

CASE REPORT

Primary malignant mucosal melanoma of the oral cavity: A case report

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Introduction

Malignant melanoma is a malignant neoplasm of melanocytes or of melanocytic precursors¹. Primary mucosal melanoma of the head and neck is a rare entity, accounting much less frequently than their cutaneous counterparts. Oral mucosal melanomas are highly malignant tumours with the tendency to metastasize or locally invade tissues more readily than other malignant tumours of the oral cavity². Due to the rarity of this entity, data on epidemiology, tumour behaviour, treatment, follow up and survival of patients are mainly based on single case reports, hence merits reporting.

Case Report

A 55 years old male presented with the complaints of a gradually increasing swelling in the oral cavity for 1 year duration. There was no significant past or family history. Clinical examination of the oral cavity revealed a pigmented, irregular, large swelling on the hard palate. The mass was black, polypoidal and non-ulcerated. It was arising from the hard palate with bilateral extensive involvement, causing partial obstruction of the oral cavity and the oropharynx (Figure 1).

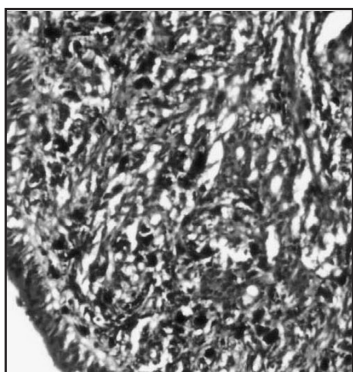


Figure 1: Black colored, polypoidal growth and non-ulcerated arising from the hard palate with bilateral extensive involvement, causing partial obstruction of the oral cavity and the oropharynx

Some pigmented macules of various sizes were also noted growing at the periphery of the lesion. It did not bleed on probing and was insensitive to touch.

On presentation, presumptive clinical diagnosis of mucosal melanoma was made. A punch biopsy was done and the tissue was sent to histopathologic analysis. The histological features were consistent with the clinical diagnosis of oral mucosal malignant melanoma (Figure 2). There was no other similar pathology in the skin. There was no apparent neck swelling and on palpation cervical lymph nodes were not enlarged. A thorough search was made for distant metastasis but the results were negative. The bilateral extensive involvement of palate rendered the case inoperable so a course of radiotherapy was planned. A hypofractionation regimen i.e. large dose per fraction of radiation was given to the patient. During the course of therapy, the patient developed distant liver metastasis for which palliative chemotherapy was given.



Figure 2: Histopathological slide showing highly cellular malignant tumour composed of sheets of epithelioid melanocytes having pale cytoplasm and open nuclei with prominent nucleoli. Sheets & fascicle of spindle cells were also present. Neoplastic cells were studded with abundant melanin pigment.

Discussion

Oral melanomas are extremely uncommon and are primarily involved in fewer than 1% of melanomas. They usually occur on the hard palate and less frequently in the buccal and labial mucosa, gingiva and alveolar ridge³. Chaudhary et al, 1958, in their study of 105 cases found that 80% cases of oral melanoma originated in maxilla, 51% limited to hard palate, 26% to alveolar ridge, 8% to soft palate and 15% of them belonged to more than one location¹. The preference of the upper jaw mucosa is in sharp contrast to the squamous cell carcinoma which commonly affects tongue and floor of mouth. Pain is a relatively uncommon feature and is generally found in advanced cases. The duration between onset of symptoms and diagnosis can be from several weeks to years¹. The delay in the diagnosis can be sometimes due to the margins of the ulcerated lesions. Unlike squamous cell carcinoma, pain is not a prominent feature, and lack everted or rolled borders because the atypical melanocytes exhibit a pagetoid mode of spreading resulting in uniform epithelial thickening. Induration is absent because of prolonged radial growth phase. Despite their cytological similarities, tumors occurring in the skin and oral cavity demonstrate a distinct difference in their immunochemical response. Most cutaneous melanomas show S-100 protein positivity and only somewhat decrease response to α and β subunits. All oral mucosal melanomas fail to stain to any degree from β subunit.

Oral melanomas may present with a variety of morphologic and macroscopic characteristics that could render the clinical diagnosis extremely difficult. The tumour usually appears as smooth, flat/raised, nodular/polypoid, intact/ulcerated, tan dark lesion. The differential diagnosis includes melanotic macule, smoking associated melanosis, post inflammatory pigmentation, melanoplakia, melanoacanthoma, nevi, Addison's disease, Peutz-Jeghers syndrome, amalgam tattoo, Kaposi sarcoma and many other conditions sharing some macroscopic characteristics. In general oral mucosal melanomas have a poor prognosis. Lymphatic spread occurs usually to sub maxillary and upper jugular lymph nodes.

Various modes of treatment have been employed in the management of these tumors. In advanced lesions, regardless of location, surgical intervention is the treatment of choice¹. Electrodisection has been useful in treating superficial melanomas of the palate without sacrificing the integrity of the palatal structures or creating a fistula. Cryosurgery has been performed occasionally. Underlying bone should be excised in lesions reaching the periosteum. Removal of maxilla may be total if antral mucosa is involved otherwise resection of a portion of palate, alveolar ridge and anterior surface of maxilla is sufficient. There are however several factors that preclude complete surgical resection of maxillary tumors. These are skull base or nasopharyngeal involvement, massive bilateral involvement, extensive involvement of face, distant metastasis, widespread or inoperable tumor, debilitation, old age and poor surgical risk patients.

The enbloc resection decreases the local recurrence rate, with little effect on metastasis and survival. Therapeutic neck dissection is to be done in cases of palpable neck nodes but there is disagreement over neck dissection should be done in absence of clinically positive lymph nodes.

Chemotherapy is generally of little value, it is occasionally used to decrease the size of the tumor and to improve surgical control. This modality has been effective as adjuvant treatment preoperatively in patients with advanced tumors and in postoperative course. It has also been employed in the treatment of local recurrences or regional systemic metastasis, but has resulted in low response rate.

Immunotherapy with bacilli Calmette Guerin (BCG) which sometimes is used with the intent of activating the host immune response has also been used but with little success. Other immunotherapeutic drugs include interferon and cimetidine, which when used together is believed to attack killer T cells and inhibit suppressor T cells, resulting in reduction in tumor size. Interferon injections have been of benefit in patients with some cutaneous and other metastatic melanomas, but the response of oral melanomas remains uncertain.

Although previously thought to be of little value owing to inherent resistance of melanomas, radiotherapy is now considered as an important adjuvant in achieving local control and may even have merit as a primary therapeutic modality⁴. Furthermore, primary irradiation is considered a viable alternative to surgery for inoperable cases¹. It has also been used as adjunctive treatment for recurrences, palliative treatment, or postoperatively when the margins are doubtful. Evidence exist that large dose per fraction radiotherapy is of greater benefit than conventional fractionation⁵.

The incidence of metastasis associated with oral melanomas is higher than that occurring in either the nasal or pharyngeal cavities. Spread takes place through blood vessels or lymphatics. Chaudhary et al, 1958, found that more than half of their patients had clinical evidence of nodal involvement and 20% demonstrated clinical or radiographic evidence of generalized dissemination¹. Visceral spread is to lungs, liver, brain, bones, skull base and skin.

While the recommended treatment is the ablative surgery with tumour free margins in combination with chemotherapy and to a lesser extent, immunotherapy or irradiation, there is a recognised need for an evidence based treatment protocol. Multimodal therapy may be proven more effective in the treatment of oral mucosal melanoma. It is apparent however that, oral melanomas are much more aggressive than their cutaneous counterpart². The more aggressive biological behavior has been attributed to angioinvasion, anatomical relations that preclude adequate surgical removal, and delay in diagnosis, tendency to early

ulceration owing to repeated trauma, in turn, may establish readily accessible avenues for metastasis, and the high rate of regional and systemic spread¹.

Conclusion

Oral melanomas are much more aggressive than their cutaneous counterpart and may present with a variety of morphologic and macroscopic characteristics. A clear understanding of the pathophysiology of this disease may yield more specific immunotherapy and chemotherapy techniques. A multicenter study is required to assess objectively the optimal treatment regimen.

References

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