Rhinophymatous Amelanotic Melanoma

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Amelanotic melanomas are well-known to mimic other dermatologic lesions and often result in delayed diagnosis and treatment. We report a case of an unusual presentation of amelanotic melanoma with an appearance similar to rhinophyma.

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Amelanotic melanoma represents a small percentage of the total number of melanomas that present yearly to physicians. These melanomas are well-known to mimic other dermatologic lesions, resulting in delayed diagnosis and delayed treatment.[1] We report a patient with an unusual presentation of amelanotic melanoma that had a rhinophymatouslike appearance. Despite an extensive literature search, we were unable to discover a similar presentation.

Case Report

A 65-year-old white man presented to the dermatology department for evaluation of a lesion on his nose. Three years prior, the patient had a black spot on the right side of his lower nose. The spot had been removed twice but had not been sent for pathologic evaluation. Six months prior to presentation, the patient stated that he developed scaly firm red bumps that began on the right side of his lower nose and subsequently spread to involve nearly his entire nose. He denied a history of skin disease, rosacea, rhinophyma, or skin cancer. He reported no pain, but the lesions had begun bleeding spontaneously over the past few weeks. He also noted difficulty breathing but did not experience weight loss, fever, chills, or night sweats. The patient's history was significant for 60 years of smoking. He had served as a firefighter for 42 years and was currently retired.

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Examination revealed 35 to 40 scaly erythematous papules and nodules that coalesced into a plaque involving the majority of the patient's nose. There were erosions in the central area of the lesion. The plaque extended onto both alae and almost to the base of the columella. Small areas on the bridge of his nose and the lateral aspects of both alae were spared. The appearance of the nose was strikingly similar to a rhinophyma (Figure 1). The preauricular and cervical lymph nodes were nonpalpable and nontender bilaterally. The working differential diagnosis at the time was sarcoid versus squamous cell carcinoma (SCC) or lupus vulgaris. Two incisional biopsies, one from the right nasal sidewall and the other from the tip of the nose, were submitted for dermatopathologic evaluation. The patient was then referred to his primary care provider for further assessment of his breathing difficulties.

Dermatopathologic evaluation revealed melanocytic proliferation with marked atypia. There was pagetoid spread throughout the epidermis and a lymphocytic reaction in the dermis. The neoplasm reached the superficial dermal margin but not the deep margins of the biopsy specimens, and invaded to a depth of

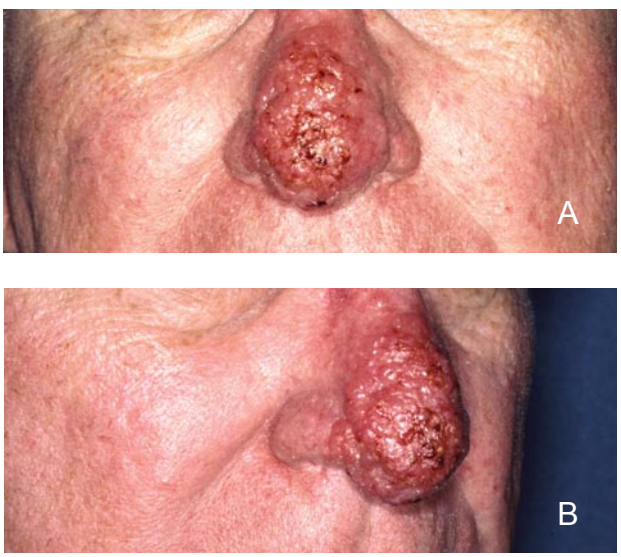

Figure 1. Erythematous papules and nodules that coalesced into a plaque with central erosions involving the majority of the patient's nose (A and B).

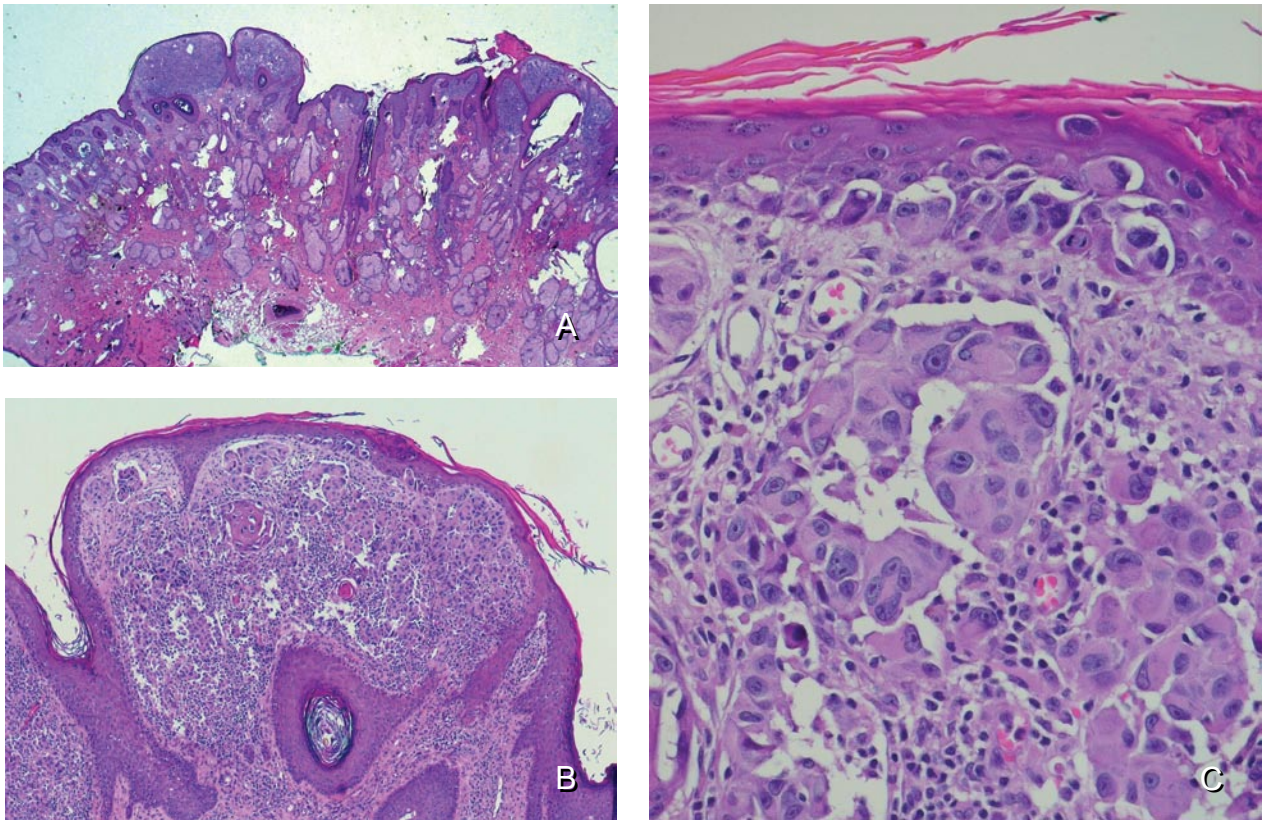

Figure 2. An extensive nodular malignant melanoma, Clark level IV, with a Breslow thickness of 2 mm. A vertical growth phase, a high mitotic index, and a brisk lymphocytic response were present. There were rare foci of microscopic epidermal ulceration but no definitive evidence of regression or satellitism. Also noted were epidermal pseudoepitheliomatous hyperplasia and pilosebaceous hyperplasia (A, B, and C)(H&E; original magnifications ×10, 40, and 200, respectively). Photographs courtesy of Cloyce Stetson, MD, Texas Tech University, Department of Dermatology, Lubbock.

1.5 mm. There was no ulceration or lymphovascular invasion. Of note was the presence of mild epithelial hyperplasia, numerous keratin cysts in the dermis, and moderate solar elastosis. The final pathologic diagnosis was a superficial spreading malignant melanoma, Clark level III, with a Breslow thickness of 1.5 mm and involvement of dermal margins.

During the course of the staging evaluation of the patient's melanoma, a mass was found in the patient's left lower lung. The mass was resected and determined to be a T2 N2 M0 SCC and nonmetastatic melanoma. After resection of the lung mass, the patient was referred to plastic surgery for removal of the melanoma. The patient underwent a partial rhinectomy; the lesion was resected with margins, while the bony cartilaginous framework of the nose was maintained. The tissue was sent for pathologic evaluation, and a full-thickness skin graft was obtained from the supraclavicular fossa and placed over the defect. Immediately after the nasal reconstruction, bilateral sentinel lymph node biopsies were taken of the upper jugular/submandibular areas.

The surgical pathologic examination revealed an extensive nodular malignant melanoma, Clark level IV, with a Breslow thickness of 2 mm (Figure 2). There was a vertical growth phase, a high mitotic index, and a brisk lymphocytic response. There were rare foci of microscopic epidermal ulceration. There was no definitive evidence of regression or satellitism. Also noted were epidermal pseudoepitheliomatous hyperplasia and pilosebaceous hyperplasia. The peripheral and deep margins were free of melanoma, with the nearest margin of 0.5 mm at the columella and right ala towards the columella. The pathologic results of the 2 sentinel lymph nodes obtained from the right and left upper jugular/submandibular areas were both free of metastatic melanoma on routine hematoxylin and eosin stain step sections and immunohistochemical stains for Melan-A. After more than 42 months of follow-up, the patient remains free of melanoma and continues to have quarterly appointments with the plastic surgery department (Figure 3).

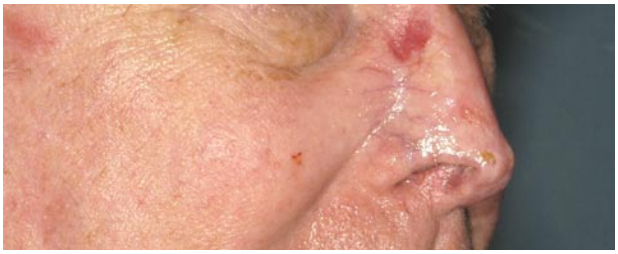

Figure 3. Six months postoperative, the full-thickness skin graft was well-healed, and the patient remains free of melanoma after more than 42 months of follow-up.

Comment

Amelanotic melanoma compromises 1.8% to 8.1% of all melanomas.[1] It can represent either a primary cancer on initial presentation, recurrence, or metastatic process. The name *amelanotic melanoma* is based on the clinical appearance of the lesion, as many amelanotic melanomas produce trace pigment that is often detectable with special immunohistochemical stains such as S100 and HMB-45.[1] One of the more commonly recognized clinical presentations of amelanotic melanoma is a lesion appearing on sun-exposed skin surfaces,[1,2] particularly the head and neck, with epidermal changes or skin-colored nodules or plaques. Another common presentation of amelanotic melanoma is an exophytic and often eroding nodule.[1] Features such as nonhealing[3] or delayed healing, ulceration, asymmetry, and brisk growth rate[4] also raise the suspicion of a malignant process. Early amelanotic melanomas, less than 1 mm thick or Clark level I, might present as asymmetric pink to red macules with either well-defined or ill-defined borders. These early lesions also might have a slight amount of pigmentation at the periphery. Although not pathognomonic for amelanotic melanoma, one study found that dermatoscopic evaluation of these early lesions demonstrated a pattern of red dots on a background of white, pink, or red. The small red dots, representing blood vessels running perpendicular to the skin surface, can facilitate an early diagnosis of amelanotic melanoma.[5] The majority of amelanotic melanomas have histologic features similar to pigmented melanomas; however, electron microscopy has shown that that the melanosomes of amelanotic melanomas often are immature.[1]

The time from the appearance of these lesions to diagnosis often is extended. These melanomas contain trace amounts to no visible pigment, enabling them to escape clinical suspicion. One study investigating 77 cases of amelanotic melanoma found that the average time from development of the lesion to diagnosis was 13 months.[6] One of the reasons for delay was misdiagnosis.[7] Amelanotic melanoma has long been confused with other lesions, both benign and malignant, earning the nickname "The Great Pretender." Other authors reported instances where these melanomas have been confused with basal cell carcinoma, Bowen disease, keratoacanthoma, intradermal nevus, nevus depigmentosus, verruca vulgaris, pyogenic granuloma, Merkel cell carcinoma, actinic keratosis, seborrheic keratosis, dermatitis, scar tissue, granuloma annulare,[8] SCC, extramammary Paget disease,[3] verruca plantaris,[5] eczema,[1] and inflammatory plaques.[2] Other reasons for prolongation of diagnosis of amelanotic melanoma include inappropriate treatment with laser vaporization, cautery, cryosurgery,[5] or curettage,[3] which cannot provide tissue for pathologic evaluation, and treatment with various ointments or escharotic substances. Most amelanotic melanomas present as advanced tumors (Clark level IV or V),[6] which may be due to the often delayed diagnosis.[9] However, it is believed that amelanotic melanoma does not carry a worse prognosis compared with other melanomas.[1] Treatment guidelines and recommendations that apply to other malignant melanomas also apply to the amelanotic variant.[1,3]

Conclusion

We report an unusual case of amelanotic melanoma with an appearance similar to rhinophyma. Of note, our patient showed classic features of amelanotic melanoma, including a brisk growth rate (with development of a rhinophymatouslike lesion in 6 months) and ulceration. We report this case for its clinical interest and to emphasize that physicians should have a high index of suspicion and a low threshold for performing a biopsy on atypical cases to confirm a diagnosis of amelanotic melanoma.[1]

REFERENCES

1. Adler MJ, White CR. Amelanotic malignant melanoma. *Semin Cutan Med Surg*. 1997;16:122-130.
2. Tschen JA, Fordice DH, Reddick M, et al. Amelanotic melanoma presenting as inflammatory plaques. *J Am Acad Dermatol*. 1992;27:464-465.
3. Roseeuw D. The invisible melanoma. *J Eur Acad Dermatol Venereol*. 2001;15:506-507.
4. Carli P, Massi D, de Giorgi V, et al. Clinically and dermatoscopically featureless melanoma: when prevention fails. *J Am Acad Dermatol*. 2002;46:957-959.

5. Bono A, Maurichi A, Moglia D, et al. Clinical and dermatoscopic diagnosis of early amelanotic melanoma. *Melanoma Res*. 2001;11:491-494.
6. Ariel IM. Amelanotic melanomas: an analysis of 77 patients. *Curr Surg*. 1981;38:151-155.
7. Anderson WK, Silvers DN. Melanoma? It can't be melanoma! *JAMA*. 1991;266:3463-3465.
8. Koch SE, Lange JR. Amelanotic melanoma: the great masquerader. *J Am Acad Dermatol*. 2000;42:731-734.
9. Rivers JK. Unusual melanoma types. *Dermatol Clin*. 2002;20:727-733.