A Case of Dermatomyofibroma in a 2-year-old Boy

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Dermatomyofibroma is a rare benign dermal tumor, mainly found in young women. Clinically it can be confused with keloid or dermatofibrosarcoma protuberans. Typical histopathologic features of dermatomyofibroma are sufficiently distinctive to alert histopathologists to consider dermatomyofibroma in the diagnostic process. We report a case of dermatomyofibroma, presenting as reddish plaques and nodules on the buttock of a 2-year-old boy. Histopathologic examination showed fascicles of uniform spindle cells in the reticular dermis, predominantly oriented parallel to the epidermal surface. Immunohistochemical study and electron microscopy confirmed its myofibroblastic nature.

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Key Words : Dermatomyofibroma, Myofibroblast, Child

Dermatomyofibroma is a recently described rare dermal tumor. Dermatomyofibromas have been diagnosed predominantly in young adult females[1,2]. Clinically they can be confused with scar or keloid, but usually there is no history of previous trauma. Typical histopathologic features of dermatomyofibroma are sufficiently distinctive to alert histopathologists to consider dermatomyofibroma in the diagnostic process. Here we present a case of dermatomyofibroma on the buttock of a 2-year-old boy.

CASE REPORT

A 2-year-old boy presented with slightly pruritic skin lesions on his left buttock. When the boy was 3 months old, his parents noted a pinkish patch on his left buttock. Gradually reddish plaques and nodules developed within the lesion. There was no history of trauma to that site. Physical examination revealed a 9 cm × 1.5 cm red to brown arcuate plaque (Fig. 1). Partial excision of the lateral nodular portion of the lesion was performed. Histopathologic examination showed fascicles of uniform spindle cells and thin collagen fibers in the reticular dermis, predominantly oriented parallel to the epidermal surface. Adnexal structures were preserved within the lesion (Fig. 2A). The spindle cells had faintly eosinophilic cytoplasm and elongated oval nuclei (Fig. 2B). Neither cytological atypia nor significant mitotic activity was observed. Immunohistochemical study revealed that the spindle cells were reactive with vimentin and smooth muscle actin but showed negativity for desmin, S-100 protein, factor XIIIa, and CD34. These overall features were consistent with a diagnosis of dermatomyofibroma. Ultrastructural examination confirmed the myofibroblastic nature of the spindle cells (Fig. 3). They had well-developed rough endoplasmic reticulum, variable amounts of vimentin filaments and mitochondria. Many of the spindle cells also contained intracytoplasmic myofilaments with dense bodies (arrow).

DISCUSSION

Dermatomyofibroma, first described by Kamino et al. in 1992, is a distinct, benign, dermal prolifer-

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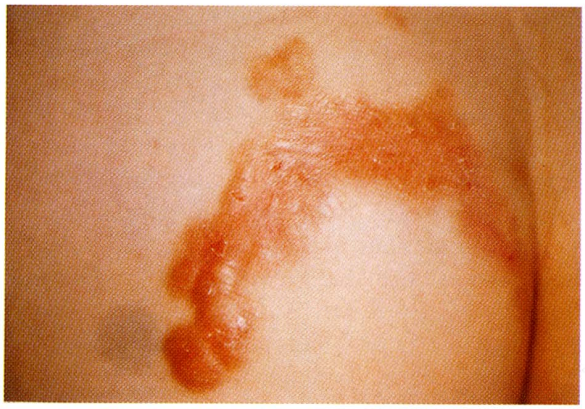

Fig. 1. Red to brown arcuate indurated plaques and nodules on the left buttock

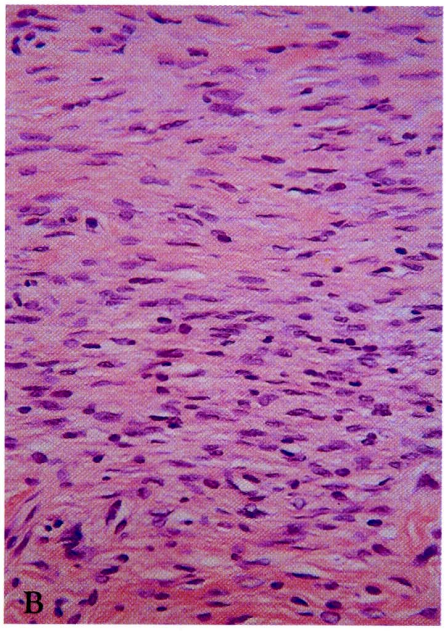

Fig. 2B. The spindle cells show elongated oval nuclei. There is neither cytologic atypia nor significant mitotic activity (H&E, × 200).

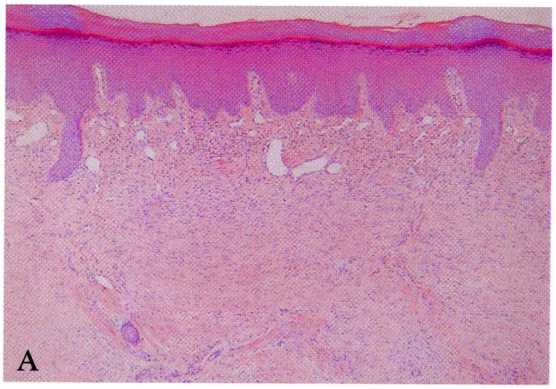

Fig. 2A. Fascicles of spindle cells and thin collagen fibers in the upper reticular dermis show parallel arrangement to the epidermal surface (H&E, × 40).

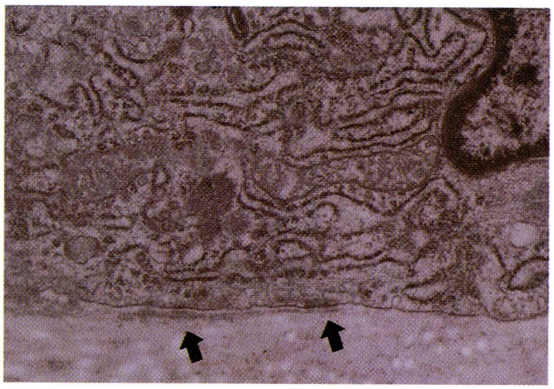

Fig. 3. Electronmicrograph shows the myofibroblastic features of the spindle cells (× 15000). Note the presence of well-developed rough endoplasmic reticulum and intracytoplasmic myofilaments with dense body (arrows).

ation of myofibroblasts and fibroblasts[1]. There is a strong 4:1 female to male predominance in the literature[1-5]. The affected males were significantly younger than the affected females. To our best knowledge, our 2-year-old boy, with a nearly 2-year history, is the youngest patient among the documented cases of dermatomyofibroma. The present case supports the earlier occurrence of this disease in male patients. Dermatomyofibromas have presented typically as a red-brown discolored plaque or nodule around axilla, shoulder or posterior neck[1,2,6]. Lesions up to 8 cm in length have been described[7], although most cases have measured from 1 to 2 cm in the greatest dimension[1,2,4,5]. It is also noticeable that the lesion of our patient was rather large and located on the buttock, a rare site. Like the present case, usually there is no history of previous trauma to the sites of involvement[1,2,6].

Histopathologically, dermatomyofibroma is a benign proliferation of spindle cells, located in the reticular dermis. Fascicles of spindle cells are usually arranged parallel to the epidermal surface. Adnexal structures are spared. The spindle cells are

positive for vimentin and variably positive for smooth muscle actin, whereas they are negative for desmin, S-100 protein and CD34. Factor XIIIa generally stains only scattered dermal dendrocytes[1,6]. Histopathologic findings and immunohistochemical profile in the present case adhered to these typical features.

Ultrastructural study confirms that dermatomyofibroma is a proliferation of both fibroblasts and myofibroblasts. Myofibroblasts share features with fibroblasts such as well developed rough endoplasmic reticulum, vimentin filaments and mitochondria and with smooth muscle cells in that they have aligned cytoplasmic myofilaments with fusiform dense body[1,8,9]. In the present case, the myofibroblastic nature pf the lesion was revealed by electron microscopy.

Clinically, dermatomyofibroma can be confused with a scar or a keloid[6]. However, there is usually a history of previous trauma or surgery in a scar or a keloid, and they show whorl-like or nodular architectures with few or absent adnexae. Dermatomyofibroma should also be distinguished from dermatofibrosarcoma protuberans. Dermatofibrosarcoma protuberans is more cellular, shows a storiform architecture, and is CD34 positive. Histological differential diagnosis of dermatomyofibroma includes dermatofibroma, piloleiomyoma and neurofibroma. Dermatofibroma is more likely to show overlying epidermal changes and a haphazard architecture. Piloleiomyoma is more cellular without the parallel array and is positive for desmin. Neurofibroma lacks the characteristic architecture of dermatomyofibroma and is S-100 positive[1,2].

Most of the patients, based on the documented cases, were treated with complete excision without recurrence or metastases[1,2,5-7]. There was a case treated with intralesional injection of triamcinolone without any improvement[3]. One-year follow-up of our case revealed no evidence of recurrence at the site of partial excision and no clinical change in remaining lesions. Pruritus, the only symptom of this case, was easily relieved with oral antihistamines. We did not carry out a complete excision in this case because the lesion was too big to be removed without permanent scar or disfigurement. Rose and Brocker[2] speculated that in males, dermatomyofibroma may regress spontaneously after childhood, whereas in females, it continues to grow, possibly under the influence of female hormones[4]. We are also doing close observation with the expectation of spontaneous regression in this case.

In summary, the recognition of dermatomyofibroma is important to prevent confusion with more aggressive disease entities, and to prevent unnecessary treatment. Although most dermatomyofibromas occur in young adult females, a number of cases have been reported in male children. As the number of reported cases increases, we will understand the histogenesis and natural history of this disease better.

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