Dermatol 1994;11:241-245.
6. Elder DE. Dysplastic naevi: an update. Histopathology 2010; 56:112-120.
7. Misago N, Takahashi M, Kohda H. Unilateral dysplastic nevi associated with malignant melanoma. J Dermatol 1991;18: 649-653.
8. Sterry W, Christophers E. Quadrant distribution of dysplastic nevus syndrome. Arch Dermatol 1988;124:926-929.
9. Marghoob AA, Blum R, Nossa R, Busam KJ, Sachs D, Halpern A. Agminated atypical (dysplastic) nevi: case report and review of the literature. Arch Dermatol 2001;137:917-920.
10. Bragg JW, Swindle L, Halpern AC, Marghoob AA. Agminated acquired melanocytic nevi of the common and dysplastic type. J Am Acad Dermatol 2005;52:67-73.

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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.[1] 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[1-3]. 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

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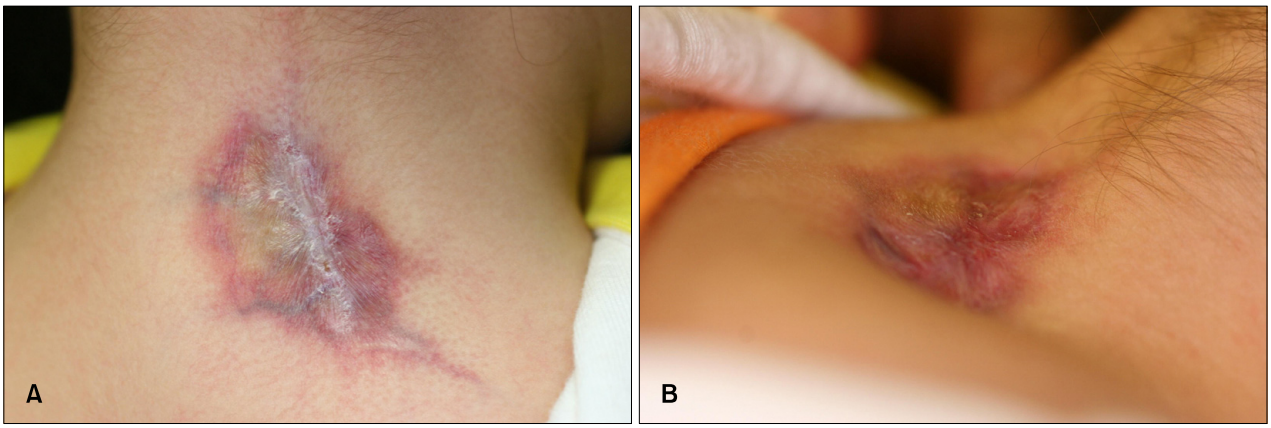

Fig. 1. A solitary, depressed, erythematous to yellowish plaque with fine wrinkles on the posterior neck. (A) Frontal view, (B) side view.

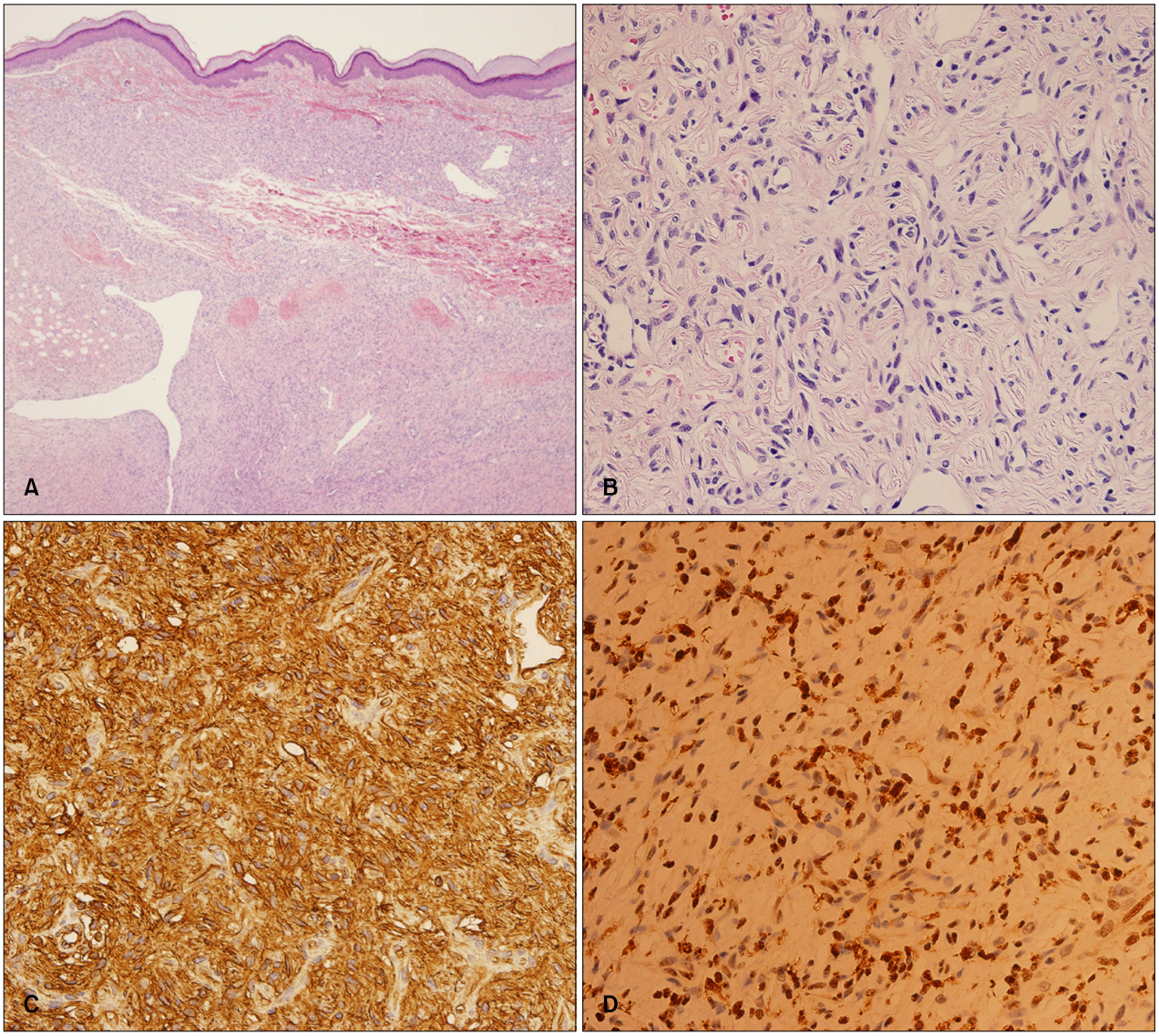

Fig. 2. (A) Full-thickness dermal cellular proliferation (H&E, ×40). (B) Proliferation of spindle-shaped or ovoid cells arranged focally in a storiform-like pattern (H&E, ×400). (C) Positive staining with CD34 (×400). (D) Positive staining with factor XIIIa (×400).

lesion. However, on re-examination of the excised specimen, the diagnosis of congenital atrophic DFSP was questioned with the presence of an ambiguous storiform-like pattern. An issue was raised to include the recently described ML-DDH in the differential diagnosis. The dermal proliferation of spindle-shaped cells extended into the subcutis and was also strongly and diffusely positive for CD34 and factor XIIIa; however, it was negative for CD1a on immunohistochemistry (Fig. 2C, D). These immunohistochemical results favored ML-DDH rather than congenital atrophic DFSP.

Recently, 3 cases of ML-DDH which were initially misdiagnosed as congenital atrophic DFSP were reported[2]. Due to the fact that ML-DDH shares some clinical and histopathological characteristics with congenital atrophic DFSP, the differential diagnosis is difficult. On the immunohistochemical stain, however, the proliferation of spindle cells in ML-DDH is positive for factor XIIIa and fascin as well as for CD34, suggesting a dermal dendritic cell property[1,2], whereas congenital atrophic DFSP is negative for factor XIIIa[4]. In some cases showing ambiguous immunohistochemical results, the fluorescence *in situ* hybridization (FISH) analysis using two-color probes for COL1A1 and PDGFB genes is a useful tool to differentiate DFSP from ML-DDH, because DFSP displays gene translocation t(17;22)(q22;q13) and chimeric COL1A1-PDGFB mRNA expression[2,5]. Nevertheless, the importance of immunochemical stains, such as factor XIIIa and CD34, should not be underestimated in the differential diagnosis between ML-DDH and DFSP.

Taken together, we emphasize the importance of a careful clinicopathological examination, including immunochemical stains such as factor XIIIa and CD34, particularly when the dermatologist encounters a solitary large congenital atrophic lesson on the neck or upper trunk with histopathology of dermal proliferation of spindle cells.

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REFERENCES

1. Rodríguez-Jurado R, Palacios C, Durán-McKinster C, Mercadillo P, Orozco-Covarrubias L, Saez-de-Ocariz Mdel M, et al. Medallion-like dermal dendrocyte hamartoma: a new clinically and histopathologically distinct lesion. J Am Acad Dermatol 2004;51:359-363.
2. Marque M, Bessis D, Pedeutour F, Viseux V, Guillot B, Fraitag-Spinner S. Medallion-like dermal dendrocyte hamartoma: the main diagnostic pitfall is congenital atrophic dermatofibrosarcoma. Br J Dermatol 2009;160:190-193.
3. Restano L, Fanoni D, Colonna C, Gelmetti C, Berti E. Medallion-like dermal dendrocyte hamartoma: a case misdiagnosed as neurofibroma. Pediatr Dermatol 2010;27:638-642.
4. Bandarchi B, Ma L, Marginean C, Hafezi S, Zubovits J, Rasty G. D2-40, a novel immunohistochemical marker in differentiating dermatofibroma from dermatofibrosarcoma protuberans. Mod Pathol 2010;23:434-438.
5. Maire G, Fraitag S, Galmiche L, Keslair F, Ebran N, Terrier-Lacombe MJ, et al. A clinical, histologic, and molecular study of 9 cases of congenital dermatofibrosarcoma protuberans. Arch Dermatol 2007;143:203-210.