“Malignant melanoma of left maxillary alveolus – a rare case report”

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Abstract

Primary oral malignant melanoma usually presents as a dark brown or black lesion. It is a rare malignancy, accounting for less than 1% of all melanomas and 1.6% of all head and neck malignancies, thus forming up to 0.5% of all oral malignancies in the world literature. In contrast to cutaneous melanoma, it does not have a defined set of clinical and pathologic classification. The oral mucosal melanoma tends to appear at a higher stage and is much more aggressive than its cutaneous counterpart. Thus prognosis of oral melanoma is poor and worse than that of cutaneous melanoma. Here we present one of such rare cases of malignant melanoma of left maxillary alveolus.

Key words: Melanocytes, Oral Malignant Melanoma, Cutaneous Melanoma.

Introduction

Melanomas are malignant neoplasms arising from melanocytes, which originate from the neural crest cells. The cells and the corresponding neoplasms arising from the neural crest are grouped under Dispersed- Neuro-Endocrine System (DNES) tumors. Melanomas are present primarily in the basal portion of the epidermis at the dermal-epidermal junction. Primary malignant melanoma has been described in virtually all sites and organ systems to which neural crest cells migrate (Sandeep Fauzdar et al., 2010).

Mucosal melanomas tend to present at later stage, they are more aggressive, and present in a vertical (nodular) growth phase of disease. Clinically, OMM may mimic other pigmented lesions. A biopsy is required in order to establish the diagnosis. The reported risk of malignant cells spreading during invasive procedures and factors such as size of the lesion or anatomical limitations may influence the diagnostic surgical procedure. Therapy of OMM is commonly based on surgical excision of the tumour, supplemented by radiotherapy, with chemotherapy and immunotherapy serving as adjuncts. Prognosis is poor, with a 5-year survival rate of approximately 15% (Hicks MJ et al., 2000).

Case report

A 66yr old male patient reported to the department of oral medicine and radiology of CDCRI (C.G), with a chief complaint of painless swelling in the left midfacial region since 1 ½ month. Patient noticed painless swelling 1 ½ month back, in the left back region of upper jaw. Past dental history revealed that, the patient underwent extraction of left upper teeth following sudden excessive mobility in a private clinic. After extraction the swelling increased in size and attained large size with fulminating growth of present size for which the patient came to our department. No significant medical history was given by the patient.

History of loss of appetite and weight loss since 1month. Extra-oral clinical examination revealed a solitary well localised dome shaped swelling in the mid-facial region on left side measuring about 5x4cm extending anterior- posteriorly from the ala of the nose, obliterating naso-labial fold to 2cm in front of the ear and superioinferiorly from 1cm below the infraorbital margin to 1cm above the lower border of mandible and elevation of ala of the nose on left side was observed. Surface appeared smooth, shiny and normal in colour with no secondary changes (Fig:1).
On palpation all inspective findings were confirmed and the temperature of swelling was not raised. Swelling was non tender, non fluctuant and hard in consistency. Swelling was not attached to skin. Left submandibular lymph node was palpable, enlarged, nontender, round in shape, 3x3cm in size and hard in consistency. The lymph node found to be fixed to underlying structures. On intra-oral examination a solitary well localized exophytic growth with nodular surface seen extending anteroposteriorly from mesial margin of 23 to distal margin of 26 and superioinferiorly from buccal vestibule adjacent to 22 to 28 to 1cm below their occlusal surfaces. Surface appeared to be nodular, black with heavy melanin pigmentation and margins were well defined but irregular. Greyish black pigmentation extending beyond the lesion to the hard palate on opposite side and posteriorly to the junction of hard and soft palate (Fig:2). Based on history, clinical findings and biological nature of tumor the diagnosis of Malignant melanoma of left maxillary alveolus was made. The clinical differential diagnosis includes Kaposi’s sarcoma, giant cell epulis, pigmented ameloblastoma and ca of maxillary alveolus was suggested.

CT Scan : Shows heterogeneously enhancing mass lesion noted in bucco-alveolar region of left side measuring approximately 5.2x4.1x5.8cms. The mass seemed to cause destruction of upper jaw and anterior wall of left maxillary antrum extending into the maxillary sinus (Fig:4). Enlarged heterogeneously enhancing lesion noted in the left submanbibular region measuring about 2.5x2.7cm with hypodense centre suggestive of necrotic lymphnodes.

Atypical melanocytes infiltrating into the deeper connective tissue. The malignant melanocytes were pleomorphic and hyperchromatic and contained brownish- black granular pigment in the cytoplasm. The lesion was diagnosed clinicopathologically as malignant melanoma. (Fig:5)

**Discussion**

Mucosal melanoma is a rare entity, representing 0.8- 3.7% of all melanomas (Manolidis S et al 1997, Panje WR et al 1996, Patel SG et al 2002). Most cases develop from the fourth decade of life, being extremely rare prior to 30 years of age. The prognosis is poorer as compared to its cutaneous counterpart, with a 5-year survival of 0-10% (Panje WR et al., 1996). Lymph node involvement at the time of diagnosis is present in 6-25% of patients. This frequency increases when the thickness of the tumor is >5 mm and 20% of patients present radiological evidence of generalized disease (Patel SG et al 2002). Melanocytes are neural crest-derived cells; reside primarily in the basal epithelial layer (Auluck A et al., 2008). The function of melanocytes in the mucosa is not fully understood, but their presence in the basal layer of the epithelium is well known. In physiologic states, the melanocytes in mucous membranes do not produce melanin. However in pathological conditions like neoplasms etc, they produce it. The clinical presentation of OMM is a painless mass with or without ulceration and bleeding. Colour may vary from black, grey, purple and red. Biopsy and histopathological examination of the lesion is confirmatory diagnostic tool. Determining whether the lesion is primary or secondary is important because cutaneous melanoma may metastatize to mucous membranes (Parvathi Devi et al., 2011). No universally accepted staging system for mucosal melanoma exists and clark’s classification, cannot be applied to oral mucosal melanoma because of the lack of histologic landmarks analogous to papillary and reticular dermis (Niti Singhal et al., 2011).

The 1995 WESTOP (Western Society of Teachers of Oral Pathology) Banff Workshop on OMM drew attention to the fact that most OMMs are discovered and biopsied in an advanced stage, which probably contributes to the heterogeneity of microscopic patterns. The 2002 (revised) TNM Melanoma Staging System of the American Joint Committee on Cancer, applying the T-symbol for the thickness and the ulceration status of the tumour, the symbol N for the regional lymph nodes, the symbol M for distant metastases and the serum lactic dehydrogenase (LDH) level does not provide specific guidelines for OMM (Marco Meleti et al 2007). A simple TNM clinical staging system for oral malignant melanoma is as follows:

**Stage I :** Primary tumor present only (Tany N0M0)
- Level I : Pure in situ melanoma without evidence of invasion or in situ melanoma with microinvasion.
- Level II : Invasion upto the lamina propria.

**Stage II :** Tumor metastatic to regional lymph nodes (Tany N1M0).

**Stage III :** Tumor metastatic to distant sites (Tany Nany M1).

Microscopically two histological patterns are described : an in situ pattern and an invasive pattern, and a combined pattern is seen in advanced lesions. Mucosal melanomas are said to have aggressive vertical growth phase. Different cell types like spindle cells, plasma cells and epitheloid tumor cells arranged in sheet-like, alveolar, neutrotropic or desmoplastic pattern may be observed. These tumors react strongly to S-100 and more specifically to HMB-45 (Niti Singhal et al., 2011). Treatment of OMM is still controversial and there is no consensus regarding the best therapeutic approach. Data from several studies...
indicate radical resection of the primary tumor as the treatment of choice. Surgery could be combined with radiotherapy, chemotherapy or immunotherapy even though the effectiveness of such therapies both as primary or in association with the surgical treatment is mostly unknown (Macro M et al., 2007; Umeda M et al., 1994).

A protocol adopted by Umeda and Shimada[9] refers to the extent of the margins:
1. Excision of the primary lesion, preferably using an intraoral approach and involving at least 1.5 cm of healthy tissue.
2. Excision of any lymph node metastases (Stage II).
3. Consider chemotherapy.

Adjuvant chemotherapy with decarbazine, platinum analogs, nitrosoureas, and microtubular toxins have been used for palliative purposes or for therapy of metastatic melanoma, but does not seem to influence survival. Umeda M and Shimada K in 1994, suggested dimethyl triazenoimidazole carboxamide (DTIC), nimustine hydrochloride (ACNU) or vincristine (VNC) as drugs of choice for postoperative chemotherapy. Anticancer therapy using IFN-gamma and anti-Fas antibody has shown positive results against OMM cells in an experimental mode (Kamei T et al., 2005). Systemic immunotherapy has been used as adjuvant or for palliation in the treatment of disseminated disease. Immunotherapy with IL-2 and other cytokines is not associated with an enhanced survival rate. The definition of the so-called cancer testis antigens (CTAs) expression profile in OMM could lead to the development of a new vaccine-based therapy. Gene therapy is still in an experimental phase (Macro meleti et al., 2007).

In this case patient noticed painless swelling 1 ½ month back, in the left back region of upper jaw. The clinico-pathological findings along with supporting findings (imaging and biopsy) suggests in favour of melanoma. The patient was then referred to oncology department for multidisciplinary approach. Based on various studies conducted by different authors it was found that oral melanomas are more aggressive than their cutaneous counter parts. Vairaktaris et al showed that 5 year survival does not exceed 5-9%, with the current knowledge given the extreme rarity of oral melanoma and wide reports of very poor prognosis and poor understanding on the medical management, surgical intervention remains the mainstay of treatment (Kamei T et al., 2005; Silvia Cristina Aguas et al 2009; GU GM et al 2003). In such cases with decreased patient appetite, and nodal involvement, estimated survival for such patient could be less than 1 or 2 years. Given such characteristics of oral melanoma more documentation and publications of report cases should be encouraged, detailing its clinical presentations, management and long term treatment follow up may help the understanding on oral melanomas and may improve prognosis but early diagnosis and early surgical treatment will continue to be the main weapon for good prognosis.

Figure1. Lateral profile of the patient showing extra oral swelling.
Figure 2. Large exophytic growth with black melanin pigmentation seen.

Figure 3. OPG view of oral malignant melanoma.

Figure 4. CT scan of oral malignant melanoma.
Conclusion

Clinically, OMM may mimic other pigmented lesions, so the oral diagnostician should be more vigilant and be thorough in oral examinations of pigmented lesions to rule out suspicious oral pigmentation to save the life and esthetics of patient.

References


Abbreviations

OMM: Oral Malignant Melanoma

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