammation. Presence of any three features is sufficient to make a diagnosis of ELPV (Simpson et al., 2013). Criteria 1, 2, and 3 were present in this patient, thus aiding a clinical diagnosis of vulvovaginal-gingival syndrome. Apart from the oral and genital lesions, there was also associated anal involvement in this patient. Hence she often experiences pain when passing urine and defecating. This condition has a high impact on patient's quality of life (Lewis & Bogliatto, 2013; Gorouhi et al., 2014) with the painful, persistent oral ulcers affecting feeding

hence resulting in nutritional deficiencies, weight loss and even depression (Amanda, 2015). This was the case observed in this patient, as she has had lesions for over a year. She couldn't feed well hence then weight loss confirmed by the patient and anemia revealed in the FBC. Her quality of life was seriously affected. Upon institution of steroid therapy (topical and systemic), which is the mainstay of treatment, patient responded well to the steroid therapy. In the acute phase, duration of initial therapy may be 4-6 weeks, with a maintenance therapy being needed for a long time due to the chronic nature of the condition, and to prevent recurrences (Amanda, 2015). This patient showed marked improvement within 5 weeks of treatment. Other drugs that have been found to be beneficial are mycophenolate mofetil, hydroxychloroquine, methotrexate, dapsone, azathioprine, tacrolimus, oral antifungals, oral antibiotics like tetracycline (Cooper et al., 2008; Raj et al., 2012; Simpson et al., 2013; Amanda, 2015;). The patient was placed on dapsone, along with the steroid therapy. Dapsone is a steroid-sparing drug that is very effective in treatment of ELP (Raj et al., 2012). The combination of dapsone and prednisolone showed satisfactory result. The patient is currently on routine follow up, on maintenance dose of 10mg prednisolone, 50mg dapsone 12 hourly till lesions are completely clear.

## CONCLUSION

Erosive lichen planus presenting with oral and genital ulcerations is an uncommon variant of lichen planus. It can be of diagnostic and treatment challenge, due to the chronic and refractory nature of the condition. Thorough clinical assessment is of utmost importance to reach accurate diagnosis and thus institute appropriate treatment. Long term monitoring of patients is very important for early detection any any malignant change.

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