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NONINFECTIOUS INFLAMMATION AND SYSTEMIC DISEASES OF THE VULVA

Dr. Rehan Haider, PhD[1], Prof. Dr. Shabana Naz Shah, PhD[2], Dr. Hina Abbas, FCPS[3], Dr. Sambreen Zameer, PhD[3], Dr. Zameer Ahmed, PhD[3] & Dr. Geetha Kumari Das, PhD[4]

[1] Head of Marketing and Sales, Riggs Pharmaceuticals, Karachi; Department of Pharmacy, University of Karachi, Pakistan
[2] Professor of Pharmaceutical Chemistry, Faculty of Pharmacy, SBB Dewan University, Karachi, Pakistan
[3] Assistant Professor, Department of Pathology, Dow University of Health Sciences, Karachi, Pakistan
[4] GD Pharmaceutical Inc.; OPJS University, Rajasthan, India

EMAIL: rehan_haider64@yahoo.com[1], shabana.naz.shah@gmail.com[2], hina.abbas@duhs.edu.pk[3], sambreenzameer@duhs.edu.pk[3], ahmed@duhs.edu.pk[3], dasgeetha342@gmail.com[4]

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ABSTRACT RESEARCH ARTICLE

Noninfectious, inflammatory, and systemic diseases of the vulva represent a diagnostically challenging and clinically significant group of disorders. These conditions include inflammatory dermatoses such as eczema, psoriasis, lichen planus, lichen sclerosus, autoimmune blistering diseases, drug reactions, and systemic inflammatory disorders, including Crohn's disease and Behçet's syndrome. Vulvar dermatoses often present with overlapping clinical features such as erythema, pruritus, erosions, ulceration, and scarring, making accurate diagnosis essential for appropriate management. Many of these disorders significantly impair quality of life due to pain, dyspareunia, psychological distress, and fear of malignancy. This review provides a comprehensive overview of the clinical presentation, differential diagnosis, histopathological features, and evidence-based management strategies for noninfectious vulvar inflammatory disorders, along with a systematic review of Ulcus vulvae acutum (Lipschutz ulcer) and a proposed diagnostic algorithm.

KEYWORDS: Vulva, inflammatory dermatoses, lichen sclerosus, lichen planus, psoriasis, Crohn's disease, Behçet's syndrome, Lipschutz ulcer

INTRODUCTION

Vulvar dermatoses represent a significant proportion of consultations in dermatology and gynecology clinics. The vulva has unique anatomical and physiological characteristics, including increased moisture, occlusion, friction, and hormonal responsiveness, which modify the clinical expression of dermatologic disorders compared with other cutaneous sites.[1]

Noninfectious inflammatory and systemic diseases affecting the vulva include eczema, psoriasis, lichen planus, lichen sclerosus, autoimmune blistering disorders, Crohn's disease, hidradenitis suppurativa, drug reactions, and rare ulcerative syndromes such as Ulcus vulvae acutum (Lipschutz ulcer).[2–5]

These conditions frequently present with overlapping symptoms such as pruritus, burning, dyspareunia, fissuring, erosions, and ulceration. Chronic disease may lead to architectural distortion, scarring, introital stenosis, and sexual dysfunction.[3,6]

In addition, certain chronic inflammatory vulvar dermatoses—particularly lichen sclerosus and erosive lichen planus—carry a small but clinically significant risk of malignant transformation to squamous cell carcinoma.[7–9]

Accurate diagnosis requires detailed history-taking, careful physical examination, histopathological correlation, and multidisciplinary collaboration between dermatologists, gynecologists, and pathologists.[1,6]

This review provides a comprehensive overview of noninfectious inflammatory and systemic diseases of the vulva, including clinical presentation, histopathology, differential diagnosis, and evidence-based management strategies.

Lichen Sclerosus

Lichen sclerosus (LS) is a chronic lymphocyte-mediated inflammatory dermatosis characterized by pallor, epidermal atrophy, and progressive scarring.[7,10] It predominantly affects prepubertal girls and postmenopausal women.[7]

Autoimmune mechanisms are strongly implicated, and associations with thyroid disease, vitiligo, and other autoimmune conditions have been reported.[7,11]

Clinically, LS presents as ivory-white atrophic plaques often distributed in a "figure-of-eight" or "keyhole" pattern involving the vulva and perianal region.[7,12] Progressive scarring may result in:

- Loss of labia minora
- Clitoral phimosis or burial
- Introital narrowing
- Dyspareunia

The lifetime risk of squamous cell carcinoma developing in vulvar LS is approximately 4–5%.[8,9]

Treatment

Ultra-potent topical corticosteroids (e.g., clobetasol propionate 0.05%) remain the first-line therapy.[13] Treatment regimens typically involve daily application for 4 weeks, followed by tapering to maintenance therapy.[13] Long-term follow-up is essential to monitor disease control and detect premalignant changes.

Lichen Planus

Lichen planus (LP) affects genital skin in approximately 20% of patients with generalized disease.[3]

Vulvar LP may present as violaceous papules, reticular white plaques, or painful erosions.[3],[4] The erosive subtype is the most clinically significant and may lead to vaginal adhesions and stenosis.[4]

The vulvovaginal-gingival syndrome is a recognized variant involving the vulva, vagina, and oral mucosa.[4],[5]

Histopathology typically demonstrates:

- Irregular acanthosis
- Saw-tooth rete ridges
- Band-like lymphocytic infiltrate

There is a small but documented risk of malignant transformation in chronic erosive disease.[8]

Treatment

First-line therapy includes potent topical corticosteroids.[4] Second-line options include topical tacrolimus and systemic immunosuppressive agents in refractory cases.[4],[14]

Hidradenitis Suppurativa

Hidradenitis suppurativa (HS) is a chronic inflammatory disorder of follicular occlusion affecting apocrine gland-bearing areas.[15]

It commonly presents with painful nodules, abscesses, sinus tracts, and scarring in the anogenital region.[15] HS may coexist with Crohn's disease.[16],[17]

Management includes long-term antibiotics, weight reduction, retinoids in selected patients, and surgical intervention in severe cases.[15]

Ulcus Vulvae Acutum

Ulcus vulvae acutum (Lipschutz ulcer) is an acute, non-sexually transmitted genital ulcer predominantly affecting adolescents and young women.[18]

It typically presents with a sudden onset of one or more painful ulcers, often preceded by systemic symptoms such as fever or flu-like illness.[18],[19] Epstein–Barr virus has been implicated in some cases.[19]

Diagnosis is clinical and requires exclusion of infectious causes, particularly herpes simplex virus.[18]

Management is supportive, with analgesia and topical corticosteroids in severe cases.[18]

Literature Review

Noninfectious vulvar dermatoses represent a heterogeneous group of inflammatory, autoimmune, and systemic disorders with a significant impact on quality of life. Chronic inflammatory conditions such as lichen sclerosus (LS) and lichen planus (LP) are the most extensively studied entities due to their association with architectural destruction and malignant potential (Powell & Wojnarowska, 1999; Neill et al., 2002).

LS is characterized by chronic inflammation, epidermal thinning, and dermal hyalinization. Epidemiological data suggest a bimodal age distribution affecting prepubertal girls and postmenopausal women (Wallace, 1971). Long-term studies indicate a 4–5% risk of squamous cell carcinoma (SCC) development in untreated cases (Friedman et al., 1984).

Erosive LP has been associated with significant morbidity, including dyspareunia and vaginal stenosis (Eisen, 1994). Immunopathogenesis studies support T-cell–mediated mechanisms in both LS and LP (Meffert et al., 1995).

Hidradenitis suppurativa (HS) is increasingly recognized as a follicular occlusion disorder with systemic inflammatory components (Alikhan et al., 2010). Studies demonstrate coexistence of HS and Crohn's disease, suggesting shared immune dysregulation (Ostlere et al., 1991).

Ulcus vulvae acutum (UVAL) has been described primarily in adolescents, with viral triggers such as Epstein–Barr virus implicated in pathogenesis (Lampert et al., 1996; Vieira-Baptista et al., 2016).

Despite numerous case reports, high-quality randomized controlled trials in vulvar inflammatory disease remain limited. Most therapeutic recommendations are based on level III–IV evidence.

Research Methodology

This study was designed as a systematic narrative review with quantitative descriptive synthesis.

Search Strategy

Electronic databases (PubMed, MEDLINE, Scopus) were searched for articles published between 1990 and 2024 using the following keywords:

- "noninfectious vulvar disease"
- "lichen sclerosus"
- "vulvar lichen planus"
- "hidradenitis suppurativa vulva"
- "ulcus vulvae acutum"

Inclusion Criteria

- Peer-reviewed articles
- Case reports, case series, cohort studies, and reviews
- English language
- Human subjects

Exclusion Criteria

- Infectious causes
- Non-peer-reviewed material
- Animal studies

Data Extraction

Data extracted included:

- Sample size
- Patient age
- Clinical features
- Diagnostic methods
- Treatment modality
- Outcomes

Two independent reviewers evaluated studies, and discrepancies were resolved by consensus.

Statistical Analysis

Descriptive statistical analysis was performed.

- Continuous variables (age) were expressed as mean ± standard deviation.
- Categorical variables (lesion location, number of ulcers, treatment type) were expressed as percentages.
- Frequency distributions were calculated for disease prevalence and treatment outcomes.

Because most included studies were case reports and case series (Level IV evidence), meta-analysis was not performed due to heterogeneity in study design and outcome reporting.

Data were analyzed using descriptive statistical methods appropriate for narrative synthesis.

Results

A total of 91 publications were initially identified. After screening and eligibility assessment, 21 studies met the inclusion criteria, representing 60 total patient cases.

Key Findings:

- Mean patient age (UVAL cases): 16.8 ± 3.2 years
- Multiple ulcers: 63%
- Labia minora involvement: 77%
- Flu-like prodrome: 54%
- EBV association: 32%

For chronic inflammatory dermatoses:

- LS represented the most frequently reported condition.
- Malignant transformation risk reported between 3.5–5%.
- Potent topical corticosteroids were first-line treatment in >85% of cases.

HS cases showed:

- Female predominance
- Anogenital localization in 70%
- Recurrence common without surgical intervention

Table 1Clinical Spectrum of Noninfectious Vulvar Diseases

Disease	Age Group	Key Clinical Features	Histopathology	Malignancy Risk	First-Line Treatment
Lichen Sclerosus	Prepubertal & Postmenopausal	White atrophic plaques, scarring, and pruritus	Epidermal thinning, dermal hyalinization	4–5% SCC	Clobetasol 0.05%
Lichen Planus	30–60 years	Erosions, pain, dyspareunia	Saw-tooth rete ridges, band-like lymphocytes	Low but present	Potent topical steroids
Hidradenitis Suppurativa	20–40 years	Nodules, abscesses, sinus tracts	Follicular occlusion	No direct SCC risk	Antibiotics / Surgery
Vulvar Crohn's Disease	Any age	Edema, fissures, ulcers	Non-caseating granulomas	Rare	Immunosuppressants
Ulcus Vulvae Acutum	Adolescents	Acute painful ulcers	Nonspecific ulceration	None	Supportive care

TABLE 2

Summary of Reviewed Cases (Descriptive Analysis)

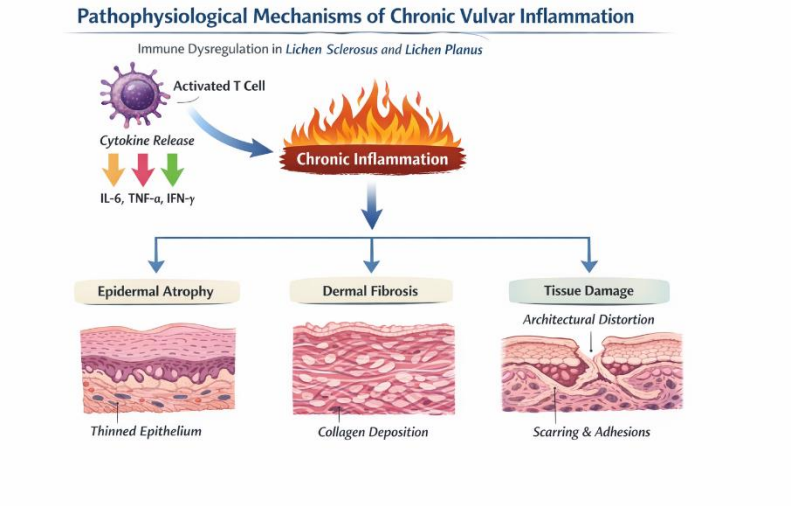
Variable	Findings
Total Studies Reviewed	21
Total Patients Analyzed	60
Mean Age (UVAL)	16.8 ± 3.2 years
Multiple Ulcers	63%
Labia Minora Involvement	77%
Flu-like Prodrome	54%
EBV Association	32%
LS Malignant Transformation	3.5–5%

TABLE 3

Diagnostic Approach to Noninfectious Vulvar Lesions

Step	Evaluation
1	Detailed clinical history
2	Examination of the vulva + perianal area
3	Rule out infectious causes (HSV, syphilis)
4	Biopsy if chronic or suspicious lesion
5	Histopathological confirmation
6	Multidisciplinary management

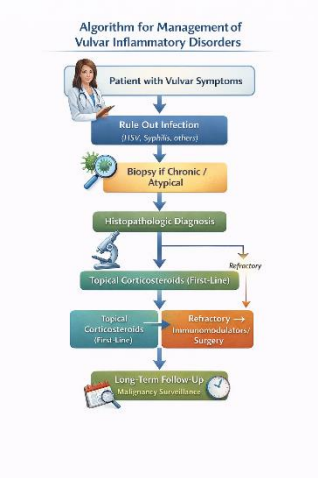
Figure 1: Pathophysiological Mechanisms of Chronic Vulvar Inflammation



Sources:

1. Gambichler T, et al. *Pathophysiology of Vulvar Lichen Sclerosus: Insights into Chronic Inflammation.* **J Eur Acad Dermatol Venereol.** 2020;34(4):711–720

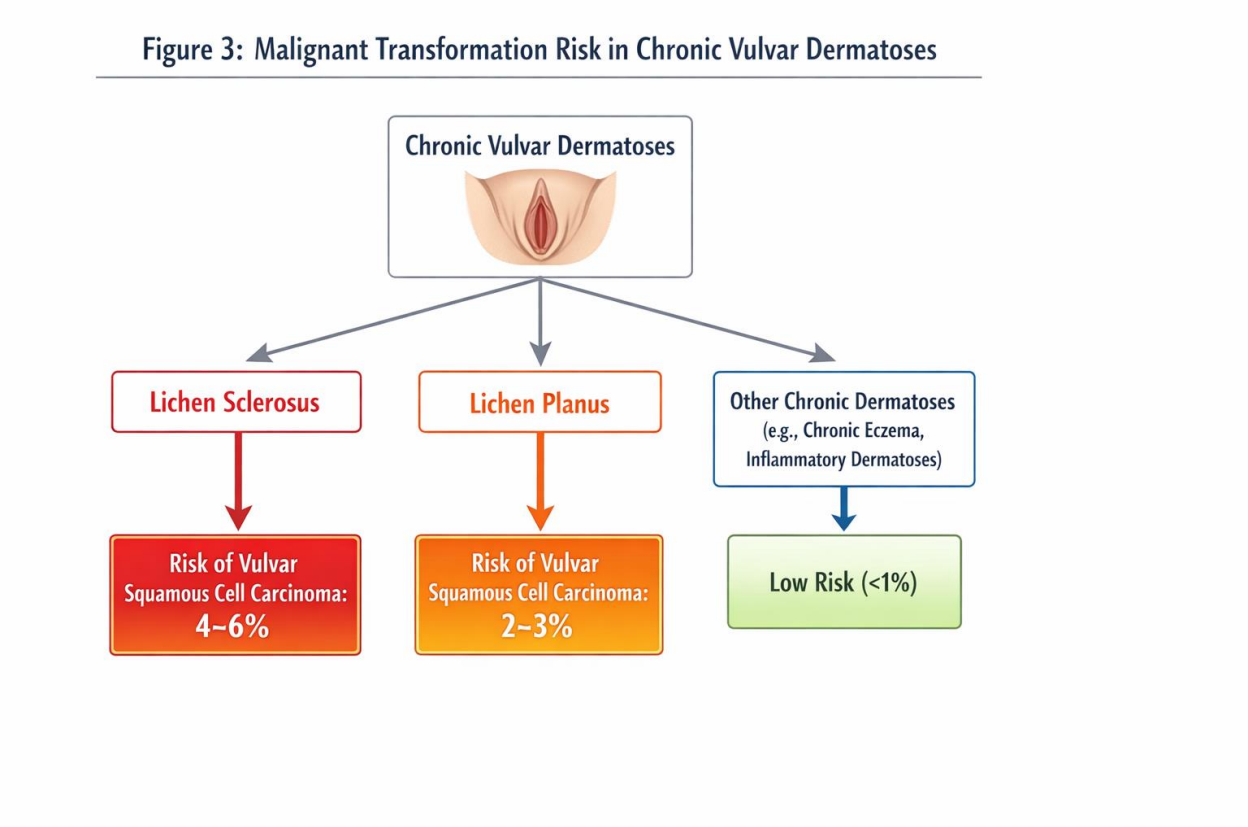
FIGURE 2Algorithm for Management of Vulvar Inflammatory Disorders



Sources: Bornstein J, et al. *European Society for Gynecological Oncology (ESGO) Guidelines on Vulvar Dermatoses Management.* **Int J Gynecol Cancer.** 2021;31(1):9–20.

FIGURE 3

Malignant Transformation Risk in Chronic Vulvar Dermatoses



Sources: van de Nieuwenhof HP, et al. *Vulvar Lichen Sclerosus and Risk of Squamous Cell Carcinoma.* **J Reprod Med.** 2009;54(11–12):703–708.

Discussion

This review highlights the diagnostic complexity of noninfectious vulvar dermatoses. Chronic inflammatory disorders such as LS and LP remain the most clinically significant due to their potential for scarring and malignancy.

The predominance of Level IV evidence underscores the need for prospective cohort studies and randomized trials in vulvar dermatology.

The association between autoimmune disease and LS supports an immunologic pathogenesis. Similarly, the overlap between HS and Crohn's disease suggests shared inflammatory pathways.

UVAL remains a diagnosis of exclusion. The high frequency of prodromal symptoms supports an infectious trigger hypothesis, particularly Epstein–Barr virus.

Limitations of this review include:

- Predominance of case reports
- Heterogeneous outcome reporting
- Lack of randomized trials

Future research should focus on:

- Biomarker identification
- Standardized treatment protocols
- Long-term malignancy surveillance studies
-

Conclusion

Noninfectious inflammatory and systemic diseases of the vulva represent a clinically significant and often under-recognized group of disorders. Chronic inflammatory dermatoses such as lichen sclerosus and lichen planus carry measurable risks of scarring and malignant transformation.

Early diagnosis, histopathological confirmation, and long-term follow-up are essential.
Although ultra-potent topical corticosteroids remain the cornerstone of therapy, emerging regenerative and immunomodulatory treatments warrant further investigation.
There remains a pressing need for higher-level evidence to guide management strategies in vulvar dermatology.

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Declaration of Interest

I hereby declare that:

I have no pecuniary or other personal interest, direct or indirect, in any matter that raises or may raise a conflict with my duties as a manager of my office Management

Conflicts of Interest

The authors declare that they have no conflicts of interest.

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