REVIEWS

The Vulvar Dermatoses

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ABSTRACT-

Introduction. Dermatologic diseases of the vulva may cause dyspareunia. These disorders may be overlooked by gynecologists and urologists because of lack of residency training experience. Dermatologists who are most familiar with these diseases are infrequently trained in vulvovaginal examination. As such, these disorders are often improperly diagnosed and treated.

Aim. To describe the presentation and management of the major vulvar dermatoses including irritant and allergic contact dermatitis, lichen sclerosus, lichen simplex chronicus, and lichen planus.

Main Outcome Measure. Data from a peer review literature search on the topic of vulvar dermatoses.

Methods. The literature for this review article was obtained through a Medline search. Appropriate dermatology textbooks were utilized for additional information.

Results. A comprehensive survey of the vulvar dermatoses.

Conclusion. Vulvar dermatoses must be considered a part of the differential diagnosis of any woman with a sexual pain disorder. As such, healthcare providers who evaluate and treat women with dyspareunia must become familiar with the most common dermatologic disorders of the vulva. **Burrows LJ, Shaw HA, and Goldstein AT. The vulvar dermatoses. J Sex Med 2008;5:276–283.**

Key Words. Vulvar Dermatoses; International Society for the Study of Vulvovaginal Disease, Dyspareunia

Introduction

A lthough the vulva is the most visible female genital structure, it has received the least attention in the medical literature and has even been referred to as "the forgotten pelvic organ" [1]. There are many ways in which the keratinized skin and mucocutaneous surfaces of the vulva differs from skin on the rest of the body: it is the only area of the human body where epithelium from all three embryologic layers coalesce. In addition, because the vulvovaginal tract contains foreign proteins and antigens necessary for reproduction, this area of the body has a unique immunologic response [2]. Lastly, the subcutaneous tissue of the labia majora is looser, allowing for considerable edema to form [3].

Vulvar dermatoses may present in a variety of ways, ranging from asymptomatic to chronic dis-

abling conditions, which are often difficult to treat and severely impact a woman's quality of life. All of these conditions may present with dyspareunia. Other associated symptoms may include pain, itching, fissuring, and bleeding after intercourse. The differential diagnosis includes vulvar intraepithelial neoplasia, a vulvar malignancy, vulvar Crohn's, vulvar ulcerations because of a sexually transmitted disease, plasma cell vulvitis (a very rare disorder), and other dermatoses that may be found elsewhere on the body such as psoriasis. Patients may be embarrassed by the disfiguring changes that may occur and avoid sexual intimacy. Because of their personal nature, patients are frequently reticent to discuss their symptoms with a healthcare provider. Additionally, many clinicians feel challenged with respect to the management of vulvar disease. These factors may result in women receiving suboptimal treatment, resulting in

Table 1Genital care for women

What is it?

Genital care means the way in which women keep their genital area healthy. This part of the body (the vulva)* is made up of skin, moist areas, and glands. Secretions (moistness) from the vagina keep it clean and healthy and these secretions are normal. These secretions protect the vagina and the skin.

Are there any problems with washing the genitals?

Yes. The skin and moist surfaces of this part of the body are very delicate. It is important not to wash with harsh chemicals that may irritate the area. Washing too often, or rubbing too hard when drying, can irritate this skin. If you have problems in this area, washing with plain, lukewarm (not hot) water is best. Using soap, shower gels, and some cleansers can make the problems worse. Your healthcare provider may be able to suggest a soap substitute.

What is the best way of keeping myself "clean"?

Gently separate the outer "lips" and bathe the inner skin with plain water, using your hands only. Gently pat dry the outer skin. Do not use a hair dryer.

What about clothing?

Wear well-fitting clothing and avoid thongs, girdles, tight jeans, and hose. Wash underclothes in a mild detergent and avoid fabric softeners.

What is best to use for my period?

Disposable menstrual pads and tampons can be used. The best ones are natural cotton or hypoallergenic products. Remember to use ones that fit properly and change them regularly.

What else should I know?

It is not necessary to wash the vulva every day and it should not be washed more than once a day. Do not wash the vagina. Do not use wipes, deodorants, douches, or other cosmetic and cleansing products. Women with a problem in this area should use only treatments prescribed by their healthcare provider.

*You can find more information about the vulva in the information leaflet 'The Normal Vulva' on this website. International Society for the study of Vulvovaginal Disease Patient Information Committee, June 2006.

persistent symptoms and/or avoidance of sexual relationships altogether.

The International for the Study of Vulvovaginal Diseases recently devised a new classification system for vulvar dermatoses (Table 1) [4]. The purpose of this review is to discuss the diagnosis and treatment of the more common vulvar dermatologic conditions which may be associated with dyspareunia.

The History, Physical Examination, and Vulvar Hygiene

All patients with compliants of dyspareunia should undergo and comprehensive history and physical examination. Additionally, any patient with a vulvar complaint should be instructed on routine vulvar care measures including using all cotton underwear washed in hypoallergenic soap (no thong underwear), avoidance of allergens and irritants (soaps, perfumes, and feminine hygiene products such as Vagisil and douches), use of unscented undyed menstrual pads and tampons, and washing with water after urination.

History

To accurately diagnose any vulvar condition, a thorough history must be elucidated. Details regarding the onset and duration of symptoms, (including any possible systemic symptoms), identifying what makes the patient feel better or worse, current sexual practices, a complete sexual history (including any sexually transmitted diseases), and a list of all and previous medications, particularly topical and over the counter applications are essential. Exercise regimens should be reviewed as these can impact vulvar health and sexual function [5]. The clinician should also elicit any history of genital surgery, including labiaplasty, a procedure being performed with increasing frequency [6]. A complete medical history and any relevant family history may be helpful as well.

Physical Examination

Examination of the vulva must be thorough. We recommend performing the examination in the same way for each patient so as to not miss anything. Proper lighting is crucial and some patients prefer to hold a mirror to observe the examination and help communicate where their symptoms are located. We recommend vulvoscopy after the application of 3% acetic acid for most patients at the initial visit. Magnification allows the clinician to see the disease process in greater detail, aiding an accurate diagnosis and ruling out a malignant or premalignant condition. The authors find that photographs at the initial visit are especially useful for documenting baseline physical examination findings as well as progress with treatment.

The vulvar examination should begin with the patient in the dorsal lithotomy position. Specifically, the vulva should be thoroughly examined for atrophy, tenderness, erythema, induration, fissures, lichenification, ulceration, erosions, hypopigmentation, scarring, phimosis of the clitoris, and narrowing of the introitus. The vulva and vulvar vestibule should then be carefully palpated with a cotton swab. The swab can help elicit decreased sensitivity of the vulva and clitoris that may be evidence of vulvar vestibulitis syndrome or atrophic vestibulitis. A speculum exam should be performed to look for ulcerations or synechiae in the vagina which can be a sign of erosive lichen planus (LP). In addition, the vagina should be examined for a loss of vaginal rugae, vagina pallor, and petechiae which are evidence of atrophic vaginitis. While the speculum is in the vagina, a sample of the vagina discharge should be obtained for pH, wet mount, and culture as vulvovaginitis (candidiasis and bacterial vaginosis) are frequent comorbid conditions of the vulvar dermatoses.

The authors strongly recommend vulvar biopsy whenever abnormalities are seen on vulvoscopy. A 5-mm punch biopsy should be performed using sterile technique and the biopsy site should be closed with one or two stitches of absorbable suture such as *Vicryl-Rapide*. A biopsy is extremely useful in differentiating many dermatologic disorders which can present in similar ways. Immunofluorescence is required to diagnosis the immunobullous diseases, but these are rare. Working with an experienced dermatopathologist can help to ensure the accuracy of biopsy results.

A general examination of the skin, eyes, and mouth (including gingival margins) should be performed paying particular attention to the presence of ulcers or erosions at sites other than the vulva as some diseases may exist elsewhere on the body.

General Treatment Guidelines and Vulvar Care

Patient education and open communication are critical in treating vulvar disease. Patients should feel that the clinician understands the stress, anxiety, and physical discomfort they feel. The disease(s) they have should be explained to them and a plan for treatment should be reviewed. Written information describing the disease, the symptoms, and treatment not only help to validate the patient's feelings and concerns, but are also very educational. An important part of the initial discussion with a new patient is to define realistic expectations for treatment. Many patients will not be cured of their disease, but will achieve symptomatic relief. Thus, many conditions will be managed for a lifetime.

Additionally, any patient with a vulvar complaint should be instructed on routine vulvar care and hygiene, as detailed above. The International for the Study of Vulvovaginal Diseases website (http://www.ISSVD.org) has an excellent handout on genital care.

Irritant and Allergic Contact Dermatitis

Contact dermatitis is an inflammation of the skin because of an external agent acting as an irritant or allergen (Figure 1). An irritant is any substance that causes inflammation, erythema, and induration upon exposure to the dermis, whereas an allergen is any substance that stimulates a type IV delayed hypersenstitivity reaction in sensitized individuals [7]. The incidence of vulvar contact dermatitis in the general population is unknown; however, the reported incidence in a vulvar clinic in the United Kingdom was 20-30%, and in an Australian vulvar clinic, the incidence was 15% [8,9]. It is generally accepted, however, among specialists in vulvar disease that as women increasingly apply products to the vulva, the incidence of contact dermatitis is increasing. Common products containing chemicals that may cause contact dermatitis include soaps, menstrual pads, panty liners, toilet paper, diapers, fabric detergents, fabric softeners, feminine sprays, cosmetics, spermicides, pessaries, and condoms. Additionally, medications that frequently cause contact dermatitis include benzocaine, hormonal creams, corticosteroids, topical antifungals, and antibiotics [7].

Women with contact dermatitis present with burning, itching, dyspareunia, and fissuring around the introitus [8]. Physical examination findings may range from mild erythema to weeping, lesions. Histologically, findings are nonspecific. One may see spongiosis, acanthosis, parakeratosis, and a dermal inflammatory infiltrate. The lax tissues of the vulva predispose to marked dermal edema. Continued exposure to the irritant and chronic rubbing or scratching may eventually lead to lichen simplex chronicus (LSC).

The diagnosis of contact dermatitis of the vulva is made by taking a detailed history and careful physical examination. One should have a low threshold to perform a biopsy and rule out coexisting conditions. The differential diagnosis includes candidiasis, psoriasis, sebhorreic dermatitis, irritant contact dermatitis, LSC, and extensive extra mammary Paget's disease [7]. Patch testing may be helpful in making the diagnosis.

The cornerstone of treatment of contact dermatitis is identification and removal of the causative irritant or allergen. Details of the patient's daily routine for hygiene (the use of soaps, deter-



Figure 1 Contact dermatitis is an inflammation of the skin because of an external agent acting as an irritant or allergen.

gents, douches, antifungal treatments, cleansing cloths, sprays, creams, and lotions) should be discussed as well as their practices around the time of menses and intercourse. Proper vulvar care should be reviewed with these patients (and all patients with any vulvar disorder).

Topical steroids are used to decrease inflammation (triamcinolone 0.1% ointment twice a day for moderate cases and clobetasol 0.05% ointment once a day may be prescribed for severe cases). Ice packs (or bags of frozens peas) and antihistamines such as hydroxyzine can be used to help alleviate pruritus. Low-dose tricyclic antidepressants such as amitriptyline can be used to help women stop scratching in their sleep. All patients should be instructed in proper vulvar hygiene.

The duration of treatment and any changes to a treatment regimen should be guided by the patient's symptoms. Patients should be seen 1 month after initiating treatment. Topical steroids and tricyclic antidepressants should be tapered after the patient has been symptom-free or has had tolerable symptoms for a few months and feels comfortable discontinuing the medication. Superimposed candidal infections should be treated with fluconazole (the duration of treatment and choice of antifungal may vary depending on the species) and bacterial infections (such as staphylococcal) should be treated with an oral antibiotic. For patients who report minimal or no improvement, the diagnosis should be reevaluated and the clinician should consider other or additional etiologies for the patient's symptoms.

LSC

Lichen simplex chronicus (Figure 2) of the vulva is an eczematous disorder characterized by itching, scratching, and lichenification [10]. The condition is the end stage of an itch-scratch-itch cycle and is also known as neurodermatitis, pruritus vulvae, squamous hyperplasia, and hyperplastic dystrophy. The etiology of the initiating pruritus that leads to LSC includes numerous irritative and infectious disorders including candidiasis, atopic dermatitis, contact dermatitis, and eczema [2]. The intense, chronic itching caused by these conditions leads the patient to repetitively rub and scratch the



Figure 2 Lichen simplex chronicus of the vulva is an eczematous disorder characterized by itching, scratching, and lichenification.

affected area. The skin responds by thickening and developing a coarse texture, with increased skin markings called lichenification. The skin may also have variable pigmentation and feel leathery. The skin may appear edematous and excoriated with or without fissuring and the pubic hair may be broken or absent. Excoriations may have superimposed infection with yeast or bacteria. Because the vulva is moist, crusts and scales commonly seen with dermatitis elsewhere on the body may not be seen on the vulva. Histopathological examination shows hyperkeratosis, spongiosis, acanthosis, and a chronic dermal inflammatory infiltrate [11].

Women with this condition are frequently "aware of" their vulva and may feel self-conscious with respect to physical intimacy. As numerous vulvar disorders may present with pruritis, a biopsy should be performed to distinguish LSC from lichen sclerosus (LS), LP, or neoplasia.

Treatment consists of patient education to break the itch-scratch-itch cycle. As in irritant and allergic contact dermatits, it is essential to eliminate all irritant/allergen exposure. Second, it is necessary for women to stop scratching. Often this can be difficult as many women only scratch in their sleep. A combination of oral amitriptyline (10–25 mg) at bedtime combined with application of ice very successfully relieves nighttime pruritus. Lastly, topical application of mid to high potency corticosteroids or topical calcineurin inhibitors are used to decrease the underlying inflammation [12]. The authors have found it useful if women soak in warm water for 15 minutes prior to the application of the corticosteroids as this softens the lichenified skin and allows better penetration of the topical corticosteroid ointment. Concomitant infections may be managed with oral antibiotics and/or fluconazole. The long-term management of these patients is similar to those with contact dermatitis.

LS

Lichen sclerosus is a chronic, lymphocyte mediated cutaneous disorder affecting approximately one in seventy women [13]. There is a bimodal peaked incidence in premenarchal girls and in menopause women with the average age of diagnosis being 51 years of age [14]. Extra genital lesions may occur in 11% of female patients [15]. While the etiology of LS has not been completely elucidated, it is most likely that LS is an autoimmune disorder as it is highly associated with other auto-immune disorders including autoimmune thyroid disease, alopecia areata, viteligo, pernicious anemia, and LP. In addition, there are high levels of circulating autoantibodies in patients with LS [16–18]. Women with LS have a 4–6% risk of developing vulvar carcinoma [13,15]. LS has been found in greater than 60% of cases of squamous carcinoma of the vulva [19].

Clinically, while some patients are asymptomatic, most give a history of pruritis or pain [14]. One study that focused on the impact that LS has on a women's sexual satisfaction showed that women with LS were found to be less likely to be sexually active (vaginal intercourse, oral intercourse, and masturbation) than control groups. Furthermore, 79% of women with LS reported chronic vulvar pain [20].

On physical examination, one may see white atrophic plaques ("cigarette paper"), depigmentation, submucosal hemorrhage and because of the chronic inflammation associated with this condition, scarring with narrowing of the introitus and distortion of the vulvar architecture (Figure 3). With time, scarring and decreased elasticity of the skin may predispose to fissuring of the posterior fourchette. LS may involve the labia minora and inner portion of the labia majora, interlabial sulcus, clitoris, and the perianal region, but almost never involves the vagina. Sometimes, scar tissue forms between the clitoral prepuce and the glans clitoris leading to "phimosis" of the clitoris [21]. A biopsy specimen should be obtained to confirm the diagnosis as the histopathologic changes of LS are distinctive and make biopsy a very useful diagnostic tool. Characteristic pathologic finding include hyperkeratosis of the epidermis, epidermal atrophy with loss of rete ridges, homogenization of the collagen in the upper dermis, and a lichenoid (band-like) inflammatory infiltrate in the dermis. In addition, it is essential to obtain the biopsy prior to starting corticosteroids as the pathonumonic changes described above can resolve with the application of corticosteroids [22].

After histologic confirmation of the diagnosis and the biopsy site has healed, the mainstay of treatment is ultrapotent topical corticosteroid ointment, such as clobetasol propionate ointment applied daily until all active disease has resolved [23]. To facilitate the absorption, patients may be instructed to soak in warm water for 15 minutes and pat the skin dry before applying the medication. Patients should be seen 2–3 month after initiating therapy to confirm improvement. Areas of ulceration that do not resolve after appropriate treatment with corticosteroids must be biopsied to rule out vulvar intraepithelial neoplasia or carci-



Figure 3 On physical examination, one may see white atrophic plaques ("cigarette paper"), depigmentation, submucosal hemorrhage, and because of the chronic inflammation associated with this condition, scarring with narrowing of the introitus and distortion of the vulvar architecture.

noma. Once improvement has been demonstrated, the frequency may be tapered down to once or twice per week. It is important to counsel patients that this is a chronic disease and that treatment only when symptomatic is not sufficient as there can be active disease without symptoms. Thus, patients require lifetime therapy for this disorder. Frequently, they can be managed with a onceweekly application of clobetasol and should have an annual physical examination.

LP

Lichen planus is an inflammatory, autoimmune, mucocutaneous disorder, with multiple clinical variants that may involve both keratinized skin and mucosal surfaces. There are three clinical variants of vulvar LP. The most common form, erosive LP, affects the vulva and vagina. Papulosquamous LP affects the vulva and hypertrophic LP involves the perineum and perianal area. Lichen planus affects approximately 1% of all women, and the most common site of involvement is the oral mucosa [24]. Approximately 25% of women with oral LP also have vulvovaginal involvement [25].

Erosive LP is characterized by glassy, brightly erythrematous erosions associated with white striae (Wickham's striae) [26]. The disease may involve the labia minora and vestibule while sparing the vulva, or may be associated with loss of the labia minora, narrowing of the introitus, and obliteration of the vagina (Figure 4). Vaginal involvement has been reported in up to 70% of patients with erosive LP [27,28]. Patients frequently have copious vellow discharge composed of lymphocytes and parabasal cells (immature epithelial cells of the vagina) [26]. In severe cases, intravaginal synechiae may form, causing partial or complete obliteration the vagina. Vulvovaginal-gingival syndrome (plurimucosal LP) encompasses the triad of erosive (desquamative) vulvitis, vaginitis, and gingivitis.

Histologically, erosive LP is characterized by hyperkeratosis in areas of keratinized skin, irregular



Figure 4 Erosive lichen planus may involve the labia minora and vestibule while sparing the vulva, or may be associated with loss of the labia minora, narrowing of the introitus, and obliteration of the vagina.

acanthosis with a sawtooth appearance of rete ridges, a prominent granular layer, and basil cell liquefaction. Apoptotic eosinophilic basal and prickle cells (colloid bodies) are sometimes present, as is a band-like dermal infiltrate composed primarily of T cells [29]. It is important to note that there are no pathonemonic histologic features of LP. To distinguish erosive LP from immunobullous diseases (mucous membrane pemphigoid, pemphigus vulgaris, and linear IgA bullous disease), a biopsy taken from normal tissue at the edge of an erosion should be sent for direct immunofluorescence. A positive result excludes the diagnosis of LP.

Papulosquamous LP may present as small, intensely pruritic, violacious papules arising on keratinized skin. Usually the papules on the vulva and perianal skin are poorly demarcated, pink, and opaque [26]. Papulosquamous LP may be confused with genital warts or molluscum contagiosum and can be differentiated from these two diseases by histologic examination. Hypertrophic LP presents as hyperkeratotic, rough lesions involving the perineum and perianal area. They may resemble squamous cell carcinoma vulvar intraepithelial neoplasia, or LS [26].

Women with LP of the vulvovaginal region may present with itching, burning, vulvovaginal discomfort, dyspareunia, postcoital bleeding, vaginal discharge, and destruction of the vulvovaginal architecture. On physical examination, the vulvar skin and vaginal mucosa are friable, and bleed easily upon insertion of a speculum. In severe cases, there is narrowing or obliteration of the vaginal canal.

The differential diagnosis of LP includes other inflammatory vulvar dermatoses, lichenoid drug reactions, and erythema multiforme. It is commonly misdiagnosed as LS. However, the lesions of LP do not exhibit the classic "cigarette paper" appearance of LS. Furthermore, vaginal involvement in LS is very rare; however, LS and LP may be found in the same patient.

In general, vulvar and vaginal LP is not readily treated as these lesions frequently are impervious to current therapies. Some authors have recommended the initial use of topical medications, reserving systemic treatments for patients who fail topical treatments or those with extensive disease affecting multiple areas of the body [29].

First-line treatment of vulvar LP are daily topical potent or ultrapotent corticosteroid ointments such as fluocinonide 0.05% or clobetasol propionate 0.05% [26]. A warm sitz bath before application may allow better penetration through keratinized lesions. Vaginal LP is treated with intravaginal hydrocortisone suppositories. Commonly used formulations are those used to treat hemorrhoids [30]. Other treatment options include potent corticosteroid ointment applied to a vaginal dilator and inserted into the vagina. This treatment also helps in preventing obliteration of the vagina.

Tacrolimus, a topical macrolide immunosuppressant has recently been described for the treatment of a vulvovaginal LP [31,32]. When applied with a vaginal applicator, serum levels are high enough to achieve systemic immunosuppression. However, when applied intravaginally with a finger or with a suppository, serum levels are negligible [26]. Topical cyclosporine has also been described for the treatment of oral and vulvovaginal LP. However, it is of limited utility because of its irritative properties and high cost.

For women whose disease is refractory to topical treatments, oral corticosteroids such as prednisone will usually control the disease. In general, patients with LP should be seen within 1 month of initiating treatment. The frequency of office visits thereafter will be dictated by the patient's symptoms and severity of disease. Topical corticosteroids should be used until all active lesions have resolved. The frequency of application is then tapered.

Conclusion

Sexual dysfunction, specifically dyspareunia, is common in women and the etiology may be multifactorial. All of the vulvar dermatoses may contribute to dyspareunia and should be considered in the differential diagnosis. A detailed history, careful physical examination, and appropriate laboratory testing will aid in establishing the diagnosis. A compassionate and empathetic approach is essential to treating these patients.

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Statement of Authorship

Category 1

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- (a) Drafting the Article Howard A. Shaw; Lara J. Burrows
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Category 3

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