An Amelanotic Malignant Melanoma of the Lip: Unusual Shape and Atypical Location

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Practice Points

- Amelanotic malignant melanoma is difficult to clinically diagnose because of the lack of melanin pigment typically found in melanomas.
- Amelanotic malignant melanoma generally occurs on the trunk and lower extremities; it rarely is located on the lip with various clinical features.
- The clinician should be aware of the possibility of melanoma, especially when an amelanotic nodular lesion is present on the lip.

Amelanotic malignant melanoma (AMM) is characterized by little or no visible pigment. The diagnosis of AMM is a challenge for clinicians because it is a rare entity that presents with various clinical features. We describe a case of AMM on the lower lip in a 63-year-woman, which manifested as an erythematous mass that resembled grouped papules.

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Amelanotic malignant melanoma (AMM) is a rare subtype of cutaneous melanoma, characterized by little or no visible pigment. Clinicians have difficulty diagnosing this lesion because its various clinical features tend to mimic a variety of other less serious diseases.[1] Amelanotic malignant melanomas often occur on the trunk and lower extremities, but any site on the body can be involved.[2,3] According to a PubMed search of articles indexed for MEDLINE using the medical subject headings (MeSH) *melanoma*, *amelanotic* and/or *lip*, there is only 1 report in the literature of AMM on the lip.[4] We describe a case of an atypical AMM on the lower lip.

Case Report

A 63-year-old woman presented with an erythematous mass with an uneven surface located on the lower lip of 1 year's duration without associated symptoms. The patient had been treated with intermittent steroid injections in a local clinic for several months and the lesion had been growing. Physical examination revealed a 2×2-cm erythematous mass that resembled grouped papules (Figure 1). On physical examination, there was no lymph node enlargement,

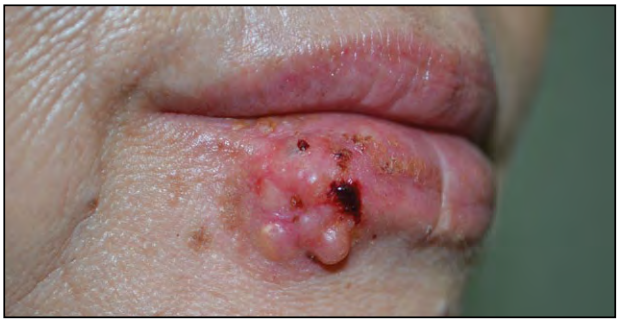

Figure 1. Well-demarcated erythematous nodule with crusting on the lower lip.

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and the patient had no remarkable medical or family history. Under suspicion of basal cell carcinoma, squamous cell carcinoma, sarcoidosis, and verruca vulgaris, a skin biopsy was taken from the nodule, which showed numerous nests comprised of atypical tumor cells with abundant mitoses in the dermis. The tumor cells had enlarged hyperchromatic nuclei with prominent nucleoli but no melanin pigment (Figures 2A and 2B). In an immunohistochemical study, the tumor cells stained positive for HMB-45 (human melanoma black) and S-100 protein (Figures 2C and 2D) but were negative for p63. From these findings, AMM was diagnosed. Under general anesthesia a wide local excision was performed followed by a skin graft. Histologic evaluation of the excised specimen was consistent with the AMM diagnosis and the Breslow thickness was 9 mm. Further evaluation including chest radiograph and whole-body positron emission tomography–computed tomography findings was unremarkable. The final diagnosis was AMM stage IIB (T4aN0M0) and the patient was transferred to the department of internal medicine for chemotherapy; however, she refused additional treatment and was lost to follow-up.

Comment

Amelanotic malignant melanoma is rare, accounting for 2% to 8% of all cutaneous melanomas seen by dermatologists.[3] The clinical presentation of AMM is variable, and a high index of suspicion is required for its diagnosis. Pizzichetta et al[2] suggested that clinical features such as peripheral pigmentation, ulceration, and asymmetry may help in the diagnosis of AMM; however, despite these efforts, AMM is still a diagnostic challenge for clinicians due to the absence of pigmentation. It often is misdiagnosed as verruca vulgaris, dermatofibroma, seborrheic keratosis, basal cell carcinoma, and squamous cell carcinoma.[3]

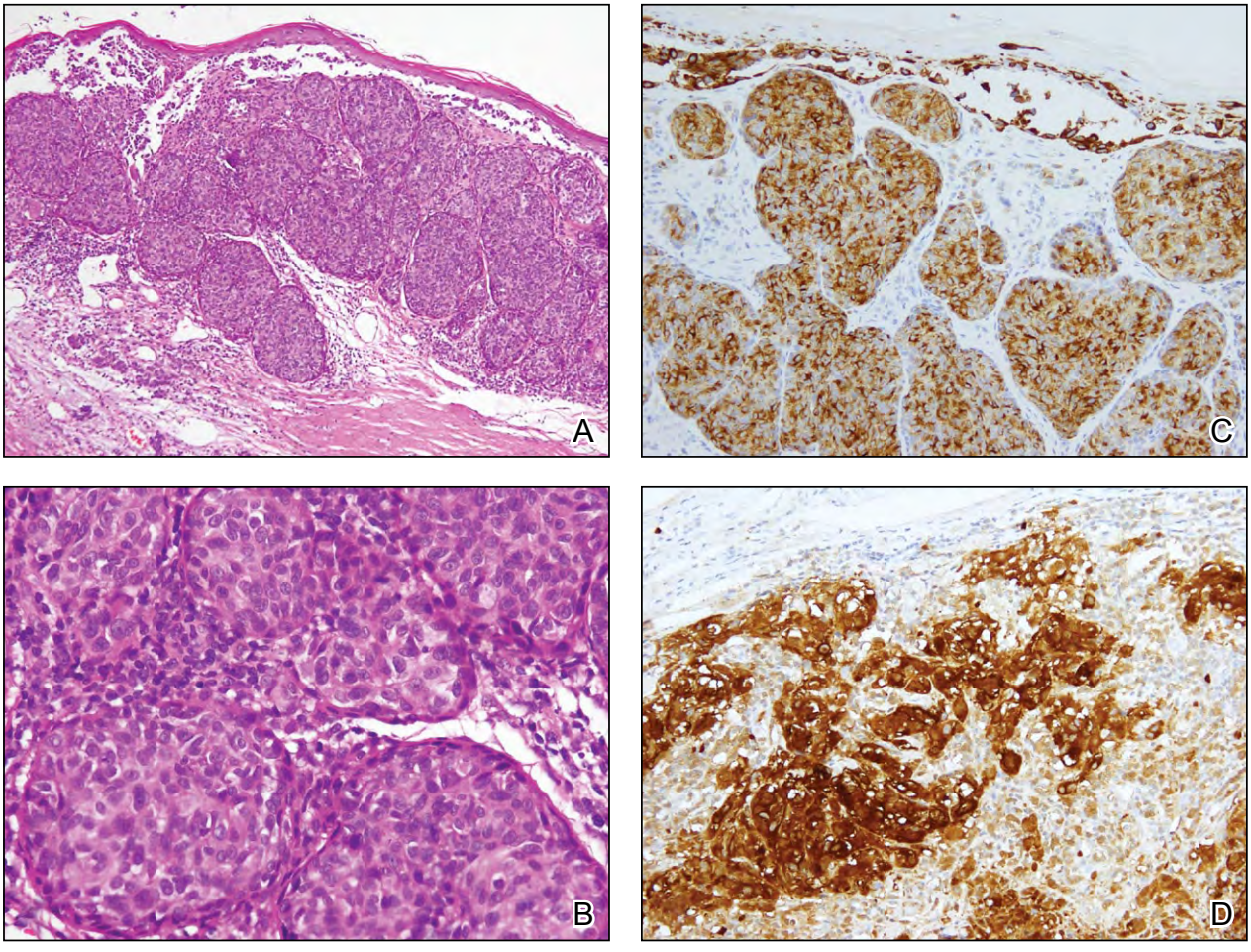

Figure 2. Histopathology showed a diffuse dermal nodule composed of atypical tumor cells devoid of pigment (A) (H&E, original magnification ×100). The tumor cells possessed enlarged hyperchromatic and pleomorphic nuclei (B) (H&E, original magnification ×400). Immunohistochemical staining was positive for HMB-45 (human melanoma black)(C) and S-100 protein (D)(both original magnifications ×200).

In a review of studies, AMM was reported to be most commonly located on the trunk and lower limbs.[2,3] Involvement of the lip is extremely rare, with 1 case in a patient with Rothmund-Thomson syndrome.[4] In our case we did not initially suspect AMM because of its rarity with unusual clinical features and distribution.

The histopathologic findings of AMM are similar to pigmented melanomas, except for the lack of pigmentation. Immunohistochemical staining including HMB-45 and S-100 protein is helpful in making an accurate diagnosis.[1,5] In our case, these histopathologic and immunohistochemical findings provided a diagnosis of AMM. Once AMM is diagnosed, treatment follows the same guidelines as pigmented melanomas.[1] However, AMM has a worse prognosis than pigmented melanomas, presumably because of the delay in diagnosis.[1-3]

Conclusion

Our case underlines the fact that physicians should be aware of the possibility of AMM, which is a great masquerader when encountering an amelanotic lesion on the lip.

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