



## Review Article

# Myopericytoma -A Perivascular Myoid Cell Tumor with Review of Literature

Authors

**Kalpna Mangal<sup>1\*</sup>, Bhawna Bansal<sup>2</sup>, Alka Mittal<sup>3</sup>**

<sup>1\*</sup>Assistant Professor, <sup>2</sup>PG Resident (2<sup>nd</sup> year), <sup>3</sup>Sr. Demonstrator

Department of Pathology, SMS Medical College & Hospital, Jaipur (Rajasthan), India 302004

Corresponding Author

**Dr Kalpna Mangal**

204, Sidha Apartment, Devi Marg, Bani Park, Jaipur, Rajasthan, India, 302016

Tel No.: 9887056209, Email: [dr.kalpna867@gmail.com](mailto:dr.kalpna867@gmail.com)

## ABSTRACT

*Myopericytomas are rare perivascular neoplasms, often mistaken as solitary myofibroma or other PMC (perivascular myoid cell)/PEC (perivascular epitheloid cell) tumors, due to their histomorphological overlapping. Their most common site is superficial soft tissues of extremities particularly distal portions of lower limb followed by head and neck in adults. Men are more commonly affected than women. The recent concept of phenotypic recognition as PMC and PEC phenotype has changed the concept of angiogenic tumors. Here we are reporting a case of sub cutaneous myopericytoma of sternal region in a 21yr male with complete review of literature. The patient had noticed a well defined nodularity of 1.5 x 1.2cm in midsternal region. Total excision was performed. On gross examination it was unencapsulated, well circumscribed reddish brown nodule with firm consistency. Cut surface grey brown homogeneous. Histologically it comprised of numerous thin walled vessels, proliferating spindle to oval cells having eosinophilic cytoplasm, blend to mildly pleomorphic nuclei, arranged in concentric perivascular pattern like hemangiopericytoma and focal glomoid nodular proliferation. Immunohistochemically tumor cells were positive for SMA (Smooth Muscle Actin), calponin and Bcl2, negative for cytokeratins, CD-31, CD-34, S-100, melanosome, CD-99 & KIT. MIB score was 25-30 %, p53 was mildly positive. Mostly Myopericytomas usually behave in a benign fashion, Local recurrences & metastasis may occur in atypical & malignant neoplasms. The patient is doing well without recurrence in thirty months after surgery.*

**Keywords:** Myopericytoma (MPC), Myofibroma (MF), Hemangiopericytoma (HPC), PEC, PMC, IHC.

## INTRODUCTION

Myopericytoma are rare perivascular benign neoplasms. According to updated classification of vascular tumors it has got place amongst tumors of perivascular cells.<sup>[1]</sup> After recent recognition of perivascular myoid cell phenotype and

perivascular epitheloid cell phenotype, angiogenic tumors have been described as PMC tumors e.g. Myopericytoma, myofibroma, myofibromatosis, angioleiomyoma and glomus tumors. Another category is PEC tumors, that comprises melanocytic lineage & gives rise to PECOMA,

clear cell sugar tumors, angiomyolipoma and lymphangiomyomatosis.<sup>[2]</sup>

Myopericytomas are rare perivascular well circumscribed slow growing painless tumors, most commonly develop in superficial soft tissues of extremities followed by head and neck of adults. Although often they have been noticed since birth or early adulthood<sup>[3]</sup>. Men are more often affected than women. Tumors can be solitary or multiple sometimes painful; recur locally in 10 – 20% of patients<sup>[4]</sup>

We aim to present here an unusual case of myopericytoma in 21year averagely built and nourished male presented with complaints of painful sternal nodularity, numbness and weakness of upper extremities. On palpation there was a well defined nodularity (1.5 x1.2cm), firm & partially fixed, present in mid sternal region.

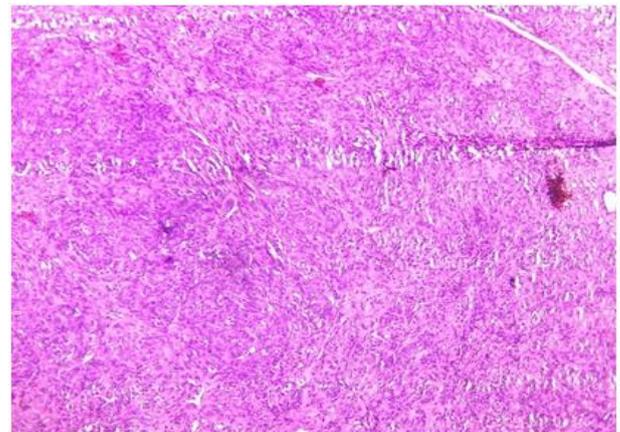
There was no relevant past or family history. On X ray examination it showed a subcutaneous soft tissue swelling without any bony involvement. Pre-operative CBC, ESR & CRP were within normal limits. The surgical excision of nodule was planned and performed, sent for histopathological evaluation. After 3 weeks of surgery patient reported relief from pain and numbness. In thirty months, Post surgical follow up, patient going well with no recurrence. We aim to discuss this rare entity and its unusual presentation, with complete review of literature to the best of our knowledge.

## **PATHOLOGY**

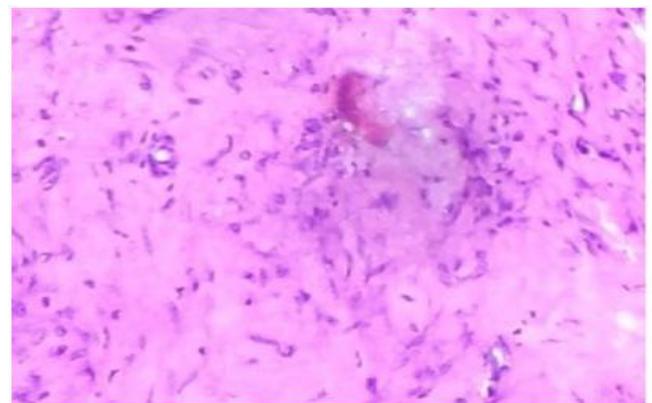
Excised lesion demonstrated an uncapsulated well circumscribed reddish brown nodule which was firm in consistency. Cut surface was greyish brown, homogenous. Total of 4 sections were taken and processed, following standard histopathological protocols.

Microscopic examination of H & E stained sections was performed. It comprised of proliferating spindled to oval cells with eosinophilic cytoplasm arranged around prominent thin walled branching to staghorn vessels (fig.1-A) & there were focal areas of

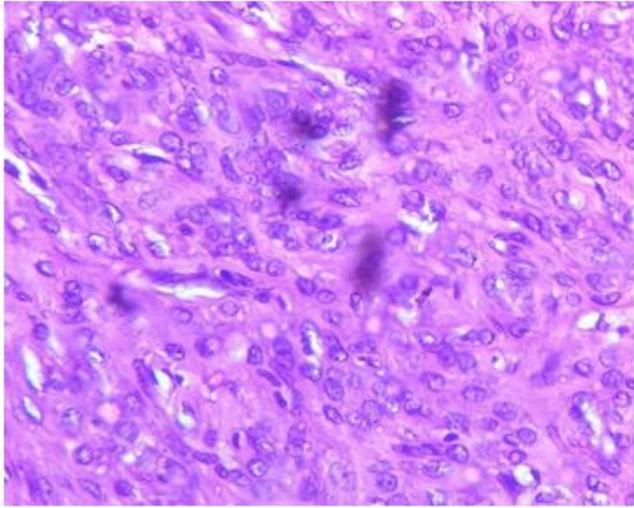
hyalynization & necrosis(Fig.1-B). Another foci showing a concentric perivascular arrangement (glomoid pattern) of the cells giving truly pericytic appearance (fig. 2) .Mild nuclear atypia & < 2 mitotic figures per 10 HPF were observed. These histomorphological findings can also be found in other PMC tumors (including MF). Immunohistochemical analysis were imperative in reaching the correct diagnosis. On IHC diffuse & strong immunoreactivity of tumor cells for SMA (Fig.3), h-caldesmon & calponin (Fig.4) was observed. Focal positivity shown for Desmin (Fig.5) while tumor cells were negative for CD-34 (Fig.6), CD-31, Vimentin (Fig.7) & S-100 protien. MIB score was 25-30% (Fig.8). The diagnosis of myopericytoma of low to intermediate grade was made on the basis of the prominent histomorphological & immunoreactive findings.



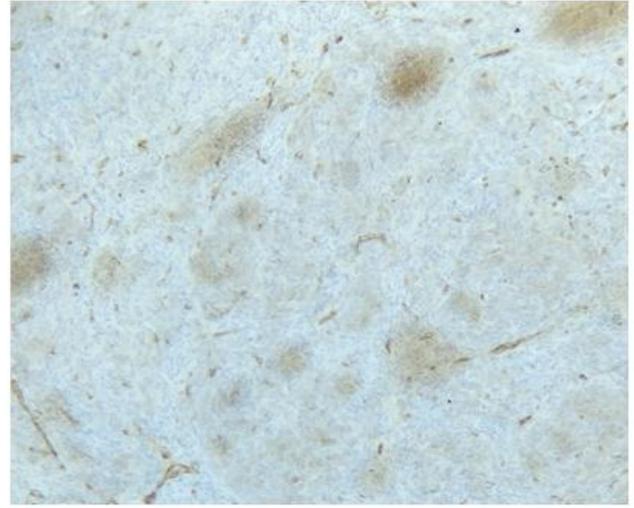
**Fig.1(A).** Numerous thin walled vessels , concentrically surrounded by proliferative spindled to ovoid myoid cells (H&E staining ,original magnification X 100).



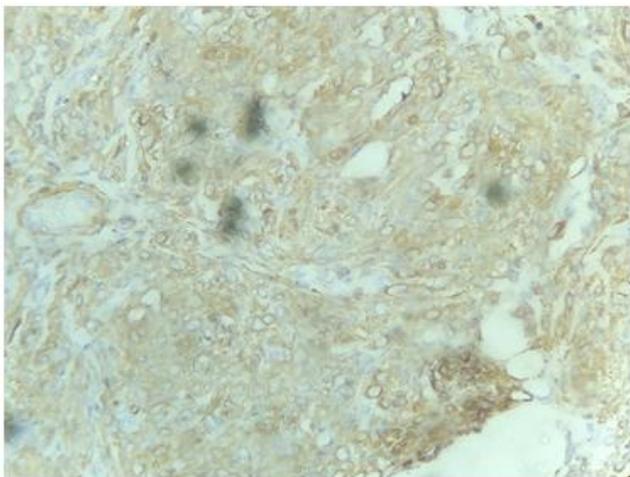
**Fig.1 (B).** Foci of hyaline degeneration in central zone (H&E staining ,original magnification X 400).



**Fig.2** Concentrically arranged myoid cells giving glomoid pattern (H&E staining ,original magnification X 400).



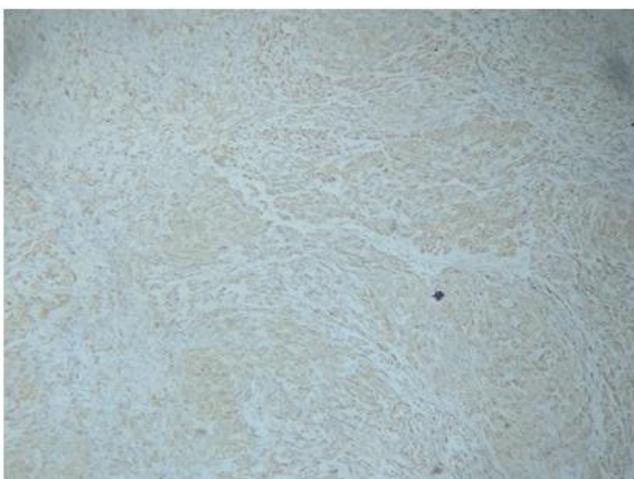
**Fig.5** Focal positivity of tumor cells for Desmine (IHC original magnification X 100)



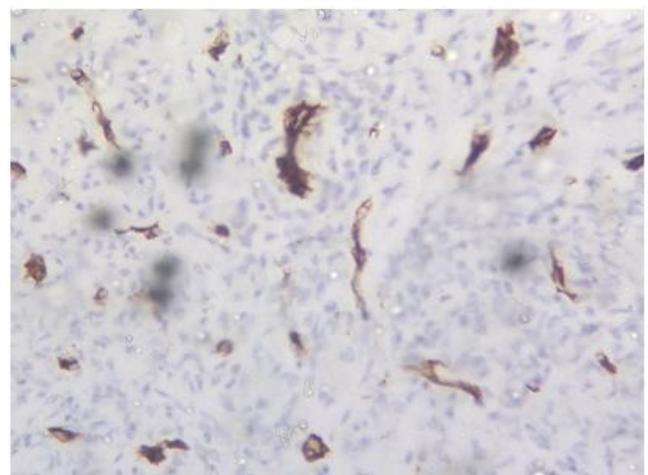
**Fig.3.** Strong immunoreactivity of tumor cells for SMA (IHC original magnification X 400 ).



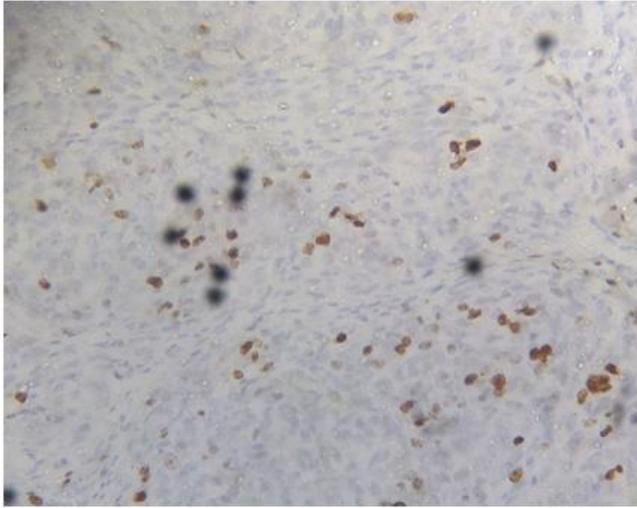
**Fig.6.** The tumor cells are negative for CD-34 while positivity shown by vasculature (IHC original magnification X 100).



**Fig.4** Positive staining of tumor cells for calponin (IHC original magnification X 100).



**Fig.7.** Negative staining of tumor cells for Vimentin (IHC original magnification X 400).



**Fig.8.** M I B score of tumor cells 25-30 % ((IHC original magnification X 400).

## DISCUSSION

Myopericytoma is relatively a new entity and listed as a perivascular tumor in WHO classification of soft tissue tumors [5]. They are usually benign lesions although a few recurrence or malignant cases have been described [3,6,7]. Cytological atypia & increased mitotic activity i.e. > 4 MF per 10 HPF with or without necrosis are the criterias for malignancy. Malignant myopericytomas are commoner in immunodeficient patient with Epstein bar virus (EBV) positivity. Unusual location, multiplicity of lesion & history of rapid growth are the usual findings in EBV patients [8].

MPC shares a very interesting histogenic and morphologic relationship to other similar tumors namely glomus tumors or glomangiopericytomas, MF and HPC. Ultra structurally, origin of this tumor is from pericytes which are perivascular stem cells and considered to be pluripotent and capable of differentiating into smooth muscle cells and glomus cells.[7]

The term myopericyte coined by Dictor et al [9] was used to denote atypical pericytes surrounded by bundles of sclerotic smooth muscle abutting on staghorn vessels. Myo pericytes represent a transitional form between pericytes and smooth muscle cell of vessels [10]. Progenitor cells for myo pericytes include the myofibroblast or the pericytes both of which exhibit the properties of

modified smooth muscle cell[6]. The myofibroblast is a spindle shaped cell with an elongated nucleus and pale eosinophilic cytoplasm. Due to overlapping histological features, misdiagnosis of myopericytoma is frequent. Thorough microscopical evaluation of myopericytoma revealed, numerous proliferating vascular channels, some are staghorn which gives HPC like pattern along with foci of concentrically arranged plump ovoid to spindle pericytic cells giving typical characteristic glomoid pattern. No zonal phenomena was found, which differentiate it from myofibromas. It is usual to find some foci of overlapping morphology with HPC, MF & glomus tumor within the same tumor, hence create diagnostic confusions. Now IHC plays imperative role in final diagnosis.

Tumor cells in MPC shows positive staining for SMA (smooth muscle actin), h-caldesmon & calponin. While negative for CD-34, CD-31, CD-99 & S-100. Focal positivity for Desmin & Bcl-2 was found. Negative staining for S-100 protein & melanosome ruled out the other possibilities including PEC tumor group (as rest give positivity for these).

We have searched the possible literature via pubmed using the term myopericytoma and found a total of 15 different papers and 16<sup>th</sup> will be our case (literature describes total of 76 cases via 15 case reports). Many of them are cited [11-17].

Male:female ratio found was 1.5:1 and age ranged from 13 to 87 years; with median age of presentation 47 years. Most common presentation was asymptomatic nodule however painful nodule was also the feature in some of the cases. The most common site affected is found in lower extremities followed by upper extremities, head and neck region and the trunk. In our case it was solitary painful nodule in midsternal region & both the later findings are unusual.

Most of the MPC,s were solitary & 5 were multiple; involving single or varied anatomical locations. On gross examination all the lesions were circumscribed, firm, nodularity of greyish white to haemorrhagic red brown colour.

In most of the cases lesions were confined to the dermis and superficial part of the subcutaneous tissue. 9 cases arose within vessels showing an attachment to the vascular wall.<sup>(18-20)</sup> and in 3 cases lesion developed secondary to trauma or previous scar.

Histologically these lesions were well circumscribed, with exception of 4 cases that had ill defined border and an infiltrative pattern. These lesion contain numerous thin walled blood vessels, ovoid plump spindle to round myoid lesional cells with eosinophilic cytoplasm, round to spindle nucleus and concentric perivascular growth pattern.

On IHC staining 100% positivity was seen for alpha smooth muscle actin & 91% positivity was for calponin and h-caldesmon. Desmin was negative or focally positive. Our case was positive for SMA and h-caldesmon indicating the HPC related tumor entity and belongs to PMC tumor group based on IHC findings including myopericytoma<sup>(3,12,13)</sup>, myofibroma, myofibromatosis, angioleiomyoma and glomus tumors. Of these the present case was myopericytoma because it was negative for S 100 protein and melanosome, while rest give positivity. As the present case was negative for S100 and melanosome, it does not belong to PEC tumor group which includes PECOMA, clear cell sugar tumors, angiomyolipoma and lymphangiomyomatosis<sup>(2)</sup>. Finally the possibility of Myofibroma has ruled out due to positive staining for h-caldesmon in the present case. MIB score was 25-30% and p53 was mildly expressed, suggesting a low to intermediate malignant potential of present case.

Wide local excision is the treatment of choice and is mandatory to check for any recurrence or atypical changes. Recurrences and malignant transformation have been described specially in deep seated or multinodular tumors. Pleomorphism and high mitotic rate are the determining factors of aggressive behaviour<sup>(7)</sup>. Hence myopericytomas are great mimickers amongst PMC AND PEComas and can arise in

varied locations. The diagnosis of myopericytoma requires the judicious use of immunomarkers and exclusion of all other related entities by morphological and IHC findings.

Multiplicity of lesion, its unusual location and history of rapid growth should be taken with great suspicion for possible malignant behaviour and possible co-association with immunodeficiency.

The prognosis after excision was good in most of the cases, only 2 cases showed local recurrence. No recurrence was found in the present case, even after thirty months following surgery. Remote metastases were negative in all the cases and all the patients are doing well after excision as per literature<sup>(21)</sup> & present follow-up. Myopericytoma is a newly proposed entity by WHO, which has overlapping histomorphology with PMC & PEC tumor groups, creates diagnostic confusions. To solve the mystery IHC is mandatory. Hence it is important for both clinician & pathologist to be cognizant for this rare & new entity.

#### ACKNOWLEDGEMENT

Authors wish to express and convey their sincere thanks and gratitude to Mr.Subhash Gupta for the support given in completion of this Review Article.

#### CONFLICTS OF INTEREST

Authors declare that we did not get any grants for this study; required funds were generated by ourselves.

The Authors also declare that there are no conflicts of interest.

#### REFERENCES

1. J.Eduardocalonje, Christopher D M Fletcher .Pericytic tumors . In : Fletcher Diagnostic histopathology of tumors 4<sup>th</sup> Edition volume 1,2013;(42,43)
2. Armah , H.B. and Parwani , A.V. Perivascular epitheloid cell tumor. Arch Pathol Lab Med. 2009; 133:648-654
3. Mentzel T, Dei Tos AP, SapiZ ,Kutzner H 2006 Myopericytoma of skin and soft

- tissues: Clinicopathologic and immunohistochemical study of 54 cases. *Am J Surg Pathol* 30:104-113
4. J. Eduardo calonje , Christopher D. M. Fletcher. Vascular tumors. In : Fletcher Diagnostic histopathology of tumors 4<sup>th</sup> Edition volume 1 , 2013 ; (78,79)
  5. Fletcher CD, Unni KK, Mertens F, editors. World Health Organisation classification of tumors. Pathology and Genetics of Tumors of Soft Tissue and Bone. Lyon , France : IARC press ; 2002. P138-9.
  6. Terada T. Myopericytoma of low grade malignancy in the oral cavity. *Rare tumors* 2012;4:e9
  7. MC Menamin ME, Fletcher CD. Malignant myopericytoma : Expanding the spectrum of tumors with myopericytic differentiation. *Histopathology* 2002; 41:450-60.
  8. Lau PP, Wong OK, Lui PC, Cheung OY, Ho LC, Wong WC, etal. Myopericytoma in patients with AIDS : A new class of Epstein - Barr virus - associated tumor. *Am J Surg Pathology* 2009;33:1666-72.
  9. Dictor M, Elnor A, Andersson T, etal :Myofibromatosis like hemangiopericytoma metastasizing as differentiated vascular smooth muscle and myosarcoma: Myopericytes as a subset of ‘myofibroblasts’. *Am J SurgPathol*1992; 16: 1239.
  10. Vasenwala SM , Afroz N , Ansari HA, Khan AH , Basari R, Rehman S. Myopericytoma of lip : A rare lesion in an unusual location . *Indian J pathol Microbiol* 2015 ; 58 : 211-3.
  11. Martignoni,G., Pea , M., Regheilin, D., Zamboni , G., and Bonetti , F. PEComa : The past , the present and the future. *Virchows Arch.*2008;452 :119-132.
  12. Dray , M. S., Mc Carthy, S.W., Palmer , A.A., Bonar, S.F., Stalley, P.D., Marjoniemi, V. et al. Myopericytoma : a unifying term for a spectrum of tumors that show overlapping features with myofibroma. A review of 14 cases. *J Clin Pathol.* 2006 ;59 : 67-73
  13. Scott , R.S. , Blank , K.L., Proffer L.H., Kraus,E.W., and Heim – Hall , J. Perivascularmyoma of myopericytoma and myofibromatosis – type arising in a chronic scar. *J Cutan Pathol* .2006 : 33 :231-235.
  14. Laga, A. C. ,Tajirian , A. L.,Islam , M.N., Bhattachaya, I., cohen, D. M., Plamondon, C.J. etal. Myopericytoma : report of 2 cases associated with trauma . *J Cutanpathol.* 2008;35 : 866-870.
  15. Mc Menamin ME, Calonje E: Intravascular myopericytoma . *J Cutan Pathol* 2002; 29:557-561.
  16. Mimami ,Y.,Shiomi,T., and Manaba , T. Perivascularmyoma : case report and immunohistochemical and ultrastructural studies . *Pathol Int.* 2002;52 : 69 -74.
  17. Terada ,T. Minute myopericytoma of the neck : a case report with literature review and differential diagnosis. *Patholoncol Res.* 2010;16 :613-616.
  18. Park H J, Lee DR , Park MY ,etal : A case of intravascular myopericytoma . *J ClinPathol*2010 ; 63 : 847-848.
  19. Ko JY, ChoiWJ, Kang HS, etal: Intravascular myopericytoma : an interesting case of a long standing large, painful subcutaneous tumor. *PatholInt*2011 ;61 : 161-164.
  20. Woollard AC, Southgate C , Blair JW : Intravascular myopericytoma of the superficial palmar arch. *J Hand SurgEur* 2007 ;32:475.
  21. Tadashi Treada . Myopericytoma of soft tissue ( thigh) . *Human Pathology : Case Reports* (2016) 3, 37 - 40.