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CASE REPORT

Primary Mucosal Melanoma: Report of Two Cases

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ABSTRACT

Primary mucosal malignant melanoma is of unusual occurrence and poor prognosis. We report two cases of primary mucosal melanoma, one in a 70-year-old male in the palate and the other in a 73-year-old female in the maxillary sinus. Early diagnosis and proper clinical and cytologic evaluation is necessary to institute therapeutic intervention. These cases are reported on account of their rarity of occurrence and difficulty of treatment.

Keywords: Melanoma, Mucosa, Palate, Maxillary sinus, Case report.

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INTRODUCTION

Primary mucosal melanomas of the head and neck region comprise only about 0.2% of all melanomas.[1] Approximately 1000 patients with this disease have been reported in the literature so far. Weber from Germany in 1856, first described mucosal melanoma and Lucke in 1869 recognized it as a distinct clinical entity.[2] Holdcraft and Gallagher have reported a 5-year survival rate of only 13%.[3]

Here, we report two cases of primary mucosal melanoma.

CASE REPORTS

Case 1

An edentulous male patient aged 70 years, reported with severe pain in the palate with increasing black discoloration over the whole of the hard palate. He had a pre-existing black macule since childhood. The lesion started increasing in depth of pigmentation and became painful since one month. Slight elevation of the macule was also noticed since 2 weeks. The patient was poorly built and nourished. He was under treatment for diabetes mellitus which was under control.

Clinical examination showed a velvety black lesion of 5 × 6 cm in size with well defined, irregular borders extending from buccal vestibule covering one half (right) of the hard palate was observed. The surface appeared smooth and intact. The other half showed asymptomatic melanosis. On palpation, the lesion was extremely painful and firm in consistency. Regional lymph nodes were not palpable.

Fine needle aspiration cytology (FNAC) was positive for melanoma. Histopathology was consistent with malignant melanoma, superficial spreading type. Bleaching of slides was done as the cells were heavily pigmented.

At his next visit after 10 days, the lesion showed mild nodularity with a few satellite lesions of 1 mm diameter (Fig. 1). Maxillectomy was proposed to be done but the patient was deemed unfit to undergo surgery due to poor general health. The patient further developed chest infection which was diagnosed as chronic obstructive pulmonary disease (COPD) and treated. Three weeks after the initial visit of the patient, submandibular lymphadenopathy was noted. He was treated with palliative radiotherapy.

Investigation for distant metastasis was done by whole body positron emission tomography (PET). Distant osseous metastasis was noted 4 months after initial diagnosis and

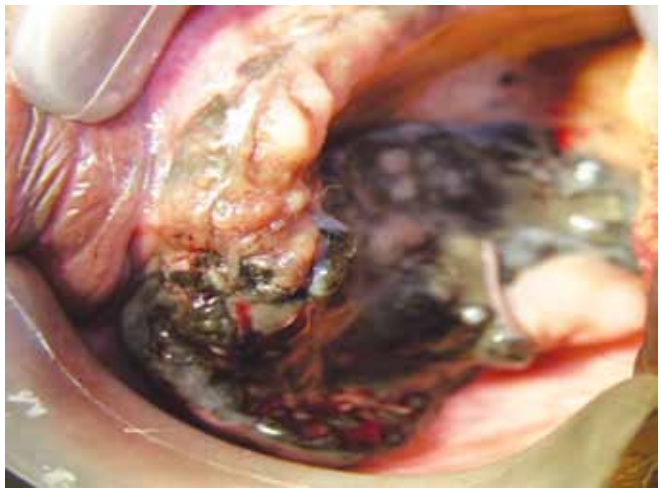


Fig. 1: Oral melanoma—clinical picture showing nodularity

confirmed by computed tomography (CT) and by pap smear (Fig. 2A) followed biopsy (Fig. 2B). Lesion was staged as Clark's III, showing high mitotic index and Breslow index 2 mm. The patient succumbed to the disease 5 months later.

Case 2

A female patient aged 73 years, presented to the Ear Nose Throat department with a complaint of swelling in the right side of the face. The swelling was diffuse and firm in consistency. Regional lymph nodes were palpable on the ipsilateral side. The patient was of moderate build and had no systemic disease.

Whole body PET and CT scan showed regional bone involvement. FNAC of involved lymph nodes revealed melanocytes. Incisional biopsy from the maxillary sinus showed melanoma, superficial spreading type with Clark index II, fair mitotic index and Breslow index 1.5 mm (Fig. 3).

Maxillectomy was done followed by radiation and chemotherapy (Fig. 4). The patient has been under follow-up for 6 months since initial diagnosis and has not shown evidence of nodal or distant metastases.

DISCUSSION

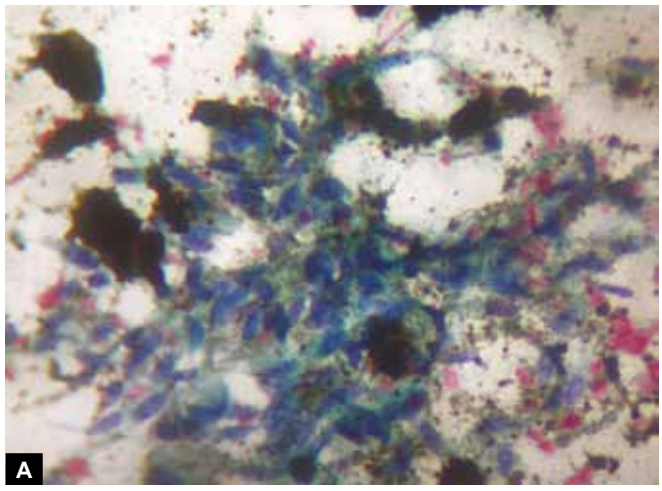
These two cases of mucosal melanoma are reported on account of their rarity of occurrence and the increased necessity of early diagnosis. There is marked difference in both behavior and disease progression between mucosa and cutaneous melanoma.[4,5] Mucosal melanomas have a worse prognosis compared to cutaneous melanomas. A contrasting difference with skin melanoma is that local metastasis does not affect survival.[2] Also exposure to sunlight which is invariably present in cutaneous melanoma does not apply in the etiology of mucosal melanoma.[6]

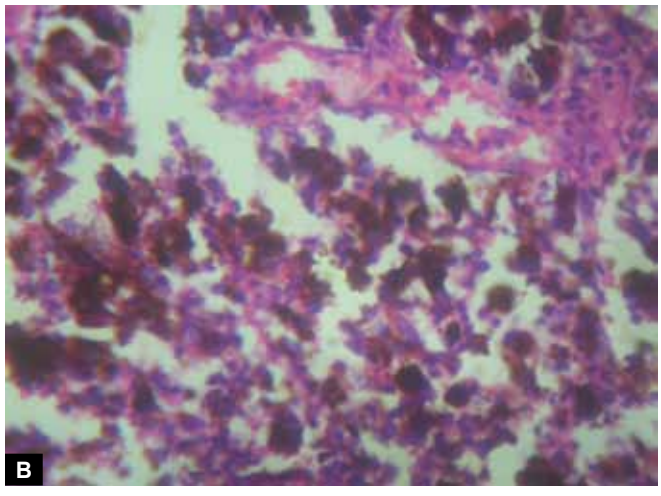
Etiology

Mucosal melanoma is derived from melanocytes of the regions where ectomesenchyme is present. There are fewer incidences in mucosa derived from endoderm. The use of tobacco and exposure to formaldehyde have been implicated but without substantial evidence.[2]

Age, Sex and Race

The neoplasm usually appears in persons after the age of 50 years[4,7] with a male to female ratio of 2:1.[1,4,6,7] It is uncertain whether the incidence of this specific type of melanoma is higher among Asians and blacks or whether the ratio of mucosal melanoma to skin melanoma is different due to a lower incidence of cutaneous melanoma in these racial groups.[2] Typical occurrence of sinonasal mucosal





Figs 2A and B: (A) Pap smear of oral melanoma and (B) H&E section of oral melanoma (high power view)

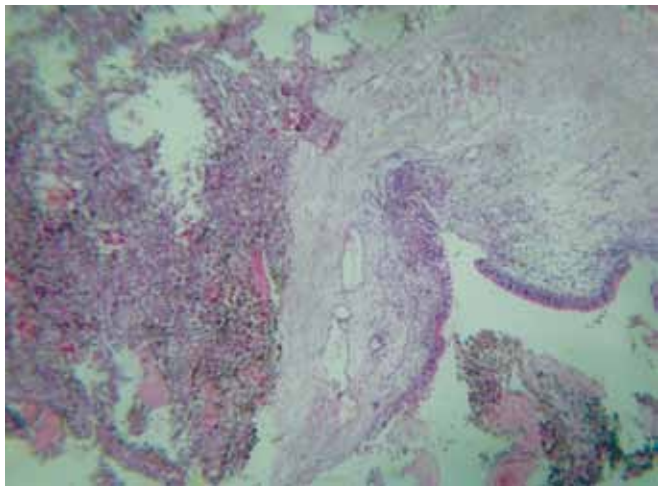


Fig. 3: H&E section of sinus melanoma—low power view

melanoma is 2 decades later than that of skin melanoma and is therefore a disease of the elderly.[8] However, oral cavity melanoma occurs at a younger age than the former.[9]

Site

The hard palate and the maxillary alveolus are the leading sites.[2] Buccal mucosa, mandibular gingiva, tongue, base of the oral cavity and the lips are the other sites in decreasing

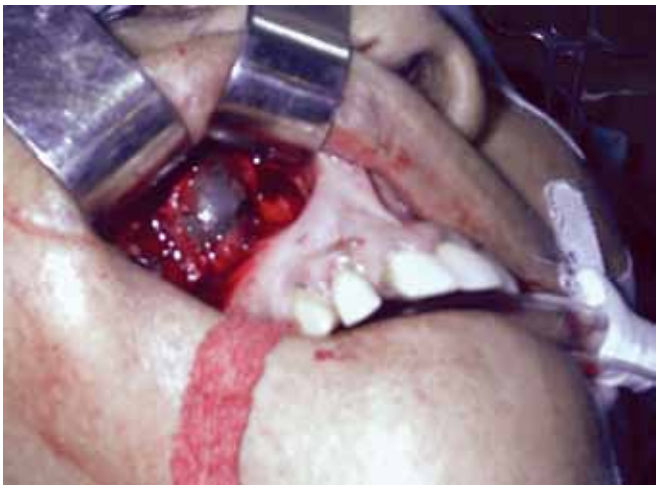


Fig. 4: Sinus melanoma—peroperative view with sinus opened

frequency.[7] The sites of sinonasal melanoma have not been accurately documented.[2]

Signs and Symptoms

The presenting complaints of the patients were unilateral obstruction and epistaxis in 85 to 90% of cases.[10] Pain and facial deformity were encountered in advanced cases.[2]

Asymptomatic oral melanomatosis preceded mucosal melanoma in 30 to 37% of cases. The melanosis represents the radial phase of the growth of the tumor and precedes the vertical component by years. No survival advantage has been demonstrated for patients with pre-existing melanosis.[9]

Diagnosis

Diagnosis of melanoma is done by the identification of intracellular melanin in routine hematoxyllin and eosin, conventional staining with stains such as the Fontana stain and enzyme immunohistochemistry (dopa reaction to demonstrate tyrosinase activity). This requires fresh tissue and also it is not always positive for amelanotic melanomas.[11] Immunohistochemistry is used widely for confirmatory diagnosis where there is cause for doubt. Positive reaction is elicited with antivimentin and NKI/C-3 antibodies and with HMB-45.[11] There is strong reaction with S-100, which is a calcium-binding protein, found in neural tissues. Oral mucosal melanoma stains specifically with antibodies against the alpha subunit of S-100.[12]

Three different cellular types of malignant mucosal melanoma are identified: spindle cell, polygonal cell, and mixed cell. However no correlation between cell type and survival has been established. The oral cavity melanomas resemble acral lentiginous melanoma histologically and clinically.[13]

There is the conspicuous absence of any lymphoplasmacytic reaction to mucosal melanoma.[14] Another feature of mucosal melanomas is angioinvasiveness and the early metastatic spread to distant sites is attributed to this.[2] The rate of survival is inversely proportional to the depth of invasion but has no correlation with size of the lesion.[2] Local failure of surgical treatment is due to the frequent occurrence of multicentricity.[15]

Staging

Staging is as follows: Localized disease (Stage I), metastases to regional lymphatics (Stage II), and distant metastatic disease (Stage III).

Pathologic description and leveling system cannot be applied for mucosal melanomas. The critical factor that determines the prognosis of melanoma of skin is vertical invasion depth; however, the Breslow classification is not a valuable prognostic determinant for oral cavity tumors.[4,5]

Metastasis

Oral melanoma has a predilection for metastasis to the lungs, liver, brain and bones.[4] Lymph node metastasis is seen in approximately 19% of patients at the time of presentation and another 16% develop lymph node metastases after treatment, whereas 10% present with distant metastasis. Local treatment failure occurs in more than 50% of patients and invariably leads to distant metastases. Salvage treatment is effective in only 25% of this cases.[3]

Treatment

Wide local excision is the accepted treatment of choice for oral malignant melanoma.[7] In our case, complementary investigation showed metastatic lesions in one patient. Systemic chemotherapy is used in patients with advanced stage III (nonresectable regional metastases) or stage IV (distant metastases).[1,6] Radiation is seen to offer only a transient response.

CONCLUSION

Primary mucosal melanomas are rare entities in the oral and maxillofacial region. They exhibit a different clinical course and presentation from that of the more common cutaneous melanomas. There are also variations between oral and sinonasal melanomas in the age of occurrence and the difficulties encountered in diagnosis. Distant metastasis occurs mostly due to unsuccessful local treatment.

CLINICAL SIGNIFICANCE

Mucosal malignant melanomas are unpredictable and more aggressive than cutaneous melanomas. Besides, the complex anatomy of this area makes complete surgical excision difficult. Thus, early diagnosis and treatment are of paramount importance.[4]

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