Recurrent amelanotic lentigo maligna melanoma: a case report[1]

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Amelanotic melanoma occurs in about 2% of melanoma patients. It displays a slight male predominance and appears in the head and neck area. The average age of onset is about 47 years. Amelanotic lentigo maligna melanoma (ALMM) is a rare variety of amelanotic melanoma. A profile of the patient at risk for ALMM is offered, based on this case and published reports. ALMM shows a marked female predominance with an average age of onset of 62. Sites of involvement include the face, upper body, and upper extremities. Histologic evaluation of persistent pruritic erythematous lesions is suggested in patients fitting the ALMM risk profile.
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Melanoma is a malignant tumor consisting of melanocytes and is often lethal. Although prognosis is statistically predictable, in any given case it may be uncertain. Recently, the medical and lay press have expressed increased interest in this tumor. Melanoma represents 1% of all cancer deaths and 1.8% of all new cancers, except for amelanotic skin carcinoma.[1] Studies by Magnus[2] and Elwood and Lee[3] indicate that the incidence of melanoma is rising and that mortality is increasing at a rate of 3%–9% a year. In addition, the tumor is occurring in a younger age group with rising morbidity and mortality.

Most melanomas occur initially on the skin. As many as 88% of these tumors can be identified as superficial spreading melanomas (60%–70%), nodular melanomas (12%–16%), or lentigo maligna melanomas (5%–10%), in accordance with the classification established in 1973.[4] Lentigo maligna and lentigo maligna melanoma have been characterized as small freckle-like lesions occurring on sun-exposed areas, primarily on the neck and face, and gradually expand with time. The average age of onset is 47 years.[5] According to Clark et al,[6] the lesion occurs nearly equally in men and women.

Case report

A 57-year-old white woman of Celtic extraction was seen in 1978 for treatment of a recurrent melanoma on the left anterior chest wall. The patient first noted an oval red patch surrounded by a white halo on her chest in 1969. After several years, the lesion expanded laterally and became pruritic. She denied having had a pigmented lesion on the site previously; clinical examination failed to reveal pigmentation. The malignancy was excised with wide margins, and the resultant defect was closed by a skin graft in 1975. The lesion was characterized histologically as a lentigo maligna melanoma by the Armed Forces Institute of Pathology. In 1976, small asymptomatic erythematous macules appeared on the superior and inferior borders around the skin graft. The lesions slowly expanded across the graft and peripherally. After two years, the patient sought further care and was referred to the Department of Dermatology at the Cleveland Clinic. At this time, the lesion was clinically amelanotic. Examination with magnifying loupes showed no evidence of pigmentation.

The graft and involved borders (*Fig. 1*) were removed with a wide excision. The excised tissue showed marked atypical melanocytic hyperplasia at the dermal-epidermal junction (*Figs. 2 and 3*) with focal invasion of atypical melanocytes into the papillary dermis (*Fig. 4*) surrounded by a marked lymphocytic host response. The tumor was classified as Clark's II and Breslow's level of invasion, 0.16 mm. Following surgery, a metastatic work-up was performed.

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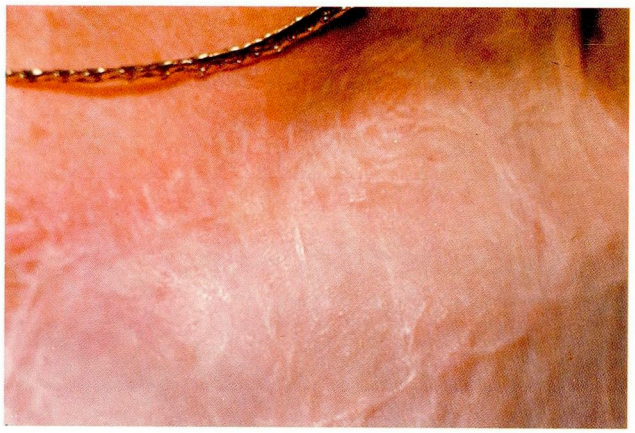

Figure 1. Erythematous macular lesion at the margin of the skin graft, representing recurrent amelanotic lentigo maligna melanoma.

The sternal bone marrow was free of melanoma. Computed tomographic scans of the head, liver, and spleen, as well as a bone scan and chest roentgenogram, were normal. Serum protein electrophoresis, a complete blood count, an electrolyte profile, a blood chemistry screen, quantitative measurement of immunoglobulins, urinalysis, and T and B cell counts were within normal limits. The delayed hypersensitivity response was normal. A subsequent follow-up evaluation did not reveal tumor recurrence.

Discussion

Amelanotic melanoma is an uncommon entity and occurs in about 2% of all melanoma patients. Cochran[7] could detect no differences in the prognosis between melanotic and amelanotic melanomas. Huvos et al[8] believed that the prognosis was worse for amelanotic melanomas and that the tumor was more aggressive when compared to the pigmented variety of the same level and stage. They speculated that the amelanotic tumor was a more anaplastic form of melanoma or that nonpigmentation prevented early detection, and hence, early intervention.

Huvos et al[8] showed that a predominant number of amelanotic melanoma patients are women. In 1981, Ariel[9] reviewed 77 cases of amelanotic melanomas occurring in 3305 melanoma patients from 1935 to 1967 and showed that the tumors occurred in two thirds as many women as men. Also, the combined average age of the patients was 44 years. Nearly one half of the amelanotic tumors were on the head and neck. Most of the remaining malignancies were on the trunk and lower extremities; fewer than 10% of the tumors occurred on the upper extremities. The lesions were all nodular and many had ulcerations. Pruritus was a common feature and often caused the afflicted individual to seek medical care. The study further suggested increased aggressiveness of amelanotic melanoma based upon higher rates of metastases and poor survival rates.

Amelanotic lentigo maligna and amelanotic

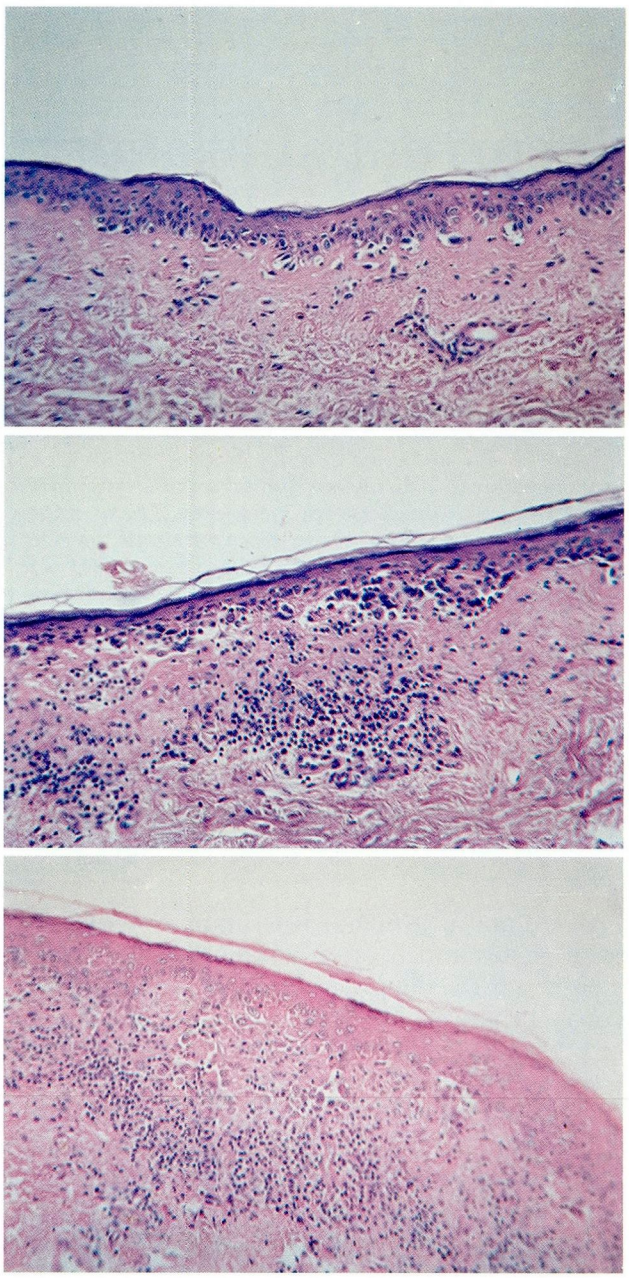

Figure 2. Atypical hyperchromatic melanocytes along the dermal-epidermal junction in a lentigo maligna pattern (hematoxylin and eosin stain, ×20).

Figure 3. Atypical melanocytes singly and in junctional nests with moderate lymphocytic dermal reaction consistent with premelanoma (hematoxylin and eosin stain, ×20).

Figure 4. Invasion of the papillary dermis by atypical cells breaking through the dermal-epidermal junction, with marked host response consistent with lentigo maligna melanoma (hematoxylin and eosin stain, ×20).

lentigo maligna melanoma are rare. In 1967, Mishima[10] proposed a dual developmental route to explain the differences in behavior between lentigo maligna melanoma and the other forms of melanoma. He believed that the amelanotic counterpart for lentigo maligna melanoma did not exist. Burket[11] described a case of amelanotic lentigo maligna melanoma in 1979; Su and Bradley[12] probably reported the first case of amelanotic lentigo maligna in the English literature in 1980. In both cases, the malignancies occurred in areas where pigmented lesions had previously been removed. Paver et al[13] reported three cases of primary amelanotic lentigo maligna thought initially to be nevus depigmentosus, superficial basal-cell carcinoma, and nonspecific dermatitis, respectively. The authors further summarized available case reports. Since then, an additional case has been reported by Lewis[14] that represents the second report of an amelanotic lentigo maligna melanoma in the literature.

After compiling the data from our case of recurrent amelanotic lentigo maligna melanoma with the previously reported cases of lentigo maligna and lentigo maligna melanoma (amelanotic types), a crude profile can be constructed. Of the 10 patients, only 2 were men (both in their seventies); the lesions were located on the scrotum and had been present for three and eight years, respectively. Prior to biopsy, extramammary Paget's disease was suspected. In 2 of the 8 women, the lesions were located on the chin or cheek; in 3, on the left arm; and in one each, on the leg, neck, and chest. The average age of the women was 62 years when a diagnosis was made. Duration of lesions before biopsy was classified as (1) amelanotic lesions resembling Bowen's disease, basal-cell carcinoma, or other premalignant diagnoses present only 2–3 years prior to accurate diagnosis; and (2) malignancies ressembling actinic damage, neurodermatitis, or other inflammatory conditions present 7–12 years before diagnosis.

Conclusion

Although rare, amelanotic lentigo maligna and amelanotic lentigo seem to occur predominantly on the trunk and upper extremities of women who are slightly older than those afflicted with the pigmented counterparts. Because these tumors can mimic a number of entities, persistent erythematous pruritic areas should be examined with caution and a histologic evaluation should be obtained if malignancy is suspected.

Addendum

Following submission of this article to the *Quarterly*, another case of amelanotic lentigo melanoma was seen by Borkovic and Schwartz on the right shoulder of a 72-year-old woman.[15]

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