Plaque With Central Ulceration on the Abdomen

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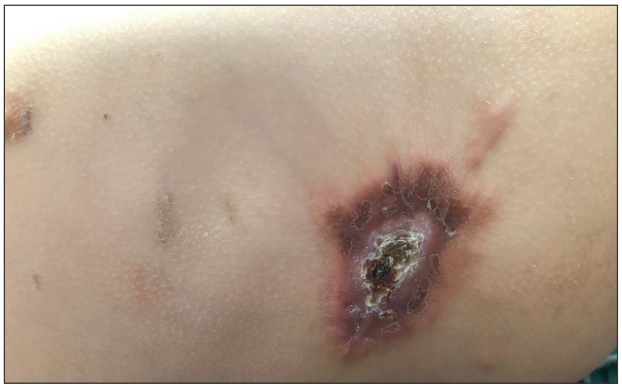

A 14-month-old girl presented to the dermatology department with a firm asymptomatic lesion on the abdomen of 6 months' duration. The lesion started as a flesh-colored papule and developed slowly into an indurated plaque that darkened in color. The patient had no history of trauma to the area. Physical examination revealed a dark reddish–brown, indurated, irregularly shaped plaque with central ulceration and elevated borders on the right abdomen. The plaque measured 2×3 cm with a few smaller satellite nodules distributed along the periphery. Abdominal ultrasonography revealed a multinodular proliferation in the dermis and subcutis of the right abdomen.

WHAT'S YOUR **DIAGNOSIS?**

a. dermatofibrosarcoma protuberans
b. dermatomyofibroma
c. multiple clustered dermatofibromas
d. plaquelike myofibroblastic tumor
e. proliferative fasciitis

PLEASE TURN TO PAGE 119 FOR THE DIAGNOSIS

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THE **DIAGNOSIS:** Plaquelike Myofibroblastic Tumor

An incisional biopsy of the plaque demonstrated a hypercellular proliferation of bland spindle cells in the dermis that infiltrated the subcutis. The overlying epidermis was mildly acanthotic with both ulceration and follicular induction. There was trapping of individual adipocytes in a honeycomb pattern with foci of erythrocyte extravasation, microvesiculation, and widened fibrous septa (Figure 1). Immunohistochemistry was positive for vimentin, actin, and smooth muscle actin (SMA) (Figure 2A). Variable positivity for Factor XIIIa antibodies was noted. CD68 staining was focal positive, suggesting fibrohistiocytic lineage. Expression of CD31, CD34, S100, and anaplastic lymphoma kinase was negative, and Ki-67 was present in less than 10% of cells (Figure 2B).

We reviewed the case in conjunction with soft-tissue pathologist (Y.L.), and based on the clinical and immunophenotypic features, a diagnosis of plaquelike myofibroblastic tumor (PLMT) was made. The patient's parents refused further treatment, and there was no sign of disease progression at 6-month follow-up.

Plaquelike myofibroblastic tumor is an unusual pediatric dermal tumor that was first described by Clarke et al[1] in 2007. Clinical manifestation of PLMT on the right abdomen was unique in our patient, as the lesions typically present as indurated plaques on the lower back, but the central ulceration in our case resembled a report by Marqueling et al.[2] Ulceration and induration of PLMT developing at 8 months of age can suggest an aggressive disease course corresponding with deep infiltration and is seen mostly in children.

The histopathologic features of PLMT include an acanthotic epidermis and follicular induction, which also are characteristic of dermatofibroma (DF). The proliferation of spindle cells extended deep into the fat with foci of erythrocyte extravasation and microvesiculation of the stroma similar to nodular fasciitis and proliferative fasciitis. The presentation of infiltrating and expanding fibrous septae and trapping of individual adipocytes in a honeycomb pattern is similar to dermatofibrosarcoma protuberans (DFSP). Most cases of PLMT are positive for SMA. Factor XIIIa typically is variably positive, and in one report, 31% (4/13) of cases showed positive staining for calponin.[3] Rapid growth, ulceration, and recurrence emphasize that PLMT can be locally aggressive, similar to DFSP.[4]

The main differential diagnoses include DF and its variants, dermatomyofibroma, DFSP, and proliferative

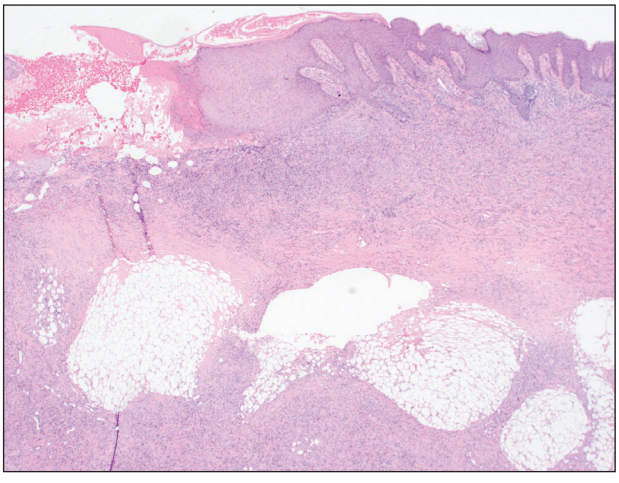
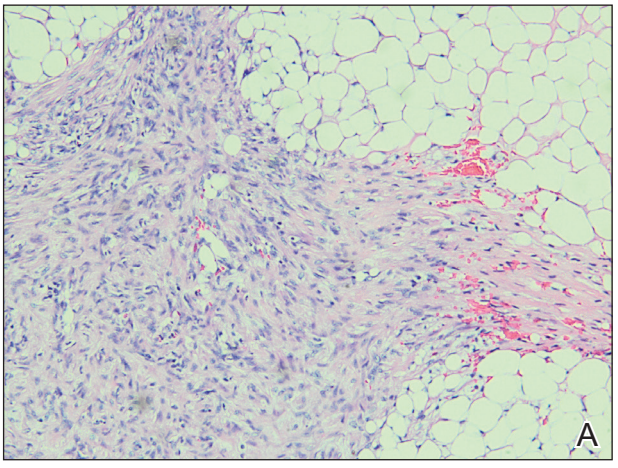


FIGURE 1. Histopathology of the plaquelike myofibroblastic tumor revealed overlying acanthosis and follicular induction resembling a dermatofibroma (H&E, original magnification ×40).



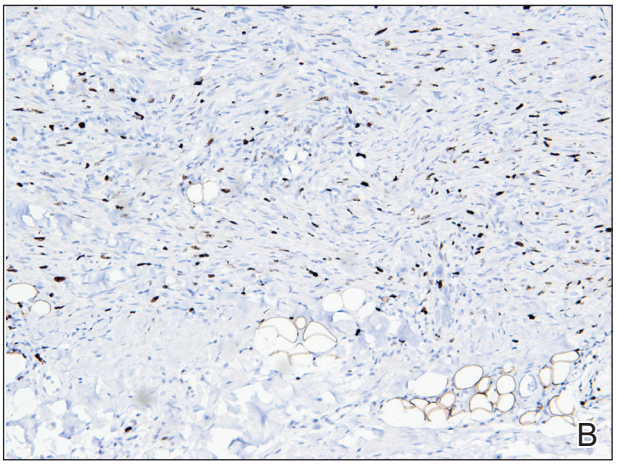


FIGURE 2. A, Histopathology also revealed a proliferation of spindle cells that extended deep into the fat with foci of erythrocyte extravasation and microvesiculation of the stroma (H&E, original magnification ×100). B, Ki-67 was present in less than 10% of cells (original magnification ×100).

fasciitis.[3,5] In the cases mentioned above, microscopic features were similar with a relatively well-circumscribed proliferation of spindle cells arranged in short fascicles through the entire reticular dermis, and the overlying epidermis was acanthotic.

Dermatofibroma commonly manifests in adults as a minor nodular lesion (commonly <1 cm), and usually is located on the legs. It has several clinical and histologic variants, including multiple clustered DF (MCDF)—a rare condition that has been reported in children and young adults and generally appears in the first and second decades of life. Of the reported cases of MCDF, immunohistochemical staining for SMA was performed in 8 cases. All these cases showed negative or minimal staining.[3,4,5] Smooth muscle actin staining in DFs is negative, or weak and patchy, unlike in PLMT where it is diffuse, uniform, and strong.

Dermatofibrosarcoma protuberans typically occurs in young adults and manifests as dermal and subcutaneous nodular/multinodular or plaquelike masses, with rare congenital cases. Immunohistochemical staining for CD34, which typically is firmly and diffusely positive, is the most reliable marker of DFSP.[6] Factor XIIIA in DFSP typically is negative for focal staining, mainly at periphery or in scattered dendritic cells. The prognosis of DFSP generally is excellent, with local recurrences in up to 30% of cases and extremely low metastatic potential (essentially only in cases with fibrosarcomatous transformation).[6] Dermatomyofibroma is another rare benign dermal myofibroblastic tumor that typically manifests with indurated hyperpigmented or erythematous plaques or nodules on the shoulders and torso.[6] This condition occurs mainly in adolescents and young adults, unlike PLMT. The most striking features of dermatomyofibroma are the horizontal orientation of the spindle cell nuclei and the pattern of the proliferation concerning the adnexal structures, especially hair follicles. The hair follicles have a normal appearance, and the proliferation extends up to each follicle, then continues to the other side without any displacement of the follicle. Tumor cells are variably positive for SMA in dermatomyofibromas and are negative for muscle-specific actin, desmin, S100, CD34, and Factor XIIIA.[6]

Immunohistochemistry can be very useful in differentiating PLMT from other conditions. Neoplastic cells stain positively for CD34 but not for Factor XIIIa and SMA in cases of DFSP. Dermatofibroma and its variants always present with collagen trapping at the periphery of the lesions and may demonstrate foamy macrophages, hemosiderin, or plasma cells FXIIIA(+), CD34(−), and variable SMA reactivity. This positivity usually is less prominent in DF than in PLMT. Neoplastic cells in dermatomyofibroma often stain positive for calponin, but only focally for SMA. The clinical features of dermatomyofibroma include early onset, large size, multiple nodules, and plaquelike morphology. Moulonguet et al[4] hypothesized that, although MCDF and PLMT appear to show some distinctive clinical and histologic features, they also show similarities that could suggest they form part of the myofibroblastic spectrum. Furthermore, Moradi et al[7] also considered them as part of the same disease spectrum because of their overlapping clinical, histologic, and immunohistochemical features.

The microscopic features in our case are notable, as the lesion demonstrated overlying acanthosis and follicular induction, resembling DF. The stroma contained microvesicular changes and erythrocyte extravasation, characteristic of nodular or proliferative fasciitis. Additionally, densely packed spindle cells infiltrated deep into the subcutaneous adipose tissue, similar to DFSP.[2,3] Our findings expand on the reported histopathologic spectrum of this tumor to date.

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