Oral Mucosal Melanoma With Extensive Nodal And Visceral Metastasis

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ABSTRACT

Mucosal Malignant melanoma of the oral cavity is an extremely rare condition, constituting approximately 1-2% of all the melanomas. It has a poor prognosis. Clinicians should have high index of suspicion for any pigmented lesion in the oral cavity, which should be routinely biopsied, in order to detect this disease early, so that it can be managed appropriately. Here is an unusual case of a lady presenting with a nodule in the breast with cervical lymphadenopathy, who on detailed examination, was found to have an asymptomatic pigmented lesion on the hard palate, the biopsy of which confirmed melanoma, but on further investigation, it was found to have extensive metastasis.

Key Words: Oral Mucosal Melanoma, Metastasis

INTRODUCTION

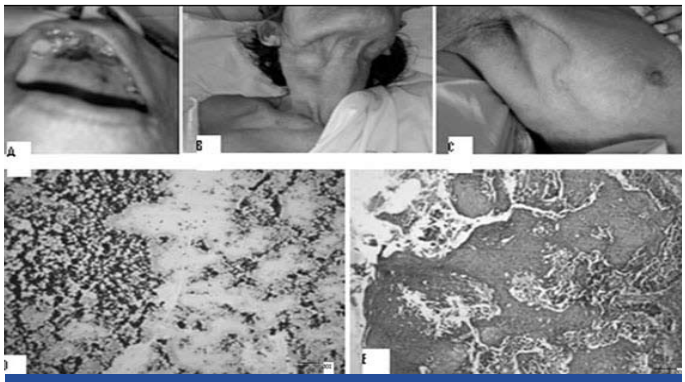
Malignant melanoma is a cancerous condition of the melanocytes. These cells are found in the basal layer of the epidermis, the mucosal membranes, and in the eyes. Mucosal melanomas represent less than 4% of all the melanomas and present a difficult challenge when distinguishing the primary from the non melanocytic lesions. Lack of pigment occurs in approximately 35% of the cases. The first description of mucosal melanomas was made by Weber et al in 1856[1]. Unlike cutaneous melanomas, mucosal melanomas do not have precursor lesions and risk factors[2]. The treatment recommendations are also not standardized. Melanomas of the oral cavity constitute 40% of all mucosal melanomas of the head and neck. The most common site for the occurrence of oral mucosal melanoma is the palate [hard and soft][3].

CASE REPORT

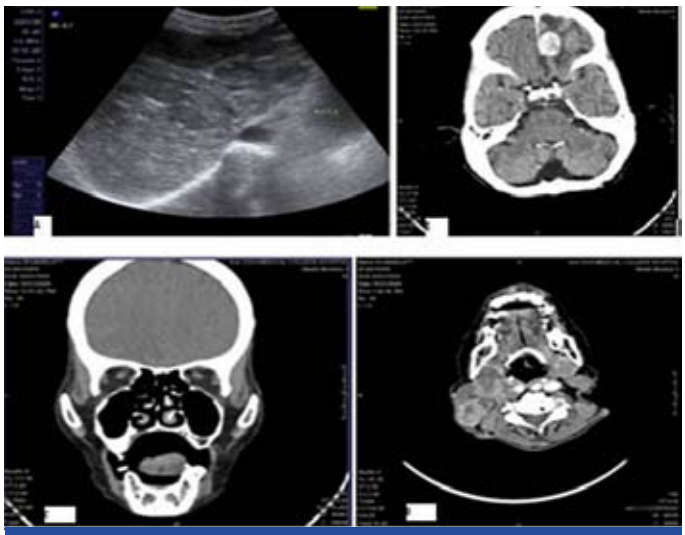
A 72 year old female patient came with a history of anorexia, weight loss approximately 10-15%, multiple neck swellings and a swelling in the right breast since 4 months. On examination, she was found to have bilateral cervical lymphadenopathy. The lymph nodes were hard. The level 2 lymph nodes were fixed. In the right breast, there was a subcutaneous nodule in the upper outer quadrant. The per abdomen examination revealed no organomegaly and free fluid. The per rectal and proctoscopic examination showed no abnormality.

Oral examination showed poor oral hygiene with a partially edentulous state. A black pigmented plaque, 3 by 3 cm, was present on the mucosa of the hard palate, extending from the palatine rugae to the alveolar margin.

The fundus examination was normal. Fine needle aspiration cytology of the cervical lymphnodes and the subcutaneous nodule on the breast showed features of malignant melanoma. A punch biopsy of the patch also showed features which were suggestive of malignant melanoma. Cytopathology showed large hyperchromatic cells which were arranged singly and in clusters. The cells showed prominent nucleoli and eccentric nuclei. A fair amount of pigment was seen. Histopathology showed tissue enclosing nests and groups of malignant cells with scanty cytoplasm and hyperchromatic nuclei, with some of them showing the intracytoplasmic melanin pigment.



[Table/Fig: 1] A) Oral Mucosal Melanoma of Hard Palate. B) Cervical lymphnode metastasis. C) Metastatic subcutaneous nodule over upper outer quadrant of right breast. D) Photomicrograph of Haemotoxilin and Eosin stained fine needle aspiration cytology of lymphnode. E) Photomicrograph of Haemotoxilin and Eosin stained punch biopsy of hard palatal mucosa



[Table/Fig: 2] A) Abdominal ultrasound showing liver metastasis. B) Contrast enhanced computed tomogram showing brain metastasis. C) Erosion of palatine process of maxilla. D) Cervical lymphnode metastasis.

X-raychest showed features of lung metastasis. Abdominal ultrasonography showed features which were suggestive of liver

metastasis. Computed tomography of the brain showed features which were suggestive of brain metastasis. Due to the advanced age, extensive metastasis, and the poor performance status of the patient; the decision for surgery or radiotherapy was deferred and the patient was discharged on symptomatic treatment.

DISCUSSION

Mucosal melanoma of the oral cavity is a rare malignancy. It has no known predisposing factors and is difficult to diagnose and manage. Differentiating it from a metastatic melanoma is often challenging.

On gross appearance, the tumours on the palate are usually flat, with a varying degree of thickness. The colour is usually dark blue black. Microscopically, the tumour cells present themselves as densely packed, large epithelioid cells with eosinophilic cytoplasm. The melanin pigment which is located intra or extracellularly, may be abundant, but may be sparse or even absent at the light microscopic level. The prognostic value of various levels of invasion, as established in the Clark's classification, does not apply for mucosal melanoma because of the absence of histological landmarks which are analogous to the papillary and reticular dermis.[4]

The staging systems that are applied to cutaneous melanoma are not applicable to mucosal melanomas. The American Joint Committee on Cancer does not have published guidelines on the staging of oral malignant melanomas. The generally followed guideline is a clinical classification [5]; stage I-clinically localized disease, stage II- regional lymph node disease and stage III –distant disease. The tumour thickness is a reliable prognostic indicator for survival. In this case, the patient was in stage III.

Distribution[6]	Mucous membranes
Head and neck	55.4%
Female genital	18.0%
Anal / Rectal	23.8%
Urinary	2.85%

[Table/Fig 3]: SPercentages of the distribution of mucous membrane melanomas in different mucous membranes.

The majority of patients lack early symptoms, thus leading to a delay in diagnosis. They usually present with ulceration and bleeding. The ulceration lacks induration and rolled up edges. There is usually lack of pain. As the tumour enlarges, the patient may notice a swelling, loosening of teeth or ill fitting dentures. One third of all oral mucosal melanomas lack pigment and one third have pre existing pigmentation or melanosis [7].Oral melanomas have to be differentiated from a number of benign conditions like amalgam, melanocanthoma and melanotic macule[8].

A thorough physical examination, including a search for regional adenopathy was done. A careful and complete skin examination should be performed, to rule out a primary cutaneous melanoma that has metastasized. A biopsy should be taken. It is generally not feasible to apply the concept of an excisional biopsy, as it would entail a major surgical procedure in almost all cases, an incisional biopsy is therefore generally accepted for the establishment of the diagnosis [9]. An oral endoscopy should be done. A computed tomography scan is helpful. A metastatic workup including x ray chest and abdominal ultrasound should be done.

Differential diagnosis of oral malignant melanoma includes: addison's disease, blue nevi, ephlides(freckles), kaposi's sarcoma and oral nevi.

The treatment modalities for head and neck mucosal melanomas have ranged from a conservative approach by using radiation therapy to radical resection with extensive lymph node dissection. Regardless of the treatments, the survival has remained poor. The general consensus in treatment recommendations is primarily surgery with the surgical excision of the primary site, if at all possible, followed by radiation therapy for residual disease or nodal involvement. The nature of the surgical procedure to be carried out depends on the site and extent of the primary tumour. The majority of the oral melanomas are located on the palate and the maxillary gingiva, and as tumours at these sites do not have to invade very deeply before they reach bone, the underlying bone must be removed in these cases [10]. Nodal metastasis plays a role in survival; however, neck dissection is only recommended if there is a clinically positive nodal disease or if resection of the primary tumour requires entry into the neck. By itself, neck dissection does not affect the local and regional control rates or patient survival [11]. Mucosal melanomas have generally been considered to be radio resistant and the role of radiotherapy in treating the primary disease has been controversial. Favourable responses have been reported by irradiation [12]. The recommended doses were 30 Gy in five fractions for 2.5 weeks. In very old patients, primary treatment with radiotherapy is considered as an alternative to major surgery. Brachytherapy is a good alternative in areas where external beam radiation is unable to administer high enough doses of radiation to a small area, while reducing the exposure to the surrounding structures. The conclusions were that mucosal melanomas have a response rate that exceeds that seen in cutaneous melanomas, and that neither surgery nor radiotherapy as a single modality can achieve the local control rates in most of the patients. Recurrent disease at the primary site -a variety of treatment modalities, either alone or in combination, can be useful. These are radiation therapy alone or in combination with hyperthermia, cryotherapy, carbon dioxide laser surgery, and electrodesiccation [13]. The treatment modalities can also include decarbazine and interleukin; also, some studies have shown a higher success rate with treatment with interferon –alpha. Chemotherapy and immunotherapy had no much role in the improvement in the survival or locoregional control [14].

Treatment	Cutaneous	Mucosal
Surgery alone	92%	56%
Radiotherapy	<1%	8%
Chemotherapy	<1%	2%
Surgery+Radiotherapy	1%	19%
Surgery+ Chemotherapy	1%	2%
Radiotherapy+ Radiotherapy	<1%	2%
Surgery+ Chemotherapy	<1%	3%
No Treatment	3%	6%

[Table/Fig 4]: SPrognosis of Melanoma with different modalities of treatment [15].

CONCLUSION

Mucosal Melanoma is a rare tumour of the oral cavity which can metastasize rapidly .The purpose of this article is to emphasize early diagnosis. Since this is a disease which cannot be easily diagnosed, awareness amongst the population must be achieved. Hence, the presence of a pigmented lesion in the mucous membranes of the oral cavity should raise the suspicion of malignant melanoma and all such lesions should be histopathologically examined routinely after biopsy.

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