Adult Multiple Myofibromas on an Atrophic Patch on the Thigh

Heesang Kye, In Hyuk Kwon, Soo Hong Seo, Hyo Hyun Ahn, Young Chul Kye, Jae Eun Choi

Department of Dermatology, Korea University College of Medicine, Seoul, Korea

Dear Editor:

Myofibroma is an uncommon, benign myofibroblastic neoplasm of vascular origin, mostly occurring on the skin. It can also involve skeletal muscles, bones, or viscera, which can be fatal. As about 90% of myofibromas develop before the age of 2 years, it was previously termed infantile myofibroma. Adult-onset myofibroma is rare and is distinguishable from the infantile form. It is exclusively solitary and confined to the dermis and subcutis. However, a few cases of adult multiple myofibromas have been reported^{1,2}. Herein, we report a rare and unusual case of adult multiple myofibromas occurring on a preexisting atrophic patch.

A 62-year-old healthy woman presented with multiple firm subcutaneous nodules under a preexisting atrophic patch on the left thigh (Fig. 1). The patch was soft and heterogeneous, composed of atrophic and hypertrophic portions, and was >10 years old. During the last 3 years, multiple firm nodules developed under the patch. She had no history of trauma, burns, or treatment. Two incisional biopsies were performed on the nodule and the patch. Histopathological examination of the nodule showed well-circumscribed, biphasic tumors composed of intersecting fascicles of spindle cells and a vascular compartment resembling a hemangiopericytoma with polygonal cells (Fig. $2A \sim C$). The tumor cells were positive for smooth muscle actin (SMA) staining and were negative for CD34, desmin, and S-100 staining (Fig. 2D, E), consistent with a myofibroma. Histopathological examination of the patch showed mild epidermal and dermal atrophy; however, these findings were not diagnostic. A small tumor suggestive of myofibroma was incidentally found in the dermis; this tumor was not clinically noticeable (Fig. 2F). She is currently being observed without excision, showing no remarkable change for 1 year.

Myofibromas are histologically well-circumscribed, biphasic-patterned tumors consisting of spindle and vascular portions; however, monophasic variants can occur. A myofibroma shows an arrangement of interlacing fascicles of spindle-shaped cells resembling myofibroblasts, and vascular foci resembling hemangiopericytoma with polygonal cells. Spindled cells express vimentin and SMA and are usually negative for desmin³. In our patient, the preexisting



Fig. 1. Multiple firm subcutaneous nodules under the atrophic and telangiectatic soft patch on the left thigh.

Received June 23, 2014, Revised November 14, 2014, Accepted for publication November 21, 2014

Corresponding author: Jae Eun Choi, Department of Dermatology, Korea University Anam Hospital, 73 Inchon-ro, Seongbuk-gu, Seoul 02841, Korea. Tel: 82-2-920-5470, Fax: 82-2-928-7540, E-mail: grace79@ korea.ac.kr

This is an Open Access article distributed under the terms of the Creative Commons Attribution Non-Commercial License (http:// creativecommons.org/licenses/by-nc/4.0) which permits unrestricted non-commercial use, distribution, and reproduction in any medium, provided the original work is properly cited.



Fig. 2. Skin biopsy of the nodule showed well-circumscribed, biphasic tumors (A: H&E, \times 12.5) composed of intersecting fascicles of spindle cells (B: H&E, \times 200) and a vascular compartment with polygonal cells (C: H&E, \times 200). The tumor was positive for smooth muscle actin stain (D: \times 40) and negative for desmin (E: desmin, \times 40), S-100, and CD34 stains. The skin biopsy of the patch showed nonspecific mild epidermal and dermal atrophy with a small tumor of myofibroma in the dermis (F: H&E, \times 12.5).

patch was clinically suspected to be smooth muscle hamartoma; however, the pathologic results were not diagnostic. The patch included a small myofibroma in the dermal layer, which implied that multiple myofibromas would continue to develop under the patch. Kim et al.⁴ previously reported a case of multiple myofibromas with congenital smooth muscle hamartoma on the thigh of a woman, which had very similar clinical manifestation as our case. The pericytes in vascular foci are considered to be the origin of spindle-shaped neoplastic cells that show myofibroblastic differentiation. However, the pericytes can also differentiate into other mesenchymal cells; their differentiation into smooth muscle cells was demonstrated in ultrastructural studies of hemangiopericytomas⁵. Therefore, smooth muscle hamartoma might develop from multipotent pericytes that later create myofibromas. Although the patch was not diagnosed in our case, we suggest the possibility that the preexisting patch and the myofibromas might have originated from the same parental cells before differentiation. Further investigations may help find their possible relation.

REFERENCES

- Granter SR, Badizadegan K, Fletcher CD. Myofibromatosis in adults, glomangiopericytoma, and myopericytoma: a spectrum of tumors showing perivascular myoid differentiation. Am J Surg Pathol 1998;22:513-525.
- Martin AA, Blanchard LE 3rd, Hudgins EM. Adult-onset multiple myofibromas. J Am Acad Dermatol 2011;64:610-611.
- Guitart J, Ritter JH, Wick MR. Solitary cutaneous myofibromas in adults: report of six cases and discussion of differential diagnosis. J Cutan Pathol 1996;23:437-444.
- Kim DS, Lee HJ, Kwon YS, Lee JH. Multiple adult myofibromas with congenital smooth muscle hamartoma. Clin Exp Dermatol 2009;34:254-256.
- Nunnery EW, Kahn LB, Reddick RL, Lipper S. Hemangiopericytoma: a light microscopic and ultrastructural study. Cancer 1981;47:906-914.