Benign Diseases of the Vulva

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Abstract

Many benign diseases can affect the vulva. These varied conditions can present similarly, often with pain, burning, pruritus, vaginal discharge, and dyspareunia. Physical exam findings can be nonspecific, with erythema, excoriations, and lichenification. Contact dermatitis is a frequent cause of vulvar pruritus and pain, and nearly all topical vaginal products can cause irritation. Removal of common irritants usually results in disease resolution. Lichen simplex chronicus is a condition characterized by severe pruritus leading to a persistent itch-scratch cycle. Lichen planus is a chronic, immune-mediated dermatosis presenting with pruritic papules on the vulvar skin and white lacy striae on mucosal surfaces. Lichen sclerosus is an immune-mediated disorder that results in waxy white "cigarette paper" changes to vulvar skin and can also affect the perianal region resulting in a classic "figure of eight" appearance. Patients with lichen sclerosus should be followed closely due to an increased risk of vulvar squamous cell carcinoma. Psoriasis is a dermatologic condition which can manifest as painful and pruritic vulvar erythema. Topical corticosteroids are the first-line therapy for lichen simplex chronicus, lichen planus, lichen sclerosus, and vulvar psoriasis. Vulvar intraepithelial neoplasia (VIN) is a premalignant condition which can present variably with raised or flat lesions, ranging in color from white to red or black. It can be associated with carcinogenic human papillomavirus (HPV) infections as well as with vulvar dermatoses. All cases should be treated, and options range from excision, to laser ablation, to topical medical therapy with Imiquimod. Patients remain at risk for recurrence and require continued surveillance.

Keywords

Vulva • Dermatoses • Contact dermatitis • Lichen simplex chronicus • Lichen planus • Lichen sclerosus • Psoriasis • Vulvar intraepithelial neoplasia • VIN

Contents

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1 Introduction

This chapter focuses on the diagnosis and management of common benign vulvar diseases. Benign vulvar disease encompasses numerous disorders, including infections, autoimmune diseases, topical irritation, dermatoses, neoplasms, cysts, and masses. Many of these varied diseases can present quite similarly. Patients often complain of vulvar irritation, burning, pain, pruritus, dysuria, dyspareunia, and vaginal discharge. On exam many vulvar conditions appear similar, with erythema, edema, signs of excoriations leading to ulcerations, and lichenification in cases of chronic scratching. This review will focus primarily on dermatoses commonly affecting the vulvovaginal region, including contact dermatitis, lichen simplex chronicus, lichen planus, and lichen sclerosus, as well as psoriasis and vulvar intraepithelial neoplasia.

Diagnoses are often made clinically after assessing for a history of medication and product use and ruling out common infections including vulvovaginal candidiasis. Treatment is usually initiated empirically, and the majority of vulvar dermatoses are treated with topical corticosteroids. However clinicians should have a low threshold to obtain a biopsy in cases of unusual appearing lesions or in instances of treatment failure.

Disease prevalence, common symptoms, physical exam findings, recommended diagnostics, treatments, and prognoses of common vulvar dermatoses will be reviewed here. Given that many of these conditions can become recurrent or chronic, it is important that physicians remain mindful of the potential impact on patients' quality of life that these diseases can have and continue to work with patients to achieve good symptom control.

2 Vulvar Dermatoses

2.1 Contact Dermatitis

Contact dermatitis affects approximately 15–30% of the population (Crone et al. 2000; Moyal-Barracco and Wendling 2014). Common causes of vulvar contact dermatitis are irritants and allergens. Fragrances, topical medications, and preservatives are the most frequent culprits (Hoang et al. 2014; O'Gorman and Torgerson 2013). Acute cases of contact dermatitis can present with erythema, edema, and even vesicles or erosions. Chronic contact dermatitis often presents as erythema with lichenified or excoriated areas. Patients complain of pruritus, pain, vulvovaginal dryness, burning, and dyspareunia (Fischer et al. 1995; Hoang et al. 2014).

Irritant contact dermatitis is the most common subtype. Factors including moisture from sweat or urine, heat, friction from clothing or scratching, estrogen deficiency, or enzymes can damage the skin's barrier properties and exacerbate irritation due to topical products (Margesson 2004; Moyal-Barracco and Wendling 2014). The list of potential irritants is extensive and essentially includes anything that comes in contact with the vulvovaginal region. Common irritants include soaps and detergents, dyes, perfumes, deodorants, lotions, condoms, spermicides, lubricants, sanitary products including tampons and pads, topical medications including anesthetics, antibacterials, antifungal preparations, corticosteroids, and bodily fluids including semen, saliva, and urine (ACOG 2008; Margesson 2004; Marren et al. 1992).

Allergic contact dermatitis is an immune-mediated type IV hypersensitivity reaction to an allergen which occurs in sensitized patients (Margesson 2004; Moyal-Barracco and Wendling 2014). Numerous allergens have been identified; however, topical medications, including lidocaine, corticosteroids, and neomycin, as well as perfumes and metal such as nickel, are known to be causative agents (Marren et al. 1992; Moyal-Barracco and Wendling 2014). Patch testing in conjunction with a dermatologist can be considered if the etiologic agent cannot be identified (Crone and Stewart, 2000).

Patients with vulvar contact dermatitis often complain of burning or pruritus, and the course can be either acute or chronic. Poorly defined erythema is often noted on exam, and associated findings including edema, erosions or ulcers, and papules or vesicles can be found, especially in the

area covered by diapers or sanitary napkins. If pustules or crusting are noted, a superimposed bacterial infection may be present. In chronic cases excoriation, changes in pigmentation and lichenification may be noted (Margesson 2004; Moyal-Barracco and Wendling 2014). On biopsy, contact dermatitis is characterized by histopathological findings of intraepidermal edema, spongiosis, acanthosis, parakeratosis, and infiltrating lymphocytes and eosinophils (Hoang and Reuter 2014; Moyal-Barracco and Wendling 2014).

In cases of contact dermatitis, use of all topical products should cease, and patients should use only water to clean the vulva. Resolution of symptoms is expected once the causative factor is removed. Patients may feel greater symptomatic relief with the addition of petrolatum for moisturization, and systemic antihistamines can be added for relief of pruritus (Margesson 2004; Moyal-Barracco and Wendling 2014).

2.2 Lichen Simplex Chronicus

Lichen simplex chronicus is a common dermatosis which causes severe vulvar pruritus. Initial symptoms may have been triggered by a topical irritant, candidiasis, moisture, heat, or friction, or concurrent lichen sclerosus (ACOG 2008; Virgili et al. 1997). Subsequent scratching alleviates the itch but damages the skin, over time resulting in a thickened epidermis which can appear leathery with areas of hyper- and hypopigmentation. Patients complain of a chronic recurrent pruritus, which often becomes worse at night. Frequent scratching results in erosions with accompanying pain and burning (ACOG 2008; Moyal-Barracco and Wendling 2014; Virgili et al. 2001).

Lichen simplex chronicus predominantly affects the hair-bearing skin on the labia majora. Skin often appears thickened and may be erythematous, pigmented, or pale, and excoriations are frequently noted. This condition often exhibits a chronic, relapsing pattern; however, it has not been associated with an increased risk of squamous cell carcinoma (Moyal-Barracco and Wendling 2014; Stewart 2010).

Potent topical corticosteroids are the mainstay of treatment for lichen simplex chronicus. Daily application for 3–4 weeks is recommended initially, followed by a taper or switch to a less potent topical steroid for the following 3–6 months to prevent recurrence (Lynch 2004; Moyal-Barracco and Wendling 2014). Topical calcineurin inhibitors such as tacrolimus or pimecrolimus ointment applied once or twice daily for 6 weeks, then tapered, are second line therapies for patients who do not tolerate or do not respond to topical corticosteroids (Goldstein et al. 2007; Moyal-Barracco and Wendling 2014).

2.3 Lichen Planus

Lichen planus is an inflammatory condition which can affect the vulvovaginal region as well as oral mucosa. The epidemiology of lichen planus is not well studied, but prevalence estimates range from 0.22–5%, affecting both males and females (Gorouhi et al. 2014; Shiohara and Kano 2008). Vulvovaginal lichen planus most often affects peri- or postmenopausal women, though there have been reports of affected children (Gorouhi et al. 2014; McPherson and Cooper 2010).

Lichen planus is thought to be a T-cell-mediated inflammatory autoimmune disease. Studies have suggested possible genetic susceptibility as well as environmental triggers associated with lichen planus. Associations have also been noted between lichen planus and coexisting stress, anxiety, and depression as well as hepatitis C infection, other autoimmune illnesses, dyslipidemia, and other viral infections (Gorouhi et al. 2014; Shengyuan et al. 2009; Vallejo et al. 2001).

Several subtypes of lichen planus have been described, based on morphology as well as the primary site of involvement (Gorouhi et al. 2014). Oral involvement is the most common subtype of mucosal lichen planus, with vulvovaginal involvement being the next most common and esophageal, laryngeal, and conjunctival involvement being rare (Eisen 1999; Gorouhi et al. 2014). In addition to affecting the mucous membranes of the mouth, vulvovaginal and anogenital region, gastro intestinal tract,

conjunctiva, and mouth, lichen planus can affect the skin, nails, and hair. Among patients with vulvar lichen planus, 43–100% also have oral lesions, and 25–57% of patients who initially present for oral lesions are found to also have vulvar lesions (Belfiore et al. 2006; Eisen 1999; Gorouhi et al. 2014). Approximately 17–22% of patients with vulvovaginal disease will also have skin lesions (Gorouhi et al. 2014, Simpson et al. 2012).

Lichen planus can have a variable appearance. Classic dermatologic presentations include the "6 Ps": "Pruritic, Purple, Polygonal, Planar, Papules, and Plaques" (Gorouhi et al. 2014; Lazar and Murphy 2009). When present on mucosal surfaces, the classic description is of white, lacy, or fernlike striae known as Wickham striae. Pruritic papules ranging in color from pink to violaceous can also be observed (ACOG 2008; Rogers and Eisen 2003).

Vulvovaginal lichen planus has three major subtypes: erosive, papulosquamous, and hypertrophic, with erosive being the most common (Gorouhi et al. 2014). Erosive vulvar lichen planus affects only the mucous membranes and can lead to vulvovaginal scarring, agglutination and resorption of the labia minora and clitoral hood, formation of synechiae, introital stenosis, and vaginal obliteration in extreme cases (Fig. 1). Affected women often complain of dyspareunia, burning, vaginal discharge, and postcoital bleeding and can have significant psychological distress due to the condition (Gorouhi et al. 2014; Lewis 1998).

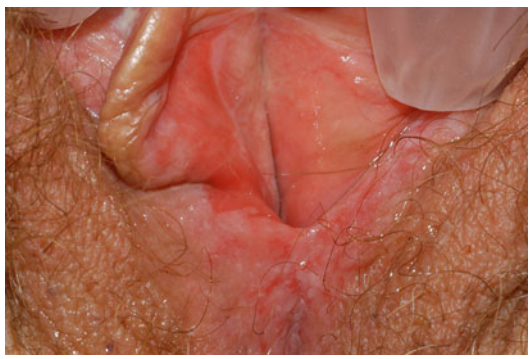

Fig. 1 A case of erosive lichen planus with early vaginal agglutination. (Reprinted from American Journal of Obstetrics and Gynecology, Vol 214, Issue 2, Fairchild PS, Haefner HK, Surgical management of vulvovaginal agglutination due to lichen planus, 289.e1-2, 2016, with permission from Elsevier)

Vaginal pH is frequently increased to 5 or 6 in lichen planus, and a wet mount often reveals numerous inflammatory cells as well as immature parabasal and basal epithelial cells (ACOG 2008; Rogers and Eisen 2003). Biopsy of the lesion confirms the diagnosis, and immunofluorescence staining can be used to distinguish between lichen planus and other similarly presenting immunobullous diseases. A dense continuous band of infiltrating lymphohystiocytes at the dermal-epidermal junction in the upper dermis is the classic histologic finding (Fung 2016; Gorouhi et al. 2014; Ramer et al. 2003). Other findings include hyperkeratosis, areas of wedge-shaped hypergranulosis, spongiosis in the spinous layer, squamatization, as well as elongated and rounded rete ridges. However, erosive vulvovaginal lichen planus can often have nonspecific histopathological findings, lacking the classic markers (Fung 2016; Gorouhi et al. 2014).

Topical application of super potent corticosteroids is the first-line treatment of vulvovaginal lichen planus with up to 55% of patients experiencing symptomatic improvement with topical corticosteroids alone, while some patients will require oral therapy with prednisolone or a combination of topical and oral corticosteroids (Schlosser and Microwski 2015). Topical corticosteroid therapy has been shown to be effective in treating symptoms including burning, pruritus, discharge, and dyspareunia; however the symptoms caused by vaginal stenosis were not significantly improved by topical corticosteroid therapy (Anderson et al. 2002; Schlosser and Microwski 2015). A variety of systemic immunomodulators have been studied with limited benefits observed and often with significant adverse effects noted. As such, systemic treatments are often limited to use for severe disease or refractory cases (Lotery and Galask 2003; Schlosser and Microwski 2015).

Vaginal dilators can be helpful in both treatment and prevention of adhesion and synechiae

formation, which can occur in cases of inflamed, ulcerated vaginal mucosa. Dilators are often used in conjunction with topical corticosteroids, estrogen creams, or topical calcineurin inhibitors and can be used twice daily initially, then taped down to three times per week. Dilator therapy has been shown to be beneficial in helping patients resume sexual intercourse (Lotery and Galask 2003; Schlosser and Microwski 2015). In addition to dilators, use of vaginal hydrocortisone suppositories at a dose of 25 mg, initially used twice daily, then tapered down to twice weekly maintenance therapy, has been demonstrated to be effective for controlling symptoms of lichen planus (Anderson et al. 2002). If synechiae do form, surgical therapy should not be attempted until the inflammation due to lichen planus is well controlled. Lysis of adhesions and careful tissue dissection should be performed under general anesthesia, and vaginal dilators along with topical corticosteroids, with the addition of topical estrogen therapy in certain cases, must be used during the healing process to prevent scarring and additional adhesion formation (Kortekangas-Savolainen and Kiilholma 2007; Schlosser and Microwski 2015).

Regarding prognosis, cutaneous lichen planus can resolve in about 6 months to 1 year; however some types of oral lichen planus can become chronic and progressive. Erosive lichen planus, which is the most common subtype affecting the vulvovaginal region, can display a chronic, waxing and waning, cyclic pattern. Risk of malignant transformation of lichen planus has been suggested, but studies have been inconclusive, and further study is required (Cooper et al. 2004; Gorouhi et al. 2014).

2.4 Lichen Sclerosus

Lichen sclerosus is one of the most common inflammatory dermatoses of the vulva. It is a chronic autoimmune inflammatory condition affecting the vulvovaginal region, thought to be mediated by lymphocytes. It is more common in women than men (Schlosser and Microwski 2015). Lichen sclerosus exhibits a bimodal age distribution. It occurs commonly in prepubertal girls with a prevalence of 1 in 900 and a mean onset at 5 years of age. Perimenopausal to postmenopausal women are usually affected between ages 45 and 55; however, it can also present in women of reproductive age. There is often a delay in diagnosis of approximately 2 years in young patients and 5–15 years in older women. Cutaneous lesions outside of the genital region can be present in 6–15% of patients affected by lichen sclerosus (Cooper et al. 2004; Schlosser and Microwski 2015).

Reports suggest a familial predisposition, though the mode of inheritance has not been identified. Several genes have been studied with possible protective as well as predisposing genes under investigation. There is also a significant link between lichen sclerosus and autoimmune disorders, including autoimmune thyroid disease, as well as vitiligo, alopecia areata, pernicious anemia, and diabetes mellitus, among others (Meyrick et al. 1988; Schlosser and Microwski 2015). Additionally the role of estrogen and androgens in lichen sclerosus has been investigated but remains a subject for further study (Schlosser and Microwski 2015; Taylor et al. 2008).

Patients commonly present with vulvar pruritus which can be more severe at night. They can develop painful vulvar and perianal erosions and fissures which lead to dysuria, dyspareunia, and dyschezia. Patients can develop urinary retention and constipation secondary to pain. As such, stool retention is commonly the presenting symptom of lichen sclerosus in young girls. Disease progression and labial agglutination are concerns for young girls diagnosed with lichen sclerosus, and abnormal voiding with weak urinary stream can result from labial agglutination (Powell and Wojnarowska 2001; Schlosser and Microwski 2015).

Lichen sclerosus typically affects the labia majora and minora, clitoris and clitoral hood, and the posterior fourchette. The perianal area is affected in 30–60% of women, often with a classic hourglass or "figure of eight" pattern when both the vulva and perianal region are affected. The vaginal mucosa is usually spared. A white, hypopigmented vulvar skin with a waxy or

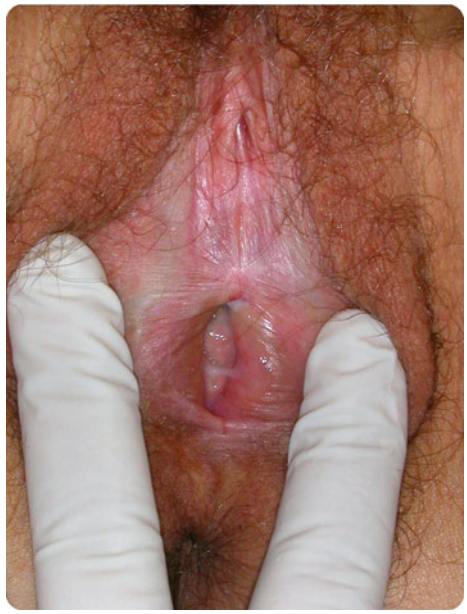

Fig. 2 Scarring of the vulva from lichen sclerosus. (Reprinted from the American Journal of Obstetrics and Gynecology, Vol 196, Issue 2. Goldstein AT, Burrows LJ. Surgical treatment of clitoral phimosis caused by lichen sclerosus, 126.e1-4, 2007, with permission from Elsevier)

wrinkled texture, classically described as appearing like "cigarette paper," is the typical clinical exam finding. Patients may also have fissures, erosions, and ulceration as well as hyperkeratosis or purpura, and clitoral hood edema (Schlosser and Microwski 2015; Virgili et al. 2014). Scarring can occur to varying degrees, most commonly affecting the clitoral hood and labia minora as well as the posterior fourchette and introitus (Fig. 2). Resorption of the labia minora has been observed, and scarring can result in stenosis of the vaginal introitus or urethral meatus to the point that intercourse and voiding are impaired. Lichen sclerosus can also result in areas of hyperpigmentation which can be difficult to differentiate from potential melanoma, and as such, biopsy is recommended. Extragenital lesions in cutaneous lichen sclerosus can also be observed and predominantly affect the trunk, though rare cases affecting the face, scalp, mouth, and nails have been reported (Schlosser and Microwski 2015).

Treatment with topical corticosteroids is recommended if clinical signs are suggestive of lichen sclerosus. Biopsy is indicated if lesions do not respond to topical therapy; if there is concern for malignancy, including melanoma; or if ulcerated lesions are present (Schlosser and Microwski 2015). Hyperkeratosis, an atrophic epidermis, loss of rete ridges, and band-like lymphocytic infiltrate are classic histopathological signs of lichen sclerosus (Schlosser and Microwski 2015).

Lichen sclerosus is associated with a nearly 300 times increased risk of development of vulvar squamous cell carcinoma, with a period of 4–10 years between diagnosis of lichen sclerosus and diagnosis of vulvar squamous cell carcinoma. It is unclear if successful treatment of vulvar lichen sclerosus decreases the risk of vulvar squamous cell carcinoma; however, carcinoma is seen typically in elderly patients and has not been reported in pediatric patients. Age greater than 70 years has been associated with treatment failure and disease recurrence (Carli et al. 1995; Schlosser and Microwski 2015).

Topical treatments in ointment formulations are preferable as they have improved potency and absorption. Ointment formulations also have emollient properties and are less likely to contain vulvar irritants including preservatives, alcohol, and propylene glycol. Topical superpotent corticosteroids are the mainstay of therapy for vulvar lichen sclerosus, and clobetasol propionate 0.05% ointment has demonstrated significant efficacy in randomized trials (Schlosser and Microwski 2015). An initial twice-daily application is recommended. As symptoms improve, the application can be tapered and a less potent corticosteroid can be used. Maintenance therapy over a period of 6–12 months is commonly recommended. Side effects of long-term topical corticosteroid therapy include atrophy and thinning of the skin as well as development of striae and telangiectasias. Long term use can also increase susceptibility to superimposed candida infections and can be associated with reactivation of herpes simplex viral infection (Schlosser and Microwski 2015).

Topical estrogen therapy may be beneficial in postmenopausal patients who experience

significant atrophy with topical corticosteroid use. In addition, oral antifungals and antihistamines can be used to combat side effects rather than utilizing their topical formulations. Topical creams may exacerbate symptoms due to their potential to act as topical irritants. Second-line therapy includes a twice-daily application of topical calcineurin inhibitors, such as tacrolimus 0.1% ointment, which may be chosen for use in cases which demonstrate a lack of response to topical corticosteroids or in patients who develop side effects from corticosteroid therapy; however, their use has been associated with a reportedly increased risk of dermatologic malignancies (Fischer and Bradford 2007; Schlosser and Microwski 2015).

Systemic retinoid therapy such as acitretin can be useful for patients with hyperkeratotic or hypertrophic vulvovaginal lichen sclerosus refractory to superpotent topical corticosteroids. Oral acitretin regimens of 20–30 mg per day over a course of 16 weeks have been studied; however, as retinoids are teratogenic, they should not be used in women who might become pregnant. There is evidence that acitretin can reduce the incidence of squamous cell carcinoma in at-risk patients such as those who have undergone organ transplantation. It is suggested that there may also be a similar protective effect against squamous cell carcinoma in patients with lichen sclerosus; however, further study into this area is necessary (Bousema et al. 1994; Schlosser and Microwski 2015).

Surgical therapy for severe vulvar and introital scarring affecting function should only be undertaken after resolution of inflammation with at least a 6-month, disease-free period. Superpotent topical corticosteroids must be used to prevent a postoperative lichen sclerosus recurrence due to the Koebner phenomenon (the development of new lesion areas secondary to trauma) as well as to prevent recurrence of labial agglutination and stenosis of the introitus (Schlosser and Microwski 2015).

Additional supportive measures can be undertaken to help manage the pruritus, pain, and irritation. Patients should wash the vulvar area with water only, then pat dry and avoid rubbing. Sitz baths as well as ice packs and cool compresses can be utilized to ease burning symptoms. Petroleum jelly or A and D ointment can act as soothing emollients and decreases friction in the affected areas. Some patients develop a secondary vulvodynia which may be treated with topical 5% lidocaine ointment, while other patients may benefit from systemic treatments for neuropathic pain including amitriptyline and gabapentin (Schlosser and Microwski 2015).

In most patients disease control should be attained within 3–4 months. Patients should then be followed every 6–12 months to monitor for disease recurrence as well as for the development of atrophy, scarring, or potential malignant changes. Cases that do not improve despite appropriate therapy should be reassessed, and alternate diagnoses such as contact dermatitis, a superimposed infectious process, or undiagnosed malignancy should be considered and further evaluated. Suspicious or unresponsive areas must be biopsied. A multidisciplinary approach should be considered for patients with refractory or multifocal disease as well as to help manage common psychological effects secondary to severe or persistent disease (Schlosser and Microwski 2015).

2.5 Psoriasis

Vulvar psoriasis affects approximately 5% of women and 15% of girls with vulvar dermatosis. This chronic dermatosis is generally diagnosed clinically and has several presentations, including the classic form with well-demarcated thick scaly plaques, discrete pustules in the pustular form, or as erythema with minimal scaling in the inverse form which presents predominantly in the anogenital area and in flexural folds. Vulvar psoriasis affects the hair-bearing cutaneous regions of the vulva, especially the mons pubis and labia majora, and generally spares the mucosa (Hoang et al. 2014; Kapila et al. 2012). Biopsy frequently is nondiagnostic, however areas of parakeratosis with neutrophils, known as Munro's microabscesses, as well as spongiosis are a common finding (Hoang et al. 2014).

Treatment of vulvar psoriasis should focus on relief of symptoms and the minimization of Koebnerization. Use of soothing emollients and avoidance of topical irritants is recommended. Flares can be managed with topical mid- to high-potency topical corticosteroids. Topical corticosteroid therapy can then be followed with topical tar preparations, retinoids, calcineurin inhibitors, and low potency topical corticosteroids for maintenance therapy. In rare cases of severe refractory disease, systemic therapy with methotrexate may be considered; however limited efficacy has been demonstrated with this treatment (Kapila et al. 2012).

3 Neoplasms

3.1 Vulvar Intraepithelial Neoplasia

Vulvar intraepithelial neoplasia (VIN) is a premalignant squamous lesion with a peak incidence of 5 per 100,000 women which is observed primarily in Caucasian women in their forties (Preti et al. 2014). VIN was historically divided into three grades. However, after studies revealed that VIN 1 was predominantly a self-limited disease due to human papillomavirus (HPV) infection, the International Society for the Study of Vulvovaginal Disease (ISSVD) revised the classification system in 2004 such that now only a high-grade disease is classified as VIN, and what was previously called VIN 1 is now classed and treated as condyloma (Sideri et al. 2005). VIN is now subdivided into usual-type VIN and differentiated VIN. Usual-type VIN is the most common type and is linked to carcinogenic HPV genotypes as well as states associated with persistent HPV infection including being immunocompromised and smoking. This grouping includes warty, basaloid, and mixed VIN. Differentiated VIN accounts for only 2–5% of cases of VIN. It is more commonly associated with vulvar dermatoses such as lichen sclerosus, a risk factor for squamous cell carcinoma, and is not associated with HPV infection (Preti et al. 2014; Sideri et al. 2005).

There is no screening test specifically for VIN, which is diagnosed clinically. VIN has a variable presentation. Color can range from gray or white to red, brown, or even black. The majority of lesions are raised (Fig. 3); however, they can be flat as well. Vulvar colposcopy can be used to aid visual inspection of the area after applying cotton pads soaked with 3–5% acetic acid. Any suspicious or pigmented lesion, any areas refractory to treatment, or lesions found in a postmenopausal woman should be biopsied (ACOG 2011; Preti et al. 2014).

All cases of VIN should be treated. If malignancy is suspected, VIN lesions should be surgically managed with wide local excision, using either a cold knife or loop electrode excision. In rare cases, such as in immunocompromised women with extensive disease, a skinning vulvectomy may be considered. Gross margins of 0.5–1 cm are preferred when performing wide local excision, though adjustments can be made to avoid structures such as the urethra, clitoris, and anus. Negative margins on pathologic examination are preferred, however risk of recurrence remains. If suspicion for malignancy is low, CO_2 laser ablation or topical therapy with imiquimod 5% for 12–20 weeks, cidofovir 1%, or photodynamic therapy in conjunction with the photosensitizing agent 5-aminolevulinic acid (ALA) can be utilized (ACOG 2011; Preti et al. 2014).

The quadrivalent HPV vaccine reduces the risk of developing VIN as it confers protection from HPV genotypes 6, 11, 16, and 18 (Muñoz et al. 2010). Smoking cessation should be stressed, as it is associated with usual-type VIN, and the diagnosis and treatment of vulvar dermatoses may reduce the risk of differentiated VIN and development of squamous cell carcinoma (ACOG 2011).

Despite treatment, recurrence rates can be as high as 30–50%, and rates are even higher if positive margins were noted on excision. After treatment, patients should be re-examined at 6 months and 1 year and can then resume yearly monitoring if no new lesions are noted (ACOG 2011), though some studies suggest following patients more closely, with follow-up every 3 months for the first 2–3 years, then every 6 months subsequently (Preti et al. 2014).

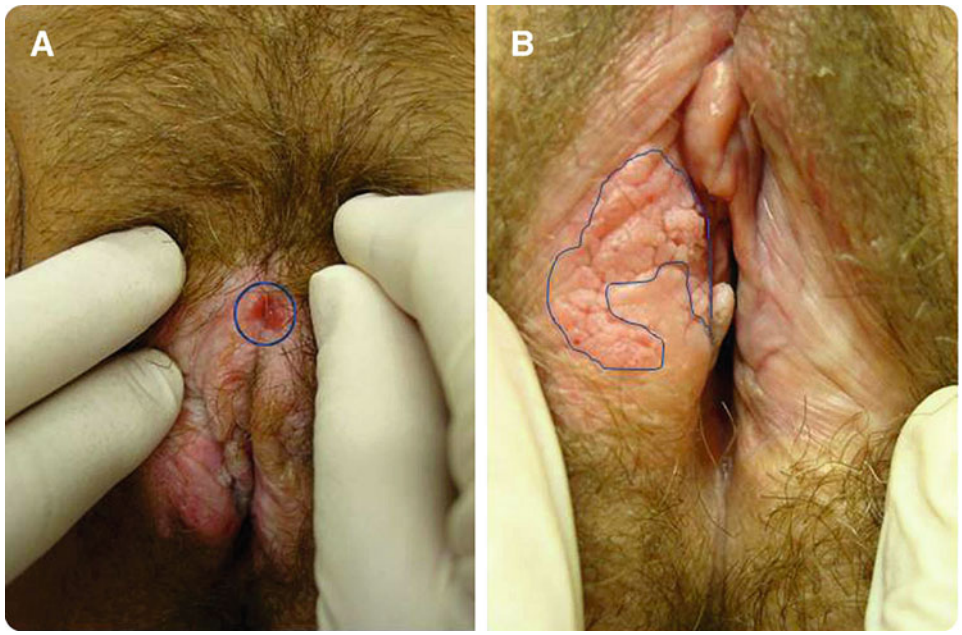

Fig. 3 (**a**) Vulvar SCC on the left labium minus, 9 mm diameter and with a depth of invasion of 4 mm. (**b**) Lesion on the right labium minus, which turned out to be VIN on histologic examination. (Reprinted from American Journal of Obstetrics and Gynecology, Vol 203, Issue 2, Simons M, Van De Nieuwenhof HP, Van Der Avoort IA, Bulten J, De Hullu JA, A patient with lichen sclerosus, Langerhans cell histiocytosis, and invasive squamous cell carcinoma of the vulva, e7-10, 2010, with permission from Elsevier)

4 Conclusion

As reviewed above, numerous conditions can manifest as vulvar disease. When evaluating vulvar complaints, a thorough history and physical examination are essential, as well as an awareness of the wide range of potential diagnoses. The first steps in evaluation often involve assessment for and removal of potential irritants as well as testing for suspected infections. Discontinuation of topical products is often a crucial component in the management of symptoms, as nearly any product in contact with the vulvovaginal region has the potential to cause irritation.

The common dermatoses reviewed in this chapter, including contact dermatitis, lichen simplex chronicus, lichen planus, lichen sclerosus, and psoriasis, are often treated with topical corticosteroids as a first-line therapy. However, providers should have a low threshold to obtain a biopsy for tissue diagnosis in refractory cases or unusual presentations in order to rule out underlying malignancy or alternate diagnoses.

In severe and chronic cases of vulvar dermatoses, a long-term follow-up is often indicated to monitor for disease recurrence or progression, potential malignant transformation, as well as vulvovaginal scarring, labial agglutination, or introital stenosis. Cases of VIN must be monitored closely due to high rates of recurrence as well as risk of malignancy.

Depending on the etiology of the vulvar disease, patients may have multifocal disease affecting more than one organ system. As such, a multidisciplinary approach should be taken to coordinate therapy. Additionally, patients can experience significant morbidity from vulvar dermatoses, which may have wide impacts on various activities ranging from normal sexual activity to basic voiding function. These morbidities can have significant psychological impacts on patients which should be addressed as a component of comprehensive gynecologic care.

5 Cross-References

- Benign Vulvar and Vaginal Pathology
- Diagnosis and Treatment of Vulvovaginitis
- Management of Chronic Recurrent Vulvovaginitis
- Management of Vaginal and Vulvar Lesions in the Older Woman
- Preinvasive Epithelial Disease of the Vulvar

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