

Case Report

Primary mucosal melanoma of maxillary gingiva with metastasis: A rare case report with brief review of literature

Jimsha Vannathan Kumaran, Mariappan Jonathan Daniel, Mithunjith Krishnan¹, Divakar Seetharaman

Department of Oral Medicine and Radiology, Mahatma Gandhi Postgraduate Institute of Dental Sciences, ¹Department of Conservative Dentistry and Endodontics, Indira Gandhi Postgraduate Institute of Dental Sciences, SBV University, Puducherry, India

Abstract

Primary malignant melanoma of the oral cavity is a rare neoplasm. Over 90% of melanoma occurs in skin, but they may also arise from the mucosal surfaces or other sites where neural crest migrates. Intraorally, the most common sites are the palate and gingiva followed by mandibular gingiva and lip mucosa. Oral melanoma is initially asymptomatic. It may develop as a slowly growing mass and be present for months or years before being noticed. Mucosal melanoma of the head and neck has poor prognosis, with the survival of a patient with mucosal melanoma being less when compared with cutaneous melanoma, so early diagnosis is important.

Keywords: Melanocytes, metastasis, primary malignant melanoma

Address for correspondence: Dr. Jimsha Vannathan Kumaran, Department of Oral Medicine and Radiology, Mahatma Gandhi Postgraduate Institute of Dental Sciences, Puducherry, India.
E-mail: dr.jimsha.mds@gmail.com

INTRODUCTION

Melanoma is a malignant neoplasm of melanocytic origin. It is one of the most biologically unpredictable and deadly of all human neoplasms, and the most common forms of melanoma are the cutaneous and the ocular. It is the third most common cancer of the skin. Oral mucosal melanoma is a rare malignancy with the tendency to metastasize and locally invade tissues more readily than other malignant tumors of the oral cavity. Oral malignant melanoma was first described by Weber in 1859.^[1,2] Later, it was named as “melanotic sarcoma” by Lucke in 1869.^[3] The World Health Organization has defined mucosal malignant melanoma as a malignant neoplasm of melanocytes or of melanocyte precursors. Mucosal melanoma involving the sinonasal cavity, oral cavity, pharynx, larynx, and upper esophagus is extremely rare

and accounts for only 0.5% of all oral neoplasms. Nearly 80% of oral melanomas (OMs) arise in the mucosa of the upper arch, with the majority occurring on keratinizing mucosa of the palate and alveolar gingivae.^[3,4] It occurs between 30 and 90 years of age, with a higher incidence in the sixth decade with a mean age of 56 years.^[5] It is having a higher prevalence in yellows, blacks, Japanese, and Indians of Asia due to more frequent finding of melanin pigmentation in oral mucosa of these races. The delayed detection may be the cause for the poor prognosis, with a 5-year survival being between 15% and 38%.^[6,7] This case report is of a mucosal melanoma of the maxillary gingiva with distant metastasis to the liver and lung which has been discussed with detailed radiological investigations including contrast-enhanced computed tomography (CECT) and histopathology to emphasize the necessity for early diagnosis and treatment of this lesion.

Access this article online	
Quick Response Code:	Website: www.ijcpc.org
	DOI: 10.4103/ijcpc.ijcpc_11_17

This is an open access article distributed under the terms of the Creative Commons Attribution-NonCommercial-ShareAlike 3.0 License, which allows others to remix, tweak, and build upon the work non-commercially, as long as the author is credited and the new creations are licensed under the identical terms.

For reprints contact: reprints@medknow.com

How to cite this article: Kumaran JV, Daniel MJ, Krishnan M, Seetharama D. Primary mucosal melanoma of maxillary gingiva with metastasis: A rare case report with brief review of literature. *Int J Clinicopathol Correl* 2017;1:61-5.

CASE REPORT

A 57-year-old woman reported to our department with a chief complaint of pain in the upper back teeth for the past 1 year; the patient was asymptomatic 1 year back after which she developed pain in relation to the left upper posterior teeth, and she also noticed a black-colored swelling in relation to the left maxilla which progressively increased to the present size, but she did not give any attention for the same. For the past 6 months, there was a rapid increase in the size of the swelling, and the intensity of pain also increased. The patient also gave a history of difficulty in swallowing, speaking, and taking food, and there was no history of nasal stuffiness, nasal discharge, and epistaxis. The patient had been treated with radiotherapy for cervical cancer 17 years back; there is no relevant dental history. She is married and has three children. There is no history of any harmful habits.

On general examination, she was thin built and moderately nourished and the vital signs appeared to be within normal limits. Lymph node examination revealed an enlargement of both the right and left submandibular lymph nodes measuring around 1 cm in size and were firm, tender, mobile, and multiple in number. On intraoral examination, a diffuse blackish exophytic pigmented growth was present in the left maxillary gingiva measuring around 4 cm × 4 cm in size approximately extending from distal aspect of 24 till the maxillary tuberosity, extending to palatal gingiva [Figure 1]. The growth was sessile with rough surface and irregular borders, and there was a mild evidence of necrotic slough. On palpation, all inspectory findings confirmed that the growth was firm to hard in consistency and severely tender with indurated margins, and there was no discharge. The teeth in the vicinity of swelling were Grade 2 mobile and depressible into their socket. On the basis of history and clinical findings, a provisional diagnosis of malignant melanoma in the left side of the maxillary gingiva was given. Blood investigations revealed a slight increase in the eosinophil count, and other parameters were within normal limit. Thermal vitality test was performed, and the teeth 26, 27, and 28 were found to be nonvital. Maxillary lateral occlusal radiograph revealed radiolucency in the left side of the maxilla extending posteriorly to the distal aspect of 28 with displacement of 27 and 28 [Figure 2]. Orthopantomogram reveals ill-defined noncorticated radiolucency extending from distal aspect of 26 till the tuberosity. A breach in the continuity of the floor of the maxillary sinus was evident with complete loss of crown in relation to 28 and bone surrounding the 27 and 28 giving a tooth-floating appearance [Figure 3]. Paranasal

sinus view revealed haziness in relation to the left maxillary sinus when compared to contralateral side [Figure 4]. CT scan of the head and neck showed a hypodense mass in the left maxillary alveolus extending to the surrounding area with complete destruction of bone in the molar area [Figure 5]. Enlarged enhancing bilateral Level I, II lymph nodes were also seen.



Figure 1: Intraorally, a blackish exophytic growth in the left side maxillary gingiva

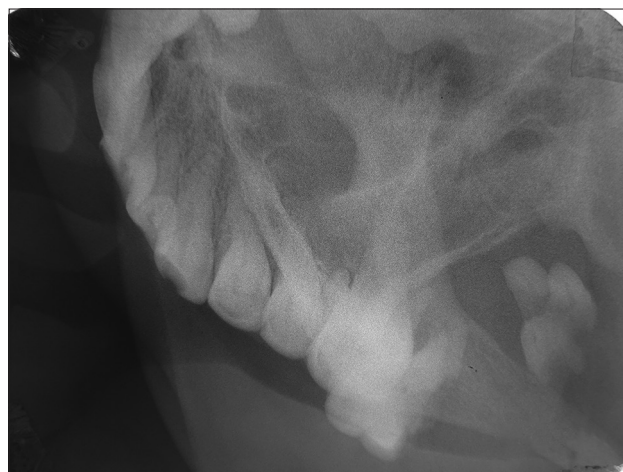


Figure 2: Maxillary lateral occlusal view – well-defined radiolucency in the left side maxilla extending to posterior part of palate



Figure 3: Orthopantomogram – well-defined noncorticated radiolucency in the periapical region of 27 extending posteriorly involving the maxillary tuberosity

CECT of thorax [Figure 6] and abdomen [Figure 7] showed multiple scattered ground glass nodules of varying sizes in bilateral lung fields, with feeding vessel sign impression of lung metastasis, multiple scatters hyperdense lesions in both the lobes of liver postcontrast portal phase lesions appeared iso attenuating and masked with surrounding parenchyme suggestive of liver metastasis. Thoracolumbar spine and pelvis showed osteopenia. An incisional biopsy was performed under local anesthesia, and the lesional tissue was sent for histopathologic examination. The malignant cells were in nest or cluster and in groups in an organoid fashion. The malignant epithelioid melanocytes showed vesicular nuclei with prominent nucleoli, nuclear pleomorphism, hyperchromatism, and loss of cohesiveness of the cells, confirming the diagnosis of malignant melanoma. Based on the clinical examination and radiologic and histopathologic features, a final diagnosis of malignant melanoma of the maxillary alveolar gingiva with distant

metastasis to the lung and liver was arrived. Medical information was provided to the patient and his family regarding the diagnosis, staging, therapeutic options, and prognosis. The patient was referred to the regional cancer center for further management.

DISCUSSION

OM is a rare malignant disease, which represents only 2%–8% of all melanomas. This tumor is of melanocytic origin. Melanocytes are the neural crest-derived cells that are present within the basal cell layer of the skin and mucous membrane. OM is thought to arise either *de novo* from melanocytes or from the preexisting oral pigmentations.^[8,9] The cytogenetic alteration acquired within the oncogenes, tumor suppressor genes, and DNA repair genes of the melanocytes is said to play a role in the etiopathogenesis of melanoma. The inadequate regulation of biosynthesis of melanin by the melanocytes leads to increased oxidative



Figure 4: Paranasal sinus view – haziness in the left side maxillary sinus



Figure 5: Axial computed tomography section reveals hypodense mass in the left maxillary alveolus with destruction in the bone



Figure 6: Contrast-enhanced computed tomography thorax – multiple scattered ground-glass nodules of varying sizes are noted randomly in bilateral lung fields

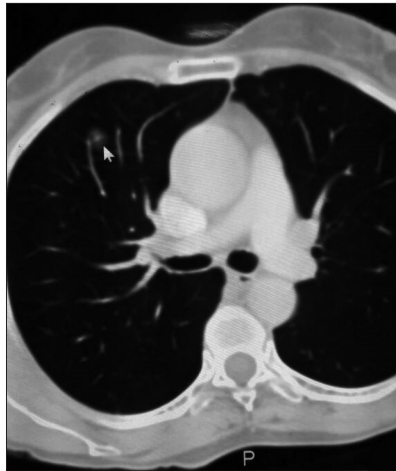


Figure 7: Contrast-enhanced computed tomography abdomen – multiple scattered hypodense lesions in both the lobes of the liver

stress and metabolic by-products that cause DNA damage in the affected melanocytes and thus favor cancer initiation and progression.^[10] Epidemiological studies suggest that incidence is increasing in patients, especially males older than 45 years, but melanoma most commonly occurs in women of childbearing age, and the youngest case reported to date involved an 8-month-old African-American girl with sinonasal melanoma.^[11] Clinically, mucosal melanomas present as a painless, dark brown or black colored, macular or nodular growth. The ABCDE rule of melanomas is followed where A stands for asymmetry; B is for borders that show irregular blurred, notched, or ragged edges; C is for color, usually nonuniform, pigmentation varies from brown, black, tan, red, white, or blue; D stands for diameter of lesion that is usually >6 mm; and E is for elevation or a raised surface.^[12,13] Clinical symptoms vary from swelling, pigmented growth, bleeding, pain unhealed extracted socket, and mobility of teeth. Clinically OMs are classified into five types, i.e., pigmented nodular, nonpigmented nodular, pigmented macular, pigmented mixed, and nonpigmented mixed.^[14]

There are four main clinic pathologic subtypes of melanoma such as superficial spreading melanoma, lentigo maligna melanoma, acral lentiginous melanoma, and nodular melanoma. In the first three types, initial growth is characterized by radical extension of tumor cells. These lesions show good prognosis when they are diagnosed at the early stage but in the nodular type there is a vertical invasion in to the underlying connective tissue.^[15] According to the American Joint Committee on Cancer staging primary mucosal melanoma of the head and neck in this case, it was stage 4-C (any T, any N, M1) in the area of advanced diseases. Common sites for metastasis are lung, bone, brain, and liver with a widespread involvement occurring in advanced diseases.^[16] The treatment of OM is still controversial excision of primary lesion, preferably using an intraoral approach, and involving at least 1.5 cm of healthy tissue is recommended. Surgery can be combined with radiotherapy, chemotherapy, or immunotherapy. Postoperative radiotherapy is generally recommended if poor prognostic pathological features are present. Even though melanoma is regarded as poorly radiosensitive, other irradiation modalities such as intraoral mold, intraoral electron beam, or interstitial brachytherapy have also been used.^[17,18] Initially, melanoma is totally asymptomatic, and sometimes, pigmentation may not be noticed by the patients because of its biologically aggressive nature and delayed diagnosis; the prognosis of these lesions is very poor. The 5-year survival rates range from 6.6% to 20.0%.^[19] Various clinical parameters, including tumor site, age of the patients >60 years, being male, and regional or distant

metastasis also, are predictive of poor prognosis. In this case, the patient is already having distant metastasis to the liver and lung, so the prognosis is poor.^[20,21]

CONCLUSION

In the present case, an undiagnosed macular blackish pigmentation preceding the development of the malignant melanoma and the late diagnosis with evidence of distant metastasis led to poor prognosis. Hence, early detection and treatment is essential for better prognosis in the case of malignant melanoma and the need for clinicians to meticulously examine the oral cavity and make a careful observation. Follow-up of any pigmented lesion is mandatory.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Pour MS. Malignant melanoma of oral cavity. *J Dent* 2007;4:44-51.
2. Ullah H, Vahiker S, Singh M, Baig M. Primary malignant mucosal melanoma of the oral cavity: A case report. *Egypt J Ear Nose Throat Allied Sci* 2010;11:48-50.
3. Elneel Ahmed MA, Karrar MA, El-Siddig AA, Zulfu A. Case report oral malignant melanoma: A rare case with unusual clinical presentation. *Pan Afr Med J* 2015;1-5:86-8.
4. Cockerell CJ. The pathology of melanoma. *Dermatol Clin* 2012;30:445-68.
5. Gu GM, Epstein JB, Morton TH Jr. Intraoral melanoma: Long-term follow-up and implication for dental clinicians. A case report and literature review. *Oral Surg Oral Med Oral Pathol Oral Radiol Endod* 2003;96:404-13.
6. Delgado Azañero WA, Mosqueda Taylor A. A practical method for clinical diagnosis of oral mucosal melanomas. *Med Oral* 2003;8:348-52.
7. Green TL, Greenspan D, Hansen LS. Oral melanoma report of a case. *J Am Dent Assoc* 1986;113:627-9.
8. Twyana R, Hong Z. Malignant melanoma in the oral cavity: A rare case report with review of literature. *J Nepal Dent Assoc* 2015;15:52-8.
9. Pai A, Prasad S, Patil BA, Dyasanoor S, Hedge S. Oral pigmentation: Case report and review of malignant melanoma with flow charts for diagnosis and treatment. *Gen Dent* 2012;60:410-6.
10. Hendzel OW, Słowińska M, Olszewska M, Rudnicka L. Melanoma of the oral cavity: Pathogenesis, dermoscopy, clinical features, staging and management pathogenesis epidemiology. *J Dermatol Case Rep*

Kumaran, *et al.*: Primary mucosal melanoma of maxillary gingiva

- 2014;3:60-9.
11. Ravid JM, Esteve JA. Malignant melanoma of the nose and paranasal sinus, juvenile melanoma of the nose. *Arch Otolaryngol* 1960;72:431-44.
 12. Biradar V, Latturiya R, Biradar S, de Nascimento JS, Taylor AM, de Almeida OP, *et al.* *J Clin Exp Pathology* 2014;4:1000197.
 13. Rani GS, Kumar TV, Kolasani B, Begum MR, Priya Srinivasan A. Case report primary malignant melanoma of maxilla: Report of a case with discussion. *Case Rep Dent* 2014;2014:624306.
 14. Tanaka N, Amagasa T, Iwaki H, Shioda S, Takeda M, Ohashi K, *et al.* Oral malignant melanoma in Japan. *Oral Surg Oral Med Oral Pathol* 1994;1:81-90.
 15. Khalifa H, Abdullah S, Sallam K, Khalil H, Moneim IA, Elaffandi A, *et al.* Primary malignant melanoma of the tongue. *Can J Surg* 2009;52:E309-11.
 16. Lopez-Graniel CM, Ocholo-Carrillo FJ. Malignant melanoma of the oral cavity diagnosis and treatment experience in Mexican population. *Oral Oncol* 1999;35:425-30.
 17. Hicks MJ, Flatiz CM. Oral mucosal melanoma epidemiology and pathobiology. *Oral Oncol* 2000;36:152-69.
 18. Umeda M, Shimada K. Primary malignant melanoma of oral cavity its biological classification and treatment. *Br J Oral Maxillofac Surg* 1994;32:39-47.
 19. Shibuya H, Takeda M, Matsumoto S, Hoshina M, Suzuki S, Takagi M, *et al.* The efficacy of radiation therapy for a malignant melanoma in the mucosa of the upper jaw: An analytic study. *Int J Radiat Oncol Biol Phys* 1993;25:35-9.
 20. Nascimento JS, Taylor AM, Paes de Almeida O, de Andrade BA. Primary oral melanoma: A case report with immunohistochemical findings. *J Clin Exp Pathol* 2014;4:6-8.
 21. Biradar PM, Latturiya R, Biradar S. Late diagnosis of oral mucosal melanoma: Case report. *J Dent Allied Sci* 2012;1:85-7.