

Case Report

International Journal of Health Policy Planning

Myopericytoma: A Rare Perivascular Neoplasm – Case Report

Devi Shanker Malik^{1*} , Mahipal Singh², B S Dhakad³ and Srujana Kushtagi⁴

¹Director, General and Laparoscopic Surgery, Eternal Hospital, India

²Director and HOD Radiology, Eternal Hospital, India

³Attending consultant, General and Laparoscopic Surgery, Eternal Hospital, India

⁴Senior Resident, General and Laparoscopic Surgery, Eternal Hospital, India

*Corresponding Author

Devi Shanker Malik, Director, General and Laparoscopic Surgery, Eternal Hospital, India.

Submitted: 2026, Sep 02; Accepted: 2026, Sep 24; Published: 2026, Sep 30

Citation: Malik, D. S., Singh, M., Dhakad, B. S., Kushtagi, S. (2026). Myopericytoma: A Rare Perivascular Neoplasm – Case Report. *Int J Health Policy Plann*, 5(2), 01-05.

Abstract

Introduction: Myopericytoma is a rare, typically benign perivascular tumour originating from myopericytes. While it shares morphologic features with other myoid neoplasms, its distinct concentric growth pattern and specific immunohistochemical profile are diagnostic hallmarks. We present a case of myopericytoma to highlight its clinical presentation and the diagnostic rigor required to differentiate it from its mimics.

Case Presentation: A 69-year-old male presented with a slow-growing, painless subcutaneous mass in the distal part of left leg for last 5-6 months. Clinical findings were non-specific, so, initially diagnosed as a soft tissue tumour. His ultrasound demonstrated a well-defined heterogeneously hyperechoic lesion with mild internal vascularity seen in subcutaneous planes, superficial to Tendo-Achilles (closely abutting it) at posterior aspect of distal leg. A benign lesion?

Surgical excision of this soft tissue tumour was done under regional anaesthesia.

Histopathology reveals a circumscribed lesion composed of interlacing bundles of smooth muscle cells with elongated, blunt ended nuclei and eosinophilic cytoplasm. Numerous thick-walled vascular channels are interspersed within the smooth muscle bundles. Endothelial lining of vessels is intact. Suggestive of Spindel cell neoplasm.? Vascular Leiomyoma (Angioleiomyoma). Immunohistochemical analysis showed diffuse positivity for Smooth Muscle Actin (SMA) and h-Caldesmon, while Desmin, CD34, and S100 were negative, confirming the diagnosis of myopericytoma.

Discussion: Myopericytoma exists on a morphological continuum with myofibroma and angioleiomyoma. The absence of desmin staining in this case was pivotal in excluding angioleiomyoma, while the lack of a biphasic growth pattern ruled out myofibroma. Although usually benign, recognizing the myopericytic phenotype is essential for excluding malignant variants and ensuring appropriate surgical management.

Conclusion: This case underscores the importance of considering myopericytoma in the differential diagnosis of subcutaneous spindle cell tumours. A combination of characteristic "onion skin" morphology and a targeted IHC panel is essential for an accurate diagnosis and a favourable clinical outcome.

Keywords: Myopericytoma, Perivascular Tumours, Smooth Muscle Actin, Myopericyte, Spindle Cell Lesion, Glomangiopericytoma

1. Introduction

Myopericytoma is a rare, perivascular soft tissue tumor. And was first described by Granter et al., in 1998 [1]. These tumours arise from the specialized smooth muscle cells surrounding blood vessels, known as myopericytes. These tumours typically appear as slow-growing, painless nodules, commonly found in the soft tissues of distal extremities like hands, feet, or legs [2]. Unusual Site reported are the oral cavity, internal organs (colon, heart), or intracranial sites. They are usually harmless and are part of a spectrum of "perivascular" tumours that doctors look at closely to distinguish them from other similar growths like myofibroma, glomus tumours, Glomangiopericytoma and angioleiomyoma [3]. Cutaneous myopericytoma' usually presents as a solitary lesion, although multiple lesions can occur [4,5].

Histopathologically, myopericytoma is characterized by a well-circumscribed, unencapsulated nodular proliferation with numerous thin-walled vessels and a concentric, perivascular arrangement of ovoid spindle-shaped myopericytes in the dermis, subcutis or soft tissues. Mitotic activity and cytologic atypia are usually minimal. However, a rare atypical/malignant variant histologically characterized by high cellularity, significant mitotic activity, pleomorphism, necrosis, and metastasis has been reported [6]. Immunohistochemically, all cases express α -smooth muscle actin (SMA) and most also express h-caldesmon (90%).

2. Case Report

• Clinical History and Examination

A 69-year-old male presented with a 5–6-month history of a painless, slow-growing nodular swelling located on the distal part of left leg posteriorly just above the ankle joint. The patient denied any history of trauma or systemic symptoms. Physical examination revealed a firm, mobile, about 5x5 cm size subcutaneous nodular swelling. It was mildly tender on pressing. The overlying skin was unremarkable. Neurovascular status was intact distal to the lesion.

• Imaging Findings

Ultrasound: Demonstrated a well-defined heterogeneously hyperechoic lesion measuring approximately 3.0x1.3x4.2 cm (APxTRxCC) with mild internal vascularity seen in subcutaneous planes, superficial to Tendo-Achilles (closely abutting it) at posterior aspect of distal leg (Approximately. 4-5 cm superior to ankle joint). Tiny echogenic/calcific focus seen at dependent caudal aspect represents a benign lesion?

• Surgical Procedure

The patient underwent an excision of soft tissue tumour under regional anaesthesia. Intraoperatively, the lesion was found to be a well-demarcated tan-white mass measuring about 5x5 cm, with a firm consistency. There was no apparent invasion into the surrounding fascia or neurovascular bundles.

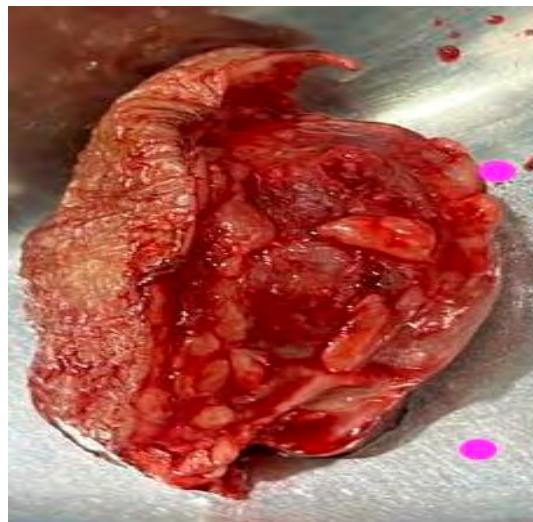


Figure 1: Operated Specimen

• Histopathological Analysis

Gross examination revealed a skin covered grey- white to grey- brown soft tissue nodule measuring 5.2x5.2 cm. Cut surface is solid, firm, and shows whorled appearance with focal vascular channels.

• Microscopic Findings (H&E)

Sections reveal a circumscribed lesion composed of interlacing

bundles of smooth muscle cells with elongated, blunt ended nuclei and eosinophilic cytoplasm. Numerous thick-walled vascular channels are interspersed within the smooth muscle bundles. Endothelial lining of vessels is intact. No atypia, necrosis or significant mitotic activity is seen. Surrounding tissue shows compressed stroma. Suggestive of Spindel cell neoplasm.? Vascular Leiomyoma, Angioleiomyoma

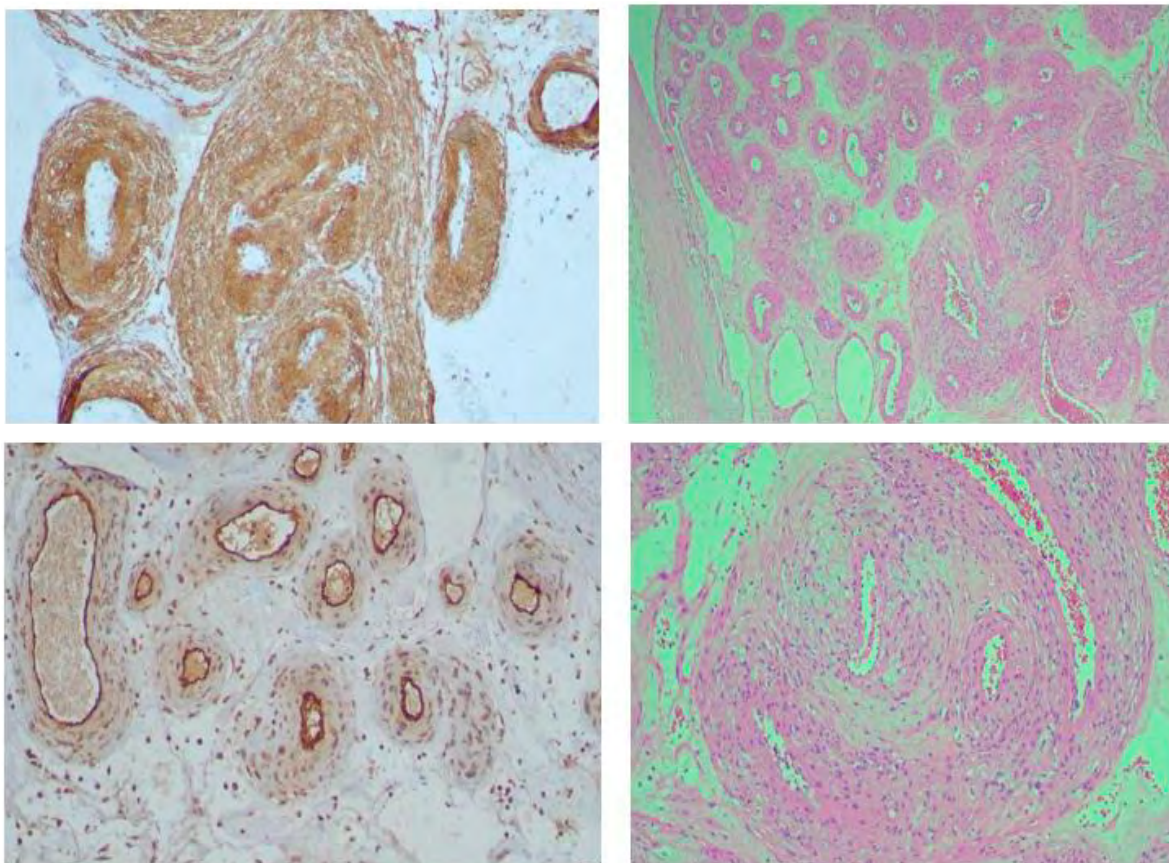


Figure 2: Microscopic Picture

- **Immunohistochemistry**

To confirm the diagnosis, IHC was performed. The tumour cells show diffuse cytoplasmic positivity for smooth muscle actin (SMA) and h-caldesmon. Desmin is negative. CD34 is negative, solitary fibrous tumour. Endothelial markers CD31 and ERG are negative, ruling out vascular endothelial origin. S100 is negative, excluding neural tumours. The ki-67 proliferation index is low, less than 5%, consistent with benign nature. Suggestive of Myopericytoma ((Benign perivascular myoid neoplasm). No evidence of malignancy.

- **Outcome and Follow-Up**

The final diagnosis was confirmed after IHC as Benign Myopericytoma. The postoperative course was uneventful. At the 3-month follow-up, the patient showed no clinical evidence of local recurrence or functional deficit

3. Discussion

The diagnosis of myopericytoma remains a challenge for clinicians and pathologists alike due to its rarity and its overlapping features with other perivascular myoid tumours. Our case highlights the classic presentation of MPC yet underscores the importance of a meticulous immunohistochemical (IHC) workup to distinguish it from more aggressive spindle cell neoplasms.

- **Cell of Origin:** These tumours arise from the specialized

smooth muscle cells surrounding blood vessels, known as myopericytes. The myopericyte is a transitional cell form between a glomus cell (pericyte) and a true vascular smooth muscle cell. The term 'myopericyte', coined by Dictor et al., was used to denote 'atypical pericytes surrounded by bundles of sclerotic smooth muscle abutting on staghorn vessels' that formed the predominant cell type in an unusual case of metastasizing 'myofibromatosis-like hemangiopericytoma' in a young boy [7].

- **Pathogenesis:** While most are sporadic, recent genomic studies have identified recurrent PDGFRB mutations (specifically in the myofibroma-myopericytoma spectrum), suggesting a potential therapeutic target in rare aggressive cases.

3.1. Histopathological Gold Standards

Diagnosis relies on identifying the "triad" of features that distinguish MPC from other spindle cell lesions:

- **Architecture:** Well-circumscribed but unencapsulated. The hallmark is the concentric (onion-skin) growth of tumour cells around thin-walled, branching blood vessels.
- **Cytomorphology:** Plump, monomorphic, spindle-to-oval cells with pale eosinophilic cytoplasm and bland, vesicular nuclei.
- **Vascular Pattern:** May show "staghorn" or hemangiopericytoma-like (HPC) branching vessels, especially in

the more cellular areas.

3.2. Immunohistochemical (IHC) Profiling
IHC is mandatory for confirming the diagnosis.

Marker	Result	Interpretation
SMA (α -Smooth Muscle Actin)	Diffuse Positive	Confirms myoid differentiation.
h-Caldesmon	Positive	Supports the transitional myopericytic phenotype.
Desmin	Negative (usually)	Key to differentiating from true Angioleiomyoma (which is Desmin+).
CD34	Negative	Differentiates from Solitary Fibrous Tumour (SFT).
S100 / SOX10	Negative	Rules out neural/melanocytic tumours.
STAT6	Negative	Rules out modern SFT (NAB2-STAT6 fusion).

Export to Sheets

3.3. Histomorphological Spectrum and Differential Diagnosis

The defining feature of MPC is the circumvascular growth pattern, where myoid spindle cells wrap concentrically around thin-walled vessels. While this "onion skin" appearance is highly suggestive, it is not pathognomonic. The differential diagnosis of myopericytoma is broad and includes other neoplasms composed of myopericytes such as myofibroma/myofibromatosis, angioleiomyoma, perivascular epithelioid neoplasm, and glomus tumour, as well as entities characterized by a vascular and/or spindled cell proliferation.

- Myofibroma: Which typically exhibits a biphasic appearance with "hemangiopericytoma-like" vascular spaces.
- Angioleiomyoma: Distinguished from MPC by its thicker-walled vessels and diffuse Desmin positivity, which was negative in our patient.
- Glomus Tumor: Which features more cuboidal cells with distinct cell membranes
- Dermatofibrosarcoma Protuberans (DFSP): Characterized by a storiform (not concentric) pattern and CD34 diffuse positivity

3.4. Molecular Insights

Recent literature has pointed toward the role of PDGFRB (Platelet derived growth factor receptor Beta) gene mutations in the pathogenesis of the myofibroma-myopericytoma spectrum. Specifically, mutations such as have been identified in both familial and sporadic cases. While molecular testing is not always required for diagnosis, it provides a fascinating window into the "driver" mutations of these pericytic growths and may explain the occasional tendency for local recurrence if the lesion is incompletely excised.

3.5. Clinical Management and Prognosis

The vast majority of MPCs follow a benign clinical course, as seen in this case. Complete surgical excision with clear margins is the treatment of choice. However, the literature describes a rare malignant variant (Malignant Myopericytoma) also, characterized by deep-seated locations, infiltrative borders, and high mitotic activity (per 10 HPF).

4. Conclusion

Myopericytoma is a distinct but often under-recognized perivascular tumour. This case illustrates that while the clinical presentation may mimic more common subcutaneous lesions (like lipomas or cysts), the diagnosis relies heavily on the characteristic concentric growth pattern on histology. We emphasize the necessity of SMA-positive and Desmin-negative IHC profiling to confirm the myopericytic phenotype. Surgeons and pathologists should maintain a high index of suspicion for this entity to ensure appropriate surgical management and to avoid the diagnostic pitfall of misidentifying it as a more aggressive malignancy.

Informed Consent

Ensure you mention "Written informed consent was obtained from the patient for the publication of this case report and accompanying images."

References

1. Granter, S. R., Badizadegan, K., & Fletcher, C. D. (1998). Myofibromatosis in adults, glomangiopericytoma, and myopericytoma: a spectrum of tumors showing perivascular myoid differentiation. *American Journal of Surgical Pathology*, 22(5), 513-525.
2. Zidane, A., Arsalane, A., Harkat, A., Traibi, A., Oukabli, M., Atoini, F., ... & Kabiri, E. H. (2010). Myopericytoma: an uncommon chest wall tumor. *Revue des Maladies Respiratoires*, 27(9), 1089-1091.
3. Dray, M. S., McCarthy, S. W., Palmer, A. A., Bonar, S. F., Stalley, P. D., Marjoniemi, V., ... & Scolyer, R. A. (2006). Myopericytoma: a unifying term for a spectrum of tumours that show overlapping features with myofibroma. A review of 14 cases. *Journal of clinical pathology*, 59(1), 67-73.
4. Jung, Y. I., Chung, Y. K., & Chung, S. (2012). Multiple myopericytoma of the face and parotid gland. *Archives of Plastic Surgery*, 39(02), 158-161.
5. Laga, A. C., Tajirian, A. L., Islam, M. N., Bhattacharyya, I., Cohen, D. M., Plamondon, C. J., & Robinson-Bostom, L. (2008). Myopericytoma: report of two cases associated with trauma. *Journal of cutaneous pathology*, 35(9), 866-870.
6. McMenamin, M. E., & Fletcher, C. D. M. (2002). Malignant myopericytoma: expanding the spectrum of tumours with

-
- myopericytic differentiation. *Histopathology*, 41(5), 450-460.
7. Dictor, M., Elnér, Å., Andersson, T., & Fernö, M. (1992). Myofibromatosis-like hemangiopericytoma metastasizing as differentiated vascular smooth-muscle and myosarcoma: myopericytes as a subset of “myofibroblasts”. *American Journal of Surgical Pathology*, 16(12), 1239-1247.

Copyright: ©2026 Devi Shanker Malik, et al. This is an open-access article distributed under the terms of the Creative Commons Attribution License, which permits unrestricted use, distribution, and reproduction in any medium, provided the original author and source are credited.