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Palatal Malignant Melanoma: A Case Report of an Unusual Site and Histologic Presentation

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Abstract **Case Report**

Malignant melanoma is a potentially aggressive tumor of melanocytic origin. Primary oral malignant melanoma is a rare neoplasm, accounting for 0.5% of all oral malignancies. We report a case of 44yr old male patient who presented with a painful pigmented nodule involving the palate and histologically showing a spindle cell type of Melanoma.

Keywords: Unusual Site Histologic Presentation Palatal Malignant Melanoma.

INTRODUCTION

Malignant melanoma is a potentially aggressive tumour of melanocytic origin [1]. That arises from a benign melanocytic lesion or de novo from melanocytes within otherwise, normal skin or mucosa [2]. Only about 1% of all melanomas arise in the oral mucosa and these account for 0.5% of all oral malignancies [2, 3]. It has been called "a silent killer", because of its highlighted aggressive behaviour and early metastatic spread [4, 5]. The most common histopathologic types of melanoma are superficial spreading melanoma (70%), nodular melanoma, lentigo maligna melanoma, and acral lentiginous melanoma (2-8%). Spindle cell melanoma (SCM) is a rare subtype of malignant melanoma composed of spindled neoplastic cells arranged in sheets and fascicles [13]. As a morphological variant of melanoma, SCM is rare and its incidence has been variably reported between 3 and 14% of all melanoma cases (including desmoplastic melanoma) [22-24].

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 [5]. Uncommonly, the tongue and mouth's floor could also be affected [9]. The onset of the disease is between 40 and 70 years, with male predominance in gender distribution [9].

The initial symptoms are often unnoticed, which lead to late diagnosis and worsening of the prognosis [4]. Patients usually seek medical help because of bleeding, pain or swelling, which are associated with vertical growth phase and progression of the disease [9, 10]. 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 [6].

CASE REPORT

A 44 – year - old male patient presented with history of painful pigmented nodule in the oral cavity. Patient's history was free of comorbidities and medications, as well as of family history for dermatologic diseases. Physical examination revealed unequally pigmented lesion, with sharply demarcated, but irregular borders, and partially ulcerated surface, covering almost the whole soft palate. A provisional diagnosis of malignant melanoma was considered and an incisional biopsy was performed and sent to the department of pathology which confirmed the diagnosis.

Gross examination

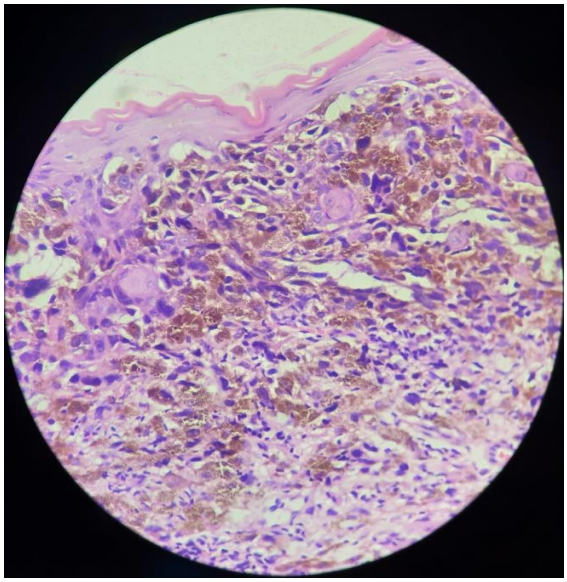
Received two grey brown soft tissue pieces collectively measuring 1x0.5x0.2cm in size. The tissue was paraffin-embedded, and the sections were stained with haematoxylin and eosin (H&E).

Microscopic Examination

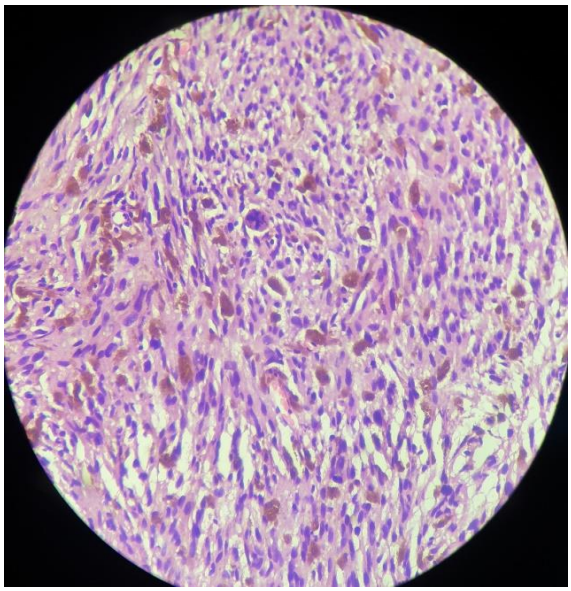
Sections examined show stratified squamous epithelial lining having increased basal cell hyperpigmentation. Tumor cells are originating from the basal cell epithelium. The tumor cells are mainly spindle shaped showing mild degree of pleomorphism

and hyperchromasia. Many scattered melanin pigment forming cells are seen. Mitotic figures identified. The differential diagnosis based on the H&E sections included melanoma, dermatofibrosarcoma protuberans and fibromatosis.

Immunohistochemistry (IHC) revealed strong positivity for HMB-45 and S-100 protein. The histopathology and immunohistochemistry confirmed the diagnosis of Spindle cell type Melanoma.



The tumor cells mainly spindle shaped showing mild degree of pleomorphism and hyperchromasia (H&E, × 40)



Scattered melanin pigment forming cells and mitotic figures noted (H&E, × 40)

DISCUSSION

Mucosal melanomas of the head and neck comprise just over 1% of all melanomas and of these about 50% arise in the oral cavity. Oral mucosal melanomas are therefore, rare representing about 0.5% of oral malignancies and less than 0.01% of all oral biopsies. SCM is rare and its incidence has been variably reported between 3 and 14% of all melanoma cases (including desmoplastic melanoma) [22-24].

The main aetiology reason for the pathologic proliferation of malignant melanocytes along the junction between the epithelial and connective tissues is not established yet [5]. 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 [7]. In contrast to cutaneous melanoma, no particular precursor lesion has been identified, and atypical melanocytic hyperplasia is considered as a proliferative phase [6]. Despite blue nevi, dysplastic nevi involve the oral cavity extremely rare, most often – the palate mucosa [8].

They arise in adults with an average age of about 55, but with a uniform age distribution from years 20 to 80 years. Very rare cases have been reported in children. In most large series there is a male predominance in a ratio of about 3:1. Prognosis for malignant melanoma in the oral region is poorer than its counterpart in cutaneous regions because of anatomic considerations and delayed diagnosis [11]. Mucosal melanomas are so rare that there are no large data bases compared to those for cutaneous melanomas; therefore, pathologic micro staging has not been possible, and the fine-tuning of the prognosis that has been useful in cutaneous melanomas (Breslow thickness) has so far not been possible in mucosal melanomas [12]. Typically, an oral melanoma is composed of sheets or islands of epithelioid melanocytes, which may be arranged in an organoid, or alveolar pattern. The cells have pale cytoplasm and large open nuclei with prominent nucleoli and occasionally they may be plasmacytoid, But Sheets and fascicles of spindle cells are seen in the rare spindle type melanoma. Spindle cell melanoma (SCM) is a rare subtype of malignant melanoma composed of spindled neoplastic cells arranged in sheets and fascicles [13]. As a morphological variant of melanoma, SCM is rare and its incidence has been variably reported between 3 and 14% of all melanoma cases (including desmoplastic melanoma) [22-24].Histologically, cytologic features of SCM are indistinct and often confused with those of other epithelial neoplasms, including sarcomas and lymphomas [18, 19]. Immunohistochemistry is a helpful tool in distinguishing SCM from other sarcomas and carcinomas [20, 21].

Despite the improvement of surgical techniques and the introduction of new chemotherapeutic agents, prognosis of this malignancy remains poor. The generally advanced stage of the tumor at initial diagnosis leads to a poorer survival of patients with mucosal melanomas as compared with

patients with cutaneous melanomas and presence of vertical growth phase are associated with median survival rate [14]. The prognosis for oral melanoma is poor with an overall median survival of about 2 years and 5-year survival of less than 20%. Stage is a predictor of survival but even localized tumors (stage I) show a 5-year survival of less than 50%. Depth of invasion (Breslow thickness and Clark's levels) is of limited value in oral lesions. Nevertheless, lesions thicker than 5 mm may have a significantly worse prognosis. Other factors associated with poor prognosis include, vascular invasion, necrosis, a polymorphous tumor cell population, and increasing age [15].

Surgery is the first choice of treatment, requiring collaboration with a maxillofacial specialist for reconstruction of the defect [16]. Carbon - ion radiotherapy is reported as an effective treatment option with acceptable toxicity in oral cavity's melanoma [17].

CONCLUSION

Oral mucosal melanomas represent about 0.5% of oral malignancies and less than 0.01% of all oral biopsies. SCM is rare and its incidence has been variably reported between 3 and 14% of all melanoma cases. Pathologically, melanoma should be considered in spindle cell lesions especially with pigment and residual nevus cells.

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