Primary melanoma with oral and sinonasal involvement: A rare case report

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ABSTRACT

Introduction: Melanoma is a rare and highly aggressive malignant neoplasm of melanocytic origin, which represents less than 2% of all head and neck malignances, being locally aggressive with poor prognosis. Case Report: A 57-year-old black man was referred for evaluation of a fast growing palatal swelling lasting five months. Medical history did not include pain or other symptoms and the patient confirmed to be an active smoker for many years. Intraoral examination revealed a large asymmetrical swelling of the right and left palate with a soft consistency and dark pigmentation. A cone beam computed tomography revealed a hypodense image located in the anterior and right side of the maxilla causing great destruction of the alveolar ridge and affecting all right maxillary sinus, the anterior portion of the left sinus and the nasal cavity. It has poor-defined aspect and irregular limits. Microscopically, sheets of atypical melanocytes invading the connective tissue were observed, with perivascular and perineural invasion. Conclusion: The final diagnosis was melanoma with oral and sinonasal involvement. The patient was submitted to surgical treatment and died few months later.

Keywords: Head and neck cancer, Melanoma, Oral cancer, Oral pathology

INTRODUCTION

Oral and sinonasal melanoma is an unusual type of cancer of melanocytic origin, representing 1–2% of all head and neck tumors, being more prevalent in males with age ranging from 20 to 83 years [1, 2], mostly affecting Caucasian people [3]. The palatal mucosa is the most affected site (32%), followed by the maxillary gingiva (16%), mandibular gingiva (7%), buccal mucosa (7%), lips (7%), and alveolar gingiva (5%) [1, 4]. The tumor usually progresses to a widespread involvement with metastasis, generally in lymph nodes, lung, and liver [1, 2]. Oral and sinonasal melanoma has a poor prognosis and rare long-term survivors reported. Most of the tumors tend to invade the adjacent tissues or metastasize [2, 5]. The oral cavity environment with rich vascular supply contribute...
to tumor infiltration. The overall 5-year survival rate is 18% for gingival and 11% for palatal tumors. The presence of lymph node metastasis reduces median rates survival to 18 months, meanwhile, the absence of lymph node involvement the overall survival rises to 46 months [2, 6]. Furthermore, the survival of these patients is less than for those with melanoma of the skin. The current treatment for oral and sinonasal melanoma is surgery and often chemotherapy or radiation therapy [2, 7]. Recurrence rates are up to 42%. The survival rate and the prognosis depend on the staging of the primary lesion and the local spread with or without metastases [8, 9].

The present case report contributes with a new case of oral and sinonasal melanoma with extremely fast development and outcome. We highlight its clinical, microscopic, and imaging aspects that must be recognized in order to improve knowledge to fast diagnosis and contribute to overall survivor.

CASE REPORT

A 57-year-old black man was referred for evaluation of an asymptomatic enlargement of the right palate noticed by his dentist and lasting five months. Patient’s medical history was unremarkable, and he reported being a former smoker for many years. Upon clinical examination a swelling in the right region of the face was evident, causing facial asymmetry. At intraoral examination a soft pigmented and ulcerated tumor was observed on the hard palate (Figure 1). The cone beam computed tomography showed the presence of an ill-defined hypodense image in the anterior and right maxilla, causing great destruction of the alveolar ridge. The lesion presented irregular limits on the remaining edge and was destructing the anterior portion of the hard palate and nasal cavity. The lesion also showed extension to the right maxillary sinus and to the anterior portion of the left sinus (Figure 2).

An incisional biopsy was performed, and microscopic evaluation showed a combined pattern of invasive melanoma with in situ component. It was observed a proliferation of pleomorphic epithelioid cells containing large portions of melanin invading the lamina propria and deep connective tissue. Mitoses, perivascular and perineural invasion, were also observed (Figure 3). The final diagnosis was primary oral and sinonasal melanoma. The patient was referred for oncologic treatment. A body screening was performed, and multiple metastatic lesions were

Figure 1: Clinical features of oral and sinonasal melanoma. (A, B) Extraoral aspect showing a large swelling at the right side of the face with displacement of right nose ala. (C, D) Intraoral aspect showing a heavily pigmented melanoma with a plaque-like component involving hard and soft palate, and a large swelling in right alveolar ridge.

Figure 2: Radiographic features of oral and sinonasal melanoma. Hypodense image located in the anterior and right side of the maxilla, causing great destruction of the alveolar ridge, with poor-defined aspect and irregular limits on the remaining edge and destruction of the anterior portion of the hard palate and anterior region of the nasal cavity. The lesion also affected the right maxillary sinus and the anterior portion of the left sinus.

Figure 3: Histopathological features of oral and sinonasal melanoma. (A) Polymorphous infiltrate of pleomorphic epithelioid cells containing large portions of melanin invading the lamina propria and deep connective tissue (HE, 400×). (B) Perivascular and perineural invasion by atypical spindled and pleomorphic pigmented melanocytes (HE, 4000×).
detected. Nevertheless, the patient was submitted to surgical treatment but died few months later.

DISCUSSION

Primary malignant melanoma of the head and neck is a rare neoplasm of melanocytic origin [1, 5, 10] and up to now its etiopathogenesis is still poorly understood. It is well known that mucosal melanoma is derived from neuroectodermal migrating melanocytes and this explains its unrarity in nonecderma-derived mucosa, such as mucosa of the nasopharynx, larynx, tracheobronchial tree, and esophagus [11, 12].

One-third of the patients are asymptomatic at diagnosis [13, 14]. The tumor may be presented as a rapid appearance and enlargement of a pigmented lesion or it may be preceded by a pigmented area for a variable period [15]. The median age of the patients with mucosal melanoma is about 60 years old, but the tumor can be found in any age. There is a slight male preponderance (1.3:1). The most common presenting symptoms are nasal obstruction and epistaxis [16].

Pigmented melanoma is usually easy to diagnose clinically, as there is often a variation in color from red to black to brown, asymmetry and an irregular outline, but amelanotic lesions have also been reported. As with nonmelanoma skin cancers, biopsy is indicated to evaluate the presence of malignant melanocytes. Unfortunately, late discovery and diagnosis often indicate the existence of an extensive tumor with metastasis [8, 17]. Diagnostic evaluation for sinonasal lesions should include fiberoptic nasopharyngoscopy and computed tomography to evaluate the local destruction and the adjacent structures affected as well as a positron emission tomography to screen for lung metastasis. A magnetic resonance imaging scan can also be helpful, especially with sinonasal tumors, that can evidence the extension of the lesion [14].

The treatment for the oral melanoma is still a matter of discussion. Surgery with clear margins continues to be the preferred choice of treatment, except for untreated tumors and cases of metastasis [14]. In these cases, chemotherapy can be used to reduce the size of the tumor so the surgery can be performed later [2, 14], therefore, the first choice of chemotherapy to melanoma metastasis is the Dacarbazine [18]. The recurrence and metastasis rates are high and most of the patients die in two years with complications of the disease [19].

CONCLUSION

This study showed the importance of the clinical, radiographic, and histopathological evaluation in order to determine the morphological aspects of oral and sinonasal melanoma to establish the final diagnosis.

REFERENCES


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Author Contributions
Thamyres Campos Fonsêca – Conception of the work, Design of the work, Acquisition of data, Analysis of data, Interpretation of data, Drafting the work, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Aline Correa Abrahão – Conception of the work, Design of the work, Drafting the work, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

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Authors declare no conflict of interest.

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