Myopericytoma: A Rare Soft Tissue Neoplasm

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Myopericytomas are rare, slow growing benign soft tissue neoplasm that arise from perivascular smooth muscles within the superficial subcutaneous soft tissue, and shares features of both glomus and smooth muscles. Myopericytomas depict one of the many related perivascular tumors of myoid lineage, with similar morphology and combined immunohistochemical profile, including positive staining for smooth muscle actin. We report a case of a fourty-seven-year-old man with a slow growing nodule on the antero-medial aspect of the left knee. An excision biopsy was performed, which confirmed a myopericytoma with no evidence of malignancy

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Perivascular tumors are soft tissue tumor that begin in the cells that wrap around blood vessels, and can be benign or malignant. This group traditionally has included glomus tumor and hemangiopericytoma. “Myopericytoma” is a recently delineated entity showing a hemangiopericytoma like vascular pattern i.e. the presence of numerous thin-walled branching blood vessels and pericellular reticulin fibers.[1]

Case Report

A fourty-seven-year-old man presented with a mass over the anterior medial aspect of left knee which had been growing slowly

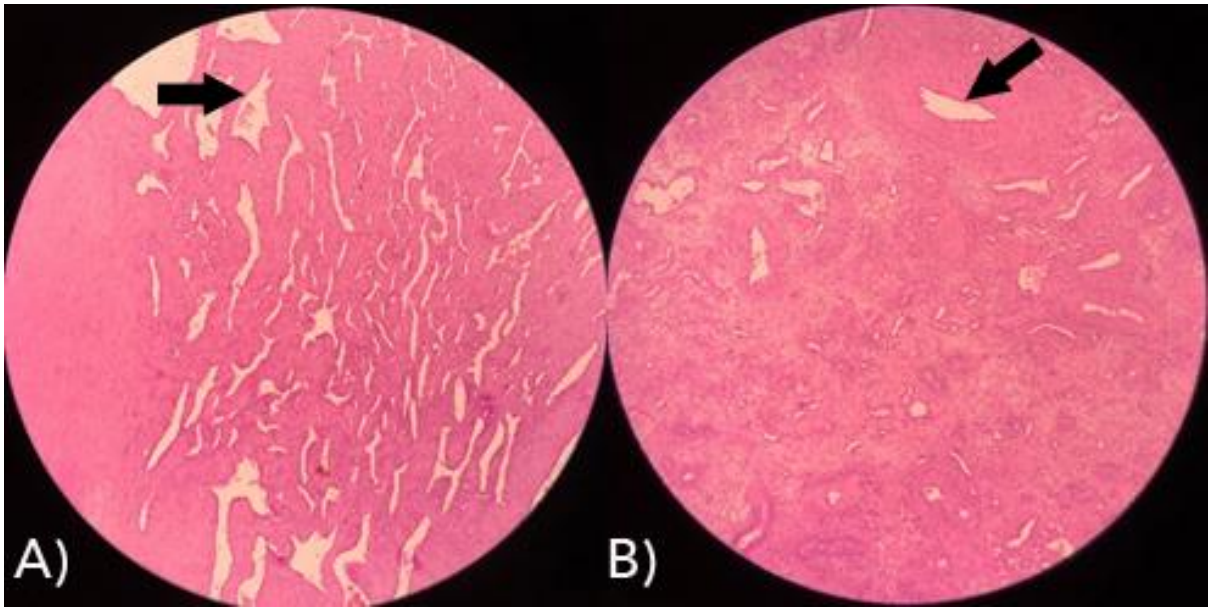

Figure 1: Photomicrograph of myopericytoma showing in blue arrow hemangiopericytoma like vasculature (A) and black arrow showing perivascular growth of myoid tumor cells, a characteristic myopericytoma morphology (B). [H&E stain; 100X]

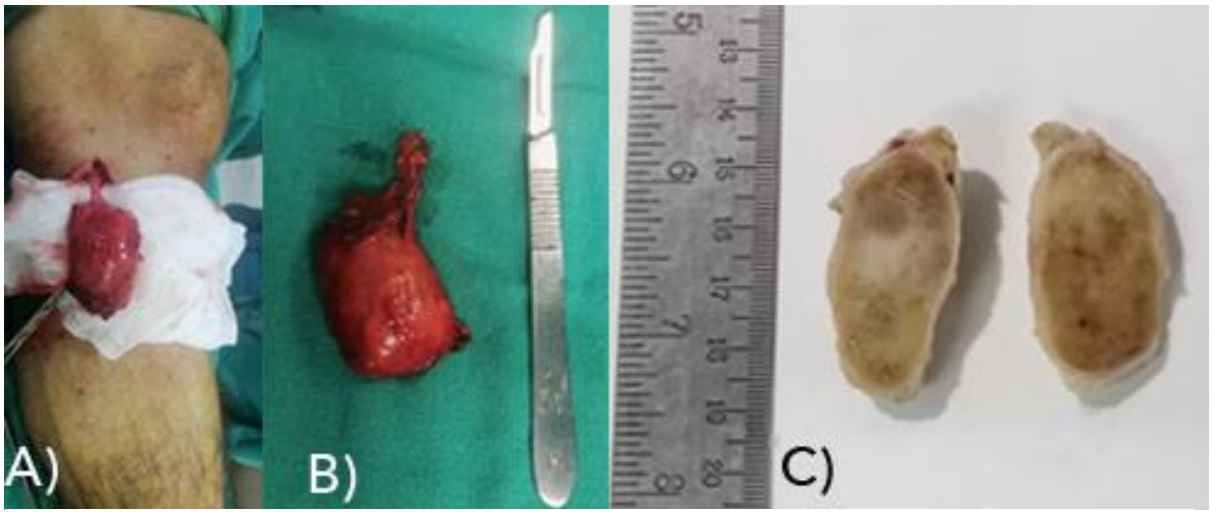

Figure 2: A) & B) Intraoperative picture showing an encapsulated mobile solid soft tissue mass with a vascular pedicle. C) Gross picture showing a circumscribed tumor with a solid, gray-white cut surface with interspersed brownish areas.

for few years, and was causing pain on movement of the knee. Physical examination revealed a 5X4 cm nodular and mobile mass, with minimal tenderness to palpation and normal overlying skin. Ultrasonography reported a well-defined hyper-vascular solid soft tissue mass with a differential diagnosis of a lipoma or a hemangioma.

A complete surgical excision was done with clear margins, and the specimen sent for histopathologic examination. Macroscopic

examination showed a 5X3 cm solid, gray-white colored capsulated nodular tissue, with a solid, gray-white cut surface with interspersed brownish areas. Histology showed a circumscribed lesion comprising of numerous thin walled variably sized, branching hemangiopericytoma-like vasculature surrounded by cytologically bland oval to spindle shaped cells with multilayered concentric growth, and a loose and edematous stroma. The lesion was surrounded by a thick pseudo-capsule with areas of hemorrhage, brownish pigment deposits and myxoid change. No evidence of malignancy was seen.

Discussion

Stout and Murray's description of hemangiopericytoma suggested that many vascular benign and malignant tumor have a similar pattern. [2] Requena et al proposed the term 'myopericytoma' for a solitary myofibroma[3] in 1996, which Granter et al adopted in 1998, and grouped myopericytoma, myofibromatosis , solitary myofibroma, and infantile hemangiopericytoma, to form a single morphological spectrum of tumors showing perivascular myoid cell/pericytes.[4] This spectrum was further expanded by McMenamin and Fletcher in 2002 with a report of 5 malignant myopericytomas that exhibited aggressive biological behavior.[5]

Myopericytoma describes a benign, usually subcutaneous tumor with, myoid-appearing oval to spindle-shaped cells with a striking tendency for concentric perivascular growth.[6] The World Health Organization officially named this tumor type as "myopericytoma" in 2002, and classified it into the group of peripheral blood cell/vascular cell tumors.[7] Myopericytomas can be multifocal involving single or multiple anatomic region,[8] and tends to occur predominantly in the skin and superficial soft tissue of the distal extremities (hand, foot, ankle, and leg), followed by the head and neck region, and the trunk. Clinically myopericytomas are often solitary and well demarcated slow growing soft tissue mass for several years. Multiple small clustered nodules termed 'myopericytomatosis' are less common. Prognosis is usually excellent with complete surgical resection. Indications for surgery include cosmesis, pain (with or without movement near a joint, as in our case), or suspected malignant transformation.[7]

Both myopericytoma and myofibroma are frequently misdiagnosed as other tumors, frequently sarcomas.[9] Most myopericytomas are benign lesions, though a few malignant cases have also been described.[4] Although malignant transformation of these usually benign lesions is rare, recurrence has been reported in 10%–20% of patients.[2] There is no role

for radiotherapy or chemotherapy. Malignant myopericytomas are rare, and typically deep seated, infiltrative tumors with nuclear pleomorphism, brisk mitotic activity, and an aggressive clinical course with or without distant metastasis disease.[5,10] Ultrasonography shows a well-defined hyper-vascular solid soft tissue mass, whereas MRI shows T1 hyperintense signal representing internal hemorrhage or fat within the lesion, and T2 hyperintense signal representing soft tissue components, thick peripheral tissue, and a varying amount of peritumoral edema that enhances avidly with gadolinium.[11]

Conclusion

Myopericytomas are uncommon benign vascular tumors that can sometimes be mistaken for a sarcoma. Malignant transformation is rare. Ultrasonography shows a well-defined vascular lesion, and surgical excision confirms the diagnosis, as well as cures the symptoms, as in our case.

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