

Case Report

Oral malignant melanoma: A case report of an unusual clinical and histologic presentation

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ABSTRACT

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. The present case occurred in a 60-year-old female patient, as a pedunculated growth involving the palate and alveolar ridge and histologically showing a desmoplastic differentiation. The article discusses the distinct clinico-pathologic presentation of this case and emphasizes on the need to identify and report such cases for further understanding of their biologic behavior.

Key Words: Desmoplastic melanoma, malignant melanoma, oral mucosal melanoma

INTRODUCTION

Malignant melanoma is a potentially aggressive tumor of melanocytic origin.^[1] that arises from a benign melanocytic lesion or de novo from melanocytes within otherwise, normal skin or mucosa.^[2] The incidence of head and neck mucosal melanomas is approximately 4/10 million population per year in the USA.^[3] Only about 1% of all melanomas arise in the oral mucosa and these account for 0.5% of all oral malignancies.^[2,4] These rare lesions may present themselves in yet uncommon ways which need to be identified and reported to understand their biologic behavior, and hence this case report is an attempt do the same.

CASE REPORT

A 60-year-old female patient reported with the complaint of a growth on the palate and inability to wear the denture since 3 months. On clinical examination, the lesional mass was seen involving the entire hard palate as a pedunculated mass, measuring approximately 3 cm × 3 cm, attached to the palatal mucosa with a narrow stalk. The base of the pedunculated mass, the palatal mucosa and the surrounding alveolar mucosa showed areas of brownish black pigmentation. The mucosa on the oral side of the lesion showed a pale whitish color [Figure 1]. On palpation the lesion was smooth, non-tender, well defined, firm to hard in consistency with induration of the posterior margins.

On computed tomography an enhancing, infiltrative soft tissue mass arising from the hard palate with subtle bony erosion noted over the mid portion of the hard palate [Figure 2] and bilateral submandibular lymphadenopathy were evident.

A provisional diagnosis of malignant melanoma was considered and an incisional biopsy was performed which confirmed the diagnosis. The patient underwent total maxillectomy with neck dissection.

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The histopathological examination of the excised tissue revealed cells with a clear cytoplasm and hyperchromatic nuclei resembling atypical melanocytes seen proliferating from dermal-epidermal junction into the connective tissue. The atypical melanocytes were epithelioid in the superficial region [Figure 3], and spindle shaped in the deeper regions of the connective tissue. The spindle cell type predominated showing an irregular branching pattern with intervening fibrous septae [Figure 4], these cells appeared fusiform to round in different orientations [Figure 5]. Atypical features like mitosis, multi-nucleation, pleomorphism, hyper chromatism, increased nuclear cytoplasmic ratio and prominent nucleoli were seen [Figure 6]. Pigmented nests of atypical melanocytes along with melanophages were seen in the superficial region. The tumor-stroma interface showed a marked inflammatory response and dense fibrosis. The evaluation the

harvested submandibular lymph node showed metastatic involvement. The histopathology confirmed the diagnosis of malignant melanoma with metastatic level I nodes. The spindle cell predominance with collagenous stroma was suggestive of desmoplastic malignant melanoma.

The patient underwent reconstruction, prosthetic rehabilitation, and has been on regular follow with no recurrence till date.

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. They arise in adults with an average age of about 55, but with a uniform age distribution from years



Figure 1: Clinical picture showing a pedunculated lesion involving the palate

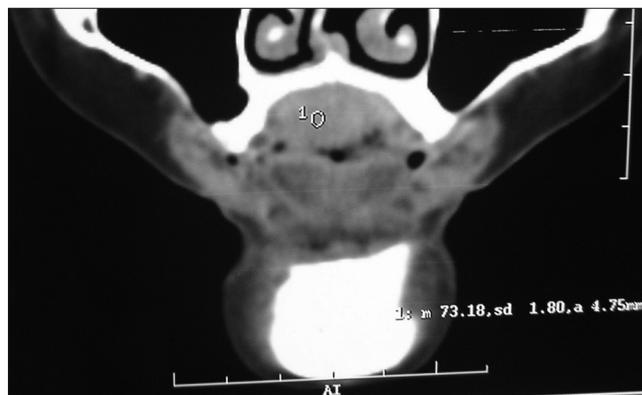


Figure 2: Computed tomography image showing subtle bony erosion noted over the mid portion of the hard palate

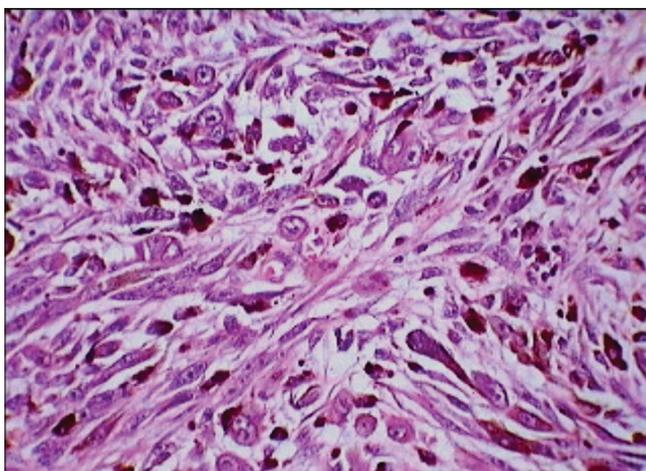


Figure 3: Superficial dermis showing pigmented epithelioid and spindle tumor cells (H and E, $\times 40$)

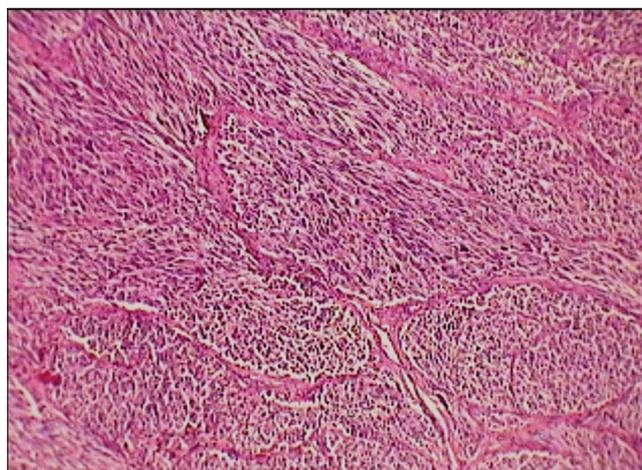


Figure 4: Deep dermis showing the tumor with predominant spindle cell type arranged in irregular branching pattern with intervening fibrous septae (H and E, $\times 10$)

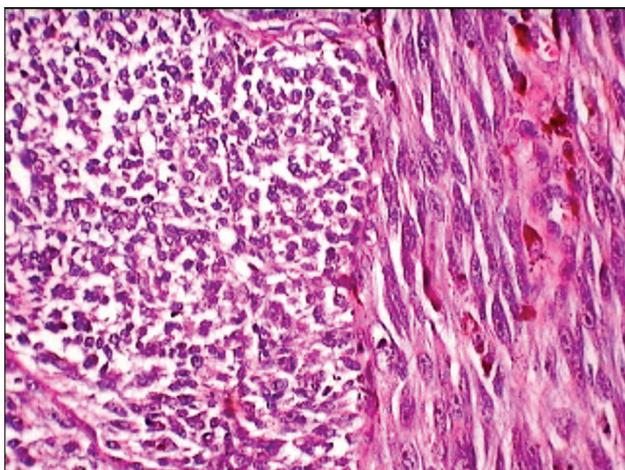


Figure 5: Spindle tumor cells appear fusiform to rounded in different orientations (H and E, $\times 40$)

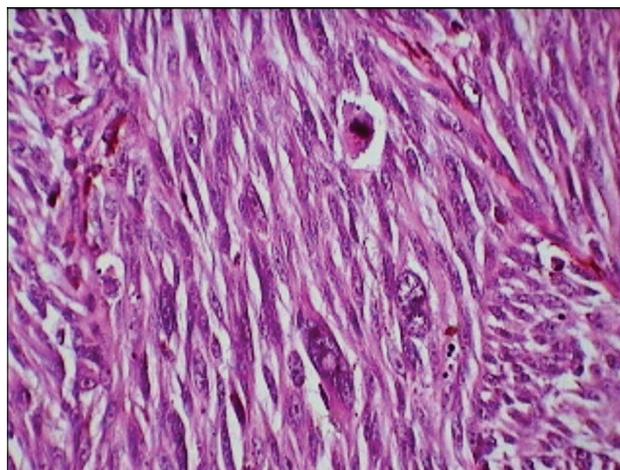


Figure 6: Tumor cells showing atypical features like mitosis, pleomorphism, hyperchromatism, increased nuclear cytoplasmic ratio and prominent nucleoli (H and E, $\times 40$)

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. No etiological factors are known to be associated with oral melanoma.^[5] Prognosis for malignant melanoma in the oral region is poorer than its counterpart in cutaneous regions because of anatomic considerations and delayed diagnosis.^[6] 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.^[7]

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. Sheets and fascicles of spindle cells may also be seen, but are usually a minor part of the lesion. Occasional lesions may be predominantly or wholly spindled.^[5]

Desmoplastic melanoma (DM) is a rare variant of malignant melanoma first described by Conley *et al.* in 1971 as an invasive melanoma composed of spindle cells surrounded by abundant collagen or fibrous stroma.^[8,9] It is a histological subtype of melanoma that has been reported to account for approximately 1% of all cases of melanoma.^[10] In DM the spindle-shaped melanocytes, which often resemble fibroblasts and are usually non-pigmented, are found in and between mature collagen bundles.

The latter may be thickened and/or associated with a mild to marked stromal fibrosis. The distribution of spindle cells is usually haphazard but occasionally they form parallel bundles or storiform areas. The overlying epidermis may be thinned or thickened. The cytological atypia of the spindle cells usually varies from mild to moderate.^[8] Similar observations were made in the present case.

It is most likely that the desmoplastic cells are derived from melanocytes that have undergone adaptive fibroplasia. Some authors have suggested that the desmoplasia occurs because of a fibroblastic stromal response and neurofibrosarcomatous differentiation of the tumor cells.^[7] Electron microscopic examination of the spindle cells shows that a few contain melanosomes and most show a fibroblastic, neurotrophic or myofibroblastic transformation. The dermal component of DM is usually negative for Melan A and Human melanoma black HMB45. Both the epidermal component and dermal atypical spindle melanocytes are positive for S100 immunostain.^[11] Immunohistochemical evaluation shows S 100 positivity in 93% cases while HMB 45 only in 7% cases.^[12]

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.^[13]

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.^[5]

DM has a propensity for neurotropic metastases, which may lead to local recurrence despite excision with apparently disease-free margins.^[14] Recurrences are common especially after incomplete excision, marginal excision < 10 mm or if neurotropism is present. The conflicting results regarding the risk of regional node field metastases and prognosis of DM patients may be due to a heterogeneity of tumors classified as DM and failure to account for tumor thickness. Regional nodal metastases appear to very uncommon in paucicellular DMs with prominent fibrosis and are associated with longer survival. Otherwise, disease free survival rates are similar to other melanomas of comparable thickness. Neurotropism, HMB45 positivity, high mitotic rate, male gender, thickness, ulceration and site all appear to affect survival which overall is 79% at 5 years. Of patients with a recurrence, 78.2% experienced it within 2 years. Wide local excision is the treatment of choice. Radiation therapy has been effective in some cases.^[8]

CONCLUSIONS

Oral mucosal melanomas are rare oral malignancies. Vigilant comprehensive analysis of published cases and recognition of new ones may be helpful in establishing definite classification and proposing clinical features that would facilitate its early diagnosis, as a prerequisite for timely treatment and better prognosis of this rare pathology.^[15]

Melanomas in the palate and the maxillary alveolar ridge may sometimes present as a polypoid mass, however the presentation as a pedunculated mass is extremely rare, with only a few reported cases, as present in this case. In addition to the pedunculated morphology, the present case showed a tumor mass, comprising of a predominant spindle cell component and fibrosis suggestive a spindle cell-desmoplastic variant of malignant melanoma. The unusual clinic-pathologic presentation of the case and the conflicting biologic behavior makes it essential that

such cases be reported and studied further to evaluate their long term behavior and prognosis.

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