

## A Rare Case of Gingivomaxillary Melanoma: Case Report and Systematic Review

Abdelghafour Jaifi<sup>1\*</sup>, Mohamed Salah Koussay Hattab<sup>1</sup>, Yassine Bennaoui<sup>1</sup>, Fadwa Mourabit<sup>1</sup>, Samir Maidame<sup>1</sup>, Houssam Ghazoui<sup>1</sup>, Zakaria Aziz<sup>1</sup>, Nadia Mansouri<sup>1</sup>

<sup>1</sup>Maxillofacial Surgery Department, University Hospital Mohammed VI of Marrakech, Morocco

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\*Corresponding author: Abdelghafour Jaifi

Maxillofacial Surgery Department, University Hospital Mohammed VI of Marrakech, Morocco

### Abstract

### Case Report

Melanomas are malignant tumors originating from melanocytic cells, primarily affecting the skin but also found in mucosal regions due to the migration of neural crest cells. Oral cavity melanoma is extremely rare, comprising 0.5% of oral malignancies, predominantly occurring in the maxillary region, particularly the hard palate and gingiva. Compared to cutaneous melanoma, oral melanomas typically manifest in older patients. Clinically, oral melanomas present as macular, nodular, or ulcerated lesions with irregular, asymmetrical borders and varied pigmentation. Symptoms such as tooth mobility, bleeding upon palpation, and delayed healing post-extraction suggest malignancy. Diagnosis is based on histopathological examination of biopsies, complicated by the tumor's architectural and cytological polymorphism, necessitating immunohistochemical studies for confirmation. Key diagnostic biomarkers include S100 protein, HMB45, tyrosinase, Melan A, and MITF. The primary treatment is surgical resection with wide margins and lymph node dissection. Radiotherapy is typically reserved for cases with insufficient surgical margins or as an alternative for elderly patients, while chemotherapy is indicated for metastatic forms. Despite treatment, oral melanomas have a poor prognosis, with a 5-year mortality rate of 95% and median survival of 1-2 years post-diagnosis, primarily due to metastases in the lungs, liver, brain, and bones. In summary, oral melanomas are highly aggressive and recurrent, underscoring the importance of early diagnosis and rapid intervention to improve survival outcomes.

**Keywords:** Melanoma, Maxilla bone, immunohistochemical study, surgery, prognosis, recurrence.

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## INTRODUCTION

Melanomas are highly aggressive malignant tumors that originate from melanocytic cells, which produce the melanin pigment. Although these tumors primarily affect the skin, they can also occur in mucosal regions due to the migration of neural crest cells. Oral cavity melanoma is an exceptionally rare type, representing only 0.5% of all oral malignancies and 0.2-8% of all melanomas. These tumors are predominantly found in the maxillary region, particularly the hard palate and gingiva. Oral melanomas exhibit unique epidemiological and clinical characteristics compared to cutaneous melanomas, necessitating specialized diagnostic and therapeutic strategies.

## CASE REPORT

50-year-old patient, diabetic DT2 on ADO, consulted the maxillofacial surgery department of the Marrakech University Hospital for a median gingivomaxillary ulcerating swelling that had been

evolving for 03 months and interfered with mouth opening, mastication and speech, associated with an altered general condition.

Endo-buccal examination revealed a pigmented, ulcerating-bourgeois swelling on the vestibular side of the upper incisor region, subtotally filling the oral cavity and bleeding on contact (Figure 1).

Facial CT revealed a lytic lesional process centred on the left maxillary bone, with locoregional infiltration and multiple bilateral laterocervical adenopathies. (Figure 2).

A biopsy was performed with anatomopathological study and immunohistochemical analysis objectifying a nodular mucosal melanoma, invading submucosal tissues associated with endolymphatic emboli.

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CT TAP revealed no secondary lesion, and the tumor was classified T4aN1MO according to American joint committee on cancer: cancer stage (Mucosal Melanoma of the Head and Neck Staging update 05 June 2018).

The therapeutic decision was an en bloc excision of the tumour via a Weber-Ferguson transfacial approach, combined with a transoral palatal incision and subtotal maxillectomy with the following borders: the pterygoid processes posteriorly, the roof of the maxillary sinuses superolaterally, the nasal septum superomedial and the upper lip anteriorly. This was combined with functional bilateral cervical lymph node dissection of lymph node chains I, II, III and IV, followed by adjuvant radiotherapy (Figure 3).

It should be noted that the patient benefited from a tracheotomy with placement of an SNG and a buzzer at the end of the surgical procedure.

Anatomopathological study with immunohistochemical analysis of the surgical specimen revealed a nodular malignant melanoma (Figure 4):

- Weak, focal cytoplasmic expression in tumor cells of the anti-PS100 antibody (Polyclonal Rabbit, Dako).
- Intense nuclear expression in 70% of tumor cells of anti-Ki67 antibody (Clone MIB-1).

- Intense, diffuse cytoplasmic expression in tumor cells of anti-Melan A antibody (CloneA103, Dako).
- A moderate and focal cytoplasmic expression of tumor cells of the anti-HMB45 antibody (CloneHMB45, Dako) –

It should also be noted that the limits of resection were clean, with the presence of 15 lymph node metastases out of the 45 received during lymph node chain analysis.

The post-operative course was marked by superinfection of the tracheostomy site, complicated by a collected cervical cellulitis requiring surgical drainage with bi-antibiotic therapy adapted to the result of the cytobacteriological examination of the pus collected, followed by weaning of the tracheostomy.

A bumblebee was kept in place to fill the loss of substance and changed every 05 days. A facial MRI was performed 2 months later with no sign of local recurrence. The patient was discharged and referred to the oncology department for adjuvant radiotherapy.

Five months later, the patient presented to the emergency department with moderate active bleeding from the surgical site associated with epistaxis, requiring hospitalization with conditioning, packing and vascular cauterization in the operating room. Despite all this, the patient died 10 months after the initial consultation.



**Figure 1: Median gingivomaxillary tumor mass filling the oral cavity**

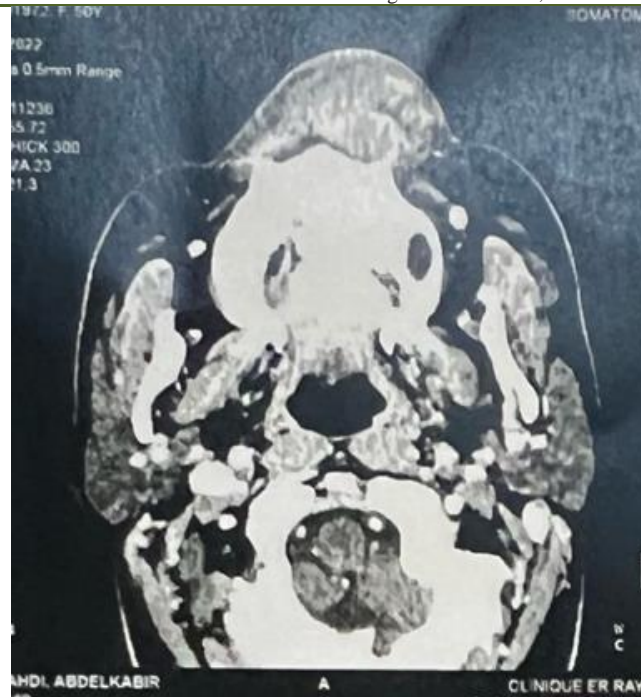


Figure 2: Facial CT scan with contrast injection reveals a tumor mass infiltrating the maxillary bone



Figure 3: A: En bloc tumor resection; B: Pigmented jugulocarotid adenopathy; C: Loss of substance classified as IIC according to Brown's classification; D: immediate post-operative appearance

## DISCUSSION

Melanomas are malignant tumors with melanocytic origins, corresponding to the cells that make up the melanin pigment in the basal layer of the epithelium. They preferentially affect the skin, but can also be found in the mucosa, as a result of the migration of neural crest cells (Femiano *et al.*, 2008) (Aguas *et al.*, 2009).

Melanoma of the oral cavity is an extremely rare tumor, representing 0.5% of malignant tumors of the oral cavity, 0.2 to 8% of all melanomas combined, and which is most common in the maxillary region (80% of cases), mainly affecting the hard palate and gingiva (Bouchareb *et al.*, 2013) (Aguas *et al.*, 2009)

Compared to cutaneous melanoma, oral involvement occurs in older patients, with a slight male predominance, and mainly affects the Asian (Japanese up to 21% of melanomas), African and North American Indian populations (Chatzistefanou *et al.*, 2016) (Femiano *et al.*, 2008) (Aguas *et al.*, 2009) (Bouchareb *et al.*, 2013).

Due to their clinical polymorphism, oral cavity melanomas can present as macular, nodular or ulcerated lesions, often with irregular asymmetrical borders (Smith *et al.*, 2016) (Saint-Blancard & Kossowski, 2006) and pigmented (black, grey, violet, red), with centrifugal color degradation associated with peripheral satellite lesions. Tooth mobility, bleeding on palpation of the lesion, or delayed healing following tooth extraction are clinical features suggestive of malignancy (Bouchareb *et al.*, 2013) (Femiano *et al.*, 2008).

Diagnosis is based on anatomical pathology, on biopsies of lesioned areas or on surgical specimens. However, this diagnosis is difficult because of a dual architectural and cytological polymorphism. The architecture of tumor proliferation is highly varied, with compact, alveolar, papillary, fusiform and desmoplastic aspects (Saint-Blancard & Kossowski, 2006).

It is therefore essential to carry out an immunohistochemical study to confirm the diagnosis and prognosis, and to rule out other differential diagnoses (Saint-Blancard & Kossowski, 2006) (Femiano *et al.*, 2008) (Aguas *et al.*, 2009).

Nevertheless, the demonstration of markers of melanocytic differentiation is the main element of diagnostic certainty. It is based on analysis of at least 5 biomarkers: S100 protein (the most sensitive marker), HMB45, tyrosinase, Melan A, MITF (Microphthalmia Transcription Factor), and even PNL-2 (Kallel *et al.*, 2010) (Femiano *et al.*, 2008) (Aguas *et al.*, 2009).

Prognostic factors depend on tumor invasion and melanoma depth at diagnosis (Bongiorno & Aricò, 2002).

Due to the extreme clinical and histological polymorphism, several differential diagnoses need to be considered. These can be summarized as follows: (Saint-Blancard & Kossowski, 2006) (Smith *et al.*, 2016).

- In the case of a pigmented lesion: a nevus, meanoacanthoma or Kaposi's sarcoma,
- A violet-red lesion: a vascular lesion, or a reactive inflammatory lesion such as epilus.
- In the presence of an epithelioid tumor, it is advisable to rule out squamous cell carcinoma, adenocarcinoma, large-cell neuroendocrine carcinoma or large-cell lymphoma (B or anaplastic).
- In the face of a proliferation of small, round cells, the following should also be considered: a primitive neuro-ectodermal tumor, lymphoma, plasmacytoma or alveolar rhabdomyosarcoma.
- In the situation of spindle cell proliferation, a spindle cell sarcoma or carcinoma should be considered.

The basic treatment for oral melanomas is surgical resection with wide safety margins (3cm) and lymph node dissection (Saint-Blancard & Kossowski, 2006) (Chatzistefanou *et al.*, 2016).

Oral melanomas are considered resistant to radiotherapy, and the indication of postoperative irradiation is recommended in cases of insufficient margins. On the other hand, radiotherapy remains an alternative to surgery for elderly patients whose surgery is contraindicated (Chatzistefanou *et al.*, 2016).

Chemotherapy is indicated in metastatic forms (Saint-Blancard & Kossowski, 2006). Immunotherapy protocols have been tested with variable results, but no studies are available (Smith *et al.*, 2016) (Femiano *et al.*, 2008) (Aguas *et al.*, 2009).

The treatment currently available is unpromising, hence the advantage of early diagnosis and rapid treatment at a potentially curable stage (Saint-Blancard & Kossowski, 2006).

Overall, oral melanomas are highly aggressive and recurrent cancers. The 5-year mortality rate for melanoma patients is 5%, with a median survival of 1 to 2 years after diagnosis. Most patients die of metastatic disease, the preferred sites being lung, liver, brain and bone (Smith *et al.*, 2016) (Chatzistefanou *et al.*, 2016) (Aguas *et al.*, 2009) (Saint-Blancard & Kossowski, 2006). (Femiano *et al.*, 2008).

## CONCLUSION

Oral melanomas are rare and aggressive. Clinical diagnosis is easy, but anatomical diagnosis remains difficult, (Mimoune *et al.*, n.d.) (Aguas *et al.*, 2009) (Bongiorno & Aricò, 2002).

Despite therapeutic advances, oral melanoma still has a poor prognosis. Surgical excision is almost always incomplete, justifying the use of additional treatment. Improving prognosis requires early diagnosis, the most complete possible assessment of locoregional extension, and an appropriate, multidisciplinary choice of treatment (Mimoune *et al.*, n.d.) (Kallel *et al.*, 2010).

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