Case Report

Dentistry

An Amelanotic Melanoma of the Oral Cavity- A Rare Entity; Case Report

DARSHANA J. PATIL, AMIT GANDHI, FRENY R. KARJODKAR, MOHAN D. DESHPANDE, SANGEETA B. DESAI

ABSTRACT

Introduction/ Objectives: Amelanotic oral malignant melanoma (AOMM) is a rare tumour that is difficult to diagnose as it lacks melanin pigmentation. The prognosis is poorer than that of pigmented melanomas because of delays in establishing the correct diagnosis and in the initiation of treatment. Amelanotic forms are also thought to be biologically more aggressive than pigmented melanomas.

Methods: A case which reported to our hospital was investigated by taking a through case history, clinical, extra oral, intraoral examination and radiological and laboratory investigations.

Results: at each stage of reporting the feature pointed to a different diagnosis, which was finally confirmed on immunohistochemical examination.

Conclusions : AOMM without radial growth phase may be misdiagnosed as epulis or squamous cell carcinoma. Questionable lesions, particularly maxillary and palatal lesions, must be biopsied for histopathologic and possibly immunohistochemical examinations followed by rapid treatment as the prognosis of AOMM is poor.

Advances in knowledge; The oral cavity being the most accessible area for examination, all the medical personnel should examine the oral cavity without fail and should be able to elicit the changes within it. Since the amelanotic variant may present diagnostic dilemma, proper investigations have to be performed to come to a definitive diagnosis and to plan the treatment thereafter.

Key Words: amelanotic, melanoma, prognosis, HMB45

KEY MESSAGE

- Oral mucosal melanomas have a 'chameleonic' presentation of a mainly asymptomatic condition. The rarity of these lesions, the poor prognosis and the necessity of a highly specialized treatment are the factors that should be seriously considered.

INTRODUCTION

Melanoma is a major health problem. When discovered early and fully excised, melanoma is highly curable. However, once the metastatic disease develops, the treatment options are limited and the survival is generally measured in months. [1]

Primary mucosal melanomas of the head and neck are a rare entity, accounting for 0.2% to 8% of all the malignant melanomas [2]

In the head and neck region, the nasal and oral cavities are the most commonly affected sites.

Oral amelanotic melanomas are rare and the prognosis is poorer than that of pigmented melanomas, because of delays in establishing the correct diagnosis and in the initiation of the treatment.

CASE REPORT

A 75 years old, debilitated, female patient presented to us in February 2011; with a history of swelling with pain and bleeding in the lower anterior teeth region since 2 months, followed by a history of extraction. The swelling had gradually increased over a period of time, leading to inability in closing the mouth.

On clinical examination, a pink, proliferative, sessile growth (5 x 5cm) with focal areas of ulceration, involving the lower alveolar ridge and the buccal and lingual vestibules, was present. [Table/Fig 1].The upper and lower anterior teeth were missing and the remaining teeth were not mobile.

The swelling was soft on palpation, with no blanching. The submandibular lymph nodes were palpable.

A provisional diagnosis of pyogenic granuloma [3] was made and the patient was referred for investigations.

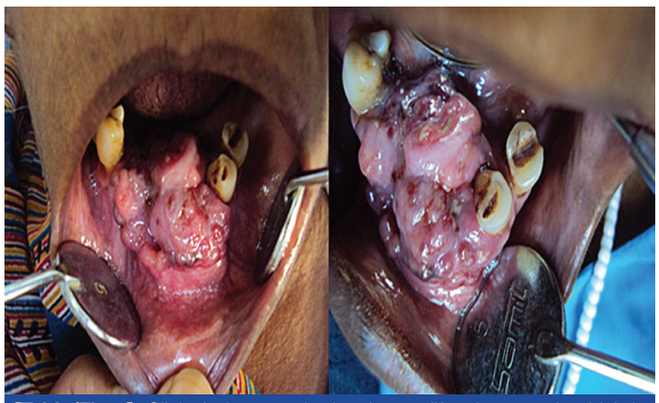
Her panoramic radiograph showed osteolytic changes with respect to the mandibular anterior edentulous region [Table/Fig 2]

Her CT scan showed an ill defined, heterogeneously enhancing, soft tissue lesion (3.29 x1.46 cm) involving the gingivobuccal sulcus, along with the destruction of the alveolar ridge. [Table/Fig 3A and B] The anterior parts of the genioglossus and the mylohyoid were lost. [Table/Fig 3C] An enlarged lymph node on the left side was compressing the left jugular vein. [Table/Fig 3D]

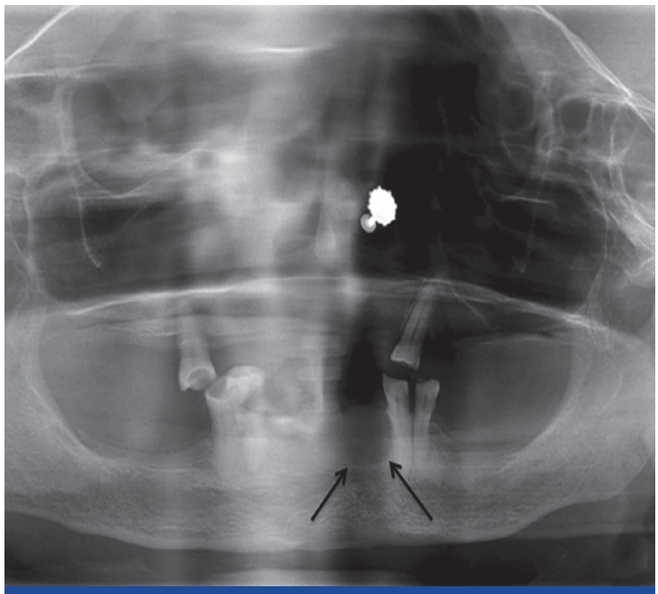
The provisional radiographic diagnosis was malignancy.

Distant metastasis was not found on the clinical, radiographic and ultra sonographic examinations of the patient.

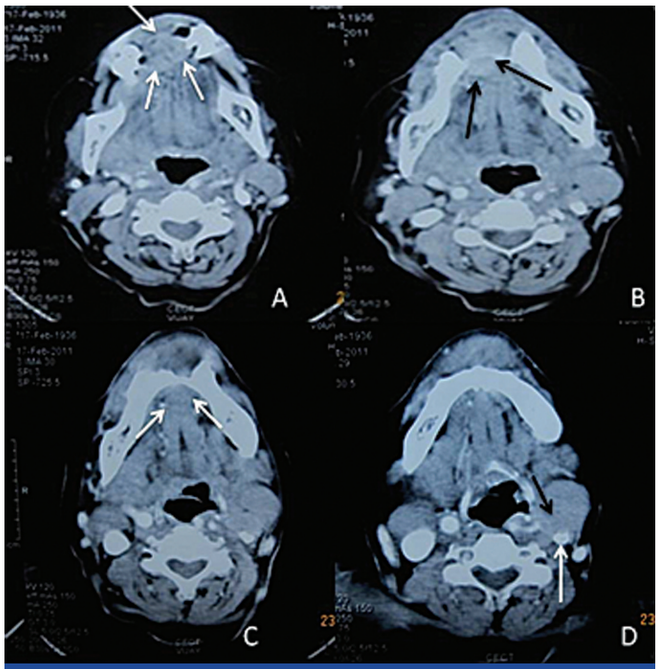
An incisional biopsy of the lesion was done under local anaesthesia, after taking the consent of the patient. The H and E stained sections



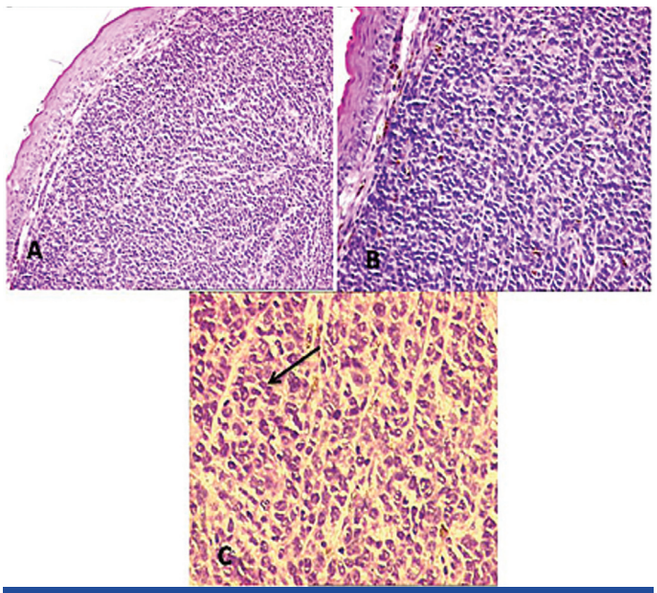
[Table/Fig-1]: Clinical photograph showing proliferative growth which is sessile, 5cm x 5cm in size, pink with focal areas of ulceration involving lower alveolar ridge, buccal and lingual vestibules.



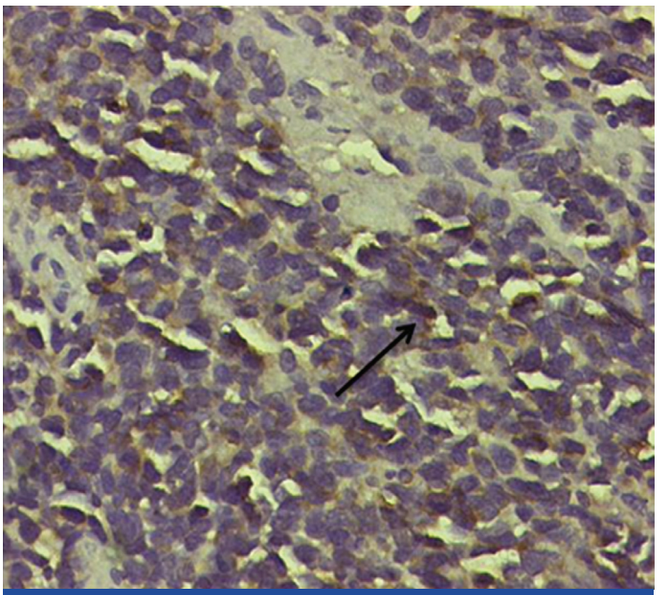
[Table/Fig-2]: Panoramic radiograph showing osteolytic bony changes (arrows) in relation to mandibular anterior edentulous region.



[Table/Fig-3]: Computed Tomogram showing; A & B shows 3.29 x1.46 cm sized ill defined, heterogeneously enhancing soft tissue lesion involving gingivobuccal sulcus along with destruction of alveolar ridge. C shows destruction of anterior part of genioglossus, mylohyoid muscles. D shows enlarged lymph node on left side (marked with black arrow) compressing left jugular vein (marked by white arrow).



[Table/Fig-4]: Histopathological picture of H & E stained section showing blue round cells with hyper chromatic nuclei and scanty cytoplasm. A, B and C showing images with 100X, 200X, 400 X, resolutions respectively. C shows blue round cells with hyper chromatic nuclei and scanty cytoplasm (black arrow).



[Table/Fig-5]: Immunohistochemistry showing HMB45 positive melanocytes (arrow).This image is with 400X resolution.

showed blue, round cells with hyper chromatic nuclei and scanty cytoplasm [Table/Fig 4], which were suggestive of Blue Round Cell Neoplasm with a differential diagnosis of

- Anaplastic large-cell lymphoma. [4]
- Poorly differentiated squamous cell carcinoma. [4] and
- Malignant melanoma.

Immuno histochemistry was performed by running a panel that showed tumour cells which were focally positive for HMB45 [Table/Fig 5] and were negative for the S100 protein, CK, LCA, CD20 and CD138.

This helped us to reach the final diagnosis of *ORAL AMELANOTIC MELANOMA*.

The patient was referred to the oncosurgeons. As the disease was in its advanced stage with the involvement of the level 2 and level 3 lymph nodes and considering the age of the patient, the

oncosurgeons opted for radiotherapy instead of surgical treatment. Unfortunately, the patient refused the treatment. At present, there is no change in her condition, but she has reported an increased bleeding from the lesion.

DISCUSSION

Oral mucosal melanoma accounts for only 0.5% of all the oral neoplasms [5]

These are highly malignant tumours with a tendency to metastasize or locally invade tissues more readily than other malignant tumours of the oral cavity [5] Rapini et al [6] found that the highest percentage of oral melanomas occurred in the age range of 41 to 60 years and that they were rare before the age of 20 years; males being more frequently affected. [6, 7] Oral melanoma can be expected to exist in the following types; Superficial spreading, Acral- lentiginous or Nodular. The most frequent site of their occurrence is the hard palate, followed by the maxillary gingival [5], the mandible, tongue, buccal mucosa and the upper and lower lips.

Intra oral malignant melanomas usually remain asymptomatic and may be detected only when there is ulceration of the overlying epithelium and/or haemorrhage [8]; this may be the reason for the poor prognosis of oral malignant melanomas, with the 5-year survival rate being between 15% to 38% [7]. In addition, the rich vascular supply which is present in the oral cavity may further contribute to the dissemination of the melanomas [9].

Rarely may melanoma present itself without clinically evident pigmentation; which is termed as amelanotic melanoma. These lesions tend to have a worse prognosis because of their delayed recognition and subsequent treatment [10]. Amelanotic melanoma accounts for 5-35% of all the oral melanomas, which appears as a white, mucosa-coloured, or red mass. Its prognosis is poorer than that of pigmented melanomas because of the delays in establishing the correct diagnosis, due to histological misdiagnosis and due to delays in the initiation of the treatment. The common sites of metastasis are the lymph nodes, the liver and the lung, with a widespread involvement occurring in advanced disease [11]

While the recommended treatment is ablative surgery with tumour-free margins in combination with chemotherapy and to a lesser extent, immunotherapy or irradiation, there is a recognized need for an evidence-based treatment protocol choice. but probably, multimodal therapy may prove to be more effective in the treatment of oral mucosal melanoma [5]

Westbury [12] describes a clinical classification as follows: I-only primary tumour present. II- metastasis present (IIa-adjacent skin involved, IIb-regional lymph nodes involved, II-ab adjacent skin and regional lymph nodes involved) and III-metastasis beyond regional lymph nodes. The patient who has been presented here falls into the classification of IIb, because satellite lesions were present at the time of the initial presentation. Melanoma is notorious for its unpredictable and widespread metastasis. The metastasis in the oral regions usually involves the soft tissues, notably the tongue [13]. The need for biopsies of infra oral melanomas cannot be emphasized. In many cases, the melanomas may appear to be relatively innocuous [7]; often an excisional biopsy can be accomplished. However, an incisional biopsy is acceptable for the larger lesions and must be performed in the darkest and thickest areas of the lesion. According to Batsakis [7], “there is no evidence that a preliminary biopsy of the primary lesion increases the risk of metastatic dissemination or that it unfavourably affects the prognosis”. Any pigmented growth that may appear to be innocuous needs to be biopsied at the earliest. Regression in melanoma is a well-recognized phenomenon [13]. The rather non-specific features of regressed melanoma (apparently inflammatory nodules, depigmented patches and flat or slightly depressed scars) are easily missed or discounted, unless the patient has noticed the regression[14] The primary mode of treatment for malignant melanoma is wide surgical resection [10]. In a review on the outcome of primary mucosal melanomas which were treated only with radiation [15], it was found that 44% of the patients survived for a period of 4.5 years of follow-up. Even though these results appear to be encouraging, radiation is most often used as a supplementary mode of treatment after surgery or after the failure of a previous management. When metastasis is not present after the examination and investigations, surgery would be the preferred option for the treatment. Advances in surgical techniques may allow a more extensive resection and reconstruction. Consideration should also be given to radiation therapy or combined therapy.

In the present case, as the patient was in a debilitated state, and as the disease was in its advanced stage with the involvement of the level 2 and level 3 lymph nodes and considering the age of the patient, the oncosurgeons opted for radiotherapy instead of surgical treatment. The prognosis was so poor (although death might not ensure for many years) that it seemed unjustifiable to embark on a massive removal, especially as the tumour had already metastasized.

CONCLUSION

The main contribution of this case report is an alarm to the oral physicians and the medical personnel who may not be able to diagnose these lesions because of their asymptomatic behaviour. The oral cavity being the most accessible area for examination, all the medical personnel should examine the oral cavity without fail and they should be able to elicit the changes within it. Since the amelanotic variant may present a diagnostic dilemma, proper investigations have to be performed to come to a definitive diagnosis and to plan the treatment.

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AUTHOR(S):
1. Dr. Darshana J. Patil
2. Dr. Amit Gandhi
3. Dr. Freny R. Karjodkar
4. Dr. Mohan D. Deshpande
5. Dr. Sangeeta B. Desai

PARTICULARS OF CONTRIBUTORS:
1. Department of Oral Medicine and Radiology, Nair Hospital Dental College, Mumbai, Maharashtra- 400 008, India
2. Department of Oral Maxillofacial Surgery, Nair Hospital Dental College, Mumbai, Maharashtra- 400 008, India
3. Professor and Head, Department of Oral Medicine and Radiology, Nair Hospital Dental College, Mumbai, Maharashtra- 400 008, India
4. Associate Professor, Department of Oral Maxillofacial Surgery, Nair Hospital Dental College, Mumbai, Maharashtra- 400 008, India
5. Professor and Pathologist Tata Memorial Hospital, Mumbai

NAME, ADDRESS, TELEPHONE, E-MAIL ID OF THE CORRESPONDING AUTHOR:
Dr. Darshana Patil
5, Vikas Nagar, Sakri, 424304
Dhule, Maharashtra, India
Mobile : 09860989148
Email : drpatildarshana@gmail.com

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