

## Palatal Melanoma: "The Silent Killer"

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### Abstract

Primary melanoma of the oral cavity is extremely uncommon tumour consisting approximately 0.2 - 8.0% of all melanoma cases and 0.5% of all oral malignancies. It has an aggressive behaviour and poor prognosis, with 5 – year - survival rate between 5 - 20%. The initial symptoms are often unnoticed, which lead to late diagnosis and worsening of the prognosis. Because of its infrequent occurrence, there is no well - defined classification and therapeutic protocol, in contrast to melanomas of another side. Early diagnosis and treatment are essentially linked to survival rate. We present a case of palatal melanoma in a 76 – year - old female patient, as we want to emphasise the importance of the early detection and accurate diagnosis of melanoma of oral cavity, to its influence of the therapeutic outcome.

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## Introduction

Primary melanoma of the oral cavity is extremely uncommon tumour consisting approximately 0.2 - 8.0% of all melanoma cases and 0.5% of all oral malignancies [1][2]. It has been called "a silent killer", because of its highlighted aggressive behaviour and early metastatic spread [1][3].

The main aetiology reason for the pathologic proliferation of malignant melanocytes along the junction between the epithelial and connective tissues is not established yet [3]. Although chronic irritation, tobacco, alcohol and formaldehyde exposure have been implicated as possible risk factors, it is considered that most of the melanomas of oral cavity arise de novo [4]. In contrast to cutaneous melanoma, no particular precursor lesion has been identified, and atypical melanocytic hyperplasia is considered as a proliferative phase [2]. Despite blue nevi, dysplastic nevi involve the oral cavity extremely rare, most often

– the palate mucosa [5]. While the hard palate is the most common side of affection of melanoma of the oral cavity, the buccal mucosa is affected in one-third of the cases, as the maxillar gingiva is more frequently involved than the mandibular one [3]. Uncommonly, the tongue and mouth's floor could also be affected [6]. The onset of the disease is between 40 and 70 years, with male predominance in gender distribution [6].

The initial symptoms are often unnoticed, which lead to late diagnosis and worsening of the prognosis [1]. Patients usually seek medical help because of bleeding, pain or swelling, which are associated with vertical growth phase and progression of the disease [6][7]. Furthermore, because of the anatomic structure of the oral cavity, bone invasion and destruction are commonly seen in cases of melanoma of the hard palate [2].

Surgery is the first choice of treatment which often require subsequent reconstruction of the defects and advanced cases [3][4]. However, the 5 -year -

survival rate in advanced cases with bone destruction varies between 5 - 20% and only 6.6% more than five years, which emphasise the underhanded aggressive nature of this kind of tumours [2][7][8].

## Case report

A 76 – year - old Caucasian female patient presented with two months history of moderate pain in the oral cavity, more pronounced on the left. Patient's history was free of comorbidities and medications, as well as of family history for dermatologic diseases. The conducted physical examination revealed unequally pigmented lesion, composed by nodular elements, with sharply demarcated, but irregular borders, and partially ulcerated surface, covering almost the whole hard palate, more pronounced in left. Satellite pigmentations with reticular characteristic were also established on the right side of the palate (Figure 1. a, b, c).



Figure 1: A, B, C - Clinical manifestation of melanoma of the oral cavity in 76-year-old female patient

The performed paraclinical blood tests established leukocytes ( $10.15 \times 10^9/l$ ) and monocytes count (8.8%) in the upper border of the normal range, slightly elevated glucose level in serum (6.18 mmol/l) and LDL - cholesterol (3.06 mmol/l). Imaging diagnostic procedures, including head and neck CT, obtained pathological lesion, affecting the mucosa, above the left half of pterygoid fossa, thickened mucosa of the left maxillary sinus (due to palatal outgrowth and lamina perpendicular), 5 - 8 mm in depth, but the preserved bone structure, without a sign of destruction. This pathologic finding was also affecting the gingiva, through the alveolar border.

Enlarged lymph nodes with changes in their morphologic structure were also detected - two in left (level 2B), measuring approximately 29/40 mm and 16/24 mm and one in the right (level 2B), measuring 10/17 mm. Pathological soft - tissue nodule 15 mm in size was established, affecting the skin and subcutaneous tissue of right cheek. Cutaneous papillomatous lesion with benign characteristics was observed on left cheek. Abdominal CT examination revealed a hiatal hernia, cholelithiasis, a parenchymal and cortical cyst in left kidney. Enlarged abdominal and pelvic lymph nodes were not detected.

The diagnosis of palatal melanoma with lymph node involvement was made, based on these findings. No data for organ metastasis spread was presented. Clinically, the patient was staged in IIb (Westbury classification) and referred to maxillofacial surgery department for palatotomy and subsequent reconstruction of the defect. Lymph node dissection was also planned. Patient's further follow up, and postsurgical screening was planned in the oncology department.

## Discussion

Although extremely rare and aggressive in behavior, due to rapid hematogenous and lymph node metastatic spread, melanoma of the oral cavity could be asymptomatic flat macule for a long time (9). Because of its infrequent occurrence, there is no well - defined classification and therapeutic protocol, in contrast to melanomas of other side [2][3]. Although several authors team have been proposed different clinical and histological classification systems, unified criteria are not accepted worldwide. Therefore, Lopez et al. classified melanoma of the oral cavity, based on its clinical appearance into: 1) pigmented nodular type, 2) non - pigmented nodular type, 3) pigmented macular type, 4) pigmented mixed - type, and 5) non - pigmented mixed type [8]. Westbury has been proposed a clinical classification, similar to that for cutaneous melanoma, as follow: I - only primary tumor, II - metastasis presented (IIa - adjacent skin involved, IIb - regional lymph nodes involved, IIc - adjacent skin and regional lymph nodes involved) and III - metastasis beyond regional lymph nodes [10]. According to these classification systems, our patients was classified as pigmented nodular type melanoma, with regional lymph node involvement. The lack of papillary and reticular dermis in oral mucosa, Clark's criteria for invasion, also could not be applied in cases of melanoma of the oral cavity [6]. Therefore, the classification of the Western Society of Teachers of Oral Pathology (WESTOP) have been implicated, based on histopathological pattern of the tumor, namely: (a) melanoma in situ, limited to the epidermis and its junction with the connective tissue; (b) invasive melanomas, which extend into the connective tissue and (c) melanomas with a combined pattern between invasive and in situ [6][11]. The level on invasion is determined as an independent predictor of survival, while the tumor thickness, vascular invasion, and necrosis have no significant influence on survival, as predictors of the survival rate, in contrast to cutaneous melanoma, which thickness determines the further diagnostic and therapeutic behavior in one hand, while vascular invasion is considered as the major predictor for distant metastatic spread [12]. Melanoma of oral cavity metastasizes early in its course in general [1]. Locoregional lymph node metastasis

occur in almost half of the cases during first 2 years of onset of the disease, while lungs, brain, bones and liver involvement affect up to 85% of the patients [3][13]. However, early diagnosis and treatment is essentially linked to survival rate. Diagnosis is usually made, based on the clinical appearance and histological findings, which could show a pleomorphic pattern that could mimic other tumours such as lymphomas, carcinomas, neuroendocrine carcinomas, sarcomas, and germ - cell tumours [14][15]. Positive immunohistochemical examination for S – 100, HMB - 45, Melan A, tyrosinase and vimentin is usually helpful for confirmation, especially in atypically presented and apigmented melanomas [15][16]. However, it is postulated that mucosal melanomas may arise not only from melanocytes but also from Schwann cells in the mucous membrane [17]. The loss of heterozygosity at 12p13 and loss of p27K1P1 protein expression contribute to melanoma progression [17][18]. Surgery is the first choice of treatment, requiring collaboration with a maxillofacial specialist for reconstruction of the defect [19]. Carbon - ion radiotherapy is reported as an effective treatment option with acceptable toxicity in oral cavity's melanoma [20].

With the presented case, we want to emphasize the importance of the early detection and accurate diagnosis of melanoma of oral cavity, to its influence of the therapeutic outcome.

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