A Case of Myopericytoma on the Lower Leg

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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 calcinosi s 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.


-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. 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. 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. Unlike previous reports, this is a typical case arising in a lower extremity, which is the common clinical feature of MP.
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. 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. MP is composed of a mixture of solid cellular areas intermixed with variable numbers of vascular channels. The cells in the solid areas are oval to spindle-shaped with eosinophilic cytoplasm and vesicular nuclei. The vascular channels are often elongated and display prominent branching resulting
in a hemangiopericytoma like appearance. A common and striking feature is the presence of concentric layers of tumor cells around vascular channels resulting in a typical onion ring appearance. Small foci were reminiscent of myofibroma, being composed of spindle cells with abundant eosinophilic cytoplasm embedded in a myxoid matrix organized in fascicles or whorls, and these areas bulged and could be invaginated into the lumina of intralobular blood vessels. 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. The novel concept of the existence of myopericytes was originally proposed by Dictor et al. 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. 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. 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 haemangiopericytoma. These tumors often show overlapping histologic features and are believed to be part of a spectrum of lesions that show apparent differentiation towards myopericytes.

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. A comprehensive study (54 cases), Mentzel et al. 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. 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. 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.

The differential diagnosis include glomus tumors, angioleiomyomas and nodular hidradenomas. 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. In addition, areas with spindle cells and abundant eosinophilic cytoplasm that mimic myofibroma are not seen in glomus tumors.

Angioleiomyomas are tumors composed of mature smooth muscle cell bundles with abundant vascular channels. These tumors are often painful and generally have a more fascicular pattern than myopericytomas. They are composed of fascicles of smooth muscle cells with cigar-shaped nuclei and abundant brightly eosinophilic cytoplasm that stain diffusely SMA and they frequently (80∼90% of cases) show desmin immunoreactivity in the smooth muscle bundles. 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. 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.

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**