Desmoplastic melanoma presenting as an alopecic patch in a young patient

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INTRODUCTION

Desmoplastic melanoma (DM) is a rare spindle cell variant of melanoma, accounting for only 1% to 4% of cutaneous melanomas.[1,2] Initially described by Conlev et al,[3] as a superficial melanotic lesion with the histology of locally invasive fibrous tumors, DM has slowly increased in incidence.[4] Epidemiologically, risk remains highest in chronically sun-exposed individuals, with a male to female ratio of 2:1, and in older individuals, with a mean age of onset of 66 years.[1]

Timely diagnosis remains challenging due to the amelanotic nature of these lesions, evading detection. Further, lesions often occur as indurated, poorly-demarcated plaques on the head and neck of photoaged skin, requiring the broad differential diagnosis of a scar or nonmelanocytic cutaneous neoplasm.[5] Histologically, DM presents as a proliferation of atypical spindle cells surrounded by collagen bundles invading the reticular dermis[6]; however, there is a variable spectrum of fibrosis and cellularity.[1] A peripheral lymphocytic infiltrate can serve as a low-power clue to the diagnosis and the lesion may be surmounted by overlying lentigo maligna. While lesions are usually positive for S100, other melanocytic immunostains, including MelanA, human melanoma black 45, and preferentially expressed antigen in melanoma, may often be negative.[2]

Presentation as an alopecic patch is an uncommon scenario, with only 4 previous reported cases to the authors' knowledge.[7-10] Here, we report a unique case of DM presenting as an alopecic patch on the scalp of a young male unresponsive to topical steroids.

Abbreviations used:

AN: alopecia neoplastica
DM: desmoplastic melanoma

CASE DESCRIPTION

A 43-year-old White male with a history of pilar cysts previously treated with topical steroids and excisions presented to his primary care provider with a 3 × 2 cm patchy area of hair loss located on the right frontal scalp for several months (Fig 1). Due to the clinical concern for alopecia areata, he was prescribed fluocinonide 0.05% topical solution. The patch failed to respond to topical steroids during the following 2 to 3 months, and he was subsequently referred to dermatology.

Upon evaluation, the patient demonstrated a 3 × 2 cm indurated skin-colored plaque with associated hair loss accompanied by the sensation of 'tingling' pain. His social history included prior sunburns and tanning booth use 3 to 4 times approximately 25 years prior. He had no personal or family history of skin cancer, including melanoma.

Two punch biopsies were obtained on the right central frontal scalp. Histopathologic examination revealed a proliferation of spindled cells with hyperchromatic nuclei positioned between thickened collagen bundles encompassing the entirety of the dermis and extending into the subcutis (Fig 2, *A* and

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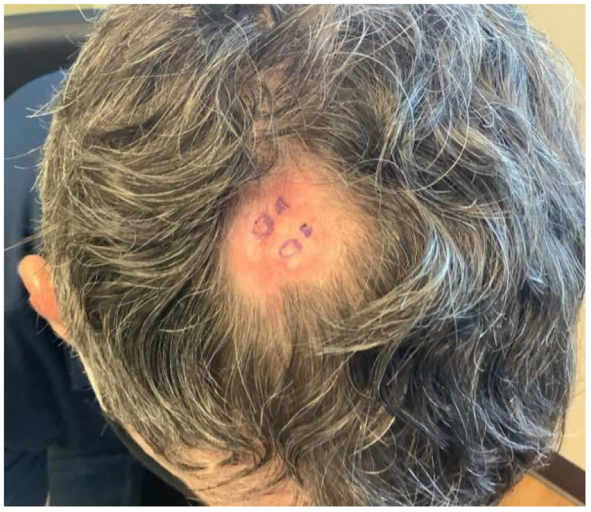

Fig 1. Clinical presentation. An alopecic patch on the right central frontal scalp present for several months and measuring 3 × 2 cm, refractory to treatment with topical steroids.

B). There was a prominent surrounding lymphocytic infiltrate with a low mitotic rate (1/mm^2) and no definitive perineural invasion. The remnant follicles displayed miniaturization, possibly secondary to underlying androgenetic alopecia, but were largely displaced by tumor and zones of desmoplasia (Fig 2, *C*). An associated junctional melanocytic proliferation was not seen. Immunostains confirmed that the spindle cells were positive for SRY-box transcription factor 10 (Fig 2, *D*) and S100, faintly but diffusely positive for preferentially expressed antigen in melanoma, and negative for MelanA and p53. These findings confirmed a diagnosis of pure DM, invasive to a Breslow depth of at least 5.7 mm.

The patient was promptly referred for definitive treatment via wide local excision of the primary site. A 2.5 × 2 cm area of tissue from the right frontal scalp was removed, and a full-thickness donor graft from the right supraclavicular region was used for closure (Fig 3). Sentinel lymph node biopsy of 2 right neck

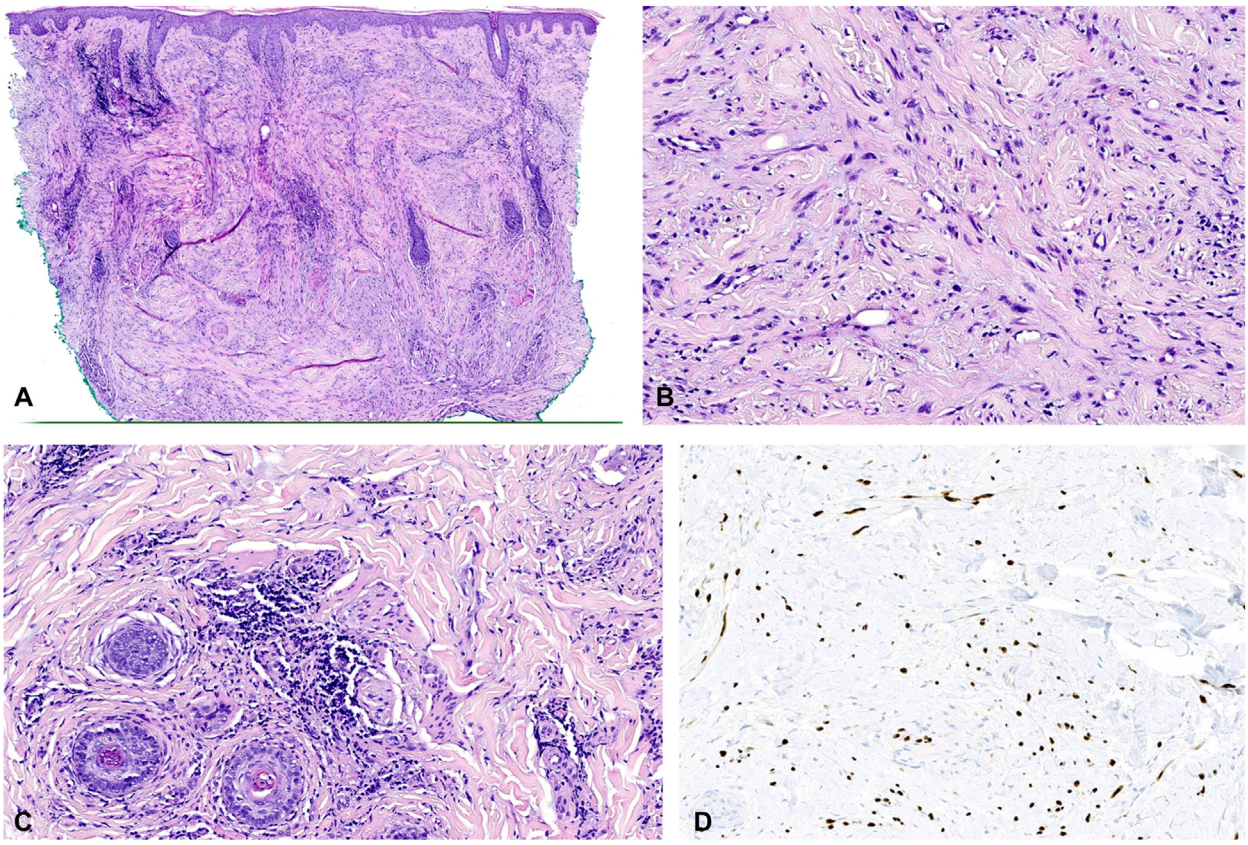

Fig 2. A, Representative section from a vertically sectioned punch biopsy containing a dense spindle cell proliferation with associated desmoplastic stroma and associated peripheral lymphoid aggregates; mitotc rate = 1/mm^2. (Hematoxylin-eosin stain; original magnification ×40) **B,** Atypical spindle cells with hyperchromatic, pleomorphic nuclei and associated desmoplastic stroma. **C,** Transverse section highlighting follicular miniaturization and obliteration by atypical spindle cell neoplasm with associated desmoplastic stroma and brisk lymphocytic infiltrate (**B** and **C**, Hematoxylin-eosin stain; original magnifications: **B**, ×200; **C**, ×200.). **D,** Atypical spindle cells display strong positivity for SRY-box transcription factor 10 (SRY-box transcription factor 10; original magnification ×200).

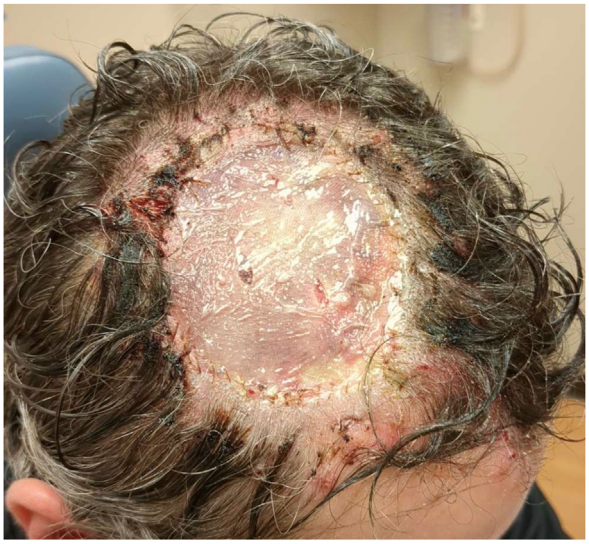

Fig 3. Treatment follow-up. Definitive treatment of the primary right scalp melanoma was performed with wide local excision, full-thickness skin graft closure with donor tissue from the right supraclavicular region, and sentinel lymph node biopsy. Follow-up depicted at 10 days postgraft placement.

lymph nodes were negative for malignant spread. Evaluation of the excisional specimen revealed a final tumor depth of 8.0 mm with final pathologic staging of IIB (pT4aN0m).

DISCUSSION

Due to its variable clinical and nonspecific morphology, DM requires high clinical suspicion. We present a rare case of DM presenting as an alopecic patch in a young individual. This case was additionally complicated by a history of pilar cysts requiring recurrent treatment at the site of hair loss, which potentially delayed the patient's concern regarding this lesion, further confounding the diagnosis.

Neoplastic processes are a rare cause of alopecia resulting from primary and metastatic tumors. Alopecia neoplastica (AN), cutaneous metastasis caused by neoplastic cells replacing hair follicles with fibrous tracts,[7] is believed to result from either direct fibrosis or the adjacent release of cytokines.[10] Commonly, AN is secondary to metastatic visceral malignancies from the gastrointestinal tract, breast, kidney, and lung.[10] Primary AN is a more infrequent phenomenon, occurring in 11 cases as of 2021[10]; among these cases, primary malignant neoplasms included angiosarcomas, hemangioendotheliomas, ectopic extramammary Paget disease, syringomatous carcinoma, amelanotic melanoma, and DM.[10] While primary AN may result from benign neoplasms including neuromas, nerve sheath myxomas, or nevocellular nevi, it is important to acknowledge malignant sources with high morbidity.

Previously, 4 cases have reported DM presenting as scar-like alopecia. The first identified a 50-year-old man with a localized erythematous patch of hair loss on the left frontal scalp.[7] Three cases presented with alopecic patches of a few months' duration[8,9]; the differential included alopecia areata, discoid systemic lupus erythematosus, and lichen planus. After failure to respond to standard therapy, 2 patients underwent biopsy followed by wide local excision of the primary lesion.[7,8] A third patient sought surgical treatment at an outside facility but was lost to follow-up,[8] while the fourth patient underwent radiation therapy vs surgery due to lesion size.[9] A few patients experienced multiple recurrences and even neurotropism in the first reported case with involvement of the trigeminal nerve, emphasizing risk of neurologic involvement.[7]

While an atypical presentation of DM further complicates a clinically challenging diagnosis, our patient lacked previous malignancy or evidence of similar lesions elsewhere that would raise suspicion for a neoplastic/metastatic etiology of his alopecia. Further, our patient had a history of pilar cysts requiring repeated incision and drainage locally and at the site of concern. Despite the high probability for a benign cause, the expedient primary care provider referral to dermatology played a formative role in the timely detection of his DM. Additionally, high clinical suspicion by his primary dermatologist led to prompt biopsy as opposed to further clinical trials of topical or injectable therapies. This underscores the importance of tissue sampling in cases of alopecia with an atypical clinical course.

Conflicts of interest

None disclosed.

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