DIAGNOSTIC DILEMMAS LEADING TO FATALITY IN MUCOSAL MALIGNANT MELANOMA OF HARD PALATE: A RARE CLINICAL PRESENTATION

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ABSTRACT

We report a case of hard palate mucosal malignant melanoma with progressively enlarging swelling on the right side of the upper neck in a 60-year-old female for which she had taken treatment from many local practitioners. Detailed history of the patient was taken. Investigations carried out are Computed tomography of maxillary region and neck, Fine Needle Aspiration Cytology of right cervical lymph node and incisional biopsy of the tissue from hard palate sent for histopathology then final diagnosis of mucosal malignant melanoma was arrived. Thus to emphasize that early diagnosis and to maintain high index of suspicion for those pigmented lesions occurring in the oral cavity could have improved the prognosis of patient.

Key words: Mucosal malignant melanoma, Cervical lymph node, Computed tomography, Hard palate, Metastasis.

INTRODUCTION

There are several subtypes of melanoma, but mucosal malignant melanoma, also referred to as oral malignant melanoma, primarily affects the oral cavity and is an extremely rare phenomenon. Oral malignant melanoma (OMM) was first described by Weber in 1859. OMM has an estimated incidence of 1.2 cases per 10 million persons per year. Primary oral malignant melanoma is rare disease representing only 0.2–8% of all melanomas. Mucosal malignant melanoma, arising from the uncontrolled growth of melanocytes is a potentially aggressive tumor of melanocytic origin present as a black macule, later it may develop as a nodule or ulceration, with asymmetry and irregular borders. Mucosal melanoma is more frequent among Japanese people and can occur in hard palate, maxillary gingival, labial and buccal mucosa of oral cavity.

The incidence of melanoma has been steadily increasing in the past several decades with an annual increase of 3-8% worldwide. It occurs slightly more often in males, 2.8:1 male to female ratio and the age range is from 20-83 years with an average age of 56 years. Oral malignant melanoma frequently exhibits an extremely aggressive behavior, high index of metastasis and has poor prognosis. Surgery, either alone or in association with radiotherapy, is the preferred treatment modality. Prognosis is poor with a 5-year survival rate varying from 0 to 55%.
CASE REPORT:

A 60 year old female reported to Department of Otolaryngology, Era’s Lucknow Medical College, Lucknow with a complaint of a painless, pigmented patch on right side of anterior hard palate from last one year which was progressively increasing and bleeds on touch. She took treatment from many local practitioners but was not relieved. Later on she developed progressively enlarging swelling on the right side of the upper neck to the present state of about lemon size [Fig.1(a)]. She gave no other history of any systemic illness or trauma to the head and neck region. Her general physical examination was insignificant and her vital signs were under normal limits. Intra oral examination showed non tender non ulcerated nodular soft tissue swelling of bluish-black pigmentation measuring 3.0 2.5 cm in dimension. It extended from right maxillary incisor to first premolar teeth not crossing the midline and on palpation borders are not well defined [Figure 1(b)]. Extra oral examination revealed solitary right upper cervical lymph node palpable about 3.0 2.0 cm in size, which was freely mobile, firm and non-tender [Figure1(a)]. Computed tomography (CT) of maxillary region and neck revealed irregular lobulated lesion of soft tissue density arising from hard palate opposite right maxillary incisor to first premolar teeth and not crossing the midline, measuring approximately 30 10 22 mm size and no bony erosion[Figure 2 (a, b)]. There is also evidence of a large necrotic right level 2 cervical lymphadenopathy measuring 32 21 mm [Fig.2 (c)]. An incisional biopsy of the lesion was performed and histopathological section [Fig. 3] shows parakeratinized stratified squamous epithelium with atypical melanocytes along with melanin pigmentation throughout the stroma proving the diagnosis of mucosal malignant melanoma. Fine Needle Aspiration Cytology of right cervical lymphnode [Fig. 4] shows dispersed population of melanin containing highly pleomorphic cells having high N/C ratio, anisokaryosis and prominent nucleoli with abundant extracellular pigment. Melanin pigment was
confirmed by Masson Fontana silver stain which confirmed it as metastatic node of mucosal malignant melanoma. Work for distant metastases (CT scan of chest, brain and abdomen) was negative.

Based on the clinical examination, radiologic and histopathologic features a final diagnosis of mucosal malignant melanoma was arrived. Medical information was provided to the patient and her family regarding the diagnosis, staging, therapeutic options and prognosis. The patient was referred to a cancer institute with facility of Radio-Chemotherapy and was called for regular follow up. According to her attended she died due to complication of metastasis with in one month of starting chemotherapy after referral.

DISCUSSION:

Primary oral mucosal malignant melanoma is a rare neoplasm, demonstrate significant heterogeneity in morphological features, developmental process and biological behavior that could render the clinical diagnosis extremely difficult and represents 0.5% of all oral malignancies. Blacks, Japanese and Asian patients tend to be disproportionately more affected than Whites.

The differential diagnosis includes melanotic macule, smoking associated with melanosis, post-inflammatory pigmentation, melanoplakia, melanocanthoma, nevi, Addison’s disease, Peutz-Jeghur syndrome, amalgam tattoo, Kaposi’s sarcoma.

Oral melanoma presents as a dark brown, bluish black mucosal discoloration. The lesions may be solitary or multiple, flat and/or elevated, borders are usually irregular, and no clear demarcation exists between the tumor and the adjacent tissues. Rarely, melanoma may present itself without clinical evidence of pigmentation, in which case it is termed as amelanotic melanoma. These lesions are fatal because they have delayed recognition and subsequent delayed treatment leading to further worsening the prognosis.

Histopathologic examination of the lesion remains the most accurate diagnostic tool. Adjunctive radiologic diagnostic methods such as CT, MRI and Positron emission tomography are sometimes useful. The histologic appearance of the tumor is an invasive pattern of growth, with the tumor cells often appearing as densely packed epithelioid or sometimes sarcomatoid cells with eosinophilic cytoplasm. Varying degrees of cellular pleomorphism, tumor invasion of blood vessels and lymphatics are seen. The special stains (Fontana silver stain and the Prussian blue stain) are accurate in only about 75% of the cases. Demonstration of the neuronal specific S-100 protein is a useful diagnostic indicator, especially if the tumor is of amelanotic type. More recently, the application of monoclonal antibody techniques HMB-45 and Mart-1 (Melan A) has increased the specificity of immunohistochemical diagnosis.

Radical surgery is the treatment of choice for oral melanoma. Elective neck dissection has also been advocated along with surgery. Radical surgery in combination with radiotherapy and chemotheraphy or radiotherapy alone is preferred in inoperable tumors or in the elderly. Immunochemotherapy has been shown to be useful as an adjuvant to surgical resection. Chemotherapy is generally reserved for proven metastatic disease. Morton et al demonstrated temporary tumor regression following the intralesional injection of cutaneous melanoma with BCG vaccine. Kirkwood et al reported that melanoma is one of the human cancers which respond to interferon anti-tumor therapy. The future promise of tumor-directed antibodies labeled with cytotoxic drugs may offer hope for improved survival. Malignant melanoma generally has been considered a poorly radiosensitive malignancy.
Primary radiotherapy appeared to be more effective than surgical treatment when the 5-year cumulative survival rates were compared. Postoperative radiotherapy could be of some use; postoperative radiotherapy using fractions of 6 Gy twice a week for a total dose of 30 Gy has to be given\textsuperscript{13}.

Five-year survival for oral melanoma is very poor with a median survival about 2 years\textsuperscript{14}. Gingival location carries better prognosis compared to palatal (median survival 46 vs. 22 months)\textsuperscript{14}. Involvement of lymph nodes affects survival considerably, with a median survival being 46 months, when lymph nodes are not involved, and 18 months when they are involved\textsuperscript{14}. As in our case patient presented with hard palate lesion along with secondary’s neck which has worse prognosis. Apart from late presentation of patients with locally advanced disease, rich vascularity and lymphatic drainage of the mouth favors earlier metastatic spread to regional lymph nodes and to distant sites such as the lungs and vertebral column\textsuperscript{10}.

Early diagnosis is essential for successful treatment and is perhaps the key factor in improving the prognosis of OMM. Surgery is the mainstay of therapy. New adjuvant immunotherapeutic modalities and chemotherapy protocols have been used to improve the survival of patients with more advanced disease.

**CONCLUSION:**

Oral mucosal melanomas are rare oral malignancies with aggressive nature and worst fatal prognosis. Oral cavity presents with many different types of benign and malignant patches which leads to diagnostic dilemma of practitioner’s. Considering various oral patchy lesions and wide range of presenting age of the patient’s, all pigmented lesions in the oral cavity should be examined with suspicion. As in our case, 60 years of age, which presented with painless pigmented lesion from last twelve month was under treatment of many practitioner’s. Earlier investigation and confirmation of diagnosis could have improved the prognosis of our patient. Vigilant comprehensive analysis of published cases and recognition of new ones may be helpful in establishing definite classification and proposing clinical features that would facilitate its early diagnosis, as a prerequisite for timely treatment and better prognosis of this rare pathology.

Hence, the purpose of this study is to emphasize on early diagnosis and to maintain high index of suspicion for those pigmented lesions for oral malignant melanoma occurring in the high risk sites such as palate.

**DISCLOSURES**

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