Dermatologic Conditions of the Vulva During Menopause

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22.1 Introduction

The aim of this chapter is to provide an overview of the vulvar skin conditions commonly found in the aging female. These disorders are underreported and may be undertreated. These conditions most commonly present with pruritus or pain [1]. Other symptoms include difficulty urinating, defecating, walking, and/or interrupted, painful relations. Skin discoloration or rash can be present. Diagnosis may be delayed due to the woman's embarrassment in presenting for gynecological care of these lesions and/or discussing the symptoms with her clinician. Many may accept these distressing symptoms as signs of aging to be expected with menopause.

Vulvar skin disorders present the practitioner with a diagnostic challenge. Although biopsy is not always required to make a diagnosis, it should be performed when lesions are present that cannot be adequately characterized. If conservative management is elected, biopsy should be performed if initial treatment fails. Treatment should also include education regarding vulvar hygiene, with removal of contact irritants when identified and topical steroid creams. Despite treatment, symptoms are often chronic and persistent, interfering with sexuality, activity level, and overall quality of life and may involve continuous active management to achieve partial relief of distress for the aging woman. Our objective is to provide the practitioner with a simple diagnostic framework and treatment plan for vulvar skin dysplasias.

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22.2 The Role of the Women's Health Provider

Women may not report symptoms of vulvar itching and pain at their annual visit. Many find the complaint embarrassing. Practitioners are often more comfortable discussing chronic medical conditions rather than sexual health complaints. Thus, the health-care provider with a focus or interest in women's health plays an important role in diagnosis and treatment of vulvar dysplasias. This includes gynecologists, mid-level providers in women's health, and primary care providers; although dermatologists and gynecologic oncologists have a key referral role when there is a refractory case, a lesion suspicious for and/or a biopsy-confirmed malignancy.

The American Cancer Society recommends that women have a full-body skin examination annually beginning at age 40 [2, 3]. In a survey of 201 female veterans, 68.7 % reported that they did not regularly undergo full-body skin exam (FBSE) [4]. Primary care physicians (PCP) are often the gatekeepers, responsible for referral within managed care organizations, and trusted with the initial medical opinion in many conditions. Those who work in the field of obstetrics and gynecology play a key role in a woman's primary health care. A cross-sectional survey of patients from primary care and dermatology clinics found that patients have a high level of confidence in their PCP's ability to treat their skin conditions [5]. Although previous reviews found primary care providers not as likely to do be as thorough as dermatologists in the diagnosis of lesions, electronic age offers the ability to bridge the diagnostic gap that may be created by lack of access to specialists. With electronic medical records, digital images may be inserted to document the lesion. In remote areas without access to specialists, telemedicine offers the ability to guide diagnostic and treatment options for dermatologic conditions.

22.3 Vulvar Skin Screening

The role of the women's health-care provider in screening for vulvar skin disorders is not limited to skin cancer screening.

The majority of vulvar skin disorders diagnosed either on exam, or when a woman presents with vulvar pruritus or pain, are not malignant. See Table 22.1 for common differential diagnoses of vulvar pruritus or pain.

As a woman ages, and particularly after menopause and its resultant hypo-estrogenism, vulvar skin and vaginal mucosa undergo progressive physical changes. Estrogen plays a key role in cellular remodeling, angiogenesis, and response to oxidative stressors [6]; therefore, decreasing estrogen levels affect the macro- and microstructure of the perineum. Estrogen receptors are located in the vagina, the vulva (to a lesser degree), and the urethra [7]. Withdrawal of estrogen leads to decreased cell turnover, blood supply, and vaginal mucosal and vulvar sebaceous gland secretions. The vaginal mucosa atrophies and becomes more alkaline. The vulvar skin thins and pigmentation decreases uniformly. The labia become less full, reflecting a loss of both fat and collagen. Turgor decreases as a result of decreased glycosaminoglycan [8]. The result is vulvovaginal atrophy, also called atrophic vaginitis, as it is believed to account for vulvovaginal pain and dryness in up to 50 % of postmenopausal women. Bothersome local symptoms may be present even in 10–25 % of women using systemic hormone therapy [1, 9].

Vulvovaginal atrophy affects both the structure and function of the vulvar skin. Thinning,

Table 22.1 Differential diagnosis of vulvar pruritus or pain

Acute
Bacterial or fungal infections
Contact dermatitis
Chronic
Dermatoses
Lichen sclerosus
Lichen simplex chronicus/squamous cell hyperplasia
Other dermatoses
Vulvovaginal atrophy
Neoplasia or preneoplastic lesions
Vulvodynia
Infection
Human papillomavirus infection
Vulvar manifestations of systemic illnesses
Crohn's disease, Bechet's

more friable skin, with the loss of the hair barrier and increased vulvar permeability, becomes more susceptible to damage from local irritants. Microvasculature changes contribute to dysfunctional response to infection. Increased vaginal pH can result in colonization with pathologic organisms.

Immune function is also a contributing factor to the development of vulvar dermatoses. In recent years, research has focused on the estrogen effects of the immune system. Aging skin has been noted to show a decrease in Toll-like receptors, which serve a primary role in cutaneous host defense [10]. Proinflammatory cytokines contribute to decreasing collagen and inflammation [11]. Thus, not only does the hypo-estrogenic vulva have a diminished barrier, but it also has a limited ability to protect against infectious microorganisms and topical irritants. Furthermore, it has a decreased response to healing after insult.

22.4 ISSD Classification of Disorders

Many nonmalignant vulvar skin disorders present with the same symptoms as premalignant or malignant vulvar conditions. A skin biopsy will result in a definitive diagnosis via histopathology; however, it requires technical skill and may result in scarring. The original classification system of nonneoplastic dermatologic conditions of the vulva by the International Society for the Study of Vulvovaginal Disease (ISSVD) was based on gross description and histopathology (Table 22.2). However, in 2011, the ISSVD presented a stepwise diagnostic approach that aims to help providers make a diagnosis based on clinical presentation. The classification was based on pathophysiology, etiology, and commonalities among clinical presentation. The goal of the ISSVD was to simplify the terminology in order to arrive at a differential diagnosis. In contrast to the original classification of vulvovaginal skin diseases, the most recent classification aims to diagnose without a biopsy. However, if a biopsy is necessary, it should be performed by a healthcare provider trained in the procedure and ideally interpreted by a dermatopathologist (pathologist with specialty training in dermatological disorders) whenever possible.

Table 22.2 Nomenclature for vulvar disease

1. Nonneoplastic epithelial disorders of skin and mucosa
(a) Lichen sclerosis
(b) Squamous hyperplasia, not otherwise specified (formerly "hyperplastic dystrophy without atypia")
(c) Other dermatoses
2. Mixed nonneoplastic and neoplastic epithelial disorders
3. Intraepithelial neoplasia
(a) Squamous intraepithelial neoplasia (formerly "dystrophies with atypia")
(b) VIN, usual type
(c) VIN, differentiated type
(d) Nonsquamous intraepithelial neoplasia
(e) Paget's disease
(f) Tumors of melanocytes, noninvasive
4. Invasive tumors
(a) VIN, Vulvar intraepithelial neoplasia
(b) Paget disease

Adapted from Lynch et al. [12]

The classification of vulvar disorders is shown in Table 22.2.

22.5 Clinical History

All women, especially postmenopausal women, should be asked about symptoms related to vulvovaginal atrophy and vulvar skin conditions. These symptoms include vulvar itching, burning, bleeding, changes in pigmentation, sores, lumps, ulcers, or pain with intercourse. When present, symptom duration, intensity, and modifying factors should be assessed. All vulvovaginal contacts should be reviewed, including over-the-counter remedies, soaps, shampoos, and sanitary products. These topical irritants should be eliminated. See Table 22.3 for a list of common irritants and possible suggestions for their removal.

For intermittent symptoms, contact/symptom diaries can be recommended. Skin conditions elsewhere, such as oral lesions or eczema should be documented. Systemic disease and all medications should be reviewed.

22.6 Physical Exam

The ISVVD 2012 publication's diagnostic approach to vulvar disorders represents a philosophical return to the physical exam. Table 22.4 offers descriptive terms useful for universal documentation of vulvar lesions and clear communication with specialists.

Physical exam of the vulva begins with visual inspection. Optimal patient positioning is in dorsal lithotomy position in gynecological stirrups, although a frog-legged position on a non-gyn examination table also may be used. Lighting should be adequate. A magnifying glass can aid visualization, but is not always necessary. Acetic acid application is not recommended because its accuracy in enhancing vulvar lesions has not been validated.

Visual inspection should include assessment of aging changes in the vulvovaginal area. Loss of hair, thinning skin, increase or loss of pigmentation should be noted. Ulcerations, excoriations, or any lesions should be noted and documented as specifically as possible in the patient chart. Localization, symmetry, and number and distribution of abnormal areas should be recorded. Border asymmetry, color, and size are also key elements to note. A cotton swab can be used to diagnose provoked vestibulodynia (through elicitation of pain with point contact through the vestibule).

The surface appearance of a lesion provides clinically helpful information. If the surface is rough, it may be due to either a crust or a scale. A crust commonly appears yellow or hemosiderin colored, and its presence implies a lesion that has involved excoriation, erosion, or any type of disruption of

Table 22.3 Tips for removing common vulvar skin irritants

Avoid scented bath products, detergents, lotions, or powders
Wear natural-fiber undergarments without dyes
At night, consider going without undergarments
Avoid tightly fitting leggings, pants or nylons
Do not scrub the vulvar area during bathing; avoid all soap to the area
Avoid activities that put tremendous pressure on the perineal area, such as biking or spin class

Table 22.4 Descriptive terminology useful for describing vulvar lesions

Blister	A compartmentalized fluid-filled elevation of the skin or mucosa
Bulla (pl. bullae)	A large (>0.5 cm) fluid-filled blister; the fluid is clear
Cyst	A closed cavity lined by epithelium that contains fluid or semisolid material
Edema	A poorly marginated area of swelling due to the abnormal accumulation of fluid in the dermis and/or subcutaneous tissue; edema may be skin colored, pink, or red
Erosion	Shallow defect in the skin surface; absence of some, or all, of the epidermis down to the basement membrane; the dermis is intact
Excoriation	An erosion or ulcer caused by scratching; excoriations are often linear or angular in configuration
Fissure	A thin linear erosion of the skin surface
Lesion	A visible or palpable abnormality
Macule	A *macule* is a flat, distinct, colored area of skin that is less than 1 cm in diameter
Nodule	A large (>1.0 cm) papule; often hemispherical or poorly marginated; it may be located on the surface, within or below the skin; nodules may be cystic or solid
Papule	A *papule* is a circumscribed, solid elevation of skin with no visible fluid, varying in size from a pinhead to 1 cm
Patch	Large (>1.0 cm) area of color change; no elevation and no substance on palpation
Plaque	Large (>1.0 cm) elevated, palpable, and flat-topped lesion
Pustule	Pus-filled blister; the fluid is white or yellow
Rash	Numerous or diffuse abnormalities (it is preferable to describe the specific abnormalities using the other terms in this list)
Ulcer	Deeper defect; absence of the epidermis and some, or all, of the dermis
Vesicle	Small blister beneath the skin

Adapted from Lynch et al. [13]

the epithelial layer. A scale is characterized by reactive keratinization, such as in eczema.

Lichenification refers to the process of cutaneous thickening that occurs secondary to chronic itching or rubbing contact. As a dermatological description, it lacks specificity for the naked eye or magnifying glass. The skin may be erythematous (red), white, or skin colored. It may or may not have excoriations. Histologically, lichenification appears as a thickened epidermal layer.

Properties that may indicate an allergic reaction include red plaques and evidence of itching, such as excoriations. A clinical history of sensitive skin, seasonal allergies, and/or asthma suggests a person with "atopy." Atopic or contact dermatitis may be causing or contributing to the chronic itching.

Inflammation typically includes the appearance of excoriations and erythema (representing reactive microvasculature). Eczematous conditions are related to atopic dermatitis but typically occur without contact irritants.

Differentiating a premalignant condition from vulvar carcinoma is difficult based on appearance alone. A biopsy is recommended in cases of nonhealing erosions, ulcerative lesions, hyperpigmented lesions, or any vulvar lesion demonstrating lack of response to treatment.

A trained health-care provider can perform a vulvar punch biopsy in the office. The site is prepped with an antibacterial solution, and 1 or 2 % lidocaine is injected subcutaneously. A punch biopsy instrument is pressed against the skin in a circular fashion to penetrate the dermis. The biopsy site should include the most abnormal appearing skin lesion as well as an adjacent potion of normal appearing skin (the very edge of the lesion). Multiple biopsy sites may be needed. Forceps and scissors are used to remove the sample. Direct pressure, Monsel solution, silver nitrate, or suture may be utilized for hemostasis. Post-biopsy care includes keeping the area clean and dry, although a topical antibiotic ointment may be prescribed.

General principles of therapy include the removal of topical irritants, focus on skin hygiene, and anti-inflammatory therapy, usually in the form of topical steroid cream. Systemic treatment for vulvar dysplasias or dermatoses is prescribed for severe, recurrent or resistant disease, or in an immunocompromised individual.

Additionally, in a patient with concomitant vulvovaginal atrophy, the addition of local estrogen therapy, in the absence of contraindications, should be considered as an adjunct to any specific therapies. Vaginal creams, rings, or tablets may be used. If VIN or cancer is present on biopsy, referral to a gynecologic oncologist is recommended.

22.7 The Dermatoses

Nonneoplastic epithelial lesions fall into one of the following three categories: (1) lichen sclerosis, (2) squamous cell hyperplasia/lichen simplex chronicus, and (3) other dermatoses.

22.7.1 Lichen Sclerosus

Lichen sclerosus has many synonyms. Pathologists may still use "lichen sclerosus et atrophicus" or "lichen albus," which refer to atrophy and whitened appearance. However, the ISSVD promotes use of the term lichen sclerosus to describe a histologically confirmed condition that typically presents with the patient complaining of pruritus in the anogenital area.

The exact incidence of lichen sclerosus is not objectively known, largely because it is believed to be underreported. In a gynecology clinic, rates are estimated to be 1.7 % [14, 15]. Patients are most often postmenopausal and Caucasian.

Lichen sclerosis likely has a multitude of mechanisms. Investigators have found a 30 % correlation between lichen sclerosus and autoimmune diseases [16]. A genetic cause has been suggested, but not confirmed [17]. The tendency of patients to be postmenopausal at presentation suggests a contribution of biological skin aging. The etiology is likely multifactorial, with changes in aging skin being critical.

Although lichen sclerosis may be asymptomatic, most women report dyspareunia, chronic itching, discomfort, or dysuria. It is thought that the inflammation affects terminal nerve fibers leading to chronic itching [15].

As the Latin alternative name, lichen albus, suggests, the skin typically has a whitish appearance. It forms a porcelain-white plaque in a figure-of-eight pattern around the clitoris and anus. Excoriations and skin thickening from chronic itching may be present. Initially, there is a loss of vulvar skin structure. Over time, the architecture is disturbed leading to retraction of the clitoris, labial adhesions, and narrowing of the introitus. This loss of architecture is a key differing feature from vitiligo, which is also characterized by depigmentation. Secondary complications due to the stenotic introitus include urethral obstruction, chronic or recurrent UTIs, and anorgasmia. The genital mucosa and cervix are spared. Diagnosis is based on histopathology to exclude premalignant or malignant lesions.

Lichen sclerosis has a controversial association with squamous cell carcinoma of the vulva. Vulvar cancer affects 4 % of women per year. Vulvar cancer in older women is typically not related to HPV, as it is in younger women [18]. Frequent surveillance for squamous cell carcinoma is recommended in all women with lichen sclerosis, with biopsy recommended for new or indeterminate lesions.

Histopathology demonstrates thickened epidermis and lymphocytic infiltrate. Nuclear abnormalities on histology may be present, representing a possible continuum from lichen sclerosus to squamous cell carcinoma [19].

Ultra-potent topical corticosteroids are the primary intervention for lichen sclerosis. Clobetasol propionate 0.05 % is most often prescribed. Clobetasol is available as a cream, gel, and ointment. Ointments are formulated to provide a barrier, are typically less irritating, and provide longer medication exposure following application. Recommended dosing is 0.05 % clobetasol propionate once nightly for 4 weeks, every other night for 4 weeks then by twice a week for 4 weeks [20]. The dosage regimen should be titrated to response. There is no agreement on the frequency of clobetasol application for maintenance. Switching to less potent corticosteroids such as hydrocortisone 1 % for maintenance is recommended.

Skin atrophy, as has been demonstrated in other parts of the body after prolonged topical steroid use, is rare in the vulvar. Although short-term overuse of potent topical corticosteroids may induce local atrophy, long-term atrophic changes have not been demonstrated. The modified mucous membranes of the labia and clitoris are relatively resistant to these changes [21, 22]. Risks of developing Cushing syndrome from prolonged topical steroid use are largely theoretical.

Testosterone and progesterone topical preparations are not recommended as multiple studies have found clobetasol propionate superior to both [23]. Studies evaluating tacrolimus, photodynamic therapy, topical retinoids, and local injection of corticosteroids were largely underpowered to determine efficacy [14, 20].

Once an adequate treatment regimen is established, reexamination is necessary every 4–6 months. These exams are intended to both monitor response and to observe for skin changes that may suggest progression to squamous cell carcinoma.

22.7.2 Lichen Simplex Chronicus

Lichen simplex chronicus (squamous cell hyperplasia) is a chronic eczematous condition characterized by a repetitive "itch-scratch" cycle. Lichen simplex chronicus can occur in any age group, including children, but is most common in older women. Women with lichen simplex chronicus experience intense itching, often with disruption of both daily activity and sleep. Clinically, the skin is erythematous and thickened (lichenified) with excoriations. The affected vulvar area may appear reddened, grayish, or whitish. There may be moderate inflammation. The affected skin area is typically symmetric.

The cornerstone of treatment for lichen sclerosis is to break the "itch-scratch" cycle. Identifying and removing chemical or environmental irritants achieve this goal. This includes evaluation for underlying yeast vulvovaginitis, with treatment if present. Mid- to high-potency topical steroids prescribed for daily use will decrease inflammation. Antihistamines will aid in reduction of the intense pruritus. Antihistamines with sedating properties, such as hydroxyzine or doxepin, will decrease nocturnal scratching and improve disrupted sleep

patterns, whereas those with nonsedating properties can be used during the day. Topical steroids should be weaned as the condition improves. A biopsy is recommended initially, if the diagnosis is uncertain or if there is a poor response to treatment. Histologically, the lesions will show irregular thickening of the epidermis with a prominent nucleated keratin layer. Rete ridges, downward projections of epidermis into the dermis, will be thickened. Although biopsy results may be nonspecific, the absence of malignant or premalignant changes is documented. In refractory, biopsy-confirmed cases, systemic steroids may be used.

22.7.3 Other Dermatoses

According to the 2006 ISSVD Classification, the "other dermatoses" include (1) inflammatory dermatoses, (2) bullous dermatoses, and (3) ulcerative dermatoses. There are similarities within the subdivisions, suggesting a continuum of illness.

22.7.3.1 Inflammatory Dermatoses: Allergic

Allergic dermatitis is diagnosed in up to 54 % of patients presenting with vulvar pruritus [24]. Allergic dermatitis may be exogenous, attributed to irritants, which causes immediate symptoms, or endogenous, an underlying atopy/allergic reaction which is typically more gradual in onset. See Table 22.5 for tips differentiating the two conditions. Clinically, the two are difficult to differentiate, although evaluation and treatment are similar. These inflammatory dermatoses may be the vulvar manifestation of eczema and appear as poorly defined, patchy, erythematous lesions.

Allergic dermatitis can be diagnosed by eliciting a history of atopy or allergies or identification of an offending allergen or irritant. Patients may have extra-genital manifestations of eczema. Vulvovaginal candidiasis must be ruled out. Biopsy is generally not required, but if performed will show nonspecific changes, such as inflammation, spongiosis, or parakeratosis. Treatment is a multifaceted and includes removal of irritants, skin hydration with emollients, and the application of mid-high-potency topical corticosteroids.

Table 22.5 Differentiating allergic and irritant contact dermatitis

Condition	Allergic contact dermatitis	Irritant contact dermatitis
Signs/symptoms	Delayed-onset pruritus, edema, vesicles or bullae	Immediate burning and stinging
Diagnosis	Patch testing by allergist	History of irritant use
Treatment	Remove offending agent(s), topical hydrocortisone, sitz baths	Remove offending agent(s), topical hydrocortisone, sitz baths

In refractory cases, systemic steroids or immune modulators may be required.

Inflammatory dermatoses may represent vulvar manifestation of psoriasis. Similar to atopic eczema, psoriasis is defined by the presence of specific lesions and usually a history of nongenital psoriasis. Psoriatic lesions are thick erythematous plaques with silvery scales Successful treatment of psoriasis may require systemic therapy with one of the several FDA-approved immunomodulating biologic agents such as infliximab, adalimumab, etanercept, alefacept, and ustekinumab [25]. For local relief, high-potency corticosteroids are often prescribed. Referral to a dermatologist or healthcare professional with experience in treating psoriasis is recommended.

Intertrigo is an inflammatory condition commonly associated with obesity. Intertrigo is characterized by an erythematous rash that often presents in the genitocrural folds. Candida (yeast) is the most common organism that presents in conjunction with intertrigo. Treatment is aimed at keeping skin folds clean and dry. Antifungal powder has both a moisture-wicking quality and antifungal properties. If inflammation is present, a topical corticosteroid may be used, although treatment of the fungal infection is first-line treatment. Diaper rash creams can also be useful as a protective barrier.

22.7.3.2 Inflammatory Disorders: Lichen Planus

Lichen planus is a mucocutaneous inflammatory disorder thought to be caused by a disturbance in the cell-mediated immune system. A genetic

component of the genital form has been postulated. Lichen planus commonly presents in women aged 50–60 and can affect the hair, nails, mouth, and/or genital area. The exact incidence of lichen planus is not clearly documented. Lichen planus has three genital manifestations – erosive, papulosquamous, and hypertrophic. The erosive type is the most common form.

The appearance of erosive lichen planus is variable, including white epithelium or red or purple papules. The classic presentation found on mucous membranes is of white, lacy, or fern-like striae, called "Wickham's striae." In the erosive form, vaginal ulcerations may be present. Commonly, the six Ps (planar [flat], purple, polygonal, pruritic, papules, plaques) are cited as common signs [26]. Unlike lichen sclerosis or lichen simplex chronicus, itching is not the most common presenting symptom. More commonly, patients present with burning and dyspareunia.

Vulvovaginal-gingival syndrome occurs when lichen planus also involves the oral mucosa. Typically, the vulva is spared in lichen planus, whereas, lichen sclerosus involves the labia majors but does not include the vagina. In long-standing disease, vulvar and vaginal architecture may be disrupted. Vaginal discharge is a common complaint in addition to burning and dyspareunia, due to the desquamation of surface (parabasal) cells. Lichen planus is especially difficult to treat in the menopausal woman as the skin changes that are typically seen with menopause may be associated with decreased immune response as well as delayed healing.

Despite the potential of delayed healing, a biopsy is recommended for diagnosis of lichen planus. Histologically a band-like lymphocytic infiltrate and colloid bodies in the basal layer of the epidermis will be seen [27].

Papulosquamous and hypertrophic lichen sclerosis are less common than erosive lichen planus. Papulosquamous lichen sclerosis is characterized by pruritic papules. Thickened (hypertrophic) plaques are common in the hypertrophic form, which may appear similar to squamous cell carcinoma.

Once diagnosis is established, treatment focuses on symptom control. Initial treatment is with high-potency steroids topically, although systemic steroids may be needed. Vaginal suppositories of 25 mg of hydrocortisone are usually effective for vaginal manifestations. They are dosed in a tapering fashion starting at twice a day over several months. Vaginal stenosis or other architectural changes from long-standing disease cannot be reversed. Oral pharmacotherapy with immunomodulators or biologic agents is initiated in persistent or refractory cases. Lichen planus is a chronic condition for which management rather than cure is the goal of treatment.

22.7.3.3 Inflammatory Disorders: Other Conditions

Other conditions that can have both oral and genital manifestations, like lichen planus, include aphthous ulcers and Bechet's disease.

Aphthous ulcers, or "canker sores," are more common in women than men. These ulcers are likely related to immunodeficiency, although no direct cause has been identified. Nutritional deficiencies have been associated with their development, namely, B12, iron, thiamine, and zinc [28]. However, vitamin supplementation is not successful in prevention or treatment of vaginal aphthous ulcers. Recurrent or severe apthous ulcers have been associated with CMV, HIV, mycoplasma, EBV, or inflammatory bowel disease. A short course of oral or topical corticosteroids can be effective, but colchicine, dapsone, cyclosporine, or thalidomide may be required in difficult to treat cases [29].

Bechet's disease may present with aphthous – appearing ulcers in the genital, oral, and ocular mucosa. Typically presenting in Asian or Middle Eastern females in the third or fourth decade of life, this systemic illness may be persistent and progressive. Bechet's disease involves systemic vasculitis that affects the GI tract, brain, joints, lungs, and large vessels as well as microvasculature. Topical steroids are recommended for treatment of vaginal lesions. Control of systemic symptoms is necessary to prevent further erosion. If Bechet's disease is suspected, a rheumatologist referral is recommended.

Hidradenitis suppurativa is a chronic condition affecting apocrine glands. The glands become blocked, an inflammatory process ensues and superinfection leads to abscess formation. The process is recurrent and leads to scarring and formation

of sinus tracts. The most commonly affected sites are the axillae, inguinal, perianal, perineal, inframammary, and retro-auricular regions.

Both genetic and environmental factors are thought to play a role in the pathophysiology of hidradenitis. A hormonal cause is postulated, likely in relation to androgens, as disease is generally from puberty until age 40. Other factors for disease development include obesity, diabetes and a likely familial predisposition. Population studies in postmenopausal women are scarce, but since androgen levels decrease in this population, the incidence of hidradenitis may be lower than in younger women.

Diagnosis is by history and physical exam. Treatment is challenging and aimed at prevention. Incision and drainage is useful for symptom relief, but frequent recurrence necessitates wide local excision of most severely affected areas. Topical clindamycin or intralesional triamcinolone is effective for treatment of local infection [30].

Medical treatments options include systemic steroids, antibiotics, antiandrogens, cyclosporine, or infliximab. Antiandrogens are largely not looked favorably upon as treatment in the aging female as testosterone levels are low in this population.

22.7.3.4 Pigmented and Depigmented Lesions

Vitiligo is a rare skin condition. The exact prevalence is unknown, but is thought to be 0.38 % in the USA [31]. Vitiligo is characterized by depigmented skin, which, histopathologically, is due to a loss of melanocytes in the epidermis. The perineal and perianal skin may be affected, with a generally symmetric distribution.

The pathophysiology of vitiligo is unknown. Familial patterns of vitiligo suggest a genetic contribution [32]. There may also be an association with autoimmune disorders. Unlike lichen sclerosis, vitiligo has no risk of progression to squamous cell carcinoma and does not cause progressive scarring. Vitiligo may however, co-exist with lichen sclerosis. Vitiligo has equal prevalence among different ethnicities. It is, however, more noticeable and possibly more disfiguring in individuals of darker skin tone.

Treatment, when desired by the patient, includes UVB phototherapy and topical immunomodulators. The most important aspect of treatment may be ruling out other conditions such a lichen sclerosis, which can produce a depigmentation but with loss of vulvar architecture. Vitiligo is not associated with pruritus or dyspareunia.

Hyperpigmentation of vulva is commonly associated with nevi. Common names include moles or skin tags. In the vulvar area, nevi should be closely monitored for progression, just as they should in any part of the body. Melanoma is the second most common vulvar malignancy after squamous cell carcinoma although both are rare primary vulvar diseases.

As with skin lesions, the ABCDs should be closely monitored on an annual basis at the minimum:

A. Asymmetry
B. Border (irregular)
C. Color (a change in color or blackish/bluish appearance)
D. Diameter (greater than 6 mm)

With electronic medical records, the feasibility of including a digital image of the lesion is increased and recommended. When a lesion changes appearance, has bleeding, pruritus, or surface skin breaks, a biopsy is recommended. The biopsy follows the same technique as previously described if it is a flat lesion. A raised lesion can be excised in the office after infiltration of local anesthetic. A dermatopathologist is optimal when available, as they usually offer the highest level of interpretation. After excision and healing of a benign lesion, there is no exact recommendation of ongoing surveillance. Yearly clinical exams are warranted, but may be biyearly in a high-risk patient, such as one with a history of a melanoma in a nonperineal location.

22.8 Systemic Diseases

Anogenital involvement in Crohn's disease is common, affecting up to 80 % of patients with bowel disease, depending on the definition of perianal involvement [33]. Although t he disease is usually diagnosed in younger women, 80 % of cases are diagnosed before age 40 [3]. External lesions secondary to Crohn's are typically ulcers,

so called "knife-cut" ulcers that appear to be stab wounds. The interlabial and genitocrural folds are especially prone to external manifestations of Crohn's. Fistulas may develop over time, which require surgical management. Treatment of the lesions is with topical corticosteroids. Control of the primary illness is paramount.

Acanthosis nigricans is a skin manifestation of hyperinsulinemia. The development of skin lesions is believed to be secondary to insulin-like growth factor stimulation of keratinocytes and dermal fibroblasts. Lesions are raised plaques with irregular contour, typically occurring in groups, most commonly located on the neck, axillae, and genitocrural folds. Patients diagnosed with acanthosis nigricans should be screened for insulin resistance and diabetes, if not previously diagnosed. Typically, the lesions regress when glucose levels are well controlled.

22.9 Summary

The aging or postmenopausal female is prone to the development of vulvar dystrophies and dermatoses. In addition to primary conditions, systemic illness also may have genital manifestations. Benign and premalignant or malignant lesions may present with similar symptoms or clinical appearance. A biopsy is not always required but is strongly recommended, especially if the lesion cannot be characterized or is not responsive to treatment. Common treatment of vulvar dystrophies and dermatoses includes high-potency topical steroids and removal of environmental irritants. Local estrogen therapy, in appropriate women without contraindications to its use, should be considered as an adjunct to other therapies. Frequent follow-up is recommended to monitor progress with treatment, and referral should be initiated as warranted.

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