Vulvar Neoplasms, Benign and Malignant

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KEYWORDS

• Melanosis • Genital nevi • Differentiated vulvar intraepithelial neoplasia (dVIN) • High-grade squamous intraepithelial lesion (HSIL) • Squamous cell carcinoma (SCC) • Basal cell carcinoma • Vulvar melanoma • Paget disease

KEY POINTS

- Many pigmented vulvar lesions are indistinguishable clinically and so require biopsy to evaluate for early melanoma.
- Simple excision is curative for dysplastic nevi and atypical nevi of the genital type.
- High-grade squamous intraepithelial lesions (HSIL; formerly vulvar intraepithelial neoplasia [VIN] 2 or 3) can be treated conservatively with imiquimod. Nonresponsive lesions need biopsy to evaluate for occult invasion.
- Differentiated VIN (dVIN) is often missed clinically and histologically. Although only accounting for 5% of vulvar dysplasia diagnoses, dVIN has a higher rate of progression to squamous cell carcinoma (SCC), shorter time interval to progression, and higher recurrence rate than HSIL.
- Prognosis of vulvar SCC correlates with lymph node invasion, size of tumor, and depth of stromal invasion.

VULVAR PIGMENTED LESIONS

Melanoma comprises 10% of vulvar pigmented lesions and occurs in the sixth to seventh decade.[1–3] The clinical goal of evaluating vulvar pigmented lesions is focused on eliminating the possibility of melanoma, which can be clinically indistinguishable from other pigmented lesions.[4]

BENIGN PIGMENTED VULVAR LESIONS

Vulvar melanoma is detected late, with a resulting poor prognosis of less than 50% 5-year survival. Detecting cutaneous melanoma early is a key component to

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reducing melanoma mortality.[5] From 10% to 12% of all cutaneous pigmented lesions occur on genital skin.[2] Vulvar compound and junctional nevi are the most common pigmented lesions of the vulva, comprising 55% of pigmented vulvar lesions. Percentages of pigmented vulvar lesions are summarized in Table 1. The small size (5 mm or less) and uniform pigmentation distinguish common nevi from melanoma. Change in an otherwise typical nevus or a nevus that appears different from the patient's other nevi, the so-called ugly-duckling sign, warrant a biopsy. Stable nevi should be followed clinically and do not require treatment.

MELANOTIC MACULES

Melanotic macules are common benign pigmented lesions located on labia minora, usually restricted to nonkeratinized skin in adult women. Melanotic macules make up 17% of vulvar pigmented lesions, and most are uniformly pigmented and of small size. If a pigmented lesion appears consistent with melanotic macule but is larger than 6 mm, biopsy should be considered because it is impossible to distinguish larger melanotic macules (>6 mm) from early vulvar melanoma. Multiple melanotic macules are seen in genetic syndromes, such as LAMB (lentigines, atrial myxoma, mucocutaneous myxomas, blue

Table 1
Percentages of pigmented vulvar lesions

Vulvar Pigmented Lesions	Percentage	Average Age (y)	Clinical Appearance	Treatment
Common nevi	55	34	1–5 mm, flesh colored, tan to dark brown, flat to dome shaped	Not indicated unless concern for melanoma exists
Melanotic macule (lentigo)	17	43	5 mm–2 cm tan to dark brown to black, flat, without elevation	Not indicated unless concern for melanoma exists
Melanoma	10	>55	Erythematous to black, solitary to multifocal, usually 6 mm or greater	Excision, staging per TNM protocol
Atypical nevus of genital type	10	21	6 mm or larger, erythematous to tan to black, may be asymmetric	Conservative excision
Dysplastic nevus	5	32	6 mm or larger, erythematous to tan to black, may be asymmetric	Conservative excision
Blue nevus	Rare	Unknown	5 mm or smaller, dark blue to black, flat to dome shaped	Not indicated unless concern for melanoma exists
Recurrent nevi	Rare	Unknown	Size varies, asymmetric color, size, irregular shape	Conservative excision
Vulvar melanosis	Rare	Unknown	Flat, asymmetric, tan to blue to black, irregular borders and size	Baseline photos, biopsy, clinical follow-up

Abbreviation: TNM, tumor, node, metastasis.