PLAQUE-LIKE MYOFIBROBLASTIC TUMOR: REPORT OF A RECURRENT PEDIATRIC CASE AND REVIEW OF THE LITERATURE

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Keywords Benign tumor, connective tissue, recurrence, skin, child.

Abbreviations **PLMT** = plaque-like myofibroblastic tumor; **SMA** = smooth muscle actin.

Case report. A 2-year-old girl presented with a history of an asymptomatic, slowly growing, 1-cm, firm, reddish-brown indurated plaque on the left scapula, which was first observed at one year of age (Fig. 1). Dermoscopic evaluation showed a brownish central area with peripheral erythema (Fig. 4).

An initial punch biopsy revealed a proliferation of spindle cells arranged haphazardly and in small fascicles within the reticular dermis, leading to a diagnosis of dermatofibroma (Fig. 5). However, during follow-up, the lesion demonstrated abrupt growth, assumed an elongated configuration, and became painful (Fig. 2). Due to clinical concern for dermatofibrosarcoma protuberans (DFSP), a complete surgical excision was performed. Histology remained identical to the biopsy; immunohistochemistry showed expression of CD38 and beta-catenin, but a lack of CD34, thereby excluding DFSP.

Two months postoperatively, several reddish-to-brown nodules developed at the scar periphery, confirming local recurrence (Fig. 3). The lesion was re-excised with wide surgical margins. An expanded immunohistochemistry panel (Fig. 6) revealed strong, diffuse, and uniform positivity for smooth muscle actin (SMA), confirming the diagnosis of plaque-like myofibroblastic tumor (PLMT). After three years of follow-up, the patient remains disease-free.

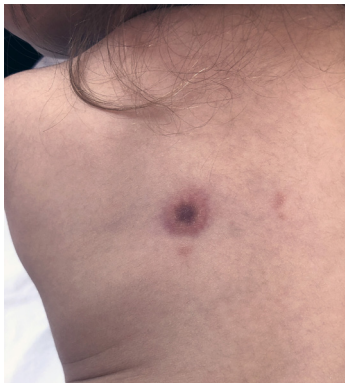

Fig. 1

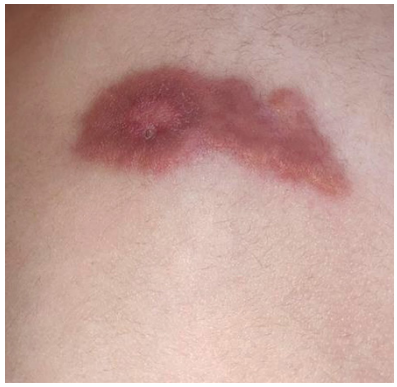

Fig. 2

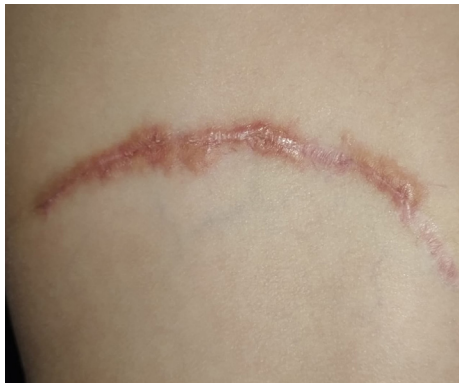

Fig. 3

Fig. 1, 2, 3: Plaque-like myofibroblastic tumor at the first (Fig. 1) and second (Fig. 2) observation; recurrence around the scar after the first excision (Fig. 3).

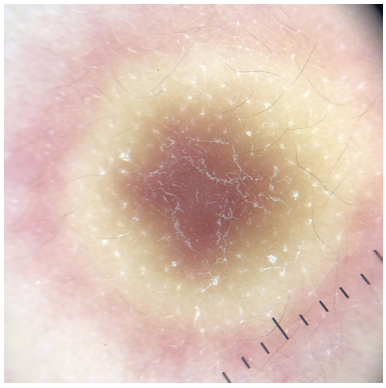

Fig. 4

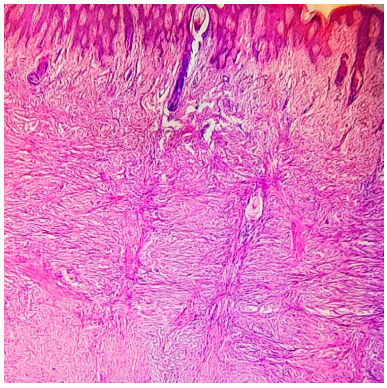

Fig. 5

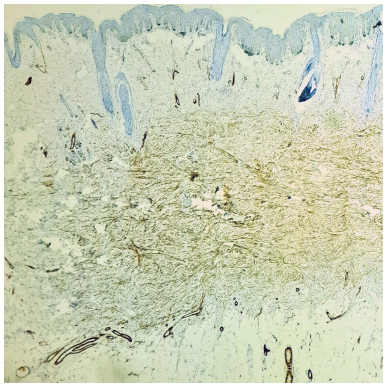

Fig. 6

Fig. 4, 5, 6: Dermoscopic appearance of the plaque-like myofibroblastic tumor at the first observation. Fig. 5 (H&E, x40) shows the histological appearance and Fig. 6 (x40) shows positivity for smooth muscle actin.

Discussion. Plaque-like myofibroblastic tumor (PLMT) was first described in 2007 by Clarke et al. (1). It typically manifests as a slowly growing, firm, indurated plaque, often composed of multiple grouped nodules. While many cases are asymptomatic, pruritus and pain — as observed in our patient — are frequent symptoms. Documented lesions range in size from 2 to 10 cm and are most commonly located on the lower back, hips, and thighs, though the scapula and flank may also be affected (2, 3).

The histopathological hallmark of PLMT is a well-demarcated nodular proliferation of bland spindle and stellate cells within the reticular and deep dermis. Key findings include a "pushing" border at the dermal-subcutaneous junction and a collagenous stroma with "wiry" bundles. Immunohistochemistry is essential for diagnosis: PLMT exhibits strong, diffuse, and uniform SMA positivity, confirming its myofibroblastic lineage. Other common markers include Factor XIIIa (moderate/diffuse positivity) and calponin. Crucially, the tumor is consistently negative for CD34, S-100, and desmin (4, 5).

PLMT is frequently misidentified as dermatofibroma, which is rare in infants and typically shows only focal SMA staining. Other differential diagnoses include: multiple clustered dermatofibroma showing minimal or absent SMA staining, dermatofibrosarcoma protuberans distinguished by a storiform growth pattern and consistent CD34 positivity, dermatomyofibroma featuring spindle cells oriented parallel to the epidermis, and infantile myofibromatosis presenting less-differentiated, round cells alongside the spindle cell component.

PLMT is benign but locally aggressive; recurrence is frequent and is strongly associated with narrow surgical margins. Complete surgical excision is curative, yielding an excellent long-term prognosis (5, 6). In selected pediatric cases, Mohs micrographic surgery may be considered to ensure optimal margin control.

Conclusion. Plaque-like myofibroblastic tumor should be considered in the differential diagnosis of any large, indurated dermal plaque in an infant or young child, particularly when rapid growth or pain conflicts with a histological diagnosis of dermatofibroma. Clinicopathological correlation and a comprehensive immunohistochemistry panel are mandatory for an accurate diagnosis and to ensure appropriate surgical management.

Conflicts of interest

The authors declare that they have no conflicts of interest.

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