

Hypertrophic lichen planus of the vulva – A missed diagnosis

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Abstract

Lichen planus (LP) is an inflammatory dermatosis which can affect the skin, nails, and all mucous membranes, including the genitalia. Lichen planus on vulvar keratinized skin can manifest with diverse clinical features, probably due to higher temperature, PH, humidity, and bacterial flora which may modify typical cutaneous features. While lichen planus (LP) may affect the vulva in isolation, it may also be part of generalized outbreak in up to 20% cases. Herein, a case of a 53 year-old female who presented with a severely pruritic plaque over labia majora Since 6 -7 months, with no response to potent topical corticosteroids is reported. Provisional diagnosis of lichen simplex chronicus was considered however, histopathology was suggestive of hypertrophic lichen planus.

Key words: Hypertrophic, lichen planus, vulva

INTRODUCTION

Lichen planus (LP) was first described by Erasmus Wilson in 1869. Lichen planus (LP) is an inflammatory dermatosis which can affect the skin, nails, and all mucous membranes.^[1] It is a T-cell- mediated inflammatory dermatosis that affects both keratinized and nonkeratinized squamous epithelium.^[2] Genitalia involvement usually comes into light only when the patient is symptomatic. Variants of vulval LP (VLP) includes classical or papulosquamous LP, erosive LP, hypertrophic LP, and lichen pilanopilaris. Erosive LP is by far the most common variant affecting vulvae while hypertrophic LP is less described. Treatment is usually challenging and unsatisfactory.

CASE REPORT

A 53-year -old female presented with itching and skin lesions over genitals since 6-7 months. There was no history of skin lesions over the body or oral mucosa. The patient had attained menopause 3 years ago. There was no associated history of dysuria, dyspareunia, vaginal discharge, or genital

ulcers. There was also no history of drug reaction. There was no specific history of surgical procedures in that area. She was not known case of diabetes mellitus or hypertension.

On examination, a single, large, well- defined hyperpigmented plaque with multiple follicular openings covering almost the whole of mucosal aspect of the right labia majora was seen [Figure 1]. There was no atrophy, depigmentation, or lichenification. Lesion was soft and non tender. Inguinal lymph nodes were not enlarged. There was no oozing or bleeding from the lesions. Per speculum examination revealed no abnormalities. The oral and conjunctival mucosa were normal. There were no similar lesions elsewhere over the body. Systemic examination revealed no abnormalities. Her blood and routine investigations were within normal limits. HIV, hepatitis B virus surface antigen, hepatitis C virus (HCV), and venereal disease research laboratory (VDRL) test were non reactive.

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Provisional diagnosis of lichen simplex chronicus was kept. Differential diagnosis considered included LP and lichen sclerosus et atrophicus. As the patient did not show response to moderately potent topical corticosteroids (TCS), the case was subjected to vulvar biopsy. Histopathological findings revealed follicular plugging with Wedge shaped hypergranulosis, basal cell degeneration, dense band-like infiltrate, pigment incontinence, and presence of melanophages, which were suggestive of hypertrophic LP of vulva [Figure 2a and b]. The patient was treated with topical potent corticosteroids (clobetasol propionate 0.05% ointment) along with antihistaminics – hydroxyzine 10 mg twice daily orally for 3 weeks. There was moderate improvement in skin condition.

DISCUSSION

Vulvar dermatoses are common, potentially debilitating conditions that can present to dermatologists and gynaecologists alike. LP on vulvar keratinized skin is a diagnostic challenge to both clinicians and pathologists. Hypertrophic lichen planus (LP) has the most dramatic clinical presentation and is the least described in the literature commonly affecting women during their 5th or 6th decade of life.^[3]

Variants of LP that can affect the vulva includes classic type or papulosquamous LP, hypertrophic LP, Erosive LP, and lichen planopilaris.^[4,5] Erosive LP is the most common variant that affects the vulva. It presents with white lacy reticulations on the medial aspect of the labia minora and vaginal orifice. It may result in an alteration of the normal architecture including the development of vaginal adhesions, stenosis, and in severe cases, complete obliteration of the vaginal canal and urinary retention. Hewitt–Pelisse syndrome (vulvo-vaginal- gingival syndrome^[6]) is a form of erosive LP; that involves the vulva, vagina, and mouth. Papulosquamous or classic type presents as small violaceous pruritic papules or plaques with overlying reticulations and involves keratinized and perianal skin. Vaginal involvement should be ruled out with speculum examination.

Hypertrophic LP of vulvae is rare, and presents with hyperkeratotic white plaques involving vulvae, perineum, and perianal area, which sometimes gives an appearance similar to squamous cell carcinoma (SCC) of vulva with incidence seen in 3% of cases.^[7] Hypertrophic LP of vulva may be associated with HCV, autoimmune disorders, alopecia areata, and coeliac disease.^[7] In lichen planopilaris,^[4] follicular keratotic papules are limited to the hair-bearing labia majora and mons pubis but can involve scalp, trunk, and extremities. Characteristically unexplained exacerbations and remissions are seen. Vulval involvement may also be seen as a part of LP on other sites in as many as 51% of cases in women. Therefore,

a thorough clinical examination should be done even in cases not complaining of genital symptoms.

Although a diagnosis of VLP can be made clinically, particularly in patients with the classic type, a biopsy may be necessary in certain situations. Patients with the hypertrophic form of VLP should undergo biopsy to rule out vulvar intraepithelial neoplasia, squamous cell carcinoma (SCC), and lichen simplex chronicus which this variant can mimic.^[8] Patients with the erosive variant should also undergo a biopsy to rule out diseases like bullous pemphigoid, pemphigus vulgaris, and mucus membrane pemphigoid.^[8,9] With erosive VLP, a biopsy should be performed at the edge of the lesion.

Treatment of vulvovaginal LP is often challenging. TCSs are widely used as the initial line of therapy in genital LP. An initially intensive regime, typically with clobetasol propionate 0.05% twice daily for 1-2 months, aims to assertively arrest the inflammatory process before tapering to a maintenance regimen of twice to thrice weekly.^[4,10] Potent or moderately potent corticosteroids ointments will control most symptoms of classical genital lichen planus, as disease is usually self-limiting.^[11]

Kirtschig *et al.*^[11] studied 44 cases with mucosal vulval lichen planus. Thirty out of 44 cases studied had oral findings and nine had cutaneous findings. No association with antibodies to hepatitis B or C virus was noted. Cases were treated with potent to very potent topical corticosteroids (TCS) however, symptoms persisted in majority of cases. In seven (16%) patients, VLP (vulval lichen planus) was in remission after a disease duration between 2 and 18 years. One patient developed vulval SCC.

Hypertrophic LP may require intralesional corticosteroids.^[12] The management of erosive LP is more difficult, as disease is persistent and will respond to potent topical corticosteroids (TCS)' application for long duration.^[4] For more resistant disease, use of topical tacrolimus, systemic immunosuppressants, and oral hydroxychloroquine.^[12] Long- term evaluation of these patients is advised because of risk of malignancy. VLP (Vulval lichen planus) can develop into a chronic condition, and monitoring for any signs of neoplastic change should be done. Various therapeutic modalities exist; but, as yet, no one treatment has consistently been shown to be of superior efficacy.

Our patient responded to potent TCS (clobetasol propionate 0.05% ointment) with relief from pruritus; however, the hyperpigmentation persisted.

Fahy *et al.*^[12] studied 100 consecutive cases diagnosed with VLP at Mayo Clinic. In 49% of cases, time to diagnosis was more than 1 year. Three cases (3%) had vulval dysplasia, including invasive SCC. Sixty-eight cases (68%)



Figure 1: Right labia majora showing large hyperpigmented plaque with multiple dilated follicular openings

had multisite LP involvement. Eleven patients (11%) had disease remission. Cases with VLP had a low frequency of disease remission.

An accurate diagnosis and follow-up of any vulvar lesion is important. Patients with cutaneous or oral LP should be examined for genital involvement. As the disease in this area may be more susceptible to malignant changes, these patients require close monitoring. Biopsy of any atypical lesion is essential to exclude the development of malignancy. Women with vulvar diseases need specific care, considering not only genital aspects but also skin changes. In this context, the dermatologist is a very well-trained professional to take good and complete care of these patients.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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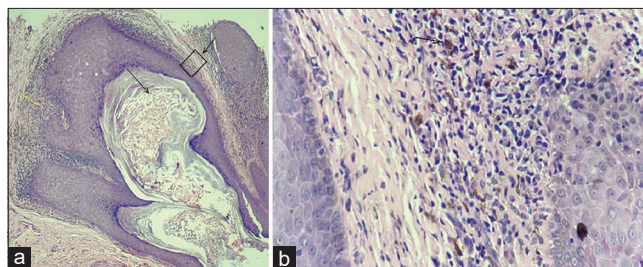


Figure 2: (a) ($\times 10$) Epidermis shows follicular plugging with Wedge-shaped hypergranulosis and basal cell degeneration. Dense band-like infiltrate visible at dermoepidermal junction. (b) ($\times 40$) Highlighted area from Figure 2a shows pigment incontinence and presence of melanophages in the dermis

Conflicts of interest

There are no conflicts of interest.

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