REVIEWS

Vulvar Dermatoses: A Primer for the Sexual Medicine Clinician

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ABSTRACT

Introduction. Vulvar dermatoses are common dermatological conditions that affect the vulva, and can cause considerable pain, irritation, pruritus, and burning, and have an adverse impact on a woman's sexual function.
Aim. To provide an overview of the clinical features, etiology, and management options for the common vulvar dermatoses, including lichen sclerosus, lichen planus, lichen simplex, contact dermatitis, and vulvar psoriasis, and briefly describe the impact of vulvar dermatoses on sexual function.
Methods. The key words "vulvar dermatoses," "lichen sclerosus," "lichen planus," "lichen simplex chronicus," "vulvar dermatitis," and "vulvar psoriasis," were utilized to search Medline and PubMed for articles, with special attention given to those published within the past 5 years.
Main Outcome Measure. Five hundred thirty-six results were generated from the literature search. Publications that were judged current and relevant to the pathophysiology, evaluation, and treatment of vulvar dermatoses were included in the review.
Results. Fifty-seven articles were selected for inclusion in this review.
Conclusions. Vulvar dermatoses can cause chronic pain, itching, and dyspareunia, and can have a profound effect on a woman's sexual expression and comfort. Delay in diagnosis is often due to hesitancy to seek treatment on the part of the patient or delay in biopsy on the part of the provider. This can result in failed prescriptive and self-treatment measures, worsening symptoms, and frustration and sexual dysfunction for the patient, and potentially the development of squamous cell carcinoma. It is imperative for sexual medicine providers, who commonly treat women with vulvar concerns, to be familiar with the presentation, diagnosis, and treatment of common vulvar dermatoses and their effect on sexual function. **Kellogg Spadt S and Kusturiss E. Vulvar dermatoses: A primer for the sexual medicine clinician. Sex Med Rev 2015;3:126–136.**

Key Words. Vulvar Dermatoses; Lichen Sclerosus; Lichen Planus; Lichen Simplex; Vulvar Psoriasis; Vulvar Dermatitis; Female Sexual Dysfunction

Introduction

Vulvar complaints are among the most frequent causes for a woman to visit a healthcare provider. The symptoms associated with vulvar discomfort, including skin changes, lesions, burning, pruritus, fissures, and dyspareunia, are often poorly managed, under diagnosed and/or treated in a "generic" fashion, without specific attention to the nuances of each clinical case. The diagnosis of vulvar dermatological disorders can represent a challenge for healthcare professionals, as disorders can be multifactorial and concomitant with other disorders. Since dermatologists may be unfamiliar with specific aspects of the vulvar examination and female sexual concerns, and general gynecologists may be unfamiliar with the management of dermatological complaints, sexual medicine specialists are often the professionals that women consult when experiencing vulvar skin discomfort that affects their sexuality.

Vulvar skin conditions irritation can affect women of any age. The etiology of vulvar complaints can be difficult to determine due to varying presentations, ranging from pruritus and burning, to changes in genital architecture. Symptoms can

be vague, intermittent, continuous, localized, or generalized [1,2]. Vulvar dermatoses can affect every aspect of a woman's life, including activities of daily living, sexuality, relationships, feelings of self-worth, and well-being. For many women, the vulva is an area of the body that is viewed as private, and any pain or irritation brings with it feelings of embarrassment or fear. As a result, women commonly self-treat and spend an exorbitant amount of money in over-the-counter products and overzealous hygiene practices, which can exacerbate symptoms. Women may fear that their vulvar symptoms are the result of sexually transmitted infections, undiagnosed cancer, poor hygiene, or that there is no medical treatment for their condition. These factors compound the profound impact vulvar discomfort has on a woman and can further lead to delay in a woman seeking care [1–3].

Vulvar dermatoses are common chronic conditions. Ongoing symptom flares require specialized skill to manage over time. Healthcare providers may feel inept or frustrated during long-term management, and this can contribute to women receiving suboptimal treatment [2,4,5]. The aim of this article is to provide an updated review of benign vulvar dermatological conditions that commonly present to clinicians working in sexual medicine, discussing etiology, diagnosis, treatment options, and comorbid sexual dysfunction.

Dermatological Evaluation

Dermatological disorders that affect the vulva often present as nonspecific pruritus, burning, or pain. Because infectious and neoplastic disease can have clinical similarities in both symptoms and appearance, it is important to conduct a detailed assessment before arriving at an empiric diagnosis. To formulate a differential diagnosis, a detailed inspection of the vulva, working inward toward the vestibule from the crural folds laterally, the mons superiorly, and the anus inferiorly, is essential. After evaluating tissues without magnification and conducting wet mount microscopic evaluation and vaginal secretion cultures to rule out infection, adding the lighted magnification of a vulvoscope/colposcope can facilitate a directed biopsy of specific areas of lichenification, fissures, hyper/hypopigmentation, plaques, or acetowhite epithelium. Histology from biopsies can distinguish between benign and malignant disease and guide the course and duration of treatment [6].

Lichen Sclerosus (LS)

LS is a complex and chronic inflammatory dermatosis that can cause significant pain, pruritus, and sexual dysfunction. LS is a common condition with incidence ranging from 1:300 to 1:1,000 in the general population, and 1 in 60 in specialty gynecological practices [7,8]. LS can occur at any age, although it is rarely found in the first year of life, and typically has a bimodal peak incidence in prepubertal girls (5–15% of cases) and in postmenopausal women (50–60% of cases) [7–10].

Theories of Causality

Multiple controversies exist regarding the etiology of LS. It is postulated that the disease state is multifactorial, and that infectious, trauma, autoimmune, and genetic factors can play a role in causality.

An infective trigger for LS has been suggested based on a correlation with the bacteria, *Borrelia burgdorferi*, in studies from Europe and Asia. This association has not been noted in the United States. Evidence that supports the theory is contradictory and remains controversial [2,7]. The Koebner phenomenon (i.e., development of skin lesions at sites of traumatized or injured skin) has been postulated in the etiology of LS. Consistent with this theory is the clinical finding that precipitating factors to plaque formation can be repetitive trauma (chronic rubbing, scratching, etc.) and irritation in the anogenital skin [7]. LS has been associated with increased circulating autoantibodies. Up to 34% of patients with LS present with coincident autoimmune disease, including thyroiditis, pernicious anemia, vitiligo, and alopecia areata. Studies suggest that LS subjects may have autoantibodies to endothelial cell adhesion molecule, BP 180 and 230, although the specific mechanism of this autoimmunity has recently been called into question [10–15]. Studies of twins and first-degree family members with LS have suggested a genetic predisposition [2,7]. Recently, LS biomarkers have been evaluated through genome expression profiling to identify molecular pathways in LS development. Researchers have identified 99 of 28,869 genes that differentially express in vulvar LS tissue. Among them, 73/99 genes are upregulated and 26/99 are downregulated. Further exploration of genetic links may further elucidate familial associations [15–17].

Symptoms

Women with LS often present with complaints of nonspecific irritation of the vulva, which progressively increases in severity, leading to pain, dyspareunia, and dysuria. Vulvar pruritus, particularly at night, is often the first presenting symptom; however, as many as 1/3 of women with LS are initially asymptomatic [3,7–10]. As the disease progresses, sclerotic changes and scratching can lead to pain and fissures. The introitus may narrow due to scarring and tissue adhesion, and fissures can occur at the posterior fourchette, due to loss of elasticity and trauma from coitus [16]. In women with perianal LS, tissue changes can lead to rectal bleeding and pain with defecation [2]. LS-related changes initially may go unnoticed, be considered a "normal" part of aging, or be misdiagnosed as vulvovaginal atrophy, potentially contributing to a delay in diagnosis [2].

In some instances, LS is identified during routine gynecological examination, but more commonly women are empirically treated or self-treat early in the disease with antifungals and/or topical estrogens, which might produce a palliative effect for irritation. As symptoms persist and worsen, most women seek a specialty consultation, during which a diagnostic biopsy is performed [3]. Biopsy-proven LS can guide directed treatment and appropriate patient education, which can improve functional outcomes and avoid complications.

Physical Examination

On physical examination, LS lesions are typically hypopigmented papules and plaques, with areas of excoriation, purpura, and ecchymosis [17–21]. LS-affected skin is described as resembling "cigarette or parchment paper" because the tissue appears wrinkled, whitened, thinned, shiny, or waxy [3,11]. The distribution of skin changes occur in a "keyhole" or "figure of eight" pattern, involving the clitoris, clitoral prepuce, labia minora, labia majora, interlabial sulci, and perineal body [2,7–11]. Architectural changes include labial resorption, agglutination, phimosis of the clitoris, and introital narrowing [16]. Chronic scratching secondary to pruritus may also result in subepithelial micro-hemorrhages [7]. In contrast to lichen planus (LP), LS does not affect the vaginal mucosa, but instead extends laterally from Hart's line [10].

Histology

Although LS can be suspected clinically, histological confirmation via biopsy should be obtained, as LS can be mistaken for other vulvar skin diseases, including squamous cell carcinoma (SCC) [3,7–11]. Pathological findings characteristic of LS include lichenoid inflammatory infiltrate in the dermis, hyperkeratosis of the epidermis, homogenization of the collagen in the upper dermis, and epidermal atrophy with loss of rete ridges [5,11,12]. It is preferable to obtain a small tissue biopsy using a Keyes punch or Tischler forceps prior to initiating topical treatment because the histological changes caused by LS can resolve post application of corticosteroids [5,10].

Treatment

The goals of treatment for LS are to alleviate a woman's symptoms, prevent disease progression and scarring, provide long-term surveillance, and prevent development vulvar cancer [2]. Treatment of LS should begin after the biopsy site has fully healed [5]. Topical corticosteroids (TCS) play a key role in the treatment of LS as they exert anti-inflammatory, immunosuppressive, and antiproliferative effects on the keratinocytes. The antimitotic effects influence the growth, differentiation, and function of cells, and inhibit cytokine production [11]. Corticosteroid ointments are preferred to creams because they are less irritating to the sensitive vulvar tissue, and they do not contain alcohol or other preservatives [2]. The gold standard for the ultra-potent class of corticosteroid treatment is clobetasol propionate 0.05% ointment, prescribed topically once daily for up to 4 weeks, then every other day for 4 weeks, then twice weekly for maintenance [8,9,22–24]. Prescribing intervals can be varied based on symptoms. To facilitate absorption of topical ointment, the patient should be instructed to soak in warm water for 15 min and pat the skin dry prior to applying the ointment, and then gently rub the topical steroid into affected tissue for 60–90 seconds [5]. Studies report that complete relief of symptoms occurs in up to 70% of LS patients who use TCS [11,16,18].

In addition to the ultra-potent TCS, less potent TCS, such as mometasone furoate and triamcinolone acetonide, have shown efficacy [24–27]. In a recent study of 58 LS patients, subjects were randomized to treatment with 0.1% mometasone furoate or clobetasol propionate 0.05%. After a 12-week treatment program, 24 (89%) patients were responders in each group.

The decrease in mean symptom and sign scores was significant compared with baseline with both treatments. No significant differences were found in efficacy end points or in toleration [26]. In a trial evaluating treatment of LS with triamcinolone 0.1% ointment, statistically significant reductions in mean symptom scores for dyspareunia, vulvar burning, and vulvar pruritus were noted between the initial and 3-month follow-up ($P < 0.05$) [27]. The potential benefit of the use of high but not ultra-potent TCS is minimization of side effects, including contact dermatitis, worsening of cutaneous skin infections, systemic effects, and skin atrophy. Clinicians should consider mometasone or triamcinolone as options for women who cannot tolerate clobetasol, for whom it is not effective or those at risk for side effects [11,23–27].

Alternatives to steroidal therapies, including pimecrolimus, tacrolimus, cutaneous lysate, processed skin cell proteins (PSP), ultrasound, and UV1, have been recently studied. Goldstein and colleagues evaluated the efficacy of pimecrolimus, a calcineurin inhibitor, in the treatment of LS [14]. Pimecrolimus acts as an immunomodulator that blocks the release of inflammatory cytokines from T lymphocytes in the skin and promotes cutaneous innate host defenses without causing skin atrophy. In a study evaluating 12 weeks of treatment in 38 patients with biopsy-proven LS, using either clobetasol or pimecrolimus, both treatment groups demonstrated improvement in pruritus, burning pain, and the Investigator Global Assessment subscales, but clobetasol was found to be superior in improving inflammation [14]. In a similar study, another calcineurin inhibitor, tacrolimus, was evaluated for LS treatment. Funaro and colleagues randomized 55 patients with LS to 3 months of treatment with clobetasol 0.05% or tacrolimus 0.1%. Study results showed clobetasol to be significantly more effective in treating signs and symptoms of LS [17].

Human fibroblast lysate cream (HFLC) inhibits pro-inflammatory cytokines interferon-gamma (IFN-γ), tumor necrosis factor-alpha (TNF-α), and interleukin-1alpha (Il-1α), (which are elevated in LS), and promotes collagen formation and wound healing. HFLC was recently evaluated against placebo in 30 patients with LS. Although there were no adverse effects with the study cream, it did not significantly affect inflammation on pre- and post-treatment biopsies [18]. Platelet-rich plasma (PRP) has been used for the treatment of a variety of medical conditions, ranging from diabetic foot ulcers to tendinopathy. It contains multiple growth factors, including platelet-derived growth factor, transforming growth factor beta, epidermal growth factor, vascular endothelial growth factor, and insulin-like growth factor, which are important modulators of mesenchymal cell recruitment and extracellular matrix syntheses during healing. In a recent non-controlled pilot study in the plastic surgery literature, 15 LS patients with labial and periclitoral vulvar plaques were treated with PRP. All study patients reported disappearance of pain and symptoms, and improved sexual function. Controlled studies of PRP for LS are currently being conducted [19]. Liming and colleagues reported on 41 cases of LS treated with high-intensity focus ultrasound (3–4.7 watts; 9–10 MHz). Thirty-four of 41 patients were completely or partially improved. Fifteen of 41 subjects underwent post-treatment biopsies in which squamous epithelium appeared normal. The inflammatory infiltrate in the dermis was reduced or absent [20]. The *JAMA Dermatology* recently published a novel trial comparing traditional treatment for LS with the use of home-based UV-A1 phototherapy 4×/week × 12 weeks. Although phototherapy resulted in significant clinical improvement, it was inferior to clobetasol in relief of symptoms and quality of life measures [21].

Despite evidence for a plethora of alternative LS treatments, the Cochrane review recommendations suggest first-line treatment for LS remains potent or ultra-potent TCS ointments. The FDA concurs, recommending pimecrolimus or tacrolimus as second-line agents for intermittent treatment in LS patients who are unresponsive to topical steroids [8,11].

LS and Vulvar Cancer

Women with LS have a 3–5% increased risk of developing genital cancer during their lifetime, with a relative risk greater than 260. An estimated 60% of all vulvar SCC arises from an area of tissue affected by LS [25–27]. Neither the duration of, nor the presence of symptoms, nor the degree of vulvar architecture change has been proven to be useful in determining malignant potential of an LS lesion [8–10].

The exact mechanism of carcinogenesis associated with LS remains uncertain. Studies suggest that the origin of LS-specific autoantibodies is the basement membrane, while antigens are associated with the intracellular matrix [8]. Nearly 50% of LS biopsies contain regulatory T cells with a monoclonally rearranged receptor γ-chain

gene. Current theories posit that antigen-driven selection of T cells and restricted T-cell receptor usage may reflect prolonged exposure of the woman's immune system to an LS-specific local antigen. The resulting dysregulation may result in a permissive environment for the development of SCC in LS patients [13].

During LS surveillance, the clinician should note new or suspicious lesions, ulcerations, nodules, or tissue that is unresponsive to treatment, and should rebiopsy to assess for malignancy. In cases of LS-associated SCC, progression to invasion can occur within 6 months, making early detection difficult. It is essential to schedule regular follow-up intervals and discuss with patients the risk of vulvar cancer. Data suggest that during surveillance, controlling inflammation is key. SCC is seen less in patients with early diagnosis and with consistent maintenance TCS treatment [2,8].

LP

LP is a painful, chronic disorder that affects the skin and mucous membranes of the vagina, vulva, conjunctiva, esophagus, urethra, and anus, as well as the scalp and nails. It affects an estimated 1–2% of the U.S. population [28–30]. The vulva and vagina may be affected in isolation or may be part of a widespread generalized skin eruption. Approximately 50% of women with LP have genital involvement [2]. It is important for providers to inquire about gum problems, alopecia, or skin rashes when LP is suspected [30]. LP is often found in perimenopausal and postmenopausal women, although it can present in premenopausal women [2]. The exact etiology is unknown; however, studies suggest that LP is likely an autoimmune disorder involving T cells that attack basal keratinocytes. Like LS, vulvovaginal LP has been associated with other autoimmune disorders, such as thyroid dysfunction [3].

Subtypes of LP

There are three main subtypes of LP that affect vulvovaginal tissue: classical papulosquamous, hypertrophic, and erosive types.

- Classical papulosquamous LP is either asymptomatic or presents as pruritus. Architecturally, Wickham's striae (white, lacy-like lesions within the dermis) are a hallmark sign of cutaneous LP, occurring on the labia minora and/or the clitoral prepuce. Papules can also be present on the labia majora and interlabial sulci. Classical LP seldom results in scarring and may resolve spontaneously [31].
- Hypertrophic LP, the least common form, typically affects the genitals and perianal area. It presents as hypertrophic lesions, which are pruritic and irritating. They can mimic malignancy, presenting as warty plaques with a purple-hued, violaceous edge [31].
- Erosive LP is the most common type of LP. It presents symmetrical erosions, located between the posterior fourchette and the anterior vestibule. Symptoms are often severe and include vulvar burning, pain, dyspareunia, apareunia, dysuria, and irritative serous vaginal discharge [29,31]. Mucosal webbing may be present on the anterior and posterior fourchette [3]. There can be loss of the labia minora, phimosis of the clitoris with midline fusion, contracture of the introitus, urethral, and/or vaginal obstruction [29–31]. In advanced disease, the vulva may lack any landmarks and be "featureless" [30]. Erosive LP can also affect the vagina. Internal synechiae and erosions can cause scarring, obstruction, dyspareunia, and contact bleeding [30]. Twenty-five percent of patients complain of irritating serous or purulent vaginal discharge with vaginal involvement [29]. Vaginal stenosis with loss of vaginal length may also be evident on exam, but may go unnoticed by women who are not attempting intercourse or tampon use [2].

Diagnosis and Biopsy Considerations

The diagnosis of LP is based on clinical findings in the examination (particularly if Wickham striae are present) and confirmation with microscopy and directed biopsy. Elevated pH, increased white blood cells, and parabasal cells can be evident on wet mount [2,3]. During vulvar biopsy, it is helpful to include the edge of erosion with adjacent, nonaffected tissue to aide in histological confirmation. Indicating the differential diagnosis on the specimen will assist the pathologist in identification. A biopsy of the vagina is generally not necessary, unless vaginal erosions are present and there are no vulvar findings [2]. The histological features of LP include hyperkeratosis and acanthosis with a "saw tooth" appearance of the basal layer. A band-like upper dermal infiltrate of lymphocytes can also be seen close to the basement membrane [31]. Approximately 1–2% of patients with erosive LP develop malignancy. As with LS, long-term

surveillance is imperative and treatment-resistant lesions require rebiopsy [26,29].

Long-term treatment of vulvovaginal LP is challenging but crucial. Management is aimed at relieving symptoms, preventing disease progression, and distortion of vulvar architecture [3,32]. First-line treatment consists of topical application of ultra-potent corticosteroids. While the scarring associated with vulvovaginal LP is irreversible, the use of clobetasol propionate 0.05% ointment can improve or resolve the symptoms in more than 90% of patients [29]. Daily corticosteroid therapy is used until all evidence of active lesions resolve, after which it is tapered to maintenance dose [5]. For internal vaginal synechiae and erosions, adjunctive use of a 25–100 mg hydrocortisone cream or suppository can be inserted daily, tapered to three times a week (TIW), and then weekly. This often results in dramatic improvement in vaginal symptoms [29]. To maintain vaginal patency, insertion of a vaginal dilator to which a small amount of corticosteroid ointment has been applied can also be used [5]. Maintenance of the dilator program and/or regular intercourse is necessary to prevent vaginal stenosis and permanent changes. With chronic use of intravaginal steroids, a maintenance dose of weekly fluconazole can be useful for prophylaxis against fungal infections [3]. In treatment-resistant cases, some authors suggest systemic oral prednisone or intramuscular triamcinolone (1 mg/kg) [30,31]. Topical calcineurin inhibitors (i.e., pimecrolimus and tacrolimus) are considered second-line treatment for LP and are used in cases when topical steroid treatment is not tolerated or fails [32,33]. Recently, Bradford and Fischer reported on long-term management (mean 6.4 years.) of 113 women with LP. In most patients, remission of symptoms was achieved in a mean of 7.5 weeks using topical ultra-potent corticosteroids alone, although 53/131 (40%) of patients used oral prednisolone as adjunctive therapy. Forty-five patients (34.3%) used a maintenance program of topical tacrolimus alternating with TCS, while 11 patients (8.5%) required low-dose weekly methotrexate. The authors concluded that long-term symptom control requires multimodal, flexible treatment programs with judicious use of oral agents [34].

Surgical Interventions

Surgery for labial fusion, vaginal scarring, and clitoral phimosis resulting from LS or LP can be performed for comfort and for restoration of urinary and sexual function. Surgery is most successful if done selectively, for patients in whom topical agents have failed and when active disease is otherwise well controlled. In a case series of 35 patients, 27 with LS and 8 with LS, researchers reported favorable results from simple perineotomy procedures (lysis of vulvar adhesions by dissection without suturing) performed for the treatment of dermatoses-associated labial fusion. At 3 months, 31 of 35 patients had no refusion. Six cases of late refusion were reported at 2-year follow-up, some of which were related to noncompliance with maintenance corticosteroid therapy [35]. Another study reported surgical treatment of eight patients with LS-associated clitoral phimosis. A lacrimal duct probe was used to lyse adhesions and the prepuce was incised in the midline. The study noted a high level of patient satisfaction, improved sensation, and sexual function. Authors stressed the importance of medical suppressive therapy postoperatively [36].

Once thought futile due to the high level of adhesion recurrence, recent studies of surgical intervention for LS and LP demonstrate a high level of sustained tissue improvement and patient satisfaction, although some studies report persistent postoperative sexual dysfunction [37].

Lichen Simplex Chronicus (LSC)

LSC is one of the most frequent causes of vulvar pruritus and is presumed to be the most common vulvar dermatoses, although the actual incidence of LSC is unknown [2]. Any pruritic vulvar skin disorder that produces scratching and chronic rubbing can lead to lichenification of vulvar tissue and can be classified as LSC [1,33]. Rather than being a distinct entity, LSC refers to any chronic, localized form of dermatitis, and by many authorities it is used synonymously with the terms *neurodermatitis*, *atopic vulvar dermatitis*, and *vulvar eczema* [1,38].

Primary vs. Secondary Subtypes

Primary LSC commonly presents in patients with a history of atopic diathesis. Atopic patients are more likely to experience itch secondary to irritation and have a personal or family history of allergic conjunctivitis, hay fever, asthma, or allergic rhinitis [1]. The inciting event for pruritus can be as minor as light touch, tight clothing, or overzealous vulvar hygiene [1]. A unique characteristic of primary LSC is that rubbing or scratching of the skin is often highly satisfying and may be

pleasurable, rather than associated with burning or pain. This can result in a self-perpetuating scratch–itch cycle, which defines LSC, and causes thickened epidermis, damage in the protective barrier of the skin, and increased susceptibility to irritants and superimposed infections [2,38]. Pruritus ranges from mild to intense, most often occurring in the evening or night. Although patients describe being able to control the itching during the day, they may be unaware of scratching while asleep and may waken with tissue beneath their fingernails [2]. In secondary LSC, patients often report that initial pruritus began with a yeast infection or during a period of stress; however, the symptoms continued upon resolution of the inciting event. Other exacerbating factors include perspiration, heat, menses, stress, friction from clothing or sanitary pads, and use of topical medications [1,34].

Physical Examination and Diagnosis

On physical exam, the vulvar skin appears thickened with the skin creases accentuated. The tissue may be pale and lichenified. Labial erythema and swelling may be noted. Darkened areas of hyperpigmentation are related to inflammation, while areas of hypopigmentation can occur due to damage to melanocytes. Erythema, ulcerations, erosions, crusting, excoriation, or fissures from scratching are often seen. Hair may be broken or absent secondary to trauma [2,39]. LSC most commonly affects the hair-bearing portion of the labia majora, but may affect the entire vulva or perianal tissue. It can be localized, bilateral, or unilateral [2,33].

During history taking, LSC can often be distinguished from other conditions causing vulvar pruritus by the characteristic report by patients that they derive intense pleasure from scratching [1]. Upon exam, biopsy confirms the diagnosis and excludes LS, LP, or neoplasia [5]. Histological examination reveals hyperkeratosis, parakeratosis, acanthosis, spongiosis, lengthened rete ridges, a prominent granular layer, and chronic inflammatory infiltrate [33].

Treatment

Treatment of LSC is aimed at disrupting the itch–scratch cycle. Treatment comprised combination therapy, including a high- or ultra-potent TCS, topical anesthetics, antihistamines, nighttime sedation with low-dose tricyclics, and as needed antifungal and antibacterial medications to treat underlying infections [1]. If left untreated, LSC can persist indefinitely, although symptoms may wax and wane [39]. Clobetasol propionate ointment 0.05% applied daily for 3–4 weeks will decrease inflammation and break the itch–scratch cycle. Steroids can then be tapered and/or changed to lower potency ointments for maintenance care two to three times a week [2,33]. Nighttime scratching can be addressed with the application of ice packs, instructing the patient to cut fingernails and wear cotton gloves at bedtime (HS) [1,5]. Topical pimecrolimus or tacrolimus can be used as adjuvants to decrease inflammation [38].

Contact Dermatitis

Allergic and irritant dermatitis are common causes of vulvar pruritus and account for up to 50% vulvovaginal skin symptoms [34]. Contact dermatitis may result from either exposure to an allergen that initiates an immune response or to an irritant that directly damages vulvar tissue. The response may be immediate, as is the case in irritant dermatitis, or delayed if due to an allergic response. Dermatitis can be acute, subacute, or chronic [37,40].

The vulva is particularly susceptible to dermatitis as the tissue may have intrinsic factors that alter its barrier to irritants. Estrogen deficiency, moisture from urine or vaginal discharge, stool residual enzymes, friction, and heat impede the barrier function of the epithelium [40]. Hygiene and hair removal practices, constricting clothing, menstruation, piercings, heat, and perspiration can contribute to vulvar irritation and delayed healing [3,41].

Subtypes of Dermatitis

Irritant contact dermatitis (ICD) results from exposure to an irritant that affects that skin by removing surface lipids, denaturing keratin, and damaging cell membranes. ICD typically occurs without any prior sensitization to the irritant [40]. Patients with ICD present with complaints of rawness, burning, stinging, or pain after contact with a substance. Itching is not typically a primary symptom [42,43]. On physical exam, erythematous plaques and patches with scale and excoriations may be present. In severe cases, vesicles, edema, erosions, and ulcerations may be noted. In cases where friction has occurred against the irritated dermis, an area may be well demarcated or symmetric (i.e., after use of irritating sanitary pads) [43]. Urine, perspiration, fecal matter, topical medications, and feminine hygiene products

frequently cause dramatic irritant contact reactions. Caustic ingredients in absorbent products include colophony, rosin, and methyldibromo glutaronitrile, and are associated with severe dermatitis, erosions, and dysuria [40,41,43].

The second subtype of dermatitis is allergic contact dermatitis (ACD). This occurs after previous sensitization to an allergen, resulting in a delayed hypersensitivity reaction [40,41,43]. The allergen may be difficult to identify due to a delay in onset of symptoms (typically 48–72 hours). In acute ACD, vulvar manifestations appear as vesiculobullous eruptions with severe pain and/or pruritus. Edema, coalescing vesicles, ulcerations, lichenified plaques, hyperpigmentation, and excoriation are often seen in chronic ACD [43–45].

The most frequently recognized allergens in vulvar ACD include topical antibiotics, corticosteroids, antiseptics, preservatives, and topical anesthetics (i.e., benzocaine, found in over-the-counter "anti-itch" products) [41]. Other common allergens include thiuram in condoms, chlorhexidine in feminine hygiene sprays, and 4-phenylene diamine in black-dyed underclothing [41,43]. Recently, "moist wipes" have gained widespread popularity as a form of vulvar hygiene. Most contain the allergen, methylchloroisothiazolinone/methylisothiazolinone (MCI/MI), found in cosmetic and industrial products [41,44].

Diagnosis

Dermatitis, whether irritant or allergic, is a clinical diagnosis based on detailed history taking. Overzealous cleansing is often a hallmark characteristic of patients with ICD and ACD [40]. Questions about new perfumes, douches, contraceptives, undergarments, topical medications, lubricants, cleansing agents, toilet paper, or detergents are key [41]. Evaluation should include a thorough examination of the skin, microscopy with pH testing, sexually transmitted infection testing, and vaginal cultures. When necessary, a biopsy can be performed. It often confirms eosinophilic infiltrate, consistent with allergic skin response [3]. Referral to a dermatologist for patch testing may also be warranted [5].

Treatment for contact dermatitis involves eradicating the offending agent or altering inappropriate hygiene practices [33]. A short course of low potency steroid ointment, oral or topical antihistamines, and/or HS low-dose tricyclic antidepressants can be used to ameliorate symptoms. The patient should be instructed on hygiene and comfort regimens, including cleansing the vulva with water only, eliminating soaps, detergents, washcloths, and douches, applying over-the-counter, unscented skin emollients, using cold compresses, and taking tepid sitz baths [4,38]. For unremitting cases, a limited course of clobetasol 0.05% ointment and/or prednisone 40–60 mg daily, tapered over 14–21 days, may be necessary to provide symptom relief [40,41,43].

Vulvar Psoriasis

Psoriasis is a chronic inflammatory proliferative skin condition affecting 4–8% of adult women. It accounts for an estimated 5% of all female patients who present with persistent vulvar discomfort. The diagnosis is often confused with lichenoid dermatoses, vulvovaginal candidiasis, or contact dermatitis [46].

Diagnosis

Women with vulvar psoriasis typically present with pruritus and burning that are bilateral and symmetrical. Vulvar tissue can appear erythematous, scaly or non-scaly with macular eruptions, and raised plaques on the labia, perineum, perianal area, and/or mons pubis. In up to 65% of patients, evidence of psoriasis is found on other parts of the body [46,47]. Skin changes can have a significant effect on the psychosexual well-being of patients, with up to 45% of patients complaining of daily pain and nearly 30% reporting dyspareunia [46]. The pathophysiology of psoriasis involves alteration in the activation of CD4+ and CD8+ T cells with anomalous proliferation of keratinocytes. The mechanism that causes the disease to present in particular zones of the body is poorly understood, although local mechanical and/or chemical irritation likely perpetuates the process [48,49].

Psoriasis begins as a clinical diagnosis. As with other chronic vulvar conditions, it is important to biopsy the affected tissue to rule out other benign or malignant conditions. Since biopsies may not demonstrate the histopathological changes characteristic of psoriasis, for example, interstitial psoriasiform spongiotic changes, cooperation between the examining clinician and pathologist may be necessary to arrive at appropriate diagnoses [46]. Fungal or bacterial coinfections are common with psoriasis, and vaginal secretions should be evaluated regularly, followed by

species-specific antifungal or antibacterial therapy [47–49].

Treatment

The mainstay of psoriasis treatment is initial use of topical high-potency TCS ointment, followed by maintenance therapy with less potent steroids and psoriasis-specific treatments, such as tar cream and calcipotriol. In a clinical audit of 194 cases of vulvar psoriasis, Kapila and colleagues report using this approach to achieve suppression of chronic psoriasis in 93.8% of patients, with a mean follow-up of 8.9 months [47]. Other authors note that systemic therapy with methotrexate can be effective, but is most often reserved for resistant severe cases [47,49]. One case report by Guglielmetti et al. documents a case of psoriasis that was resistant to traditional therapy, but improved after treatment with the anti-infective dapsone 100 mg daily × 10 months, followed by maintenance therapy with tacrolimus and calcipotriol [50].

Vulvar Skin Disorders and Female Sexual Dysfunction

Research suggests that approximately one half of women with chronic vulvovaginal symptoms experience sexual dysfunction, representing an incidence that is double that of the general population [51,52]. Studies of women with LS suggest that the disorder causes alterations in every domain of sexual functioning and is associated with significant sexual distress. In addition, women with LS have less frequent and less satisfying sexual activity than controls [53,54]. Even after effective medical and surgical treatment for LS and LP, many women continue to struggle with significant sexual dysfunction and have high levels of sexual distress [37,55]. In studies of women with LSC, chronic pruritus is related to alterations in sexual desire, arousal, lubrication, orgasm, and satisfaction (but not pain), and in women with vulvar psoriasis chronic pruritus and burning are related to dyspareunia [47,56]. Studies of women who have been treated for in situ dermatoses-related vulvar cancer suggest that survivors continue to have desire for and resume sexual activity after treatment but have lower Female Sexual Function Index scores for arousal and orgasm [52].

It is important for specialists in sexual medicine to give guidance to women who wish to be physically intimate while dealing with chronic symptoms. Specific behavioral suggestions include encouraging patients to "plan ahead" and discuss with partners possible non-penetrative sexplay options to replace coitus during flares; limiting the duration of sexplay and choosing positions that minimize friction to affected tissues, such as have one partner stand while the other partner lies supine, or rear-entry positioning with partners kneeling; applying emollient skin barriers (i.e., petrolatum, oil- and/or wax-based moisturizers) to excoriated or fissured skin before sexplay to protect against body fluids, spermicides, and lubricants; applying cool compresses or ice packs to the vulva before and after intimacy to decrease pruritus; using external vibrator stimulation to areas of clitoral phimosis to enhance arousal and orgasm; regularly using intravaginal dilators to maintain patency of the introitus and vagina, and using sexual aides, such as the Come Close ring or a custom penis sleeve to facilitate partial depth thrusting when vaginal vault length is diminished [1,2].

Conclusion

Vulvar skin conditions are a common source of chronic genital discomfort in women. Chronic vulvar dermatoses can significantly alter a woman's quality of life and negatively impact sexual comfort and function. It is essential for sexual medicine specialists to have a high level of familiarity with the classifications of vulvar disorders, to conduct detailed histories and systematic examinations. While the diagnosis of vulvar skin changes may seem intuitive, assumptions that are based on "pattern recognition" alone can be unreliable and lead to inappropriate treatment [5,6]. Vulvar biopsy, in conjunction with dermatopathology consultation, remains the gold standard for definitive diagnosis, which in turn informs competent treatment [37]. In addition, provision of verbal and written education regarding diagnosis, treatment, and strategies for managing physical intimacy can help the patient become a partner in her treatment program, can ease her fears, and contribute to her long-term vulvar and sexual wellness.

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