

Myopericytoma of Hand: A Case Report with Review of Literature

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ABSTRACT

Myopericytoma is a recently delineated entity demonstrating a hemangiopericytoma-like vascular pattern. This rare mesenchymal neoplasm arises within the subcutaneous tissue of the extremities. We report a case of a 66-year old female who presented with the complaints of swelling in the base of the right thumb. The surgical resection was done and the tissue was submitted for histopathological examination. It showed a classic concentric perivascular proliferation of bland spindle-shaped pericytic cells characteristic of myopericytoma. Despite sharing morphologic features with angioleiomyoma, myofibroma and glomus tumor, myopericytoma is thought to represent a distinct perivascular myoid neoplasm of skin and soft tissues. Though, most of the cases of myopericytoma are benign, local recurrence and malignancy has been reported.

Keywords: Haemangiopericytoma, Myopericytoma, Perivascular myoma.

INTRODUCTION

Myopericytoma (MPC) is a recently proposed subgroup of perivascular tumors in the World Health Organization classification of soft tissue tumors. The group describes tumours that originate from perivascular myoid cells and show a wide range of histological growth patterns. [1,2] The term myopericytoma was first proposed in 1996 by Requena as an alternative designation for solitary myofibroma. It was adopted in 1998 by McMenamin and Fletcher to describe a spectrum of tumours with striking concentric perivascular proliferation of spindle cells (perivascular myoid cells, pericytes). [3]

CASE REPORT

A 66 years old female presented with the complaints of swelling in the base of the right hand since the past four years. The swelling was purplish in color and progressively increasing in size associated with pain and tenderness. On clinical examination it was cystic in consistency. So a clinical diagnosis of hemangioma was given. The swelling was excised fully and sent for histopathological examination. Grossly the tumor was encapsulated and well circumscribed, firm in consistency and measuring 2.4cm x 2cm x 1.7cm. The cut surface was whitish-pink, homogeneous, without areas of hemorrhage within it.

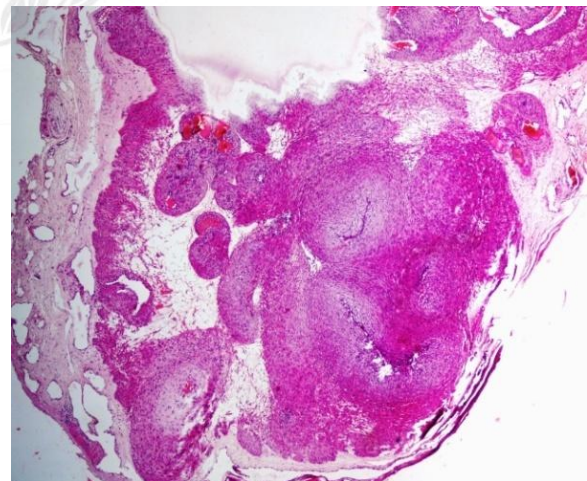


Fig1. A well-circumscribed spindle cell tumor with a fibrous capsule in the subcutaneous tissue (H&E, Scanner view).

Microscopically showed numerous thin walled blood vessels resembling an hemangiopericytoma like pattern. They were characterized by a proliferation of bland ovoid, plump-spindled, and/or round myoid tumor cells with an eosinophilic

cytoplasm and spindled or round nuclei. Characteristically, there was a distinct perivascular and concentric growth of neoplastic cells, which had Catherine wheel features or a typical spinning off from the vessel walls was seen. [Fig.1,2,3]

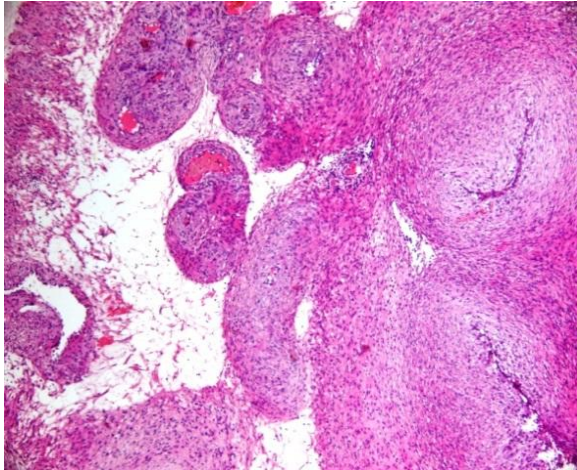


Fig.2 The tumor is composed of abundant myxoid stroma and blood vessels (H&E, 40X).

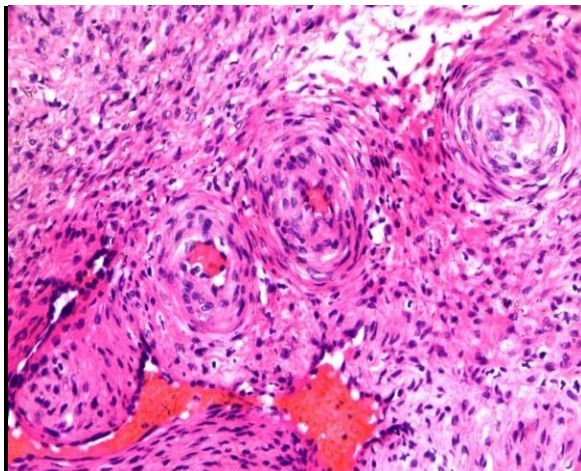


Fig3.The concentric perivascular arrangement of spindle shaped cells around blood vessel walls (H&E, 100X).



Fig 4.Smooth muscle actin immunoreactivity observed in concentric perivascular myoid cells (SMA, 200X).

Tumor stroma was fibrous. Necrosis, cytological atypia, mitoses and lymphovascular invasion was not seen. Immunohistochemically the tumour showed diffuse positivity for SMA (smooth muscle actin and vimentin but negative for S-100 protein and desmin. [Fig.4,5] Hence, the diagnosis of myopericytoma was signed out.

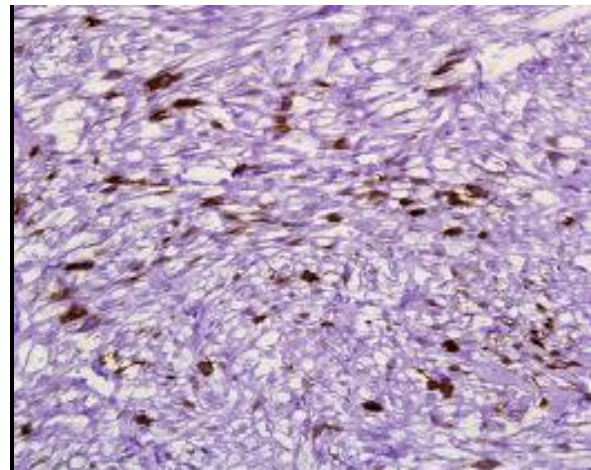


Fig5.The inner tumor cells were negative for desmin (Desmin, 200X).

DISCUSSION

Myopericytoma is a family of benign tumors that exhibit a myoid/pericytic line of differentiation. [4] It encompasses a group of tumours that originate from perivascular myoid cells. Myopericytoma can be multifocal involving a single or multiple anatomic regions, and tends to occur in dermal and superficial soft tissues of adults primarily on the extremities. [1]

Myopericytoma occurs across a wide age range from the second decade onwards, with a male predominance. It occurs in the dermis and superficial soft tissues and is most common in the lower extremity, followed by the upper extremity. Some patients may have multiple tumours. However, it can also arise at other sites including proximal extremities, head, neck and trunk. Occasional cases have also been documented in skeletal muscle, bones (skull, vertebrae, ribs, femur, and tibia), and other visceral organs. [5] Some skin lesions have a purplish hue mimicking a haemangioma, as it is in our case, whereas others present as white nodules or fixed

masses. Larger skin lesions can cause ulceration. Though the MPCs have been reported to be generally painless, our case presented with considerable pain and tenderness. The prognosis is good with marginal excision and local recurrences are uncommon. [1]

In 1942, Stout [6] described a tumour that he termed haemangiopericytoma (HPC), which characteristically occurred on the extremities, and consisted of plump spindle shaped cells arranged around prominent, thin walled, branched, “stag horn” blood vessels. Stout believed that HPC was a morphologically heterogeneous group of tumours, and recognized both myoid differentiation and an overlap with glomus tumours. This vascular pattern is very nonspecific and manifests itself in a variety of other benign and malignant tumors, including benign fibrous histiocytoma, synovial sarcoma, mesenchymal chondrosarcoma, solitary fibrous tumour, leiomyosarcoma, endometrial stromal sarcoma, and infantile fibrosarcoma. Hence, the classification of tumours solely based on their vascular pattern was found to be inappropriate.

The term myopericytoma was first proposed in 1996 by Requena as an alternative designation for solitary myofibroma. It was adopted in 1998 by McMenamin and Fletcher to describe a spectrum of tumours with striking concentric perivascular proliferation of spindle cells (perivascular myoid cells, pericytes). [3]

The use of immunohistochemistry has provided substantial evidence to classify many of these tumours that display this vascular pattern. However, a small subset still remains, probably representing examples of myopericytoma (MPC). Since the term HPC has become nonspecific, it has been emphasized to use MPC as a new term, [7] and this has now been endorsed by the World Health Organization (WHO). [8] In 1998, Granter and colleagues [7] formally used the term MPC and comprehensively described three morphological appearances

that formed a spectrum of tumours seen in the putative MPC category. These patterns/tumour types were that of myofibromatosis, MPC, and glomangiopericytoma (GPC).

In the recent WHO classification of soft tissue tumours [8] MPC and myofibroma (MF) are listed as separate entities and GPC appears as a subtype of MPC. This spectrum includes infantile type myofibromatosis, solitary myofibroma, benign myopericytoma, malignant myopericytoma, infantile hemangiopericytoma and glomangiopericytoma. Pure glomus cell tumors, glomangiosarcoma and sinonasal type hemanpericytoma are excluded from the spectrum; as the pure glomus cell tumors and the glomangiosarcomas do not show the concentric growth pattern and usually lack spindle cells. It is important to properly label this entity as MPC is frequently misdiagnosed as a sarcoma. [2]

MPC's are usually well circumscribed but unencapsulated tumors. They range in size from 11-30mm. The dermal and subcutaneous nodules of MPC are more circumscribed than deeper nodules, which can be infiltrative. [2] They are rubbery or firm and scar like in consistency, typically with a white grey or pink cut surface. There are numerous thin walled blood vessels resembling hemangiopericytoma like pattern. They are characterized by a proliferation of bland round to ovoid cells with abundant cytoplasm and arranged in a concentric perivascular pattern. These cells bear some resemblance to glomus cells, but are distinguished from them by their larger size, more abundant cytoplasm and the lack of a clearly demarcated cell border. An associated spindle cell component, resembling smooth muscle differentiation (although with less cytoplasmic eosinophilia or elongated nuclei) can also be occasionally be present. [2] Necrosis and cytological atypia are lacking and mitoses are infrequent.

In some cases, the morphological pattern resembles angioleiomyoma or be

indistinguishable from glomangioma. The less cellular areas with a prominent collagenous stroma are like fibromas. The MPCs have a spectrum of growth pattern that show some overlap with those of the myofibromas. But the myofibromas are characterized by the presence of a zonation or biphasic appearance that is quite contrary to the concentric perivascular arrangement of the plump spindle to glomoid cells in MPC. [2]

Candidates for the progenitor cell of origin for the myopericyte (and by implication MPC) include the myofibroblast or the pericyte, both of which exhibit properties of modified smooth muscle cells rather than endothelial cells. The term "myofibroblast" is applied to a spindle shaped cell with elongated nucleus and pale eosinophilic cytoplasm that usually shows a desmin negative, actin positive immunohistochemical phenotype. The pericyte is defined both by its orientation to blood vessel walls and its immunohistochemical or ultra-structural characteristics. It is a contractile arborizing cell arranged ubiquitously along capillaries and venules, with multiple extensions of the cells encircling the vasculature. It is viewed as a pluripotential resting stem cell, capable of differentiating along smooth muscle, pericyte, glomus cell, osseous, fibroblast, and adipocyte cell lines. [6,8,9] Indeed, differentiation of pericytes into myofibroblasts and smooth muscle cells has been documented. This concept accounts for the spectrum of tumours and may explain the distinctive features of each variety. [7-11]

CONCLUSION

The classification and terminology of tumors showing a so called hemangiopericytoma-like vascular pattern has been confusing and continues to evolve. The move away from the traditional HPC terminology, and the designation of myopericytoma as a distinct entity, as endorsed by the WHO [7] are useful steps forward in this evolution. Our case report supports the view that MPC is a benign,

usually subcutaneous tumor, with a predilection for the distal extremities.

Conflict of interest: Authors declare that there are no any conflicts of interest.

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