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Surgery

ORAL MELANOMA : LITERATURE REVIEW WITH TWO CASE REPORTS

KEY WORDS: melanoma, malignant

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ABSTRACT

Primary mucosal melanomas of the head and neck are a rare entity, occurring much less frequently than their cutaneous counterparts. Among those of the head and neck region, oral mucosal melanoma is extremely infrequent. It accounts for only 0.5% of oral neoplasms. Oral mucosal melanomas are highly malignant tumors with the tendency to metastasize or locally invade tissues more readily than other malignant tumors of the oral cavity. As most mucosal melanotic lesions are painless in their early stages, so delayed recognition and subsequent treatment result in worst prognosis. This paper presents two rare cases of oral malignant melanoma with significant heterogeneity in morphological features and biologic behavior with the literature review.

INTRODUCTION

Melanoma is a malignant tumor that arises from epidermal melanocytes and is most commonly occurs on skin. According to the National Cancer Database Report on cutaneous and non cutaneous melanoma, 91.2% of all melanomas arise on the skin, whereas ocular (5.2%), mucosal (1.3%) and unknown primaries (2.2%) are less frequently seen. Melanoma is a malignant neoplasm comprising of melanocytes, which are derived from the neural crest cells that constitute the melanin pigment of the basal layer of the epithelium.^[1]

Primary mucosal melanomas of the head and neck are a rare entity, occurring much less frequently than their cutaneous counterparts. Among those of the head and neck region, oral mucosal melanoma is extremely infrequent. It accounts for only 0.5% of oral neoplasms. Oral mucosal melanomas are highly malignant tumors with the tendency to metastasize or locally invade tissues more readily than other malignant tumors of the oral cavity.^[2]

The incidence of melanoma has been steadily increasing in the past several decades. This increased frequency of newly diagnosed melanomas has been observed worldwide and is of the order of 3-8% in a year. This increase in the incidence of melanoma has been sustained over time. In the United States in 1935, the lifetime risk of an invasive melanoma developing was only 1 in 500; in 1960, 1 in 600; in 1992, 1 in 105; in 1996, 1 in 88, and the lifetime risk would be 1 in 75 by the year 2000.^[3]

The etiopathogenesis of mucosal melanomas is poorly understood; however, it is well documented that the melanocytes migrate to both endodermally derived and ectodermally derived mucosa. The function of these melanocytes in the mucosa is not understood. Like their cutaneous counterparts, oral melanomas (OMs) are believed to arise either from nevus, pre existing pigmented areas, Hutchinson's premalignant lentigo or denovo (30% cases).^[4]

Here, we are reporting two cases of oral malignant melanoma with different presentations and review literature.

CASE REPORTS

CASE 1 :

A 38 year old male patient reported to the department of Oral And Maxillofacial Surgery, VYWS dental college and Hospital, Amravati with the chief complain of blackish colour lesion and pain on the left palatal region since 1 year. The lesion was rapidly progressive and pain was continuous and intense in nature. The patient gave a positive history for smoking and other deleterious habits. Extra-oral examination revealed enlargement of bilateral submandibular lymph nodes, which were tender on palpation. Intra oral examination showed the asymmetrical presentation in the form of nodular and macular gingival lesions. A black exophytic growth was seen on the left side of the hard palate. (fig 1). The lesion was extended into buccal aspect involving gingiva with maxillary molars and pre molars . Small satellite lesions were also seen between interdental gingiva around the lesions (fig 2). On palpation lesion was soft in consistency and non tender.

Radiographs were taken. Orthopantomogram (fig 3) showed displacement of maxillary molars, with destruction of interdental bone. The periodontal ligament space of left maxillary molars and second premolar was widened. The continuity of the floor of the maxillary sinus was disrupted in the maxillary left first and second molar region. ()

Incisional biopsy was performed. Histopathological studies revealed numerous atypical melanocytes within the epithelium and also invasion into the connective tissue. The pathological examination confirmed the diagnosis of malignant melanoma.

CASE 2 :

A 65 year old male reported with the chief complain of brownish black painless growth in the upper front gingivo

buccal vestibule since 1-2 months. Extra oral examination revealed normal structures except for mild localised swelling on maxillary anterior region. Intra oral examination revealed a single brownish black pedunculated growth on maxillary anterior gingivobuccal vestibule of size 2.5 *3 cm approx. Brownish black pigmentation were seen approx 1 cm surrounding swelling. It was soft and non tender on palpation with fixity to the underlying structures.(fig 4)

Orthopantomogram (fig 5) revealed splaying of maxillary central incisors and widening of periodontal ligament space of maxillary central incisors.

Excisional biopsy (fig 6) of the pedunculated mass was performed. Histopathological examinations revealed, polypoid lesion with ulceration on surface and consist of group of clusters of round ovoid or elongated cells separated by fibrous septi and many congested blood vessels. The cells shows round to oval nuclei with prominent with prominent nucleoli and clear or granular cytoplasm with brownish pigmentation (melanin) in some of the cells. The mucosa at the base of the polyp shows focal melanosis near basal layer of epithelium. All these findings revealed a malignant melanoma of gingiva buccal mucosa.

CASE 1 :



FIG 1



FIG 2



FIG 3

CASE 2:



FIG 4



FIG 5



FIG 6

DISCUSSION

Malignant melanoma was first described by Weber in 1859. It was recognized as a distinct clinical entity as “melanotic sarcoma” by Lucke in 1869 as also highlighted by Rimal et al. [6] Melanoma is a malignant tumour involving melanocytes and derived from neural crest cells. Most melanomas arise in the skin, however mucosal lesions has been also found with rare primary presentation in oral cavity. [6] No etiologic factors have

been identified for oral melanomas. Risk factors have also remained elusive. In the mouth, mechanical traumas including injury from ill-fitting prostheses and infection have been cited as possible causative factors, tobacco use and alcohol has been mentioned as possible risk factors. Ingested and inhaled environmental carcinogens at high internal body temperature may play some role. Some oral melanomas are believed to originate from junctional nevis. [7,8]

The most common presentation of oral melanomas is an asymptomatic brown , dark blue or black macule, sometimes with erythema or ulceration. A more advanced disease may take a nodular surface characteristics , they may also exhibit asymmetric, and irregular borders just like cutaneous melanomas. [9] The diagnosis of oral malignant melanoma remains difficult. Differential diagnosis includes melanocytic nevi, oral melanotic macule, Peutz-Jeghers syndrome, Addison's disease, Cushings syndrome, melanoplakia, Kaposi sarcoma. Our patient did not have any of the signs and symptoms of above diseases and complete blood investigation was normal leading to exclusion of any systemic disease.

Regional lymphadenopathy may be present as in our case and this indicates a poor prognosis. Our case exhibited signs and symptoms like enlarging, swelling, pigmentation, splaying of the teeth .Mucosal melanoma behavior, treatment and prognosis are different from cutaneous melanoma. [10,11] The biological behavior of oral malignant melanoma differ from the cutaneous melanoma. Cutaneous melanoma usually have radial growth phase if it is flat or macular and vertical growth phase if it is a mass, nodular or elevated form and vertical growth melanoma with metastasis. [12] On the other hand in oral melanoma as the radial growth phase do not tend to invade the underlying reticular corium, but are usually associated with metastasis . The surface architecture of mucosal melanomas ranges from macular (melanoma in situ) to ulcerated and nodular or invasive melanomas. [9] Clinically, oral melanomas are classified into five types: pigmented nodular, nonpigmented nodular, pigmented macular, pigmented mixed, and nonpigmented mixed. [13]

The treatment of choice is surgical excision with safety margins. According to Zitelli et al., the margins should be at least 1.5 cm for the lesion in head and neck melanoma or 2.5 cm for melanomas larger than 3 cm in diameter. Many patients with early-stage lesions are cured by surgery alone. However, adjuvant radiation therapy and immunotherapy (with interferon-alpha) can be considered. Even though melanoma is not very radiosensitive, patients in early melanomas have had good response to radiation therapy. For patients with distant metastasis vemurafenib and ipilimumab are two novel treatments but agents such as high-dose interleukin-2, dacarbazine, imatinib and paclitaxel have also been tried. These recent developments in genotype directed and immunotherapy have led to prolonged survival of patients. [14,15] Ablative surgery with tumor-free margins in combination with chemotherapy and, to a lesser extent, immunotherapy or irradiation is the recommended treatment, while there is a recognized need for an evidence-based treatment protocol choice but probably, multimodal therapy may be proven more effective in the treatment of oral mucosal melanoma. [16,17]

The 5-year overall survival rate for head and neck malignant melanoma ranges from 21% to 40% and for oral malignant melanoma it is 15% with a median survival of 25 months. Gingival melanoma has a better 5-year survival rate than palatal melanoma, with a longer median survival period (46 months versus 22 months). [7] In a study on 35 patients with oral melanoma, Tanaka et al., observed a 5-years cumulative survival rate of 35.5% in the group of patients treated with irradiation alone, while it was 15.4% for patients treated with surgery. On review of the approximately 1000 cases reported,

it was found that the 5-year survival rate was 17% and the 10-year survival rate was 5%.^[13,18]

CONCLUSION

Early Diagnosis and management of oral malignant melanoma is important for the prognosis of highly aggressive tumours. Practitioners 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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