SKIN

BRIEF ARTICLE

A Diagnostic Challenge: Desmoplastic Melanoma

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ABSTRACT

Desmoplastic melanoma (DM) is uncommon and can be a diagnostic challenge both clinically and histologically. We report a unique presentation of DM presenting as an amelanotic, depressed, indurated plaque on the forehead. The lesion was clinically concerning for a morpheaform basal cell carcinoma and with an unrevealing initial punch biopsy, an excisional biopsy with immunohistochemistry was necessary to determine a diagnosis of pure desmoplastic melanoma. When a patient presents with a depressed, indurated plaque of unknown etiology, including DM in the clinical differential diagnosis and communicating this to the dermatopathologist can potentially reduce the risk for a delay in diagnosis.

INTRODUCTION

Desmoplastic melanoma (DM) is uncommon, accounting for <4% of new melanoma diagnoses.[1] Clinically, DM is non-specific, often presenting as an amelanotic, flesh-colored nodule or plaque that clinically resembles a scar. DM can mimic both benign and malignant entities, with reports of DM presenting clinically as a neurofibroma, fibrous inflammatory hyperplasia, and sarcoma, among others.[2]

DM is most commonly located on sun-exposed skin and is more frequent in males and older individuals, with a mean age of 70 years at diagnosis.[3] On average, DM tends to be more deeply infiltrative than superficial spreading and lentigo maligna melanoma at the time of diagnosis—a feature which may be related to diagnostic delay and/or differences in tumor biology.[1,3]

CASE REPORT

A 60-year-old male with a history of basal cell carcinoma presented for evaluation of an indurated, slowly expanding, 1 cm skin-colored, depressed plaque of the right forehead. The lesion exhibited rolled borders with mild associated follicular dropout around the periphery (Figure 1). The lesion was otherwise asymptomatic, and the patient denied any history of trauma to the area.

A 4mm punch biopsy of the rolled border was performed, which revealed thickened collagen bundles with underlying spindle cells arranged parallel to the epidermis, overall felt to be consistent with scar (Figure 2). Due to the lack of obvious inciting trauma in the patient's history, the persistent clinical concern, and one focal area of fibromyxoid-like stroma identified in the punch biopsy

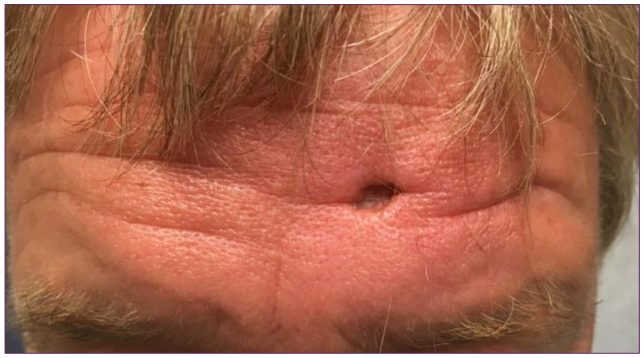

Figure 1. A 1 cm, skin-colored, depressed plaque of the right forehead. Note the apparent decrease in follicular ostia of the skin surrounding the atrophic portion of the lesion

specimen, an excisional biopsy was obtained to more definitively rule out morpheaform basal cell carcinoma. Histopathology revealed focal epidermal atrophy with an increased number of underlying parallel spindle cells and a superficial and deep lymphocytic infiltrate. SOX-10, Melan-A, and S-100 staining were performed and highlighted an increased number of cells in the basal layer as well as a focal proliferation of underlying spindle cells within the dermis (Fig. 3, S-100 staining not shown). The histopathology was consistent with a pure desmoplastic melanoma, with a Breslow Depth of 1.3 mm.

Due to the fact that the lesion extended to within 1-2 mm of the peripheral margins of the excisional biopsy specimen, the patient was referred to our Multidisciplinary Skin and Connective Tissue Oncology Clinic for discussion of further workup and treatment. The patient underwent wide local excision of the area with 1cm margins without sentinel lymph node biopsy or advanced imaging. The patient continues to follow routinely in Dermatology and has not exhibited any clinical evidence of recurrence or metastasis.

DISCUSSION

Histologically, DM is characterized by a paucicellular proliferation of spindled melanocytes accompanied by a densely fibrotic stroma and may closely resemble scar or other spindle cell tumors including atypical fibroxanthoma/pleomorphic dermal sarcoma, spindle cell squamous cell carcinoma (SCC), and leiomyosarcoma.[1]

Immunohistochemistry is essential to the diagnosis; S-100 is diffusely positive and markers of melanocytic differentiation such as Melan-A and SOX-10 are often positive but can be focally negative.[1,4] The absence of staining for epithelial markers such as cytokeratins 5/6 and muscle markers such as smooth muscle actin and desmin help to differentiate DM from spindle SCC and leiomyosarcoma, respectively. Due to these challenges, a delay in diagnosis of DM is not uncommon, particularly if a partial biopsy is taken or if appropriate immunohistochemistry is not performed due to low clinical suspicion.[4]

There are two subtypes of DM—pure DM (pDM) and mixed DM (mDM)—named by the degree of desmoplasia.[3] Mixed DM arises from and thus resembles preexisting lesions of another melanoma subtype (often lentigo maligna melanoma).[3] In contrast, pDM arises de novo.[1,4] As with all melanoma, the mainstay of treatment is wide local excision with margins determined by Breslow depth.[1,5] Compared to mDM, pDM exhibits a lower propensity for regional lymph node involvement, but a similar rate of distant metastasis, suggesting possible hematogenous spread.[4] Due to the lack of robust evidence, the utility of sentinel lymph node biopsy (SLNB) for patients with pDM remains controversial.[5] In cases of mDM, SLNB should be considered based on tumor stage as is done for conventional melanoma.[5] Although adjuvant radiation therapy is rarely used in the treatment of

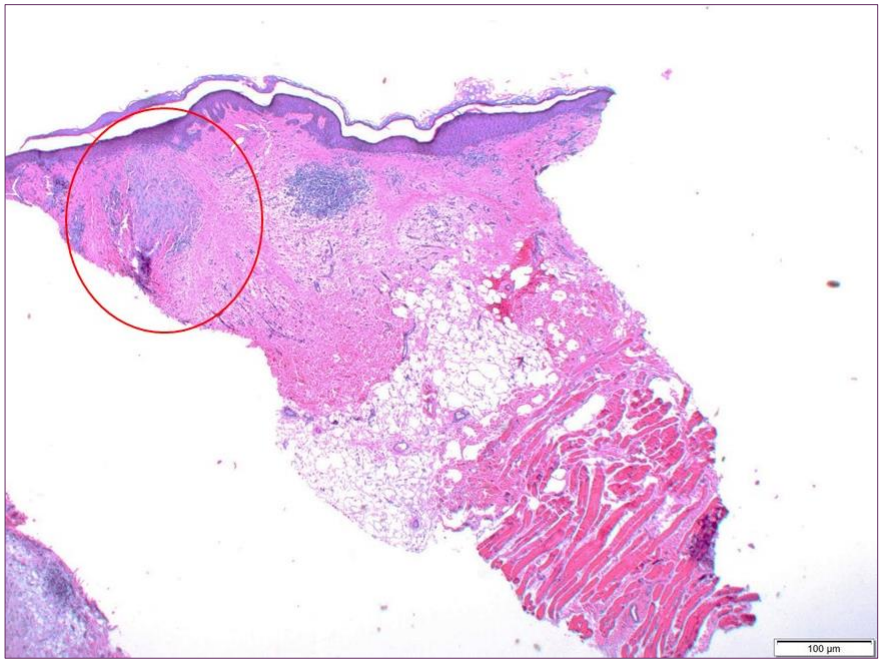

Figure 2. Punch biopsy revealing dense collagen bundles with focally increased dermal spindle cells, parallel to the dermal-epidermal junction. A focal area of fibromyxoid-like stroma is seen (circled). Hematoxylin-eosin stain (Original magnification x2)

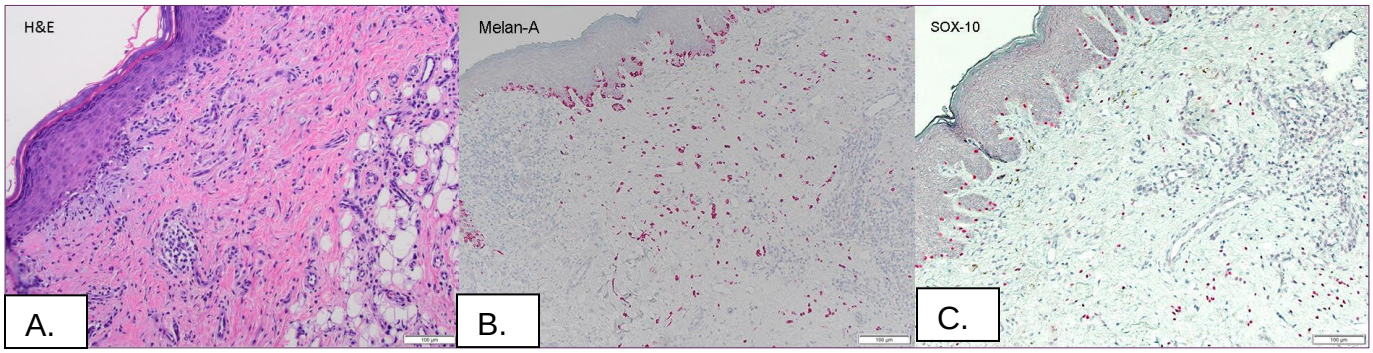

Figure 3. Excisional biopsy revealing focal epidermal atrophy with increased underlying parallel spindle cells and a slightly desmoplastic stroma. Melan-A stain reveals focal increase in basal layer melanocytes and also highlights dermal spindled cells. **A)** Hematoxylin-eosin stain (Original magnification x10) **B)** Melan-A stain (Original magnification x10). **C)** SOX-10 stain (Original magnification x10)

melanoma, it may be considered for improved local control in cases of pDM exhibiting neurotropism, especially with positive surgical margins.[5]

CONCLUSION

This case highlights that DM can be a diagnostic challenge both clinically and histologically. It is important to consider desmoplastic melanoma as part of the differential diagnosis for scar-like lesions presenting in sun-exposed areas, particularly in the absence of prior surgery or trauma.

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