Ann Dermatol Vol. 23, No. 2, 2011 DOI: 10.5021/ad.2011.23.2.201

CASE REPORT

A Case of Myopericytoma on the Lower Leg

Jun Oh Paek, M.D., Ho Song Kang, M.D., Kwang Yeoll Yeo, M.D., Hee Joon Yu, M.D., Joung Soo Kim, M.D.

Department of Dermatology, Hanyang University Guri Hospital, Hanyang University College of Medicine, Guri, Korea

Myopericytoma (MP) is a benign tumor composed of cells that show apparent differentiation towards putative perivascular myoid cells called myopericytes. It arises most commonly in the dermis or subcutaneous tissue of the extremities in adults. The most common presentation is a well-circumscribed, slow-growing painless firm mass. A 45-year-old woman presented with a 2-year history of a painless, slowly growing 0.9×0.7 cm sized firm mass in the subcutaneous tissue of the posterior side of the right lower leg. We presumed this lesion to be an epidermal cyst, pilomatricoma or calcinosis cutis and performed an excisional biopsy. The histologic examination showed that it was composed of spindle-shaped myoid-appearing cells in a concentric arrangement, intimately associated with thin-walled vascular channels. Lesional spindle cells were diffusely positive for smooth muscle actin and were negative for CD34, desmin and S100 protein. From these findings, we diagnosed this lesion as a myopericytoma. **(Ann Dermatol 23(2) 201∼204, 2011)**

-Keywords-
Leg, Myopericytoma

INTRODUCTION

Myopericytoma (MP) is one of a family of benign tumors showing a myoid/pericytic line of differentiation. It is composed of oval to spindle-shaped myoid-appearing cells with a striking tendency for concentric perivascular growth[1]. It is characterized by a well-circumscribed, slow-growing painless mass that arises most commonly in the dermis or subcutaneous tissue of extremities in adults[2,3]. In the Korean dermatologic literature, previously reported articles assumed to be related to the disease were cases that occurred in the forearm, infraorbital area and auricle of the ear[4-6]. Unlike previous reports, this is a typical case arising in a lower extremity, which is the common clinical feature of MP.

CASE REPORT

A 45-year-old woman presented with a 2-year history of a slow-growing painless nodule in the subcutaneous tissue of the posterior side of her right lower leg. She did not have underlying disease and trauma history. Upon physical examination, a 0.9×0.7 cm sized, skin-colored firm mass on the posterior side of the right lower leg was observed (Fig. 1). We presumed this lesion as an epidermal cyst, pilomatricoma or calcinosis cutis and

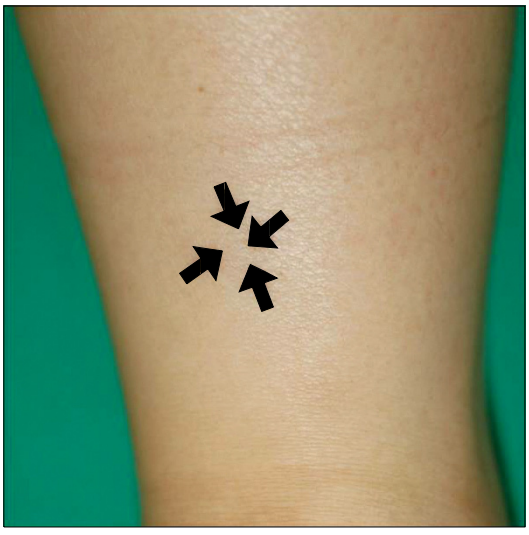

Fig. 1. A soft tissue masscovered with overlying normal skin on the posterior side of the right lower leg.

Received March 26, 2010, Revised April 7, 2010, Accepted for publication April 13, 2010

Corresponding author: Joung Soo Kim, M.D., Department of Dermatology, Hanyang University Guri Hospital, 249-1 Gyomun-dong, Guri 471-701, Korea. Tel: 82-31-560-2285, Fax: 82-31-557-4872, E-mail: tuentuen@hanyang.ac.kr

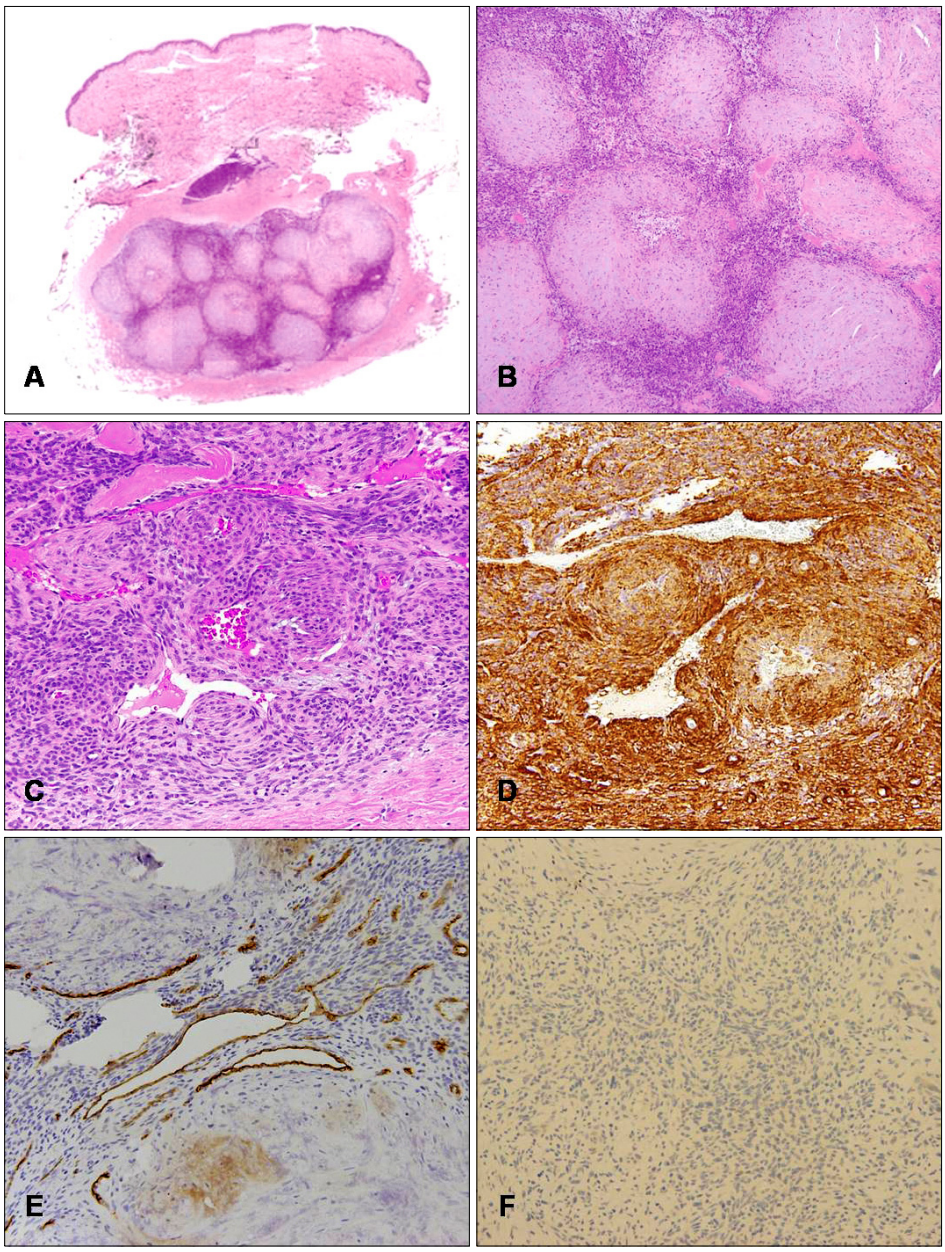

Fig. 2. (A) A well-circumscribed spindle cell tumor with a fibrous capsule in the subcutaneous tissue (scanning view). (B) The tumor is composed of abundant myxoid stroma and blood vessels (H&E, ×40). (C) The concentric perivascular arrangement of lesional spindle-shaped cells with around blood vessel walls (H&E, ×100). (D) Smooth muscle actin immunoreactivity is observed in concentric perivascular myoid cells (SMA, ×200). (E) Negative CD34 immunostaining of the perivascular myoid cells (CD34, ×200). (F) The inner tumor cells were negative for desmin (Desmin, ×200).

performed excisional biopsy.

Microscopically, the tumor was composed of a myxoid matrix and spindle-shaped myoid-appearing cells with a concentric arrangement of cells around numerous blood vessel walls (Fig. 2A∼C). In the intervascular area, the lesional spindle cells were present as sheets of cells. Mitotic figures were inconspicuous in the tumor. Nuclear atypia and necrosis were not observed. The lesional spindle cells were diffusely positive for smooth muscle actin (SMA) (Fig. 2D). In contrast, the tumor cells were negative for CD34, desmin and S100 protein (Fig. 2E, F). From these findings, we diagnosed this lesion as a myopericytoma.

DISCUSSION

MP is a recently described benign tumor composed of oval to spindle-shaped myoid-appearing cells that undergo concentric perivascular growth[2,3]. The histologic spectrum of MP is very wide and varies from lesions that are very similar to myofibromatosis to tumors that closely resemble glomus tumors and angiomyoma[7]. MP is composed of a mixture of solid cellular areas intermixed with variable numbers of vascular channels[1,2,7-9]. The cells in the solid areas are oval to spindle-shaped with eosinophilic cytoplasm and vesicular nuclei[1,2,9]. The vascular channels are often elongated and display prominent branching resulting

in a hemangiopericytoma like appearance[1,2,7,9]. A common and striking feature is the presence of concentric layers of tumor cells around vascular channels resulting in a typical onion ring appearance[2,7]. Small foci were reminiscent of myofibroma, being composed of spindle cells with abundant eosinophilic cytoplasm embedded in a myxoid matrix organized in fasicles or whorls, and these areas bulged and could be invaginated into the lumina of intralesional blood vessels[2]. The lesional spindle cells in MP were diffusely positive for SMA and negative for CD34. In addition, the tumor cells remained unlabelled for desmin, suggesting a less-differentiated smooth muscle phenotype. They were also negative for cytokeratin and S100 protein, usually positive in nodular hidradenomas[2,9].

The novel concept of the existence of myopericytes was originally proposed by Dictor et al.[10] in a report describing a tumor that involved the thyroid gland of a 5-year-old boy. This tumor showed histologic features reminiscent of myofibromatosis and hemangiopericytoma. Based on immunohistochemical analysis and electron microscopy, Dictor et al.[10] proposed that the lesional cells included a population of cells that they termed 'myopericytes'. The authors also suggested that myopericytes were the constituent cells in infantile myofibromatosis. The term myopericytoma was adopted by Granter et al.[3] in 1998 to describe a tumor that was closely related to myofibroma, with a distinctive perivascular arrangement of lesional oval to spindle cells in a concentric multilayered pattern. MP has also been proposed as the term to encompass the entities myofibromatosis, adult myofibroma, glomangiopericytoma and infantile haemangiopericyoma[2,3]. These tumors often show overlapping histologic features and are believed to be part of a spectrum of lesions that show apparent differentiation towards myopericytes[2].

The most common anatomic setting for this tumor is the skin and superficial soft tissues of adult patients. The distal extremities are frequently involved, but with increased recognition, a wider distribution has been described[1,9]. In a comprehensive study (54 cases), Mentzel et al.[9] found that the lower extremities were most commonly affected, followed by the upper extremities, the head and neck region, and the trunk. The most common presentation is a well-circumscribed and slow-growing painless nodule, although occasional cases are painful[2,9]. In our case, the patient had a painless, slowly growing nodule in the subcutaneous tissue of the lower leg.

Most cases of MP are benign lesions, although a few recurring and/or malignant cases have been described[7,11-13]. It seems that the clinical outcome of rare malignant myopericytoma is strongly associated with the depth of the neoplasm. However, more cases with expanded follow-up have to be studied to substantiate this hypothesis[12].

The differential diagnosis include glomus tumors, angioleiomyomas and nodular hidradenomas[2,7,9,13]. Perivascular arrangement of cells can be seen in glomus tumors. However, a concentric arrangement of cells that accentuates blood vessel walls is characteristic of MP, it is not seen in glomus tumors[2]. In addition, areas with spindle cells and abundant eosinophilic cytoplasm that mimic myofibroma are not seen in glomus tumors[2].

Angioleiomyomas are tumors composed of mature smooth muscle cell bundles with abundant vascular channels[13]. These tumors are often painful and generally have a more fascicular pattern than myopericytomas[2,14]. They are composed of fascicles of smooth muscle cells with cigar-shaped nuclei and abundant brightly eosinophilic cytoplasmthat stain diffusely SMA and they frequently (80～90% of cases) show desmin immunoreactivity in the smooth muscle bundles[2,13,14]. In contrast, MP has been only rarely reported to be focally immunoreactive for desmin (9% of cases), suggesting MP is composed of immature cells than angioleiomyomas[2,13]. In some nodular hidradenomas, lesional epithelial cells can show a striking concentric arrangement of cells around small ducts. However, close inspection confirms that the spaces are small ducts and not blood vessels and, if any doubt persists, a cytokeratin stain should confirm the epithelial nature of the cells[2,15].

MP is a rare benign tumor characterized by spindle-shaped myoid-appearing cell with a concentric arrangement in distended vessels. The differential diagnosis of lesions includes a number of tumors that can have a perivascular arrangement such as glomus tumors and angioleiomyomas. MP is a recently delineated benign neoplasm, so we believe this case will aid other physicians in recognizing this unique entity and improve the understanding of MP.

REFERENCES

1. Ide F, Obara K, Yamada H, Mishima K, Saito I. Intravascular myopericytoma of the oral mucosa: a rare histologic variant in an uncommon location. Virchows Arch 2007;450:475-477.
2. McMenamin ME, Calonje E. Intravascular myopericytoma. J Cutan Pathol 2002;29:557-561.
3. 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.
4. Jang KA, Ahn SJ, Choi JH, Lim YS, Sung KJ, Moon KC, et al.

Myxoid myofibromatosis-type perivascular myoma showing prominent verocay body-like formation. Ann Dermatol 2000;12:295-298.
5. Choi YS, You CE, Park MY, Park HJ. A case of intravascular myopericytoma [abstract]. Korean J Dermatol 2006;44(Suppl. 1):203.
6. Kim HY, Lim HJ, Park BC, Lee SJ, Kim BS, Lee WJ, et al. Myopericytoma-type perivascular myoma arising on the auricle of the ear. Korean J Dermatol 2009;47:1304-1308.
7. Calonje E. Vascular tumors: tumors and tumor-like conditions of blood vessels and lymphatics. In: Elder DE, Elenitsas R, Johnson BL Jr, Murphy GF, Xu X, editors. Lever's histopathology of the skin. 10th ed. Philadelphia: Lippincott-Raven, 2008:1049-1051.
8. Woollard AC, Southgate C, Blair JW. Intravascular myopericytoma of the superficial palmar arch. J Hand Surg Eur Vol 2007;32:475-476.
9. Mentzel T, Dei Tos AP, Sapi Z, Kutzner H. Myopericytoma of skin and soft tissues: clinicopathologic and immunohistochemical study of 54 cases. Am J Surg Pathol 2006; 30:104-113.
10. Dictor M, Elner A, Andersson T, Fernö M. Myofibromatosis-like hemangiopericytoma metastasizing as differentiated vascular smooth-muscle and myosarcoma. Myopericytes as a subset of "myofibroblasts". Am J Surg Pathol 1992;16:1239-1247.
11. Sapelli S, Ribas M, Martins WD, de Noronha L, Gomes AP. Myopericytoma of the lip: report of case. Head Neck 2009; 31:561-564.
12. McMenamin ME, Fletcher CD. Malignant myopericytoma: expanding the spectrum of tumours with myopericytic differentiation. Histopathology 2002;41:450-460.
13. Laga AC, Tajirian AL, Islam MN, Bhattacharyya I, Cohen DM, Plamondon CJ, et al. Myopericytoma: report of two cases associated with trauma. J Cutan Pathol 2008;35:866-870.
14. Hasegawa T, Seki K, Yang P, Hirose T, Hizawa K. Mechanism of pain and cytoskeletal properties in angioleiomyomas: an immunohistochemical study. Pathol Int 1994;44: 66-72.
15. Wilson T, Hellquist HB, Ray S, Pickles J. Intranasal myopericytoma. A tumour with perivascular myoid differentiation: the changing nomenclature for haemangiopericytoma. J Laryngol Otol 2007;121:786-789.