



CURRENT ISSVD TERMINOLOGY AND CLASSIFICATION OF VULVAR DISEASES

CLINICAL DIAGNOSIS - updated January 23, 2016

2011 ISSVD TERMINOLOGY AND CLASSIFICATION OF VULVAR DERMATOLOGICAL DISORDERS

The purpose of this classification is to assist clinicians in determining a differential diagnosis based on the morphology i.e. the clinical appearance of a lesion. Note that a single disorder may display multiple signs.

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 a yellow hue)
9. Epidermal cyst (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 (LSC) and other lichenified disease
2. Vulvar intraepithelial neoplasia

2) RED LESIONS: PATCHES AND PLAQUES

A. Eczematous & 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's disease)
6. Lichen simplex chronicus (lichenification with no preceding skin lesions)
7. Lichenification superimposed on an underlying preceding pruritic disease

B. Red patches & plaques (no epithelial disruption)

1. Candidiasis
2. Psoriasis
3. Vulvar intraepithelial neoplasia
4. Lichen planus
5. Plasma cell (Zoon's) vulvitis
6. Bacterial soft-tissue infection (cellulitis and early necrotizing fasciitis)
7. Extramammary Paget's disease

3) RED LESIONS: PAPULES AND NODULES

A. Red papules

1. Folliculitis
2. Wart (HPV infection)
3. Angiokeratoma
4. Molluscum contagiosum (inflamed)
5. Hidradenitis suppurativa (early lesions)
6. Hailey-Hailey disease

B. Red nodules

1. Furuncles (“boils”)
2. Wart (HPV infection)
3. Prurigo nodularis
4. Vulvar intraepithelial neoplasia
5. Molluscum 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)

4) WHITE LESIONS

A. White papules and nodules

1. Fordyce spots (a normal finding; may sometimes have a yellow hue)
2. Molluscum 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. Post-inflammatory hypopigmentation
4. Lichenified diseases (when the surface is moist)
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. Post-inflammatory 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 histologic 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 Part IV above)
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 (candidosis)
2. Folliculitis

7) EROSIONS AND ULCERS

A. Erosions

1. Excoriations
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's disease, etc.)
5. Vulvar intraepithelial neoplasia, eroded variant
6. Ruptured vesicles, bullae and pustules
7. Extramammary Paget's disease

B. Ulcers

1. Excoriations (related to eczema, lichen simplex chronicus)
2. Aphthous ulcers; syn. Aphthous minor, aphthous major, Lipschütz ulcer (occurring either as an idiopathic process or secondary to other diseases such as Crohn's, Behçet's, various viral infections)
3. Crohn's disease

4. Herpesvirus infection (particularly in immunosuppressed patients)
5. Ulcerated squamous cell carcinoma
6. Primary syphilis (chancre)

8) EDEMA (DIFFUSE GENITAL SWELLING)

A. Skin-colored edema

1. Crohn's disease
2. Idiopathic lymphatic abnormality (congenital Milroy's disease)
3. Post-radiation and post-surgical lymphatic obstruction
4. Post-infectious edema (esp. staphylococcal and streptococcal cellulitis)
5. Post-inflammatory edema (esp. hidradenitis suppurativa)

B. Pink or red edema

1. Venous obstruction (e.g., pregnancy, parturition)
2. Cellulitis (primary or superimposed on already existing edema)
3. Inflamed Bartholin duct cyst/abscess
4. Crohn's disease
5. Mild vulvar edema may occur with any inflammatory vulvar disease

Reference

- Lynch PJ, Moyal-Barracco M, Scurry J, Stockdale C. J Low Genit Tract Dis. 2012;16(4):339-44.