

CASE REPORT

Development of Dermatomyofibroma in a Male Infant

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Dermatomyofibroma is a rare benign cutaneous mesenchymal neoplasm of the fibroblasts and myofibroblasts. The majority of dermatomyofibromas present as red-brown discolored plaques or nodules, commonly located on the shoulder, upper arm, axilla, neck, and/or upper trunk. These lesions develop most frequently in young female patients at a mean of 28-years-of-age. Herein, a case of dermatomyofibroma is reported that developed in an infant. A 4-month-old boy presented with an ill-defined bluish firm plaque on the trunk that developed 1 month after birth. Histopathologically, there was proliferation of bland-looking spindle cells with fascicular arrangement in the dermis and subcutaneous tissue. Immunohistochemistry showed that most of the tumor cells expressed diffuse positivity for vimentin and smooth muscle actin, but were negative for S-100 protein, desmin, and CD34. (*Ann Dermatol* 23(S1) S72~S74, 2011)

-Keywords-

Dermatomyofibroma, Infant

INTRODUCTION

Dermatomyofibroma is a rare benign cutaneous mesenchymal neoplasm of fibroblasts and myofibroblasts that was first reported in 1992¹. The lesions are most common in adolescents and young adults. The mean age of diag-

nosis is 28 years and only a few cases have been reported in childhood^{2,3}. Herein, we present a case of dermatomyofibroma in a male infant.

CASE REPORT

A 4-month-old boy presented with an asymptomatic lesion on the trunk. According to the parents, the lesion was first noticed 1 month after birth and had gradually increased in size as the child grew. The family and medical histories were noncontributory. The physical examination revealed an ill-defined bluish firm plaque 2~3 cm on the left flank (Fig. 1). Histopathological examination was performed and the specimen showed dermal proliferation of spindle-shaped tumor cells parallel to the epidermis, in the reticular dermis, extending focally into the subcutaneous tissue (Fig. 2). There was no cytological atypia or mitotic figures. Immunohistochemistry showed that most of the tumor cells expressed diffuse positive responses for vimentin and smooth muscle actin (Fig. 3), but were negative for S-100 protein, desmin, and CD34. The lesion remained stable without any significant changes during 5 months of clinical observation.



Fig. 1. An ill-defined bluish firm plaque on the left flank.

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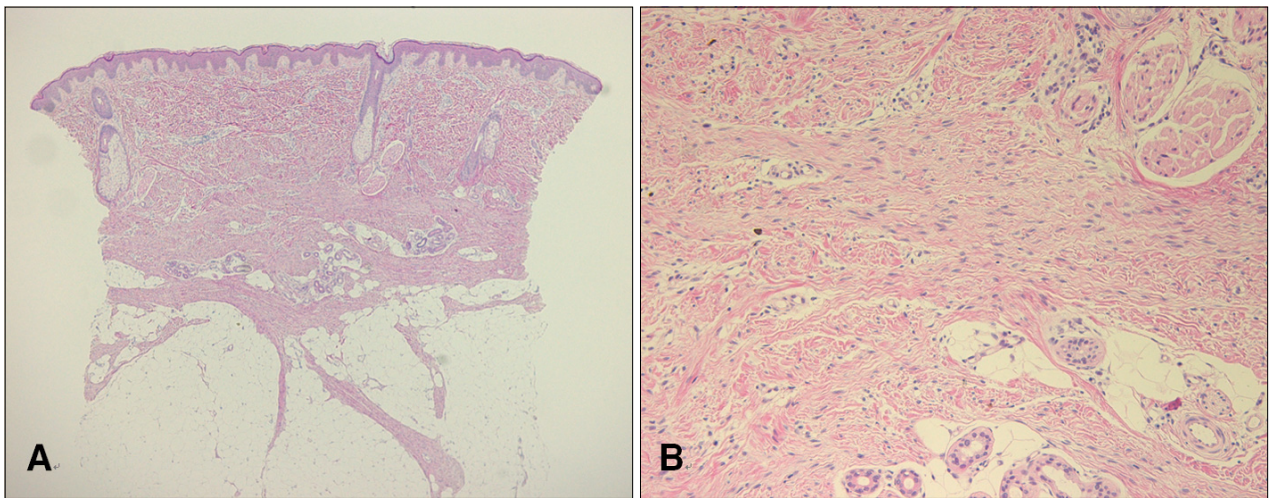


Fig. 2. Proliferation of bland-looking spindle cells with fascicular arrangement in the reticular dermis (A), extending focally into the subcutaneous tissue (B). There was no cytological atypia or mitotic figures (H&E; original magnification, A: $\times 40$; B: $\times 200$).

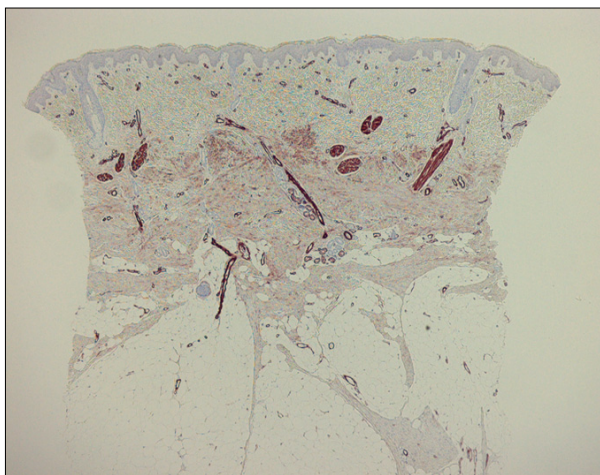


Fig. 3. Immunohistochemistry showed that most of the tumor cells expressed a diffuse positivity for smooth muscle actin (Original magnification, $\times 40$).

DISCUSSION

The majority of dermatomyofibromas present as red-brown discolored plaques or nodules⁴. Dermatomyofibroma develops most frequently in young female patients, and commonly occurs on the shoulder, upper arm, axilla, neck, and/or the upper trunk. In a recent study, female and male children and adolescent patients were reported to be affected equally, in contrast to the female dominant pattern in adults².

To date, only six cases have been reported under the age of 5 years, all of them being boys. Among those, three cases occurred in the infantile period¹⁻⁶. It has been suggested that dermatomyofibroma in male children tends to show

spontaneous regression after childhood, whereas the lesions continue to grow in female patients, possibly due to the effects of female hormones^{2,5}. However, there is no proof for this hypothesis. Further studies evaluating the expression of hormone receptors in the tissue specimen will be of value to clarify this hypothesis.

Histologically, most cases of dermatomyofibroma have a benign, ill-defined, plaque-like dermal proliferation of spindle-shaped tumor cells often arranged parallel to the epidermis^{1,2}. The tumor cells fill the reticular dermis and sometimes extend into the upper part of the subcutaneous layer, whereas the papillary dermis and adnexal structures are spared. The overlying epidermis appears normal, although sometimes a mild basal hyperpigmentation is observed⁵. The elastic fibers are preserved; this finding provides a clue to the diagnosis, because dermatofibroma and hypertrophic scars have altered elastic fibers⁵. Immunohistochemistry should be considered for cases that have non-specific histopathological features. The spindle cells stain positive for vimentin and are occasionally positive for smooth muscle actin. However, they are negative for S-100 protein, desmin, and CD34⁷.

Clinically, scar or keloid should be distinguished from dermatomyofibroma. However, frequently there is a preceding history of trauma or surgery in a scar or keloid. In addition, a whorl-like or nodular structure with no or few adnexae is observed³. The histological differential diagnosis of dermatomyofibroma includes dermatofibromas, dermatofibrosarcoma protuberans, and fibrous hamartoma of infancy. Dermatofibromas are not as plaque-like and are more likely to show epidermal hyperplasia and a haphazard architecture as opposed to the fascicles of dermato-

myofibromas, which are parallel to the epidermal surface⁸. In addition, there is a reduction or loss of elastic fibers and involvement of adnexal structures⁴. Dermatofibrosarcoma protuberans shows a more intense cellular infiltration with storiform architecture and, unlike dermatomyofibroma, it is positive for CD34⁴. In the present case, a fibro/myofibroblastic proliferation, as in fibrous hamartomas of infancy, should have been part of the differential diagnosis, since the patient was an infant. However, fibrous hamartomas of infancy could be excluded because they consist of three distinct components: the combination of fibrocollagenous trabeculae composed of bland spindle cells, small nests of loosely arranged oval or stellate mesenchymal cells set in a myxoid stroma, and mature fat².

The prognosis of dermatomyofibroma is favorable and surgical excision is the treatment of choice. No recurrence or metastasis has been reported yet. In this case, the patient was observed closely without treatment, since dermatomyofibroma in males may regress spontaneously after childhood⁶.

In summary, dermatomyofibroma is a distinctive benign dermal myofibroblastic proliferation that is rare in children, especially during infancy. However, clinicians should consider the diagnosis when characteristic myofibroblastic proliferation is present on skin biopsy.

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