Abstract

Objective. The study aimed to formulate an easy clinical approach that may be used by clinicians of all backgrounds to diagnose vulvar dermatological disorders.

Materials and Methods. The International Society for the Study of Vulvovaginal Disease appointed a committee with multinational members from the fields of dermatology, gynecology, and pathology and charged the committee to formulate a clinically based terminology and classification of vulvar dermatological disorders. The committee carried out its work by way of multiple rounds of e-mails extending over almost 2 year’s time.

Results. The committee was able to formulate a consensus report containing terminology, classification, and a step-wise approach to clinical diagnosis of vulvar dermatological disorders. This report was presented and approved by the International Society for the Study of Vulvo vaginal Disease at the XXI International Congress held in Paris, France, on September 3 to 8, 2011.

Conclusions. The authors believe that the approach to terminology and classification as well as clinical diagnosis contained in this article allows clinicians to make highly accurate diagnoses of vulvar dermatological disorders within the clinical setting. This, in turn, will reduce the need for referrals and will improve the care for women with most vulvar disorders.

Key Words: vulvar disorders, vulvar diseases, classification, terminology, nomenclature

Nomenclature (terminology) refers to the naming of diseases. Developing a consensus on the appropriate names for diseases is difficult enough even when one is dealing with a single language and a single specialty. Not surprisingly, the task becomes much harder when considering, as we are here, the needs of clinicians from multiple specialties along with clinicians from countries where English is not the primary language. To clarify the current nomenclature used in describing vulvar dermatological diseases, a committee on terminology and classification was intentionally chosen to have international expertise and representation from the fields of dermatology, gynecology, and pathology.

Classification in a medical setting refers to the orderly arrangement of diseases into groups in which the members of these groups share common characteristics. Classification may be considered from several standpoints. Most commonly, authors choose a system of classification based on a clustering of diseases according to etiology or pathophysiology using categories such as infections, endocrinopathies, malignancies, etc. This is intellectually satisfying, and for this reason, it is the approach taken in almost all medical textbooks and other publications. However, there are 2 major drawbacks with this approach. First, one must already know
the name of the disease (i.e., the diagnosis) before it is possible to look up additional material about the disorder. Clearly, this type of classification is not helpful for a clinician who is trying to establish a diagnosis for a patient with a disease that the clinician is not able to identify. Second, etiology and pathophysiology are subject to continual change owing to the development of new information, and thus, such classifications are not durable, requiring review and revision at fairly frequent intervals.

The other major approach to classification is to cluster diseases into groups with similar clinical presentations. This is difficult to accomplish when dealing with those diseases for which the diagnosis depends on a mix of history and indirect examination, but it is appreciably easier when one is dealing with dermatological disorders wherein simple visual examination provides nearly all of the information needed to arrive at a correct diagnosis. With such an approach, diseases with similar visual characteristics are grouped (classified) together. This approach then creates a relatively short list of differential diagnoses. Often, the list can be shortened even further using general medical knowledge and common sense. This leaves the clinician with just a few diseases to consider. These can then easily be reviewed in a standard textbook, after which, the clinician can usually establish a single most likely diagnosis. Not surprisingly, we have taken this approach for our classification of dermatological vulvar disorders.

The target audience for this terminology and classification of the International Society for the Study of Vulvovaginal Disease (ISSVD) is all those who participate in the care of women with vulvar dermatological problems. This includes gynecologists, dermatologists, primary care physicians, pathologists, nursing personnel, counselors, physical therapists, and students in health care fields. This classification only includes dermatological conditions characterized by the presence of visible lesions. Therefore, it does not address those patients whose vulvar disorders present without visible lesions and who instead present only with symptoms (e.g., pruritus and pain) and/or functional problems.

The objectives of this clinical approach to the diagnosis of vulvar dermatological disorders were 5-fold, (1) to accurately describe vulvar lesions; (2) to place the described lesion(s) within 1 of the 8 distinct morphological groups found in the accompanying clinical classification; (3) to formulate a short list of differential diagnoses from within that group; (4) to shorten that list by way of reading about the clinical presentation of those diseases; and (5) to use, when necessary, laboratory testing to identify the most likely diagnosis.

The clinical accuracy of this clinical approach to diagnosis depends greatly on the quality and thoroughness of one’s examination. Appropriate examination requires that the vulva be completely exposed and under adequate lighting. Patients should be placed in stirrups, lying on their back in a “frog-leg” position or otherwise situated such that the vulva can be completely visualized. The vulva should be illuminated using a slanted or horizontal lighting and is examined with the naked eye or with a 2- or 3-power magnifying lens. Currently, there is insufficient data to recommend the use of higher power magnification such as with a colposcope. Because both sensitivity and specificity are lacking, it is not recommended that acetic acid be used as a tool for routine vulvar examination.

When utilizing the new 2011 terminology and classification, please note that it does not supplant the 2006 ISSVD Classification of Vulvar Dermatoses [1]. The purpose of this new 2011 terminology and classification is to assist the clinician in arriving at a diagnosis based solely on clinical findings, whereas the 2006 classification was to help the clinician arrive at a correct diagnosis (1) when a diagnosis based on clinical examination was not possible and (2) when the microscopic findings on biopsy could only be reported as a histological pattern rather than as a single specific diagnosis. The 2006 classification remains an important tool where a specific clinical and/or biopsy diagnosis is not possible.

**DEFINITIONS, DESCRIPTION, AND DIAGNOSIS OF VULVAR LESIONS**

The diagnosis of vulvar disorders depends upon clinical history, physical examination, and, in some instances, confirmation with laboratory evaluation. The focus of this classification was on examination and description involving the following 5 steps. All aspects described later on need to be considered in formulating the description of a lesion. If more than 1 lesion is present, the most prototypical lesion should be chosen for the purposes of description. (Note that all definitions related to the eczematous and lichenified diseases are separately placed in Discussion of Terms Related to Eczematous and Lichenified Diseases of this article.)

Step 1. Define the lesion by choosing 1 or more of the following nouns. Note that, although measurements
are given in some of the definitions, these are approximate and overlap may occur between “small” and “large” 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.** Small (<1.0 cm) area of color change; no elevation and no substance on palpation.

**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.** Small (<1.0 cm) elevated and palpable lesion.

**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 (<0.5 cm) fluid-filled blister; the fluid is clear. (See Discussion of Terms Related to Eczematous and Lichenified Diseases for the definition of terms relating to eczematous and lichenified disorders.)

**Step 2.** Choose appropriate adjectives to modify the noun(s) chosen previously.

**Color.** The color of the lesion will most often be red, white, brown, blue, gray, black, or skin colored. The color of any overlying crust or scale is ignored for the purpose of describing the color of the primary lesion. Skin-colored lesions are those that match the color of the surrounding normal skin. In the mucosal portion of the vulva, skin-colored lesions will be pink or red.

**Surface.** The surface can be smooth or rough on palpation; a rough surface is due to either crust or scale. Crust is composed of serum and sometimes blood. It is usually yellow although heme pigments (red, blue, or black) may be present. Scale is composed of keratin and is usually grey, white, or silver, but palpable roughness without color is also due to scale. The presence of crust indicates that there is some disruption of the underlying epithelial barrier layer. The presence of scale indicates a hyperproliferative response of the epidermis.

**Margination.** Margination represents the transition from normal skin to lesional skin. A sharply marginated (or well-circumscribed) lesion has an abrupt transition; a poorly marginated lesion has a more gradual transition.

**Configuration.** Configuration represents the shape of the lesion as seen from above. Most lesions are somewhat round but oval, linear, angular, and annular shapes may also occur. Serpiginous lesions have a wavy or gyrate peripheral border. Annular lesions are round or serpiginous (i.e., with an arcing wavy border) and have a peripheral border that is either more elevated than is the center or has a different color from that of the center of the lesion.

**Step 3.** Formulate a list of differential diagnoses.

Using the description as indicated in steps 1 and 2, one can place an unrecognized vulvar disease into 1 of the 8 disease groups found in the following ISSVD clinical classification of dermatological vulvar disorders. Then, using one’s own general medical knowledge, a short list of the most likely diagnoses (the differential diagnoses) can be created.

**Step 4.** Reduce the number of diagnoses in the list of differential diagnoses.

Using an appropriate textbook, one can read the brief sections on clinical morphology (clinical presentation) for each of the diseases on the list of differential diagnoses. This information, along with the patient’s history, generally allows one to determine the most likely diagnosis or, at the very least, reduce the list to 2 or 3 possibilities.

**Step 5.** Confirm a clinical diagnosis.

In some instances, it will be necessary to carry out laboratory testing to confirm a suspected diagnosis. This
Table 1. 2011 ISSVD Clinical Classification of Vulvar Dermatological Disorders

1) Skin-colored lesions

A. Skin-colored papules and nodules
1. Papillomatosis of the vestibule and medial labia minora (a normal finding; not a disease)
2. *Molluscum contagiosum*
3. Warts (HPV infection)
4. Scar
5. Vulvar intraepithelial neoplasia
6. Skin tag (acrochordon, fibroepithelial polyp)
7. Nevus (intradermal type)
8. Mucinous cysts of the vestibule and medial labia minora (may have yellow hue)
9. Epidermal cyst (syn. epidermoid cyst; epithelial cyst)
10. Mammary-like gland tumor (hidradenoma papilliferum)
11. Bartholin gland cyst and tumor
12. Syringoma
13. Basal cell carcinoma

B. Skin-colored plaques
1. Lichen simplex chronicus and other lichenified disease (see definitions in Discussion of Terms Related to Eczematous and Lichenified Diseases)
2. Vulvar intraepithelial neoplasia

2) Red lesions: patches and plaques

A. Eczematous and lichenified diseases (see definitions in Discussion of Terms Related to Eczematous and Lichenified Diseases)
1. Allergic contact dermatitis
2. Irritant contact dermatitis
3. Atopic dermatitis (rarely seen as a vulvar presentation)
4. Eczematous changes superimposed on other vulvar disorders
5. Diseases clinically mimicking eczematous disease (candidiasis, Hailey-Hailey disease, and extramammary Paget disease)
6. Lichen simplex chronicus (lichenification with no preceding skin lesions)
7. Lichenification superimposed on an underlying preceding pruritic disease

B. Red patches and plaques (no epithelial disruption)
1. Candidiasis
2. Psoriasis
3. Vulvar intraepithelial neoplasia
4. Lichen planus
5. Plasma cell (Zoon) vulvitis
6. Bacterial soft-tissue infection (cellulitis and early necrotizing fasciitis)
7. Extramammary Paget disease

C. Red lesions: papules and nodules

A. Red papules
1. Folliculitis
2. Wart (HPV infection)
3. Angiokeratoma
4. *M. contagiosum* (inflamed)
5. Hidradenitis suppurativa (early lesions)

B. Red nodules
1. Furuncles (“boils”)
2. Wart (HPV infection)
3. Prurigo nodularis
4. Vulvar intraepithelial neoplasia
5. *M. contagiosum* (inflamed)
6. Urethral caruncle and prolapse
7. Hidradenitis suppurativa
8. Mammary-like gland adenoma (hidradenoma papilliferum)
9. Inflamed epidermal cyst
10. Bartholin duct abscess
11. Squamous cell carcinoma
12. Melanoma (amelanotic type)

B. Skin-colored plaques
1. Lichen simplex chronicus and other lichenified disease
2. Vulvar intraepithelial neoplasia

3) Red lesions: papules and nodules

A. Red papules
1. Folliculitis
2. Wart (HPV infection)
3. Angiokeratoma
4. *M. contagiosum* (inflamed)
5. Hidradenitis suppurativa (early lesions)

B. Red nodules
1. Furuncles (“boils”)
2. Wart (HPV infection)
3. Prurigo nodularis
4. Vulvar intraepithelial neoplasia
5. *M. contagiosum* (inflamed)
6. Urethral caruncle and prolapse
7. Hidradenitis suppurativa
8. Mammary-like gland adenoma (hidradenoma papilliferum)
9. Inflamed epidermal cyst
10. Bartholin duct abscess
11. Squamous cell carcinoma
12. Melanoma (amelanotic type)

Table 1. (Continued)

4) White lesions

A. White papules and nodules
1. Fordyce spots (a normal finding; may sometimes have a yellow hue)
2. *M. contagiosum*
3. Wart
4. Scar
5. Vulvar intraepithelial neoplasia
6. Squamous cell carcinoma
7. Milium (pl. milia)
8. Epidermal cyst
9. Hailey-Hailey disease

B. White patches and plaques
1. Vitiligo
2. Lichen sclerosus
3. Postinflammatory hypopigmentation
4. Lichenified diseases (when the surface is moist—see definitions in Discussion of Terms Related to Eczematous and Lichenified Diseases)
5. Lichen planus
6. Vulvar intraepithelial neoplasia
7. Squamous cell carcinoma

5) Dark-colored (brown, blue, gray, or black) lesions

A. Dark-colored patches
1. Melanocytic nevus
2. Vulvar melanosis (vulvar lentiginosis)
3. Postinflammatory hyperpigmentation
4. Lichen planus
5. Acanthosis nigricans
6. Melanoma in situ

B. Dark-colored papules and nodules
1. Melanocytic nevus (includes those with clinical and/or histological atypia)
2. Warts (HPV infection)
3. Vulvar intraepithelial neoplasia
4. Seborrheic keratosis
5. Angiokeratoma (capillary angioma, cherry angioma)
6. Mammary-like gland adenoma (hidradenoma papilliferum)
7. Melanoma

6) Blisters

A. Vesicles and bullae
1. Herpesvirus infections (herpes simplex, herpes zoster)
2. Acute eczema (see definitions in Discussion of Terms Related to Eczematous and Lichenified Diseases)
3. Bullous lichen sclerosus
4. Lymphangioma circumscriptum (lymphangiectasia)
5. Immune blistering disorders (cicatricial pemphigoid, fixed drug eruption, Steven-Johnson syndrome, pemphigus)

B. Pustules
1. Candidiasis
2. Folliculitis

7) Erosions and ulcers

A. Erosions
1. Excoriations (see the disorders in group 2A)
2. Erosive lichen planus
3. Fissures arising on normal tissue (idiopathic, intercourse related)
4. Fissures arising on abnormal tissue (candidiasis, lichen simplex chronicus, psoriasis, Crohn disease, etc.)
5. Vulvar intraepithelial neoplasia, eroded variant
6. Ruptured vesicles, bullae and pustules (see all of the disorders listed in group 6 “blisters”)
7. Extramammary Paget disease
usually entails a biopsy for histological confirmation by a pathologist and/or a search for an infectious etiology. Note that, in some instances, the pathologist may not be able to offer a single best diagnosis and may, instead, report the presence of a histological pattern such as, for example, a spongiotic pattern, an acanthotic pattern, or an acantholytic pattern. The ISSVD has developed and published a classification for these patterns ("pathological subsets and their clinical correlates") [1]. The use of this information together with one’s clinical findings will then allow for correct diagnosis using clinical-pathological correlation.

**DISCUSSION OF TERMS RELATED TO ECZEMATOUS AND LICHENIFIED DISEASES**

Nomenclature regarding the eczematous and lichenified diseases is particularly confusing and controversial. For this reason, our committee thought that it would be informative for users of the 2011 ISSVD classification of vulvar dermatological disorders to indicate how we believe these terms should be defined and used.

**Eczema (Adj. Eczematous)**
The term eczema (pronounced as EK zee ma) classically refers to a group of inflammatory diseases that are clinically characterized by the presence of pruritic, poorly marginated red plaques with minor evidence of microvesiculation and/or, more frequently, subsequent surface disruption (see “surface disruption” later on). Histologically, the eczematous diseases are characterized by a “spongiotic pattern” [1]. Chronic forms of eczematous disease may develop scaling and/or lichenification. The term dermatitis is used as a synonym for eczema (e.g., atopic dermatitis and atopic eczema) or is misused as a nonspecific term to describe any inflammatory skin condition.

**Surface Disruption**
Visible evidence of surface (epidermal) disruption includes one or more of the following: weeping, crusting, microvesiculation, fissuring in the folds, and erosions most often occurring as a result of excoriation.

**Lichenification**
Lichenification develops as a result of chronic scratching and/or rubbing (the “itch-scratch cycle”) and mainly involves the cutaneous portion of the vulva. Lichenification is characterized clinically by a palpable thickening of the tissue and increased prominence of skin markings. Scale may or may not be detectable in vulvar lichenification. Lichenification may be bright red, dusky red, white, or skin colored in appearance; the white color occurs as a result of moisture retention in the thickened outer layer of the epidermis. Excoriations may, or may not, be present. Histologically, lichenification is
characterized by an “acanthotic pattern” [1]. Lichenification may arise from normal-appearing skin (“lichen simplex chronicus”) or may be superimposed on some other underlying dermatological disorder such as psoriasis, lichen sclerosus, lichen planus, etc.

**Lichen Simplex Chronicus**

The term *lichen simplex chronicus* is used when the lichenification (as described previously) develops on skin that had been previously normal in appearance and where underlying skin disorders have been excluded. It is characterized by the presence of the itch-scratch cycle wherein patients scratch and rub consciously, subconsciously, and/or in their sleep. Lichen simplex chronicus is especially likely to arise in those individuals who are atopic and/or those who have had atopic dermatitis elsewhere.

**Atopic Dermatitis (Syn. Atopic Eczema)**

Atopic dermatitis is a pruritic, chronic inflammatory disease that arises in previously normal-appearing skin. It is characterized clinically by red plaques with evidence of surface disruption (notably excoriations) occurring as a result of the itch-scratch cycle. The morphological phenotype of atopic dermatitis occurs mainly, but not exclusively, in “atopic” individuals, that is, those with a personal or immediate family history of allergic rhinitis, allergic conjunctivitis, and/or asthma. The pathophysiology of atopic dermatitis involves a genetic disturbance in epidermal barrier layer function, reduced skin-surface antimicrobial peptides, and abnormalities in immune response such as the development of allergen-specific immunoglobulin E antibodies, Th2 lymphocytic response, and elevated levels of various cytokines and chemokines.

**REFERENCES**