Primary Oral Melanoma: The Great Masquerader

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Background

Primary oral melanomas are extremely rare and have a high propensity for nodal metastasis and poor survival. The most common locations include palate, maxillary gingiva and mandibular retromolar trigone. Variable appearance and absence of early signs and symptoms often complicates timely diagnosis and worsens prognosis. The likelihood of misdiagnosis and/or delayed diagnosis is compounded for amelanotic melanoma - a variant without classic pigmentation for which diagnosis relies on immunohistochemical staining. We report a patient in whom oral melanoma had distinct amelanotic and discolored components and resembled a reactive lesion.

Case Presentation

A 45-year-old white male presented to the oral medicine service for the evaluation of an asymptomatic lesion of four-months duration in his lower left jaw. He was healthy and taking no medications. Extraoral exam was negative for lymphadenopathy. Notable intraorally was an exophytic gingival mass associated with buccal and distal aspect of tooth #18. The buccal growth had normal mucosal color, slightly granular texture and spongy consistency while the distal component was reddish-grey and firm on palpation. A patchy light-brown discoloration was also present on lingual attached gingiva of teeth #18 to #21. Clinical and radiographic examination revealed a potential plaque trap on distal of tooth #19 supporting the initial clinical impression of a reactive growth possibly related to local factors. However, color variegation and indurated nature of the posterior growth were atypical and suspicious for a potentially malignant process.

Treatment and Management

The pink buccal mass biopsied and microscopic examination revealed polygonal malignant cells with variable cytoplasmic volume, pleomorphic and occasionally hyperchromatic nuclei, multiple prominent nucleoli and abnormal mitotic figures. Immunohistochemical staining showed positivity for Melan A, HMB45 and AE1/AE3 diagnosing the malignancy as mucosal melanoma. The patient underwent oncology work up, left mandibulectomy and neck dissection by the head and neck team. Final pathology confirmed primary oral melanoma with nodal metastasis (PT3N1) for which he received adjuvant radiotherapy. The patient was, later, found to have metastatic spread to multiple organs and, in spite of the start of immunotherapy, succumb to death.

Clinical Presentation

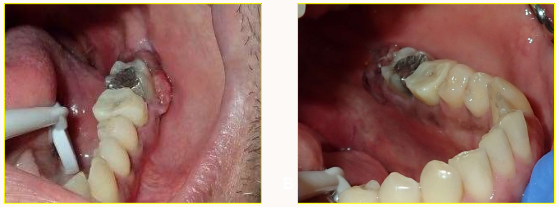

Figure 1: Clinical presentation of A) an exophytic gingival mass associated with buccal and distal aspect of tooth #18.The buccal growth had normal mucosal color & slightly granular texture while distal component was reddish-grey; B) A patchy light-brown discoloration was present on lingual attached gingiva associated with teeth #18 to #21.

Radiographic Presentation

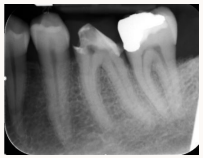

Figure 2: Periapical radiograph of the lesion area was essentially within normal limits only illustrating a potential trap for bacteria and plaque between teeth 18 & 19.

Histopathological Photograph

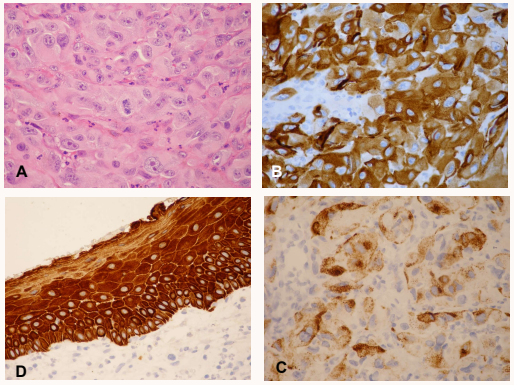

Figure 3: Histopathology micrograph of the biopsy specimen illustrating A) polygonal malignant cells with variable cytoplasmic volume, pleomorphic & occasionally hyperchromatic nuclei, multiple prominent nucleoli & abnormal mitotic figures; B) strong diffuse positive immunohistochemical staining for Melan A; C) focal positive immunohistochemical staining for HMB45; D) focal light immunohistochemical staining for AE1/AE3. These features lead to the diagnosis of malignancy as mucosal melanoma.

Discussion

Nature: aggressive malignancy with very poor prognosis with 5-Y survival rate of about 30%

Incidence: Rare (0.5% of all oral malignancies, 2% of all melanomas, ¼ of all mucosal H&N melanomas)

Racial prevalence: Higher prevalence in Japanese, AA and AI than whites

Age & gender predilection: 5th to 7th decades of life with a slight male predilection

Putative risk Factors: None except the preexistence of a mucosal field of melanin hyperpigmentation (30% develop within physiological hyperpigmentation)

Clinical Presentation: A macule or exophytic growth with brown black color and irregular borders ; most often on palate & maxillary gingiva (80%) followed by retromolar trigone in mandible

Imaging: May reveal irregular destruction; "moth eaten appearance"; MRI generally preferable to CT imaging for work up process.

Treatment: Surgical intervention +/- radiotherapy are mainstay of therapy ; efficacy of targeted chemo/immunotherapy for oral melanoma not studied.

Prognosis: Worse for exophytic ulcerated lesions with >6mm thickness, neck or distant metastasis and high mitotic rate. ; distant or regional metastasis noted in about 10% and 25% of cases at diagnosis, respectively.

Amelanotic Variant: 10-30% of oral melanomas are amelanotic characterized by the absence of melanin both clinically and microscopically. Therefore; diagnosis relies on IHC staining for a number of biomarkers. This atypical amelanotic appearance often leads to mis-/delayed diagnosis which may explain its worse prognosis. Some evidence suggests it has a higher mitotic rate & growth than pigmented melanomas.

Male gender of this patient is consistent with prior literature but he was younger than typical age and was of a race different from Japanese, AA or AI. Clinically, his lesion was biphasic in color. The non -pigmented aspect of the lesion biopsied had malignant microscopic features on H&E but no melanin. IHC staining lead to melanoma diagnosis. The patient was aware of the exophytic growth, which bled on brushing, for 4 months. Microscopically, the lesion surface was also ulcerated. These features tend to increase risk of metastasis as was the case for this patient.

Conclusion

Although rare, dental providers should be familiar with the epidemiology, site predilection and variable presentations of melanoma in the oral cavity and include it in the differential diagnosis of gingival growths even when the classic pigmentation is absent.

References

1. *Ascierto PA, Accorona R, Botti G, Farina D, Fossati P, Gatta G, Gogas H, Lombardi D, Maroldi R, Nicolai P, Ravanelli M, Vanella V. Mucosal melanoma of the head and neck. Crit Rev Oncol Hematol. 2017 Apr 112:136-152. doi: 10.1016/j.critrevonc.2017.01.019.*
2. *Feller L, Khammissa R AG, Lemmer J. 2017. A Review of the Aetiopathogenesis and Clinical and Hisopathological Features of Oral Mucosal Melanoma. The Scientific World Journal. 2017; https://doi.org/10.1155/2017/919812.*
3. *Neville B, Damm D, Allen C, Chi A. 2016. Oral and Maxillofacial Pathology. 4th Edition. St. Louis (MO): Elsevier.*
4. *Thomas N, Kricker A, Waxweiler W, Dillon P, Busam K, From L, Groben P, Armstrong B, Anton-Culver H, Gruber S et al. 2014. Comparison of Clinicopathologic Features and Survival of Histopathologically Amelanotic and Pigmented Melanomas A Population-Based Study. JAMA Dermatol. 2014;150(12):1306-1314. doi:10.1001/jamadermatol.2014.1348*