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Case Report

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Primary malignant melanoma of the lower lip-A case report

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ABSTRACT

Malignant melanoma is a malignant neoplasm composed of melanocytes or melanocytic precursors. Primary mucosal melanoma of the head and neck is a rare entity, in comparison than their cutaneous counterparts. The tumor occurs more frequently in the hard palate and gingival mucosa followed by mandibular gingiva, lip mucosa and other oral sites. The aim of this case report is to document a case of primary melanoma of lower lip with neck node secondaries in a 70year old male patient. This case is presented here for the rarity of tumor location.

Keywords: Malignant melanoma, Mucosal, Lower lip, Immunohistochemistry

INTRODUCTION

Melanoma is a malignant tumor composed of abnormal melanocytes, which are cells derived from the neural crest that constitute the melanin pigment in the basal layer of epithelium [1].Over 90% of melanomas occur on the skin, but they may also arise from mucosal surface or other sites wherein neural crest migrate, like oral and genital mucosa, nasal cavity and leptomenigeal area. About 1–8% of all melanomas arise in the oral mucosa [2]. Oral mucosal melanoma makes up approximately 0.5% of all oral malignancies [3]

Commonly involved intraoral sites are hard palate (32%), gingiva (16%), mandibular gingiva (7%), tongue (7%), buccal mucosa (7%), upper and lower Lip (7%). When the lesion is secondary or metastatic, they are more commonly present in the tongue, parotid and tonsils [4]. Overall, males

slightly outnumber females with an average age of presentation being in the middle of sixth decade.

Oral malignant melanomas demonstrate significant heterogeneity in morphological features, developmental process, and biological behaviour. Hence its a diagnostic challenge. When discovered early and fully excised, melanoma is highly curable. However, once metastatic disease develops, treatment options are limited and survival is generally measured in months. Patients with stage III melanoma (involvement of regional lymph nodes) have a 5-year survival of approximately 50 % [5].

CASE REPORT

A 70 year old patient presented with swelling in the lower lip, left side of 6 months duration to the Plastic surgery department of Stanley medical

College, Chennai. Past history was not contributory. Clinical examination revealed an ulceroproliferative lesion in the left side of lower lip of size 6x4x2cms. The growth was non tender with intact and irregular surface having well defined margins, firm in consistency and blackish

in color. Cervical lymph nodes on the left side were palpable. A complete general physical and systemic examination revealed no other primary lesion. Liver was not enlarged on examination of abdomen. Ophthalmic examination was normal.



Figure1: Ulceroproliferative pigmented growth on lower lip

FNAC of left cervical node showed single and clustered pleomorphic cells with abundant well defined cytoplasm, intracytoplasmic melanin

pigment and prominent nucleoli in a lymphoid background suggestive of metastatic melanoma deposit.

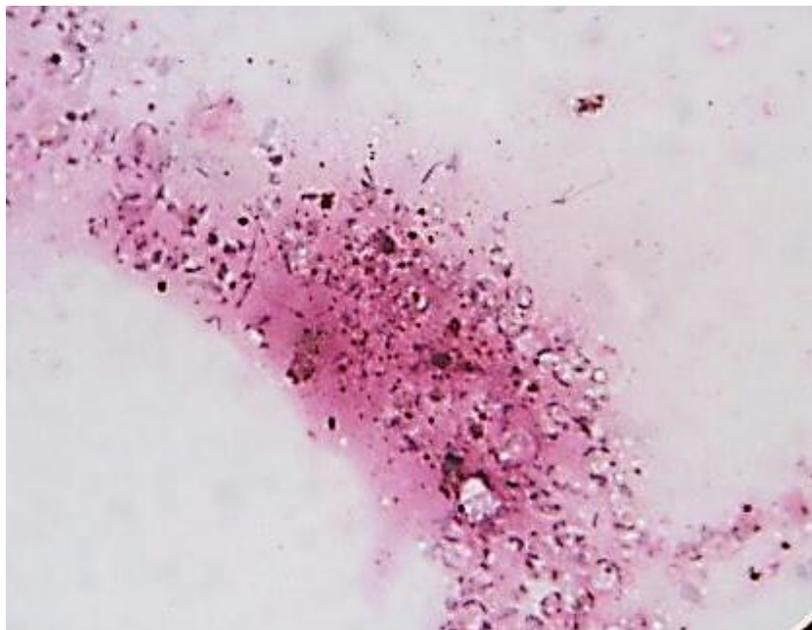


Figure 2:40x- Left cervical node showing Metastatic Melanoma deposit

Following which wide local excision of left lower lip and modified radical neck dissection left side up to level 5 was done. We received elliptical piece of skin with attached portion of lip measuring

6x4x0.5cms in dimension. Skin shows an ulcer nodular growth measuring 5x3x1cms, which was blackish and pigmented. Also received modified radical neck dissection specimen with

submandibular salivary gland. On dissection 10 lymph nodes were made out. Section studied from the tumor show squamous mucosa with junctional activity and an infiltrating neoplasm composed of polygonal and spindle shaped cells with pleomorphic nuclei, prominent nucleoli and brown

pigmentation in the cytoplasm. Submandibular salivary gland is free from tumor deposit and 5 out of 10 lymph nodes show metastatic melanoma deposits. Immunohistochemistry was done with HMB -45 which was found to be intensely positive in the malignant cells.

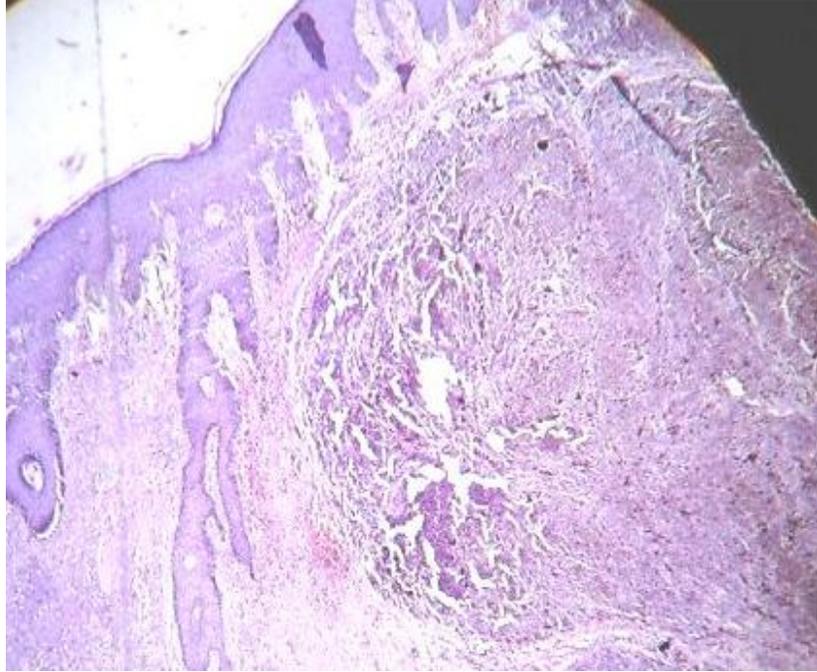
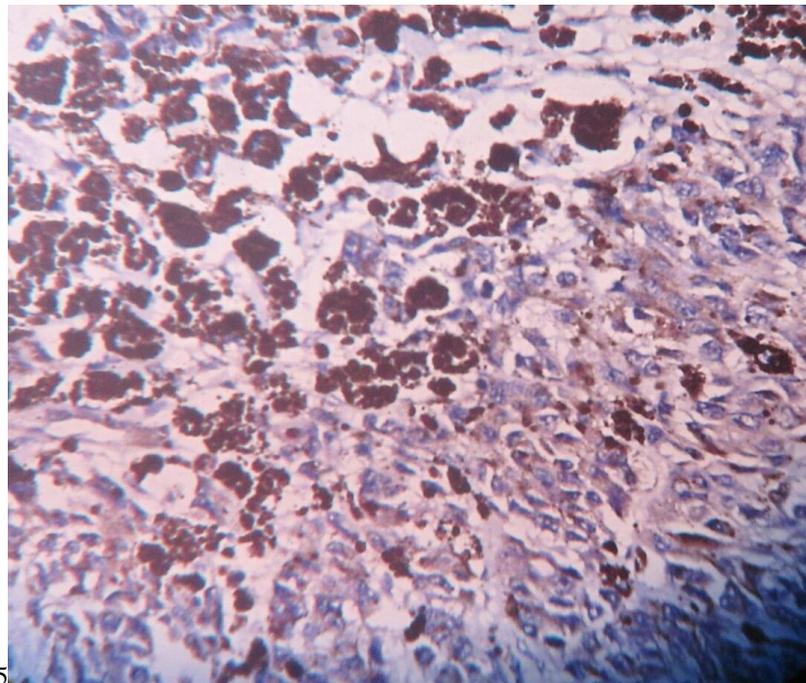


Figure 3:10x view showing clusters of neoplastic cells with junctional activity



5

Figure 4: 40x view showing HMB 45 showing cytoplasmic positivity

DISCUSSION

Malignant melanoma was first described by Weber in 1859. It was recognized as a distinct clinical entity and named as “melanotic sarcoma” by Lucke in 1869 [5]

The World Health Organization (WHO) has defined mucosal malignant melanoma as a malignant neoplasm of melanocytes or of melanocyte precursors [5]. It is characterized by the proliferation of atypical melanocytes at the epithelial-connective tissue interface, associated with upward migration into the epithelium and by invasion of the underlying connective tissues

Melanoma of the oral mucosa is a rare tumor, occurring mostly in men after the fourth decade, with a predilection for the palate and maxillary gingiva versus the predilection sites of oral squamous carcinoma, the tongue and floor of mouth. In this case lower lip was involved.

Oral melanoma is often asymptomatic, with 16% of lesions discovered incidentally. Various presentations of oral melanoma include pigmented macule, nodule or large pigmented exophytic lesion, ulceration, swelling, bleeding nodular mass, rapid enlargement or loosening of the tooth [6]. Satellite foci may surround the primary tumor. The lesions can be bluish-black, brown or amelanotic. They may exhibit asymmetric and irregular borders just like cutaneous melanomas. Oral Melanoma has an initial phase characterized by radial growth followed by a phase of invasion of the underlying tissues (the so called “vertical growth phase”). In the mouth, bony erosion is common.

The etiology of oral melanoma is unknown. Primary oral melanoma is considered only when the following criteria, described by GREENE (1953), are fulfilled: Demonstration of melanoma in the oral mucosa, presence of junctional activity, and inability to demonstrate extra oral primary melanoma [7]. Our case described here, justify the criteria to be considered as primary oral melanoma

Lopez *et al.* identified five types of oral malignant melanoma (OMM) on the basis of clinical appearance: Pigmented nodular type, non-pigmented nodular type, pigmented macular type, pigmented mixed-type, and non-pigmented mixed type [8]. Unpredictable and widespread metastasis is a well-known feature of malignant melanoma. Metastases to regional lymph nodes and distant spread to bone are encountered in end-stage

patients. Lymph nodes, central nervous system, lungs, and liver are also common regions for metastasis [9].

Grossly, tumors are usually 1.5 to 4cms in diameter with a black, macular or nodular surface. The cut surface is often homogenously black or dark pigmented.

Usually oral malignant melanoma can be diagnosed with confidence on hematoxylin and eosin–stained sections. If pigment is completely absent (amelanotic melanoma), immunohistochemical stains are of significant help. Useful markers include S-100 protein, HMB-45, and Mart-1 (Melan-A).

Treatment modality for malignant melanoma is wide surgical excision followed by radiotherapy. Recurrence is quiet common. In the present case, wide local excision with modified radical neck dissection followed by radiotherapy was given. Reconstruction of lower lip with folded deltopectoral flap was done.

Regarding the prognosis, the 5 year survival rate of primary oral melanoma is poor (15%) as compared to cutaneous melanoma (80%) [10]. Six variables are to be identified for assessing prognosis: histological regression, tumor thickness and lymphocytes infiltrating tumor, presence of satellite lesions, site, mitotic rate and sex of the patient.

There can be several reasons for poor prognosis in Primary oral Melanoma. They are: 1. Late diagnosis, 2. Anatomic limitations making radical surgery difficult, 3. Mucosal tumors show rapid invasion to deeper structures, 4. Vascularity of oral mucous membrane, 5. Mucous membrane is thinner than skin because of thinner lamina propria due to thin papillary dermis and absence of reticular dermis, thus most mucosal melanomas progress quickly to vertical growth phase and gain access to the rich vascular and lymphatic network more quickly.

A Protocol has been highlighted for approaching a patient with oral pigmentations: 1. All oral pigmented lesions that could not be clinically diagnosed should be biopsied, 2. Biopsy should be performed from the thickest & darkest region of the lesion, 3. Pathologists should be provided with complete clinical information for biopsies, and 4. Follow-up of the patient should be done which includes thorough abdominal

examination, ultrasound abdomen, CT scan neck, chest X-ray and clinical photographs

diagnosis which can be life saving cannot be overemphasized.

CONCLUSION

Oral malignant melanoma though an extremely rare malignancy is potentially very aggressive and a rapidly invasive tumor. Clinically these tumors are very silent and asymptomatic in their appearance, and hence misleading the clinician. Thus, the importance of early detection and

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Conflict of interest

None declared

Ethical approval

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