Genital Lentiginosis: A Benign Pigmentary Abnormality Often Raising Concern for Melanoma

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PRACTICE **POINTS**

- The irregular appearance of genital lentiginosis—multifocal, asymmetric, irregular, and darkly pigmented patches—often raises concern for melanoma but is benign.
- Certain genetic conditions can present with genital lentiginosis.
- Dermoscopic assessment of the lesion color is highly helpful in narrowing the differential diagnosis; seeing no gray, red, blue, or white makes melanoma less likely.
- Be aware of genital lentigines and their characteristic presentation in adulthood to avoid unwarranted concern and unneeded surgery.

To the Editor:

Genital lentiginosis (also known as mucosal melanotic macules, vulvar melanosis, penile melanosis, and penile lentigines) occurs in men and women.[1] Lesions present in adult life as multifocal, asymmetrical, pigmented patches that can have a mottled appearance or exhibit skip areas. The irregular appearance of the pigmented areas often raises concern for melanoma. Biopsy reveals increased pigmentation along the basal layer of the epidermis; the irregular distribution of single melanocytes and pagetoid spread typical of melanoma in situ is not identified.

Genital lentiginosis usually occurs as an isolated finding; however, the condition can be a manifestation of Laugier-Hunziker syndrome, Carney complex, and Bannayan-Riley-Ruvalcaba syndrome.[1-3] When it occurs as an isolated finding, the patient can be reassured and treatment is unnecessary. Because genital lentiginosis may mimic the appearance of melanoma, it is important for physicians to differentiate the two and make a correct diagnosis. We present a case of genital lentiginosis that mimicked vulvar melanoma.

A 64-year-old woman was referred by her gynecologist to dermatology to rule out vulvar melanoma. The patient had a history of hypothyroidism and hypercholesterolemia but was otherwise in good health. Genital examination revealed asymptomatic pigmented macules and patches of unknown duration (Figure 1). Specimens were taken from 3 areas by punch biopsy to clarify the diagnosis. All 3 specimens showed identical features including basilar pigmentation, occasional melanophages in the papillary dermis, and no evidence of nests or pagetoid spread of atypical melanocytes (Figures 2 and 3). Histologic findings were diagnostic for genital lentiginosis. The patient was reassured, and no treatment was provided. At 6-month follow-up there was no change in clinical appearance.

Genital lentiginosis is characterized by brown lesions that can have a mottled appearance and often are associated with skip areas.[1] Lesions can be strikingly irregular and darkly pigmented.

Although the lesions of genital lentiginosis most often are isolated findings, they can be a clue to several uncommon syndromes such as autosomal-dominant Bannayan-Riley-Ruvalcaba syndrome, which is

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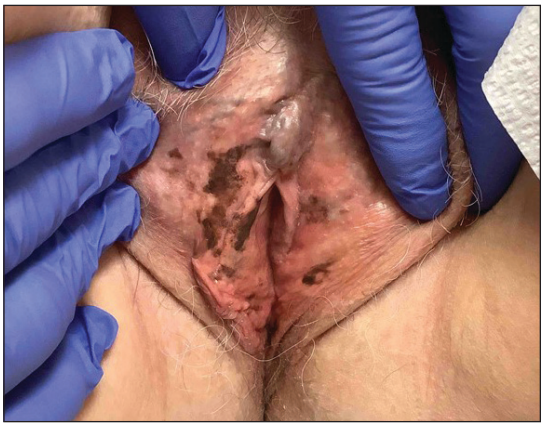

FIGURE 1. Asymmetric pigmented macules and patches of genital lentiginosis in the vulva.

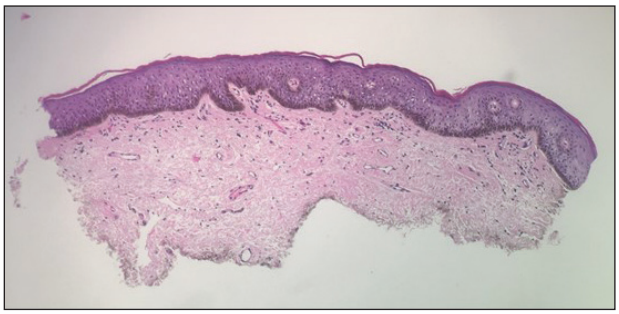

FIGURE 2. Histopathology revealed increased pigmentation limited to the dermoepidermal junction (H&E, original magnification ×100).

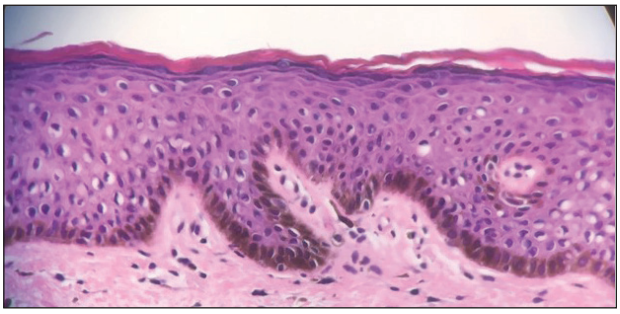

FIGURE 3. Although histopathology showed increased pigmentation, the number of melanocytes within the epidermis was not increased (H&E, original magnification ×200).

associated with genital lentiginosis, intestinal polyposis, and macrocephaly.[3] Vascular malformations, lipomatosis, verrucal keratoses, and acrochordons can occur. Bannayan-Riley-Ruvalcaba syndrome and Cowden syndrome may share genetic linkage; mutations in the tumor suppressor PTEN (phosphatase and tensin homolog deleted on chromosome ten) has been implicated in both syndromes.[4] Underlying Carney complex should be excluded when genital lentiginosis is encountered.

Genital lentiginosis is idiopathic in most instances, but reports of lesions occurring after annular lichen planus suggest a possible mechanism.[5] The disappearance of lentigines after imatinib therapy suggests a role for c-kit, a receptor tyrosine kinase that is involved in intracellular signaling, in some cases.[6] At times, lesions can simulate trichrome vitiligo or have a reticulate pattern.[7]

Men and women present at different points in the course of disease. Men often present with penile lesions 14 years after onset, on average; they notice a gradual increase in the size of lesions. Because women can have greater difficulty self-examining the genital region, they tend to present much later in the course but often within a few months after initial inspection.[1,8]

Genital lentiginosis can mimic melanoma with nonhomogeneous pigmentation, asymmetry, and unilateral distribution, which makes dermoscopic assessment of colors helpful in narrowing the differential diagnosis. Melanoma is associated with combinations of gray, red, blue, and white, which are not found in genital lentiginosis.[9]

Biopsy of a genital lentigo is diagnostic, distinguishing the lesion from melanoma—failing to reveal the atypical melanocytes and pagetoid spread characteristic of melanoma in situ. Histologic findings can cause diagnostic difficulties when concurrent lichen sclerosus is associated with genital lentigines or nevi.[10]

Lentigines on sun-damaged skin or in the setting of xeroderma pigmentosum have been associated with melanoma,[11-13] but genital lentigines are not considered a form of precancerous melanosis. In women, early diagnosis is important when there is concern for melanoma because the prognosis for vulvar melanoma is improved in thin lesions.[14]

Other entities in the differential include secondary syphilis, which commonly presents as macules and scaly papules and can be found on mucosal surfaces such as the oral cavity,[15] as well as Kaposi sarcoma, which is characterized by purplish, brown, or black macules, plaques, and nodules, more commonly in immunosuppressed patients.[16]

To avoid unwarranted concern and unnecessary surgery, dermatologists should be aware of genital lentigines and their characteristic presentation in adults.

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