Multiple pediatric dermatomyofibromas in a patient with a history of embryonal rhabdomyosarcoma

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INTRODUCTION

Dermatomyofibroma (DMF) is an uncommon benign myofibroblastic tumor most frequently seen in adult women; however, there are several reports of pediatric dermatomyofibroma.[1,2] Lesions typically appear as long-standing, asymptomatic, skin-colored to red-brown, indurated plaques.[1] DMF typically presents as a single lesion, with multiple DMFs representing an exceedingly rare presentation.[3] Here, we present a case of multiple DMFs in a pediatric patient with a history of embryonal rhabdomyosarcoma and propose a possible association between multiple DMFs and embryonal tumors.

Abbreviation used:

DMF: dermatomyofibroma

CASE REPORT

A 7-year-old girl with a history of embryonal rhabdomyosarcoma of the left maxillary sinus treated with proton beam irradiation and chemotherapy, in remission for 18 months, presented with a 6-month history of 2 slowly growing asymptomatic lesions in the posterior aspect of the neck. Examination demonstrated hypoplasia of the left maxilla at the site of the previously treated embryonal rhabdomyosarcoma and 2 0.5-1-cm rubbery superficial nodules in the posterior and left aspects of the neck (Fig 1). Punch biopsies of both lesions demonstrated proliferations of bland spindle cells arranged in intersecting bundles and fascicles parallel to the epidermis. No cytologic atypia or mitoses were noted. Lesional cells were diffusely vimentin-positive. Immunohistochemical stains for CD34, myogenin, smooth muscle-specific actin, and desmin were negative (Fig 2). She was diagnosed with multiple DMF with plans to monitor. One year after initial presentation, an additional asymptomatic lesion in the posterior aspect of the neck developed with a morphology similar to the previously biopsied lesions. At that time, the lesion was neither excised nor biopsied as *per* the patient's family's request. Two years after the initial presentation, she returned with an increase in size of all lesions, which otherwise appeared similar to their initial appearance on examination. Given the ongoing growth of the lesions, additional biopsies were performed of the new lesion along with a previously biopsied lesion. Punch biopsies revealed histopathologic and immunophenotypic features consistent with the previous biopsies. Shortly after, the patient underwent magnetic resonance imaging of the head and neck for rhabdomyosarcoma recurrence monitoring, which demonstrated no involvement of the deeper structures of the neck. Three years after initial presentation, the patient returned for monitoring with an additional lesion in the left supraclavicular region. Again, punch biopsy demonstrated findings similar to previous biopsies consistent with DMF. At 6-month and 7-year follow-ups, the patient had no new or growing lesions.

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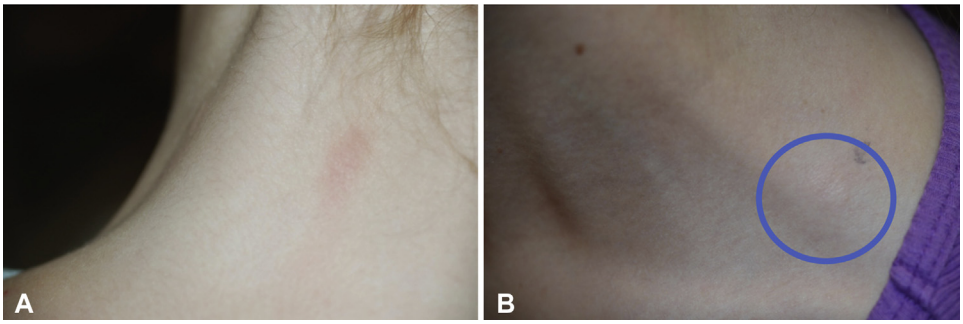

Fig 1. Clinical findings in a patient with multiple DMFs. **A,** Firm, pink, rubbery plaque in the posterior middle aspect of the neck. **B,** Firm, skin-colored to light-pink, rubbery plaque on the left medial clavicle.

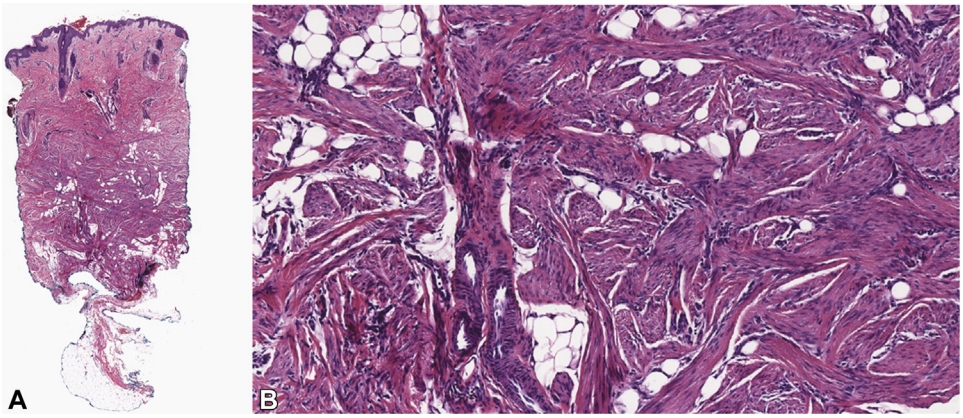

Fig 2. Histopathologic findings in DMF. **A,** Punch biopsy showing fascicles and bundles of monomorphic spindled cells oriented parallel to the epidermis, filling the mid and deep dermis. **B,** Spindle cell proliferation sparing adnexal structures. (**A** and **B,** Hematoxylin-eosin stain; original magnifications: **A,** ×40; **B,** ×200.)

DISCUSSION

Multiple DMFs are extremely rare, with only 4 reported cases to date.[3-6] Interestingly, with the addition of our case, 80% of the reported cases of multiple DMFs occurred in pediatric patients, suggesting that this phenotype may be more common in this population.[3-5] Furthermore, similarly to our case, one of the few other reported cases of multiple pediatric DMFs presented in a child with a history of embryonal "small-round-cell" solid tumor, raising the possibility of an association between multiple DMFs and embryonal tumors.[3] The reason for this possible association remains unclear but may be related to upregulation of growth factors, fibrogenic cytokines, and signaling pathways, leading to myofibroblast proliferation. While DMF is, in itself, a benign entity, it can mimic neoplasms, with locally aggressive or metastatic growth patterns and should undergo biopsy for definitive diagnosis.[2] The immunohistochemical profile of DMF is variable. A recent review of pediatric DMF identified vimentin, a nonspecific stain, which is positive in mesenchymal tumors, as the only consistently positive stain.[2] Positivity of SMA, CD34, desmin, and factor XIIIa has also been reported.[2] In male pediatric patients, DMF typically demonstrates a period of growth followed by stabilization and spontaneous regression; in contrast, pediatric DMF typically persists in female patients.[2] Given their benign nature and frequent resolution, monitoring without intervention is recommended, especially when the lesion is in a cosmetically sensitive area where full excision may have significant esthetic consequences.[2] However, given the potential association between multiple DMF and embryonal tumors reported herein, we recommend a full review of systems and elevated suspicion of internal malignancy in these patients. Further investigation is necessary to determine whether or not a definitive link between these conditions exists.

Conflicts of interest

None disclosed.

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