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Amelanotic Melanoma of the Palate: Report of Case

Yadranko Ducic, MD, FRCS and D. Allen Pulsipher, DDS, MD†*

Malignant melanoma mainly arises in the skin. It represents the leading cause of death from cutaneous malignancy in the United States today.[1,2] Fewer than 1% of all reported primary melanomas arise in the oral cavity.[3,4] The cutaneous form of this lesion normally is highly visible, with irregular borders and dark pigmentation. However, mucosal melanomas may present a diagnostic challenge for the clinician.

Rarely, melanoma may present itself without clinically evident pigmentation. Termed amelanotic melanoma, these lesions tend to have a worse prognosis because of delayed recognition and subsequent treatment.[5] However, the prognosis may improve with early detection and wide local excision.[6,7] Metastases from amelanotic melanomas have similar characteristics to their primary counterparts; they also lack pigmentation and grow rapidly. In a study of amelanotic melanoma by Huvos et al,[5] the primary site was never identified in 29% of the patients. To our knowledge, amelantotic melanoma of the oral cavity has not been reported previously.

Report of Case

A 68-year-old Caucasian female presented for evaluation of a 2-cm nodular lesion on the right soft palate found on

*Assistant Professor, Department of Otolaryngology, University of Texas Southwestern Medical Center, Dallas, TX and the Director of Otolaryngology and Facial Plastic Surgery, John Peter Smith Hospital, Fort Worth, TX.

†Chief Resident, Division of Oral and Maxillofacial Surgery, University of Texas Southwestern Medical Center, Dallas, TX.

Address correspondence and reprint requests to Dr Ducic: Otolaryngology and Facial Plastic Surgery, 1500 South Main St, Fort Worth, TX 76104; e-mail: yducic@aol.com.

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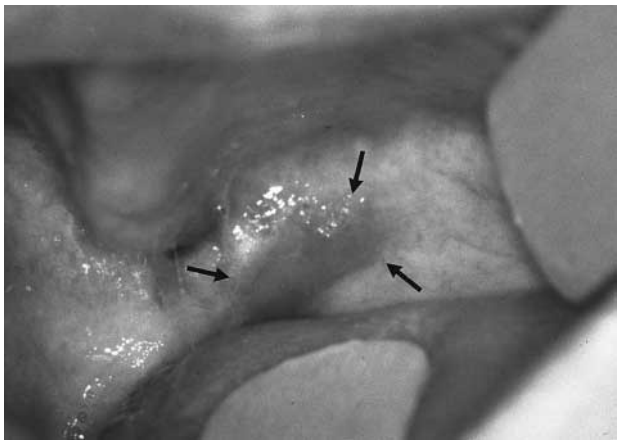

FIGURE 1. Erythematous, slightly raised, nonulcerated, nonpigmented lesion of right palate (*arrows*).

routine dental examination (Fig 1). The lesion had not been noted previously during an annual dental examination 12 months before presentation. Her past medical history was significant for a stroke, which resulted in left hemiplegia. There were no other significant medical problems. The patient had lost all of her teeth from periodontal disease; she had been wearing complete upper and lower dentures for the preceding 15 years.

One week after discovery of the lesion by the referring dentist, clinical examination revealed a nontender pink nodule measuring 2 cm in diameter on the right soft palate-hard palate junction. The head and neck examination revealed neither lymphadenopathy nor any other discernible lesions. Soft palate function was normal; there was no anesthesia or paresthesia in the area, and a neurosensory examination revealed intact responses.

An incisional biopsy was performed by the referring dentist. On clinical examination, there was no evidence of pigmentation in the specimen. Microscopic examination revealed a spindle cell-laden, fascicular growth pattern with ovoid, markedly pleomorphic cells with an increased nuclear-cytoplasmic ratio, nuclear pseudoinclusions, and paranuclear clear zones (Figs 2, 3). Both HMB-45 and S-100 immunohistochemistry revealed positive reactivity in the tumor cells. Rare foci of melanin pigmentation were evident in isolated cells.

Imaging studies were performed to rule out metastases and to determine the superior extent of the lesion. There were no other sites noted on magnetic resonance imaging of the head and neck, and the lesion was limited to the mucosal area of the soft palate. Computed tomography scans of the head, chest, and abdomen were within normal limits.

The patient was taken to the operating room for surgical excision. To provide for an adequate margin of 2 cm, an infrastructure hemimaxillectomy was performed in the usual manner via a lateral rhinotomy access incision. A split thickness skin graft was used to reconstruct the intraoral aspect of the cheek defect, providing a stable site for subsequent prosthetic rehabilitation. Gauze packing was placed in the defect to hold the graft in position. This was stabilized with the patient's maxillary denture secured to the remaining premaxillary bone with 1.5-mm titanium lag screws. The patient did well postoperatively and has undergone successful prosthetic rehabilitation. The surgical margins were clear of disease, and the patient remains disease-free at 1 year.

Discussion

The incidence of malignant melanoma has been increasing over the last 20 to 30 years.[8] Although the exact reason for this increase is unknown, it may be related to increased ultraviolet light exposure. Less than 1% of all malignant melanomas are found in the oral cavity. These occur primarily on the soft and hard palates. Because oral cavity mel-

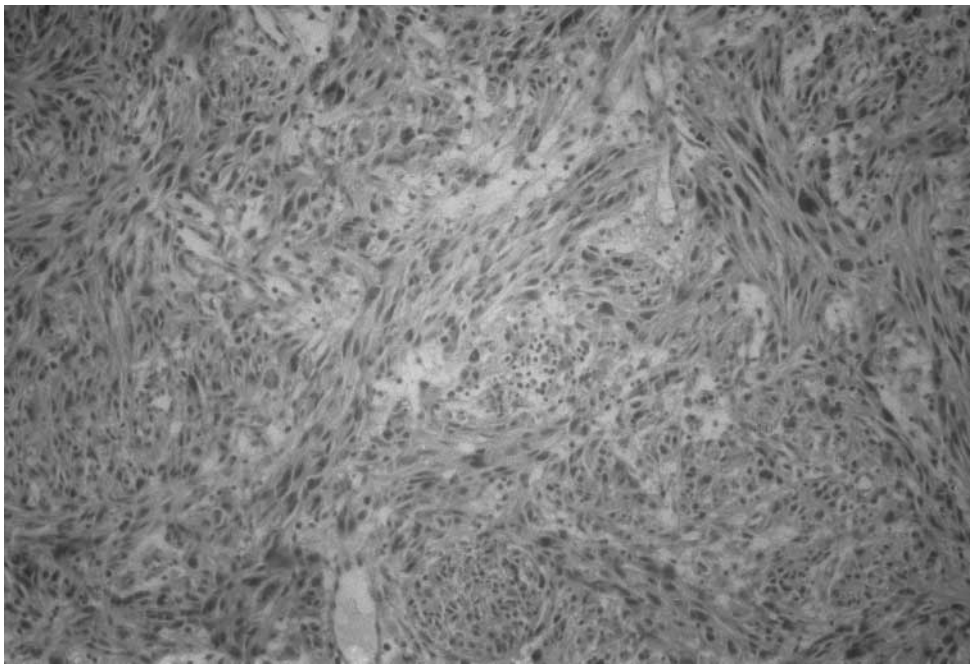

FIGURE 2. Photomicrograph showing large numbers of pleomorphic spindle-shaped cells exhibiting a fascicular growth pattern. (Hematoxylin and eosin stain, original magnification ×25.)

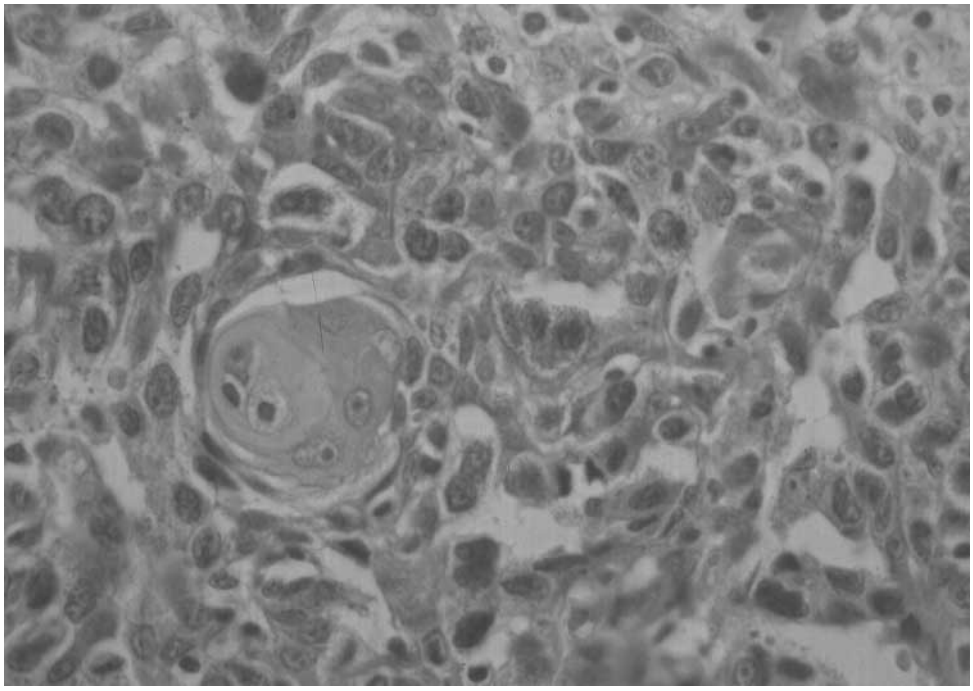

FIGURE 3. Photomicrograph at higher magnification demonstrating rare scant melanin pigment and marked cellular pleomorphism. (Hematoxylin and eosin stain, original magnification ×100.)

anomas are exceedingly rare, it is difficult to determine the optimum method of staging, prognosis, and treatment. Although statistics regarding cutaneous melanomas may not be directly applicable to oral cavity melanomas, they do serve as a useful starting point in treatment plan formulation, as with other well-described tumors occurring in unusual locations. Using Clark's microstaging criteria[9] and the expanded 4-stage staging system for melanoma,[10] prognosis and treatment criteria for these lesions may be established. Simple biopsy or curettage have shown recurrence rates of 60% to 70%.[11,12] Therefore, obtaining clear surgical margins has been shown to be necessary. Guidelines have been established for recommended excisional margins for melanomas of various thicknesses.[13,14] Melanoma in situ should have a 0.5- to 1.0-cm margin; thin melanomas (<0.76 mm) should have a 1- to 2-cm margin; and intermediate and thick lesions (>0.76 mm) should have a 2- to 3-cm margin. Such recommendations should serve as a guide when treating oral mucosal melanomas until more data have been gathered on this subtype.

Amelanotic melanoma is a rare variant of malignant melanoma comprising 2% to 8% of all malignant melanomas.[6,15,16] The primary tumor normally presents as a vascular or ulcerated nodule. However, most lesions represent metastatic disease from another primary site.

The specific cause for the lack of melanotic pigmentation in these lesions is unclear. Speece et al[17] propose that there is a deficiency in tyrosine and an enzyme required for melanin production. Others believe that this enzyme system is intact and can produce melanin, but the quantity is insufficient to be seen with histologic methods.[6,7] We favor this latter theory because electron microscopy has revealed the presence of melanosomes in all amelanotic melanomas examined to date.

The prognosis of amelanotic melanoma is much worse than that with the more common, grossly pigmented type. Retrospective studies have shown that stage I amelanotic melanoma patients have a 71% 5-year survival[5] and a 55% 10-year survival,[16] compared with stage I pigmented malignant melanoma patients, who have 80% 5-year and 70% 10-year survival.[16] Patients with stage II amelanotic melanoma have a 15% 5-year survival[5] and 17% 10-year survival.[16] Malignant pigmented melanoma has a substantially better prognosis, with a 40% 5-year survival and a 20% 10-year survival. Prognosis for stage III disease is similar, with both variants demonstrating a 0% to 2% survival at 5 years. The difference in prognosis between pigmented and amelanotic melanomas may not be biologically based. We believe that such statistics may be influenced by delays in timely diagnosis and treatment due to lack of clinical suspicion or recognition.

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