

# An Unusual Case of Desmoplastic Malignant Melanoma

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## ABSTRACT

Desmoplastic malignant melanoma is a rare variant of spindle cell melanoma, commonly seen in older adults, on sun-exposed areas. It accounts for 1-4% of all cases of cutaneous melanoma. The common location of the desmoplastic melanoma is the head and neck region, whereas, other sites are less common. Regional lymph node involvement is reported in 0 to 13.7% of the cases, which is less frequent than other cutaneous melanomas. A 75-year-old male presented with an ulceroproliferative growth on the left foot that was diagnosed as desmoplastic melanoma with regional lymph node metastasis and in transit metastasis, with extensive pulmonary metastasis.

**KEYWORDS:** Desmoplastic malignant melanoma, immunohistochemistry, in-transit metastasis, SOX 10, S-100, spindle cell variant

## INTRODUCTION

Desmoplastic melanoma was first described in 1971 by Conley *et al.* It accounts for 1-4% of all cases of cutaneous melanomas reference. Early diagnosis of desmoplastic melanoma is associated with a favourable outcome because of its slow growing nature, although most of the cases present in the later stages.

## CASE REPORT

A 75-year-old male patient was admitted with complaint of a painful ulceroproliferative growth in the left foot for seven months, with a history of fever for 10 days. He had a history of loss of appetite and loss of weight. There was no history of trauma or intermittent claudication.

Local examination showed an ulceroproliferative irregular growth on the plantar aspect of the left foot, below the second to fifth toes. The floor of the ulcer was filled with an exuberant, unhealthy, granulation tissue, with minimal serous discharge and patchy black pigmentation. A surrounding area of oedema was present. The ulceroproliferative growth was hard

in consistency and tenderness was present [Figure 1]. Lymph node examination showed multiple inguinal lymph nodes on the left side, which were hard in consistency and measured 2 × 2 cm, with in-transit metastasis on the left leg [Figures 2 and 3].

An edge biopsy from the ulcer showed elongated spindle-shaped cells surrounded by mature collagen bundles. The tumour displayed a fascicular growth pattern. There were scattered collections of lymphocytes and plasma cells. Small foci of neural transformation and neurotropism were present. More than 90% of desmoplasia was seen. It was associated with a lentigo epidermal component. Clark IV level depth of invasion



Figure 1: Ulceroproliferative growth on left foot

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10.4103/0974-2077.155093

was seen. Immunohistochemistry showed strong positivity for S-100 [Figure 4]. The *BRAF* mutation analysis was not available in our institution. The cytomorphology of the inguinal node and in-transit metastasis were similar to primary desmoplastic melanoma. Computed tomography (CT) of the thorax showed multiple nodules noted in both the lung fields in the apical segment, superobasal, posterobasal segments of the right upper lobe and right middle lobe, and anterior segment of left lower lobe. Another pleural-based nodule was seen on the right side [Figure 5].

A preoperative diagnosis was made as desmoplastic malignant melanoma, with regional and systemic metastases. As the patient had extensive metastases and was not willing to undergo palliative surgery, we started palliative chemotherapy with dacarbazine and cisplatin. In the follow-up period, the patient expired at the eighth month of follow-up.

**DISCUSSION**

Desmoplastic melanoma is commonly located on the head and neck (53.2%), followed by the extremities (26.2%) and trunk (20.6%). In the extremities, the upper

extremities represent 70% of the melanoma compared to the lower extremities. Occurrence of desmoplastic melanoma is more on sun-exposed areas.<sup>[1]</sup> Most of the desmoplastic melanoma is diagnosed when it belongs to Clark Levels IV and V.

The risk factors are male gender, old age, and ultraviolet radiation exposure to damaged skin. Lentigo malignant melanoma is a common type of melanoma associated with desmoplasia followed by superficial spreading melanoma and atypical lentiginous hyperplasia.<sup>[2]</sup> When the neurotrophic desmoplastic melanoma involves the head and neck region, it can involve the trigeminal nerve and facial nerve.

**Classification**

Desmoplastic melanoma is classified as pure desmoplastic melanoma and mixed desmoplastic melanoma based on the degree of desmoplasia present in the tumour. More than 90% of the desmoplasia occurs in pure desmoplastic melanoma, whereas, less than 10% of the desmoplasia occurs in mixed desmoplastic melanoma.<sup>[3]</sup> The classification of desmoplastic melanoma is important for its clinical, therapeutic, and prognostic significance. Pure desmoplastic melanoma is associated with better survival and fewer regional



Figure 2: Left inguinal lymph adenopathy



Figure 3: In-transit lymph node

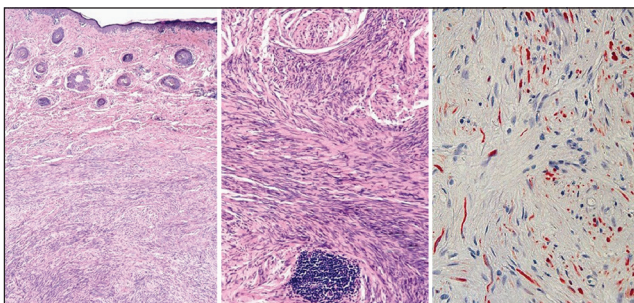


Figure 4: Histopathologic examination shows features of desmoplastic melanoma, both in low and high magnification, with eosin and hematoxylin staining. Last image shows positivity for S100 immunohistochemistry



Figure 5: CECT thorax, with pulmonary metastases

metastases and recurrences compared to mixed desmoplastic melanoma.<sup>[4]</sup>

### Clinical features

Desmoplastic melanoma presents as papules, plaques or nodules, with ulceration. More than 50% of the desmoplastic melanoma is amelanotic.<sup>[5]</sup> Desmoplastic melanoma presenting with atypical pigmentation tends to have a better prognosis, because it is often recognised earlier.

### Histopathologic features

Desmoplastic melanoma is characterised by a proliferation of the spindle cells, which are non-pigmented, spindle-shaped, melanocytes, with an abundant collagen matrix. Cellular atypia varies from near normal to moderate atypia. The nuclei of these spindle cells tend to be hyperchromatic and the cytoplasm is usually scant, with an increased nucleus-to-cytoplasm ratio.<sup>[6]</sup> When the desmoplastic melanoma invades the surrounding nerves it is desmoplastic neurotropic melanoma.<sup>[7]</sup> Neurotropism is associated with deeper tumours, greater mitotic activity, local infiltration, multiple recurrences, and more metastases than without neurotropism. Perivascular nodular clusters lymphocytes with few plasma cells, and desmoplastic melanoma, having *in situ* epidermal components, are additional important findings for the diagnosis of desmoplastic melanoma.

### Immunohistochemistry<sup>[8-11]</sup>

Immunohistochemistry is used to differentiate desmoplastic melanoma from other benign and malignant conditions. Differential diagnoses include, hypertrophic scar, sclerosing melanotic nevi, fibromatosis, atypical fibroxanthoma, dermatofibroma, malignant nerve sheath tumours, and desmoplastic spindle cell squamous cell carcinoma.

- a. SOX 10 is expressed in desmoplastic malignant melanoma, malignant peripheral nerve sheath tumours, gliomas, and certain types of breast cell carcinoma. The sensitivity and specificity of SOX 10 in the diagnosis of desmoplastic melanoma are 100 and 93%, respectively. Thirty percent show positive in malignant peripheral nerve sheath tumour. It is negative in poorly differentiated squamous cell carcinoma, atypical fibroxanthoma, and sarcoma.
- b. S100 is used for diagnosis of desmoplastic melanoma. It is expressed in desmoplastic melanoma (96%), chondrosarcoma (75%), extra-skeletal myxoid chondrosarcoma (45%), malignant peripheral nerve sheath tumour (40%), rhabdomyosarcoma (24%), Ewing sarcoma (21%), and synovial sarcoma (15%).
- c. NGFR (Nerve growth factor receptor). One-hundred percent positivity is seen in spindle

cell melanoma and desmoplastic melanoma. However, weekly positivity is seen in desmoplastic sclerotic nevus.

- d. Melan-A/Mart-1.

Melan-A expression was found in 51.4% of desmoplastic melanoma. Melan-A is weakly positive in desmoplastic melanoma, whereas, it is strongly positive in the desmoplastic sclerotic nevus.

### Prognosis

Regional lymph node involvement is reported in 0 to 13.7% of the cases, which is less frequent than other cutaneous melanoma.<sup>[12]</sup> Lymph node metastasis is significantly higher for mixed desmoplastic melanoma (24.6%), compared to pure desmoplastic melanoma (9.0%). Therefore, sentinel lymph node biopsy has a value in desmoplastic melanoma. However, in neurotropic melanoma there is sentinel significant involvement of the lymph node and sentinel lymph node biopsy is recommended.<sup>[7]</sup>

### Management

- a. Role of surgery.

Subjects having a wide local excision with margins greater than 1 cm have better survival than those undergoing excision less than 1 cm. Desmoplastic malignant melanoma excised with a margin less 1cm has a higher rate of local recurrence than those excised with a margin more than 2 cm.<sup>[1]</sup> Thus, a wide local excision more than 2 cm is recommended.

- b. Role of adjuvant radiotherapy.<sup>[13,14]</sup>

Role of radiotherapy is limited in desmoplastic melanoma, but indicated in the following situations: Deep infiltration, when located on cosmetically, structurally or functionally sensitive areas, tumour excised with narrow margins, tumour with positive margins, neurotropism.

- c. Role of chemotherapy.<sup>[15]</sup>

High-dose interferon was the approved drug for high-risk melanoma. Another trial shows a comparison between high-dose interferon and biochemotherapy consisting of dacarbazine, cisplatin, vinblastin, interleukin-2, interferon alpha 2b, and the granulocyte colony stimulating factor, given every 21 days, for three cycles. However, results show that biochemotherapy is a good alternative adjuvant treatment, but no difference is seen in the overall survival and toxicity compared to high-dose interferon.

### CONCLUSION

Desmoplastic melanoma is a rare variant of cutaneous melanoma, which needs proper study for diagnosis and management, on account of atypical presentation and

atypical histopathological features. Histopathological features such as spindle cells in a background of abundant collagen with immunohistochemistry like S-100 positive, with a nerve growth factor study can diagnose desmoplastic melanoma at an early stage. It is mandatory to differentiate pure desmoplastic melanoma from mixed desmoplastic melanoma for management and prognostic significance. Regional lymph node metastasis is unusual in desmoplastic melanoma. Surgery is the main modality of treatment, with a wide local excision, of more than a 2-cm margin, which is necessary to avoid a recurrence.

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**How to cite this article:** Javabal P, Subramanian V. An unusual case of desmoplastic malignant melanoma. *J Cutan Aesthet Surg* 2015;8:60-3.

**Source of Support:** Nil. **Conflict of Interest:** None declared.