Medallion-Like Dermal Dendrocyte Hamartoma: Differential Diagnosis with Congenital Atrophic Dermatofibrosarcoma Protuberans

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Dear Editor:

Medallion-like dermal dendrocyte hamartoma (ML-DDH) is a recently described congenital benign dermal lesion, which was first reported by Rodríguez-Jurado et al. Clinically, ML-DDH presents as a solitary, several centimeter-sized, round or oval, erythematous to yellow-brown, atrophic plaque on the neck or upper trunk. Histopathologically, ML-DDH is characterized by a proliferation of CD34+ spindle-shaped cells or ovoid cells mainly in the reticular dermis and extending into the subcutis in some cases. Only a small number of ML-DDH has been reported in English literature. Herein, we report a case of ML-DDH that was initially misdiagnosed as congenital atrophic dermatofibrosarcoma protuberans (DFSP).

A 6-year-old girl presented with symptoms of intermittently pruritic and painful, solitary, depressed, and erythematous to yellowish plaque along with fine wrinkles on her posterior neck (Fig. 1). The oval-shaped, 4.0×2.5 cm sized plaque had been present since birth. Other personal and family history was unremarkable. Routine laboratory tests were normal. The clinical impression was a scar or congenital atrophic DFSP. After obtaining an informed consent from the patient and her parents, a punch biopsy was performed on the depressed lesion. The skin biopsy specimen revealed dermal proliferation of spindle-shaped cells in a storiform-like pattern (Fig. 2A, B). The lesion was diffusely positive for CD34, but negative for S-100 protein on immunohistochemistry. Thus, the lesion was initially diagnosed as congenital atrophic DFSP. The patient was sent to a plastic surgeon for complete removal of the...
Fig. 1. A solitary, depressed, erythematous to yellowish plaque with fine wrinkles on the posterior neck. (A) Frontal view, (B) side view.

Fig. 2. (A) Full-thickness dermal cellular proliferation (H&E, ×40). (B) Proliferation of spindle-shaped or ovoid cells arranged focally in a storiform-like pattern (H&E, ×400). (C) Positive staining with CD34 (×400). (D) Positive staining with factor XIIIa (×400).