An Unusual and Rare Case of Primary Oral Malignant Melanoma Involving the Mandible in a Young Female

Abstract

Background: Primary oral malignant melanomas (POMM) are rare and have been estimated to represent 1.6% of all oral malignancies and 0.2-8% of all melanomas. Most cases of melanomas involve palate and maxillary gingiva (80%) and about 20% involve mandible. The purpose of this article is to report a very unusual and rare case of melanoma in a young female involving mandible causing extreme destruction of bone.

Methods and Findings: A 23 year-old female presented with a painful swelling over right mandible since 3 months. The growth extended from right preauricular region to submandibular region, was nodular and bluish black in colour and bled on touching. It caused displacement of mandibular premolars and molars. A provisional diagnosis of malignant osteolytic tumor was made. The patient underwent right hemimandibulectomy. Intraoperatively, the tumor was seen involving the entire mandibular body. On gross examination, right hemimandibulectomy specimen was received in pieces measuring 12 × 9.5 × 5.2 cm containing seven teeth. The tumor had perforated the lingual cortical plate. Cut surface was friable and black in colour. Microscopy showed a biphasic high grade neoplasm comprising of round to spindled tumor cells exhibiting marked nuclear pleomorphism and brisk mitosis, laden with granular brownish pigment melanin. The findings were consistent with the diagnosis of pigmented malignant melanoma and confirmed on immunohistochemistry as the tumor expressed vimentin and HMB45 and was negative for CK, EMA and p40.

Conclusion: Involvement of mandibular bone by melanoma is rare. However, when they involve the bone, they are indistinguishable from other lytic malignant tumors or osteomyelitis. The present case emphasizes the need to learn the unusual location, the possible presentation of oral melanoma and inclusion of this rare tumor in the differential diagnosis of lytic bone tumors.

Keywords: Primary oral melanoma; Mandible; Osteolytic; Junctional activity

Introduction

Melanoma is a malignancy of pigment producing cells, melanocytes, located predominantly in the skin, but also found in eyes, ears, GI tract, leptomeninges, oral and genital mucous membranes. Primary oral malignant melanomas (POMM) are rare and have been estimated to represent 1.6% of all oral malignancies and 0.2-8% of all melanomas. Rarer is the metastasis to the oral cavity which frequent to tongue, mandible and buccal mucosa [1]. Globally, oral melanomas are more common in Japan accounting for 11-12.4% of all melanomas. A higher incidence of oral melanomas is reported in India and Africa as well. Most cases of melanomas involve palate and maxillary gingiva (80%) and about 20% involve mandible. Onset is usually between 30 to 90 years of age, with higher incidence in the 6th decade. In general it does not show gender preference but some authors have reported slightly higher incidence in males [2,3].
The purpose of this article is to report a very unusual and rare case of melanoma in a young female involving mandible causing extreme destruction of bone.

**Case Report**

A 23 year-old female reported to a Regional Hospital in Ghana with chief complaint of swelling over right mandible since three months. The swelling was fast growing and associated with severe pain. Extraorally the swelling extended from right preauricular region through the angle of mandible extending anteriorly up to the midline and inferiorly extending over right submandibular region. The swelling was firm and tender on palpation. Overlying skin was stretched. Intra- orally large nodular mass was present lingually, perforating the lingual cortical plate in the region of mandibular right premolars and molar and obliterating the vestibule. The growth was nodular and bluish black in colour and bleeds on touching. The tumor caused displacement of mandibular premolars and molars.

Based on this a provisional diagnosis of malignant odontogenic tumor was made including osteosarcoma, Burkitts lymphoma, melanoma, intra-osseous salivary gland tumor and malignant odontogenic tumor.

The patient underwent right hemi-mandibulectomy and sentinel lymph node dissection as a part of treatment. Intraoperatively, it was observed that the tumor involved entire mandibular body, sparing the mandibular angle, ramus and condyle.

Grossly we received right hemimandibulectomy specimen in pieces measuring 12 × 9.5 × 5.2 cm containing seven teeth. The tumor had perforated the lingual cortical plate. Cut section of the tumor was fribale and black in colour. The tumor was extensively sampled for histopathology.

Microscopy showed a biphasic high grade neoplasm comprising of round to spindled tumor cells which exhibit marked nuclear pleomorphism and brisk mitosis. All the tumor cells were laden with granular brownish pigments melanin. The tumor ulcerated the overlying mucosa and junctional activity was noted at a few places. The anterior, posterior, medial and lateral mucosal margins were free of tumor. However, the anterior bony cut margins were positive for tumor. All 8 cervical lymph nodes were positive for metastasis and perinodal extension was noted. The findings were consistent with the diagnosis of pigmented malignant melanoma. An Immunohistochemistry panel was performed for confirmation. The tumor expressed vimentin and HMB45 and was negative for CK, EMA and p40.

**Discussion**

Although oral melanoma was first described by Weber in 1895, its etiology is still an enigma and the incidence has remained stable over decades. In contrast, the incidence of cutaneous melanomas has increased dramatically over the same period. The predilection for occurrence in the palate is still unexplained. The risk factors for oral melanomas are unknown. Chemical, thermal or physical agents such as dentures, irritation from teeth, alcohol, tobacco, poor oral health to which oral cavity is exposed have not been linked to development of melanomas. Blue nevi are more common on palate but are not reported to undergo malignant transformation [1].

Oral melanomas are thought to arise primarily from the melanocytes in the basal layer of squamous mucosa like their cutaneous counterparts. In the oral mucosa, melanocytes are observed in the ratio of about 1 melanocyte to 10 basal cells. Oral lesions may be black, brown, grey or purple. About 10% of the cases are amelanotic. Involvement of jaw bones by melanoma is rare. However, when they involve the bone, they are indistinguishable from other lytic malignant tumor or osteomyelitis [4,5].

Head and neck melanomas are more aggressive when it presents clinically as nodular growth with a vertical growth that invades the submucosa. The prognosis is poor and is based on the histological type, the depth at microscopic level of the tumor’s invasion and its localization. POMM is observed with greater frequency in the mucosa that covers the bone tissue, such as that of the hard palate and gingiva predisposing to early invasion of the adjacent bone, being an additional factor for the poor prognosis presented by this pathology [6].

The GREENE criteria in 1953 [7] for primary oral melanoma are:

- Demonstration of melanoma in the oral mucosa.
- Presence of junctional activity.
- Inability to demonstrate extra-oral primary melanoma.

The American Joint Committee on Cancer has not published guidelines for the staging of oral melanomas. However, most practitioners use general clinical stages in the assessment of oral mucosal melanoma as follows:

**Stage I:** Localized disease

**Stage II:** Regional lymph node metastasis

**Stage III:** Distant metastasis

The prognosis is poor with a 5 year survival rate generally in the range of 10-25%. The median survival is less than 2 years. Prognosis worsens when lymph node metastasis occurs. The 5 year survival rate in patients with positive nodes is 16.4% as opposed to 38.7% in patients with negative nodes [8].

Surgery remains the preferred treatment along with chemotherapy, radiotherapy and immunotherapy. The US FDA previously approved interleukin-2 and dacarbazine for use in metastatic setting. Vemurafenib is a selective inhibitor of mutant BRAF and phase I and II trials showed a relative reduction of 63% in risk of death and 74% in risk of tumor progression with improved survival rates [9].

**Conclusion**

The present case emphasizes the need to learn the unusual location and possible presentation of oral melanoma and inclusion of this rare tumor in the differential diagnosis of lytic bone tumors. Most of the pigmented lesions of the oral cavity are
benign and of no clinical consequence, dentists must maintain a high suspicion for lesions with brown-black discoloration with aggressive biological behavior and emphasize the need of histopathological examination of all pigmented oral lesions.

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Reference