CASE REPORT

Desmoplastic malignant melanoma of alveolus – A rare entity

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Summary Desmoplastic malignant melanoma (DMM) is a distinctive variant of malignant melanoma. DMM involving the oral mucosa is very rare and to our knowledge, there are only 16 cases reported in the English literature. This is a case report of DMM in a 32-year-old male patient involving the maxillary alveolus.

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Introduction

The term desmoplastic malignant melanoma (DMM) was introduced by Conley et al. in 1971,1 to describe a subset of cutaneous melanomas characterized by non-pigmented, spindle cell tumors with pronounced collagenization within an atypical melanocytic proliferation. Since then, there have been reports of 16 cases of DMM arising in the mucosa of the oral cavity.2–10 This is a case report of DMM involving the alveolus.

Case report

A 32-year-old male was referred to our institution with a chief complaint of pain and swelling of 20 days duration in the left posterior maxillary alveolus. The past medical and dental history was non-contributory. Intra oral examination revealed an extensive nodular mass of about 4 cm × 5 cm in size (Fig. 1). It was extending from 24 to 28 region, and the involved teeth were mobile. The overlying mucosa was of normal color with ulceration in few areas. The swelling was soft in consistency. There was no associated lymphadenopathy.

CT scan revealed an expansile, dense lesion involving the left maxillary antrum. The lesion was seen extending medially into the left nasal cavity eroding the lateral wall of left nasal cavity and also extending intra-orally eroding the maxilla. The lesion was extending posteriorly into the pterygopalatine fossa. There was no parapharyngeal extension. The lesion had eroded the anterior wall of maxillary antrum with extension into the subcutaneous tissue. There was thinning of the infraorbital margin. There was no evidence of calcifications or cystic degeneration (Figs. 2 and 3).
An incisional biopsy revealed proliferation of atypical melanocytes in single dispersion along the basilar layer of the overlying epithelium. In addition, widely scattered, non-pigmented spindle as well as round cells possessing moderately enlarged nuclei with coarse chromatin pattern were identified in the connective tissue. There was an associated prominent stromal desmoplasia (Figs. 4 and 5).

Immunohistochemical staining was performed for S100, Vimentin, HMB45, CD34, CD68, SMA (Dako Corporation, Carpenterica, CA) and Pan Cytokeratin (AE1/AE3; Dako Corporation, Carpenterica, CA).

Atypical junctional melanocytes, spindle and round cells in the connective tissue were strongly positive for S100 (Fig. 6) and vimentin. Cytokeratin was positive for the overlying normal epithelium but negative for tumor cells. The tumor cells did not show any reactivity to HMB45, CD34, CD68 and SMA.

According to the histological pattern of cells and the immunohistochemical findings — S-100 (+), Vimentin (+), Cytokeratin (−), HMB45 (−), CD34 (−), CD68 (−), SMA (−), the diagnosis of DMM was made.

The patient was referred for radiotherapy. A dose of 72–120 Gy was given in fractions over the tumor bed and its margins. The tumor responded favorably and the patient is under followup for the past four months.
Discussion

In 1971, Conley et al. reported a rare variant of spindle cell melanoma, generally located in the head and neck and coined the term, "desmoplastic malignant melanoma".

The histogenesis of DMM is unclear. Currently, it is believed that the desmoplastic component arises from the intra-epidermal melanocytic component and undergoes reactive fibroplasia into the spindle neoplasm. It is also believed that these melanoma cells are producing the collagen (desmoplasia) because some have found fibroblastic and myofibroblastic differentiation ultrastructurally in these cells.

DMM, a subtype of melanoma, is characterized by spindle cell proliferation in the connective tissue with a collagenous stroma. There is a wide spectrum in the cellularity and amount of stromal component varying from abundant collagen (DMM) to minimal collagen (spindle cell melanoma).

More than 80% of DMM, have an epidermal component of melanoma in situ, overlying the dermal component. If present, melanin pigment is seen in the epidermal component. Some do not have epidermal involvement and there is only a dermal melanocytic spindle cell proliferation. Nuclear pleomorphism is not typical among the spindle cells but can be seen at least focally and mitotic figures are rare.

The melanoma cells of DMM are spindle shaped and amelanotic in the vertical growth phase. However, junctional (dermal–epidermal or lamina propria–epithelial) melanocytic proliferation is not always present in DMM. In its absence, the diagnosis of DMM may be particularly difficult. Patchy, lymphocytic infiltrates of varying density can be seen in DMM.

The immunohistochemical profile of DMM differs from the usual melanomas. The conventional melanoma is

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<tr>
<td>Chen et al. – 1989</td>
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<td>Kilpatrick et al. – 1996</td>
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<td>Yu et al. – 2005</td>
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<td>Radfar et al. – 2006</td>
<td>S-100 weakly positive NGFR – Intense positive</td>
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Table 1 Summary of various immunohistochemical findings in DMM

Figure 5 Photomicrograph shows the junctional proliferation at the epithelial–connective tissue interface along with few epitheloid cells in the connective tissue (H&E), 400×.

Figure 6 Photomicrograph shows S100 positivity for spindle and round cells in the connective tissue (Dako Corporation, Carpenterica, CA), 400×.
typically positive for S100, Melan-A and HMB-45. Most DMM are S100 positive and negative for Melan-A and HMB-45.\textsuperscript{12} S100 protein immunostaining is extremely helpful in distinguishing DMM from a reactive fibroblastic proliferation. Immunostaining for cytokeratin clearly separates this tumor from most squamous cell carcinomas.\textsuperscript{14} Anstey et al.\textsuperscript{15} have proposed that positive or negative labelling for S100 protein and NSE could be useful for suggesting or excluding a diagnosis of DMM. A summary of the various immunohistochemical findings in DMM is summarized in Table 1.

Conclusion

Melanoma can present in different forms and at any anatomic site. This report documents the 17th case of DMM originating in the oral cavity. This variant of melanoma is a rare disorder with unremarkable, non-specific clinical manifestations in the oral cavity, which makes the diagnosis of this disease more difficult. Hence, an increased awareness, early suspicion and performance of appropriate biopsy could be of value in early recognition of these variants of melanoma.

References