

A Case Series of Myopericytoma: A Soft Tissue Neoplasm

NUPUR KAUSHIK¹, SHIKHA PRAKASH², GARIMA DUNDY³, JUHI SINGHAL⁴

ABSTRACT

Myopericytoma (MPC) is a rare benign mesenchymal neoplasm with perivascular myoid differentiation that most commonly arises in middle adulthood. The lesion generally involves the subcutaneous soft tissue of the lower extremities. Histologically, it is characterised by concentric perivascular proliferation of spindled myoid cells with bland elongated nuclei associated with blood vessels. The present case series emphasises five cases of MPC in relatively younger-aged female patients. Fine Needle Aspiration Cytology (FNAC) was inconclusive in all five patients. Histopathological examination showed perivascular concentric proliferation of myoid tumour cells, which were ovoid, spindled, or round cells with abundant eosinophilic cytoplasm, and no nuclear atypia, hyperchromasia, or necrosis. Immunohistochemistry (IHC) was positive for Smooth Muscle Actin (SMA). Although MPC can be diagnosed through histopathological examination, IHC is required for confirmation.

Keywords: Eosinophilic cytoplasm, Immunohistochemistry, Spindle myoid cells, Smooth muscle actin

CASE SERIES

INTRODUCTION

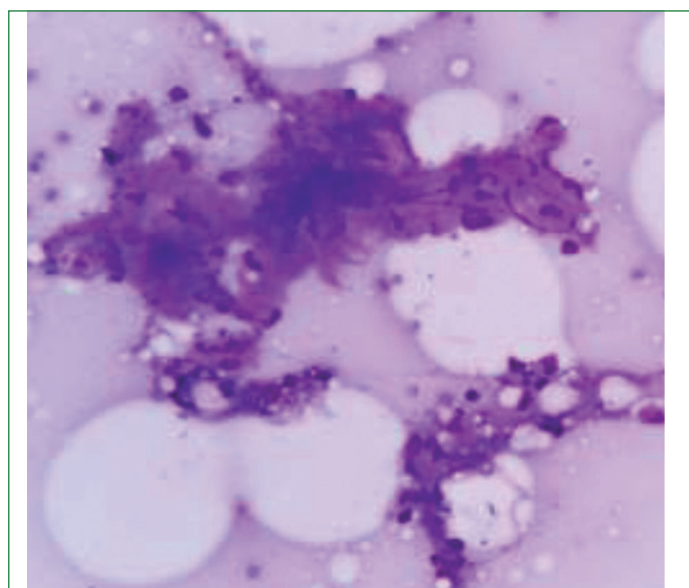
The MPC is a rare, benign soft-tissue tumour [1]. The term was endorsed by the World Health Organisation (WHO) in 2002, and the lesion was described as having myoid-like oval to spindle-shaped cells with a concentric perivascular growth pattern [2]. MPC is approximately twice as common in males compared to females, with a peak age of presentation in the fifth decade [1]. The tumour commonly presents as a slow-growing, well-demarcated, painless, solitary mass in the subcutaneous tissue of the extremities and rarely involves other sites such as the intracranial area or head and neck region [1,2]. The vascular pattern, though not characteristic, can be seen in several other neoplasms such as myofibroma, endometrial stromal sarcoma, infantile haemangiopericytoma, leiomyosarcoma, glomus tumour, and angioleiomyoma [1]. MPC is usually benign with an indolent clinical course, and only a few cases have been reported as malignant. Therefore, complete surgical excision of the affected area is the preferred and curative method [2,3]. Recurrences are rare and are most likely due to poor circumscription of the mass [3,4]. The Platelet-derived Growth Factor Receptor Beta (PDGFRB) gene encodes the protein PDGFRB, which is involved in the pathogenesis of conventional MPC [1].

Case 1

An 18-year-old female presented to the surgery outpatient department with a chief complaint of a mass on the upper medial side of her right leg for three years [Table/Fig-1]. The swelling was gradually increasing, and there was no history of trauma. On physical examination, the swelling measured 3.2×2.5×1.4 cm, was soft to firm in consistency, fixed, non tender, and showed no overlying skin changes. Fine needle aspiration cytology revealed mainly blood cells along with a few occasional bland spindle cells [Table/Fig-2]. Hence, a diagnosis of a benign spindle lesion was made, and a biopsy was advised. The swelling was excised and sent for histopathological examination. Grossly, the lesion appeared as a greyish-white mass measuring 2.4×2.0×1.0 cm [Table/Fig-3]. Histopathological examination showed an encapsulated, well-circumscribed neoplasm comprising proliferating bland ovoid to spindle cells with eosinophilic cytoplasm. The tumour cells were arranged in a concentric form around the blood vessels with no nuclear atypia and necrosis, hence a diagnosis of MPC was made



[Table/Fig-1]: Swelling on upper medial aspect of right leg.



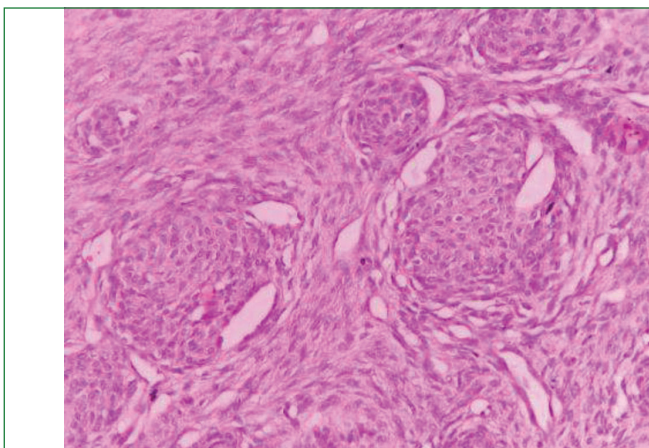
[Table/Fig-2]: Fine needle aspiration cytology smear showing cluster of bland looking spindle cells along with few lymphocyte (May-Grunwald-Giemsa (MGG), 100X).

[Table/Fig-4]. Immunohistochemistry was positive for SMA and negative for desmin [Table/Fig-5], confirming the diagnosis of

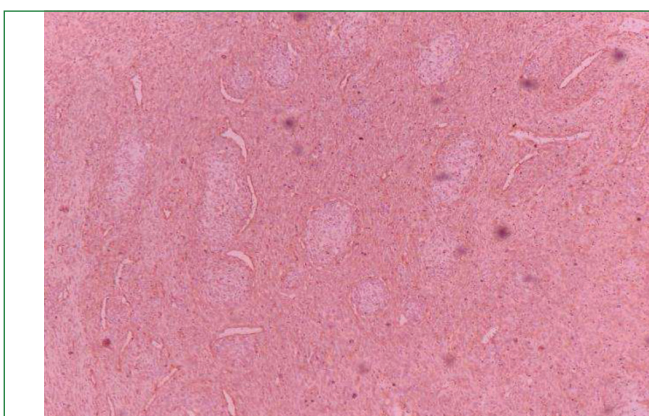


[Table/Fig-3]: Well-encapsulated and homogenous, greyish white mass.

MPC. The patient remained asymptomatic during further follow-up. Similar findings were observed in two other young female aged 17 years and 19 years respectively, who presented with mass on upper medial left leg and right leg respectively for last one year.



[Table/Fig-4]: Photomicrograph showing proliferation of bland round to ovoid cells with eosinophilic cytoplasm and indistinct cell boundaries in concentric fashion around thin-walled vessel {Haematoxylin and Eosin (H&E), 400x}.



[Table/Fig-5]: Diffuse positivity for SMA (100X, IHC).

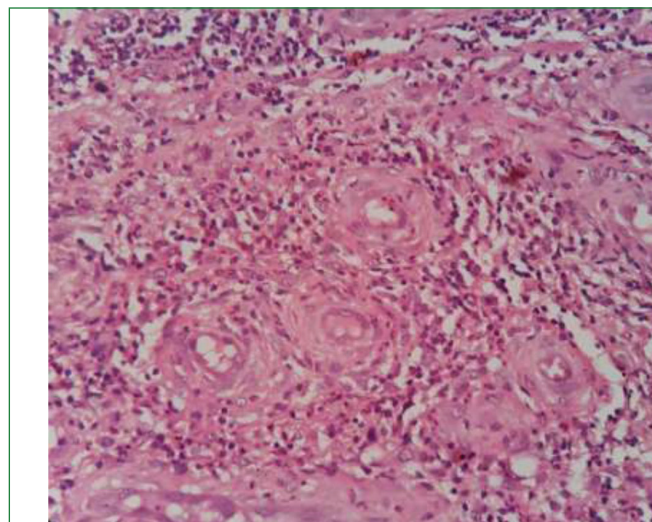
Case 2

A 28-year-old female presented with a chief complaint of a mass on her left ankle for 6 months. There was no history of any associated trauma. On physical examination, the mass measured 1.2×1.2 cm in size, was firm, non mobile, slightly tender on palpation, and the overlying skin showed hypopigmentation. Therefore, a clinical diagnosis of a ganglion cyst was made [Table/Fig-6]. Fine needle aspiration cytology yielded mainly blood cells, and a biopsy was advised. An incisional biopsy from a representative area



[Table/Fig-6]: Swelling and hypopigmentation over left ankle.

was taken and sent for histopathological examination. On gross examination, a small greyish-white tissue measuring 1.2×0.6×0.2 cm was received. Microscopy showed a non encapsulated, well-circumscribed lesion comprising myoid or spindle-shaped cells with eosinophilic cytoplasm arranged in a concentric manner perivascularly. No nuclear atypia or necrosis was observed, and a diagnosis of MPC was made [Table/Fig-7]. Immunohistochemistry was positive for SMA, confirming the diagnosis of MPC [Table/Fig-8]. The postoperative period and follow-up were uneventful.



[Table/Fig-7]: Photomicrograph showing concentric arrangement of spindle cells around thin-walled vessel with abundance of lymphocytic infiltrate (H&E, 100X).



[Table/Fig-8]: Diffuse positivity for SMA (100X, IHC).

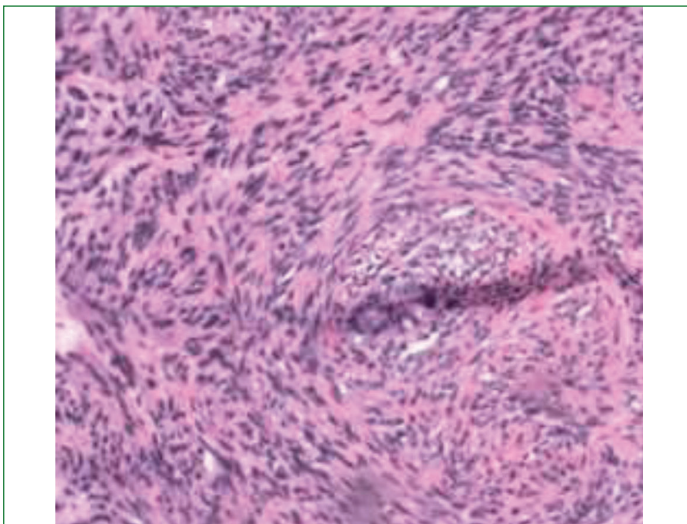
Case 3

An 18-year-old female presented with a swelling on the upper right cervical region for one year [Table/Fig-9]. There was no history of fever or organomegaly. On physical examination, the swelling measured 1.2×1.2×0.8 cm and was soft to firm, non mobile, non tender, with no overlying skin changes. On repeated aspiration, blood was aspirated with occasional bland spindle cells, hence a provisional diagnosis of a vascular lesion was made. The swelling was excised and sent for histopathological examination.



[Table/Fig-9]: Swelling over right upper cervical region.

Grossly, an encapsulated greyish-white mass measuring 1.0×0.8×0.8 cm was received. Microscopically, the lesion was well-circumscribed with tumour cells arranged in a concentric manner around the blood vessel. Individual cells had bland-looking spindle-shaped to ovoid nuclei with eosinophilic cytoplasm. There was no evidence of nuclear atypia, mitotic figures, or necrosis, hence a diagnosis of MPC was made [Table/Fig-10]. Immunohistochemistry was positive for SMA and negative for desmin [Table/Fig-11], confirming the diagnosis of MPC. The follow-up period was uneventful.



[Table/Fig-10]: Photomicrograph showing proliferated bland round to ovoid cells in a concentric fashion around thin-walled vessel (H&E, 100X).

DISCUSSION

The term MPC was first proposed in 1992 to describe an unusual tumour in a young boy that displayed features between pericytes and vascular smooth muscle cells. In 1998, the term MPC was formally used by Granter and colleagues to classify a spectrum of tumours showing characteristic concentric perivascular proliferation of spindle cells [1]. Despite the reported male preponderance in the largest series by Mentzel T et al., the authors hereby report five cases in female patients at a relatively young age [3]. MPC usually presents



[Table/Fig-11]: Diffuse positivity for SMA (100X, IHC).

as a painless, slow-growing, well-demarcated subcutaneous nodule of the distal extremities in adults [4-6], however, lesions in the head and neck have also been described [3].

Myopericytes are considered cells of uncertain position in the morphological spectrum and are considered intermediate between pericytes and vascular smooth muscle. Histomorphologically, MPC is characterised by perivascular concentric proliferation of myoid tumour cells. The cells are ovoid, spindle, or round myoid tumour cells with abundant eosinophilic cytoplasm and no nuclear atypia, hyperchromasia, or necrosis. These tumour cells usually stain positive for SMA and h-caldesmon and non reactive for muscle-specific markers such as desmin [7]. All the cases in the case series showed perivascular growth of uniform, oval to spindle-shaped cells with eosinophilic cytoplasm, with the absence of necrosis and cytologic atypia. These cells were diffusely positive for SMA.

The differential diagnosis of MPC is broad and includes haemangiopericytoma, Perivascular Epithelioid Cell tumour (PEComa), myofibroma/myofibromatosis, and glomus tumour. Haemangiopericytoma is characterised by staghorn-shaped vessels with an intervening proliferation of cells, a feature that is usually not seen in MPC. Haemangiopericytoma typically shows consistent positivity for CD34 and CD99, while immunoreactivity to SMA is rarely seen [1,2]. Myofibroma characteristically shows a distinctive biphasic pattern comprising spindle-shaped cells with pale pink cytoplasm arranged around blood vessels [8,9]. Glomus tumour can be distinguished by its specific features, including the presence of smaller cells with a rounded appearance and an arrangement in an eccentric fashion around the blood vessels, with eosinophilic cytoplasm [8,10]. PEComa can be differentiated by a morphological pattern showing epithelioid cells with eosinophilic granular cytoplasm arranged in a nesting pattern, and immunoreactivity for HMB-45 is seen in all cases [9-11].

Most MPC behaves in a benign manner; hence, surgical resection is the procedure of choice for diagnosis and treatment, and it shows an excellent prognosis after complete resection. Atypical or malignant neoplasms may show local recurrences with incomplete excision and rare metastases [1,12].

CONCLUSION(S)

The MPC is an uncommon, benign soft tissue tumour with a low rate of recurrence. The combined use of histopathological examination and immunohistochemistry is required for a correct diagnosis, and differentiation between the tumour and its mimics is essential to optimise patient outcomes.

REFERENCES

- [1] Alqassab AT, Alsadah FZ, Elsharkawy T, Alhamad M, Alsayed H.. Ankle myopericytoma: A rare case report and cytogenetic study. *Cureus*. 2022;14(1):e21307.
- [2] Valasidis A, Vahtsevanos K, Kyrgidis A, Baliaka A, Kaloutsis V. Myopericytoma of the maxillofacial region: A report of two cases. *Scientific Pages Otolaryngol*. 2017;1(1):20-24.

- [3] 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(1):104-13.
- [4] Amy DPB, Yaffe V, Kavar R, Akrish S, Abu El-Naaj. Oral Myopericytoma; A rare pediatric case report and review of the literature. *BMC Oral Health.* 2021;21:176.
- [5] Peters K, Caracciolo JT, Henderson-Jackson E, Binitie O. Myopericytoma/ myopericytomatosis of the lower extremity in two young patients: A recently designated rare soft tissue neoplasm. *Radiol Case Rep.* 2017;13(1):275-80.
- [6] Agaimy A, Bieg M, Michal M, Gedder H, Märkl B, Seitz J, et al. Recurrent somatic PDGFRB mutations in sporadic infantile/solitary adult myofibromas but not in angioleiomyomas and myopericytomas. *Am J Surg Pathol.* 2017;41(2):195-203.
- [7] Ozkent MS, Gunler T, Gonen M. A retroperitoneal myopericytoma: The first case located at the retroperitoneal area and not originates from the kidneys. *Indian J Pathol Microbiol.* 2023;66(2):369-71.
- [8] Xia L, Chen Y, Geng N, Jiang J, Yang M, Zhang W. Multifocal myopericytoma in the maxillofacial region: A case report. *Oral Surg Oral Med Oral Pathol Oral Radiol Endod.* 2010;109(2):e59-62.
- [9] Lau SK, Klein R, Jiang Z, Weiss LM, Chu PG. Myopericytoma of kidney. *Hum Pathol.* 2010;41(10):1500-4.
- [10] Dray MS, McCarthy SW, Palmer AA, Bonar SF, Stalley PD, Marjoniemi V, et al. Mtopericytoma: A unifying term for a spectrum of tumours that show overlapping features with myofibroma. A review of 14 cases. *J Clin Pathol.* 2006;59(1):67-73.
- [11] Calder KB, Schlauder S, Morgan MB. Malignant perivascular epithelioid cell tumour (PEcoma): A case report and literature review of cutaneous/subcutaneous presentations. *J Cutan Pathol.* 2008;35(5):499-503.
- [12] Mathew NK, Zhang KY, Batstone MD. Myopericytoma of the coronoid process: A case report and review of literature. *Oral Maxillofac Surg Cases.* 2015;56(2):25-28.

PARTICULARS OF CONTRIBUTORS:

1. Senior Resident, Department of Pathology, S. N. Medical College, Agra, Uttar Pradesh, India.
2. Assistant Professor, Department of Pathology, S. N. Medical College, Agra, Uttar Pradesh, India.
3. Professor, Department of Pathology, S. N. Medical College, Agra, Uttar Pradesh, India.
4. Professor, Department of Surgery, S. N. Medical College, Agra, Uttar Pradesh, India.

NAME, ADDRESS, E-MAIL ID OF THE CORRESPONDING AUTHOR:

Dr. Nupur Kaushik,
Senior Resident, Department of Pathology, S. N. Medical College, Agra-282003,
Uttar Pradesh, India.
E-mail: cool.nupurkaushik@rediffmail.com

PLAGIARISM CHECKING METHODS: [Jain H et al.]

- Plagiarism X-checker: Oct 22, 2022
- Manual Googling: Oct 17, 2023
- iThenticate Software: Oct 20, 2023 (13%)

ETYMOLOGY: Author Origin**EMENDATIONS:** 7**AUTHOR DECLARATION:**

- Financial or Other Competing Interests: None
- Was informed consent obtained from the subjects involved in the study? Yes
- For any images presented appropriate consent has been obtained from the subjects. Yes

Date of Submission: **Oct 17, 2022**Date of Peer Review: **Jan 06, 2023**Date of Acceptance: **Oct 21, 2023**Date of Publishing: **Jan 01, 2024**