Primary Mucosal Melanoma of Maxilla: A Case Report and Review of Literature

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Oral malignant melanomas are extremely rare lesions and occur commonly in the maxillary gingiva more frequently on the palate with fewer incidences in the mandibular gingiva. Malignant melanomas of oral cavity are extremely rare accounting for 0.2-8% of all malignant melanomas. A 51-year-old female reported a growth about left maxillary region with black pigmentation in anterior maxillary region and palatal region. Differential diagnosis of gingival hyperplasia, giant cell epulis, Kaposi’s sarcoma and malignant melanoma were kept. Histopathological sections showed sheets of pleomorphic spindle cells with intracytoplasmic Brown-black pigment, involving the lower part of the squamous epithelium. Tumor cells were positive for S-100 and a human melanoma black-45 immunostain, and a diagnosis of malignant melanoma was rendered. The patient underwent partial maxillectomy on left side and the case was categorized as Stages I level III. Complete excision with adequate negative margins is the treatment of choice. Malignant melanoma is relatively insensitive to radiation; hence, the role of radiotherapy is not well-defined. The tendency to invade and cause early hematogenous metastasis results in poor prognosis. The most common cause of treatment failure is Nodal involvement and multiple local recurrences.

**Keywords:** Gingiva, Hyperpigmentation, Mucosal melanoma, Radiotherapy

INTRODUCTION

Melanomas are malignant neoplasms arising from melanocytes, originating from the neural crest cells. Melanocytes are primarily present in the basal portion of the epidermis at the dermo-epidermal junction. The places where neural crest cells migrate are prone to get primary mucosal melanomas.

Primary mucosal melanoma is a rare entity in head and neck, occurring much less frequently than its cutaneous relatives. It constitutes 1% of all melanomas and 10% of head and neck melanomas. Its incidence is thought to be stable, contrary to its cutaneous counterpart, which has been rapidly increasing. Breakdown of the overlying epithelium or hemorrhage of the lesion indicates the oral melanomas. The clinical detection becomes more difficult from squamous cell carcinomas the ulcerated epithelium in both lacks the induration and rolled borders. The poor prognosis of oral melanoma with a 5 years survival rate of 15-38% is due to the delay in the detection of the early disease. Criteria for the primary oral melanoma, as described by Greene et al. Include: (1) Demonstration of melanoma in the oral mucosa (2) presence of junctional activity; (3) inability to demonstrate the extra oral primary melanoma.

Surgery remains the main treatment modality, with added radiotherapy and chemotherapy to prevent recurrence and metastasis. Despite of aggressive resection and multimodal treatment, the prognosis of outer mitochondrial membrane (OMM) remains questionable. Also, because of the rarity of this tumor, there is a lack of definite proof regarding etiology, pathogenesis, treatment protocol, and prognostic factors for OMM.

CASE REPORT

A 51-year-old female patient reported to the Department of Oral Surgery with the complaint of a growth in the left upper back region, for past 6 months (Figure 1). The growth was asymptomatic. The growth measured approximately 7 cm in diameter and was mobile. The surface over the swelling was irregular, and polypoid, and there is brownish-black discoloration that covers the mucosa. The differential diagnosis of gingival hyperplasia/fibrosis, giant cell epulis and Kaposi’s sarcoma or a melanoma was made. There is no history of smoking or tobacco chewing. The routine blood
investigations were found to be normal, and the patient was non-reactive for HIV 1 and 2. A biopsy was taken.

Histopathological sections of the biopsy showed an ulcerated polypoid growth composed of sheets, and fascicles of an oval to spindle tumor cells, displaying moderate pleomorphism with the presence of giant tumor cells and frequent mitosis. The Lower part of the squamous epithelium was also invaded by the tumor cells. Some of the tumor cells showed intracytoplasmic brown-black pigment. There was no history of a pigmented lesion at the given site or of any cutaneous lesion that had been excised or had spontaneously regressed. Based on these findings, a diagnosis of primary mucosal malignant melanoma was rendered. Following this, the patient underwent extended radical maxillectomy on the left side (Figures 2-4). The maxillectomy specimen showed a 7 cm × 6 cm brown-black growth (Figure 5) extending from the region of the upper right premolar teeth up to the right second molar that exhibited severe mobility. The sections from excision specimen showed similar histopathological features as in the biopsy. The underlying bone and all the resection limits were free of the tumor. Based on the clinical staging system for primary oral malignant melanoma, the present case was staged as Stages I level III.
DISCUSSION

Mucosal melanomas in the head and neck region occurring mainly in the upper respiratory tract and oral cavity. A few authors have reported a slight male predominance. They are more common in the elderly.

Melanocytes contain melanin pigment found in the basal layer of the epidermis are present in the mucous membrane and in the eyes. Melanocytes in the oral mucosa are found to be on the tip and periphery of the rete pegs. Melanin is usually not produced in the oral mucosa, but it can be produced under pathological conditions, as in Addison’s disease, neoplasms. Irritants and carcinogenic compounds (e.g., tobacco smoke) have been found to be responsible for the development of primary mucosal melanoma.

Malignant melanoma is a potentially aggressive tumor of melanocytic origin. About 1-8% of all melanomas arise in the oral mucosa, and these account for 0.5% of all oral malignancies. The most frequently affected oral sites are the palate and the maxillary gingiva. The age of reported patients ranges from 20 to 80 years.

OMM is an extremely rare and very aggressive tumor of melanocytic origin. Apart from oral mucosa malignant melanoma can affect mucous membranes of nose and paranasal sinuses, pharynx, and conjunctiva. As a group, mucosal melanomas invade and spread more quickly and metastasise more frequently, and are therefore, associated with much poorer prognosis than cutaneous melanomas.

Mucosal melanoma can be primary or metastatic. It is therefore, very important to rule out any other primary malignant melanoma elsewhere in the body. Greene et al. gave criteria for diagnosis of primary OMM as follows:
1. Demonstration of clinical and microscopic tumor in the oral mucosa.
2. The presence of junctional activity in the oral mucosa.
3. Inability to show any other primary site.

Tanaka et al. identified five types of OMM based on clinical appearance: Pigmented nodular type, non-pigmented nodular type, pigmented macular type, pigmented mixed type, and non-pigmented mixed type. Diagnosis of OMM can be made based on clinical presentation of the pigmented lesion with the so-called ABCD checklist (asymmetry, border irregularities, color variegation, and diameter >6 mm) that is commonly used for cutaneous melanomas. Differential diagnosis includes melanoma, melanotic macule, oral pigmented nevus, smoker’s melanosis, amalgam tattoo, and Kaposi’s sarcoma.

A simple TNM clinical staging, recognizing three stages, has shown to be of prognostic value. A recent histopathological micro staging for Stage I sub classifies it into three levels. Stage I: Primary tumor present only (N0M0).
Level I: Pure in situ melanoma without evidence of invasion or in situ melanoma with “micro invasion”
Level II: Invasion up to the lamina propria
Level III: Deep skeletal tissue invasion into skeletal muscle, bone, or cartilage
Stage II: Tumor metastatic to regional lymph nodes
Stage III: Tumor metastatic to distant sites.

Umeda et al. in their study concluded that 5 years survival rate of patients who underwent some surgical procedures, such as incision, biopsy, or tooth extraction, before definitive surgery was poor (25.9%) compared with those who did not undergo such procedures (91.7%). Similar results were shown by Rampe et al. and Austin et al. However, many authors believe that biopsy of an undiagnosed lesion, pigmented or unpigmented, occurring in high-risk sites for OMM should be done, because benefits gained by a definite diagnosis of OMM far more outweighs the risk of distant metastasis that is not yet fully established.

Tumor thickness is shown to be a strong predictor of prognosis. Usually, OMM tends to present at a more advanced stage compared with cutaneous melanomas, with 70% of Stage I and 83% of Stage II tumors presenting with a thickness >4 mm, leading to poor prognosis.

Umeda and Shimada suggested a protocol for management of OMM which refers to the extent of margins:
1. Excision of the primary lesion, preferably using an intraoral approach an involving at least 1.5 cm of healthy tissue
2. Excision of any lymph node metastasis (Stage II)
3. Consider chemotherapy.

The mucosal melanoma has a poor prognosis because of its tendency to invade and cause early hematogenous metastasis, with a 3-year mortality rate higher than 50% and a median survival time of 25 months. Gingival melanoma has a greater 5 years survival rate (18%) and a longer median survival time (46 months vs. 22 months) compared to palatal melanoma (11%). The presence of sarcomatoid and pseudo papillary architecture and undifferentiated cells are also associated with significantly poor disease-specific survival. Multiple local recurrences and early nodal involvement lead to reduced survival rate and complicates the treatment planning.
CONCLUSION

Malignant melanoma is a rare tumor of the oral cavity with very poor prognosis. Clinically these tumors are very silent and asymptomatic in their appearance that can lead to misinterpretation. Early diagnosis and aggressive management are needed to treat malignant melanoma. In spite of aggressive multimodal treatment, melanoma has a poor prognosis and very less survival rate.

REFERENCES


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