Dermatology Practical & Conceptual

A Congenital Plaque-Like Myofibroblastic Tumor

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Case Presentation

A 6-month-old boy presented with a history of congenital plaque on the right peri axillary region. Physical examination revealed an irregular violaceous, infiltrated plaque with central fine scales, measuring 30mm x15 mm (Figure 1A). Dermoscopy showed structureless whitish and violaceous areas, some dotted vessels and a thin peripheral pigmented network (Figure 1B). Histopathological examination of a skin biopsy showed a proliferation of spindle cells extending throughout the reticular dermis, arranged in a storiform pattern and separated from the epidermis by a grenz zone. The superjacent epidermis was hyperplastic with hyperpigmentation of the basal layer (Figure 1, C and D). Immunohistochemistry demonstrated diffuse positivity for smooth muscle actin (Figure 1E) while immunostaining for CD34 was negative. The diagnosis of plaque-like myofibroblastic tumor (PLMT) was made.

Teaching Point

PLMT is a rare, benign pediatric tumor, firstly described in 2007. It occurs commonly in infants and young children within the first 4 years of life [1]. This tumor typically presents as a slowly growing firm, red-brown colored, papulonodular plaque, mostly localized on the lower back [1]. Clinical differential diagnosis includes giant dermatofibroma, multiple clustered dermatofibroma and dermatofibrosarcoma protuberans. Histopathological examination shows the same features as dermatofibroma, but immunohistochemistry favors a myofibroblastic lineage [1]. Recurrence after surgical excision is reported [2].

References

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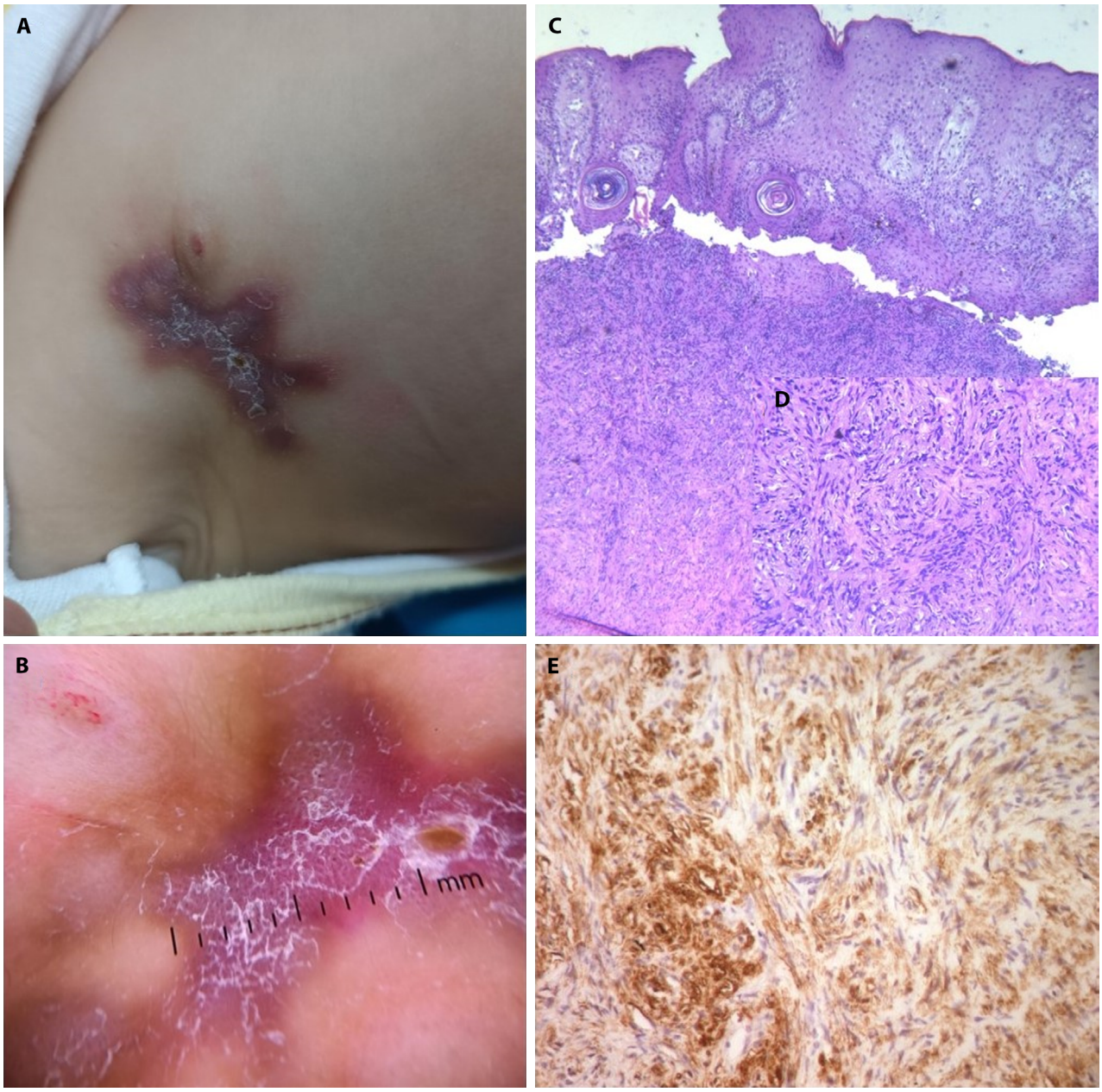

Figure 1. (A) Violaceous nodular plaque of the right peri axillary region. (B) Dermoscopy showed whitish and violaceous areas, peripheral pigmented network and dotted vessels. (C) Acanthosis, proliferation of spindle cells within the reticular dermis (H&E, ×200) arranged in a storiform pattern (D) (H&E, ×200). (E) Strong positivity for smooth muscle actin.