

CASE REPORT

Myopericytoma - a unifying term or a unique entity?

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Abstract

Myopericytoma are uncommon, slow-growing benign perivascular neoplasms that show hemangiopericytoma-like vascular pattern. We report a 52-year-male patient with a painless palpable nodule in the left thumb for the past 9 months. The mass, on the thenar aspect, was 15x12mm in size. X-ray revealed a soft tissue swelling with no bony association. The excised nodule was a non-capsulated, well-circumscribed vascular neoplasm composed of proliferating spindle to ovoid bland cells with eosinophilic cytoplasm. A concentric perivascular arrangement of the cells was seen interspersed by thin-walled, branching, staghorn blood vessels. Nuclear atypia, mitotic figures and necrosis were not observed. Immunohistochemistry revealed diffuse positivity of the tumour cells for smooth muscle actin while staining negative for CD34 and desmin - features suggestive of origin from the perivascular myoid cell. Morphological features of myopericytoma are shared with hemangiopericytoma, glomus tumors, myofibroma and solitary fibrous tumour which form the important differential diagnoses. It is a relatively newly described disease entity recognized by the World Health Organisation classification of tumours.

Keywords: soft tissue tumour, perivascular myoid cell, concentric perivascular pattern, myopericytoma, hemangiopericytoma

INTRODUCTION

Myopericytomas (MPC) are uncommon benign mesenchymal neoplasms thought to be of perivascular myoid cells. It is a recently proposed term and shows a range of histological growth patterns, recognized by the World Health Organisation classification of tumours.¹ The diagnosis is often based on the immunohistochemical and ultrastructural characteristics, demonstrating features of both smooth muscle and glomus cells, indicative of a tumour originating from the perivascular myoid cell.² We report here a left thumb swelling diagnosed as myopericytoma.

CASE REPORT

A 52-year-male patient visited the surgical outpatient department of our hospital with a complaint of a painless palpable nodule in the left thumb for the past 9 months. There was no history of trauma. The mass, on the thenar aspect, had grown to its present size of 15x12mm. There was no relevant past or family history. An X-ray

of the region showed a soft tissue swelling with no bony association. The nodule was excised and sent for histopathological evaluation.

Pathology

The excised lesion demonstrated a non-capsulated well-circumscribed vascular neoplasm composed of proliferating spindle to ovoid bland cells with eosinophilic cytoplasm. A concentric perivascular arrangement of the cells was seen interspersed by thin-walled, branching, staghorn blood vessels (Figures 1 & 2). Nuclear atypia, mitotic figures and necrosis were not observed. Immunohistochemical evaluation revealed diffuse positivity of the tumour cells for smooth muscle actin (Figure 3) while stain for CD34 and desmin was negative (Figure 4). A histological diagnosis of myopericytoma was offered.

DISCUSSION

Myopericytomas are benign perivascular neoplasms that show hemangiopericytoma-like vascular pattern and are uncommonly

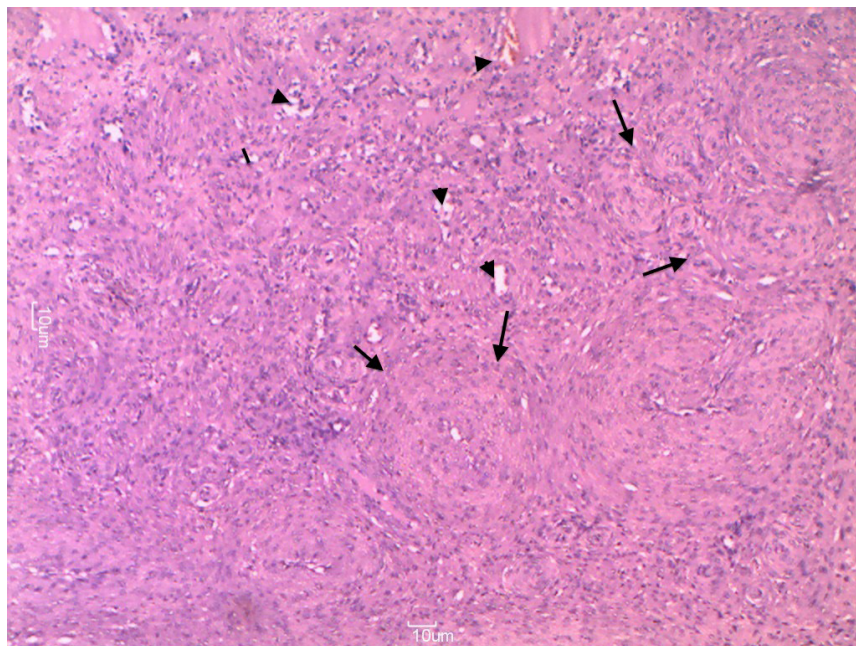


FIG. 1: The lesion showing numerous blood vessels (arrow heads) along with concentric perivascular layering of tumour cells (arrows). (H&E x100)

seen.³ They have a predilection for the distal extremities involving the skin and superficial soft tissues including the regions of trunk, head and neck.^{2,4} Generally, myopericytomas are slow-growing neoplasms, benign in behavior with heterogenous morphology composed of oval/spindle-shaped cells characterized by concentric

perivascular growth and myoid differentiation.⁴ Malignant forms, though extremely uncommon, have also been described, characterized often with recurrence locally and distant metastasis, sometimes pursuing an aggressive clinical course.⁴ Malignant histological features are characterized by mitotically active pleomorphic

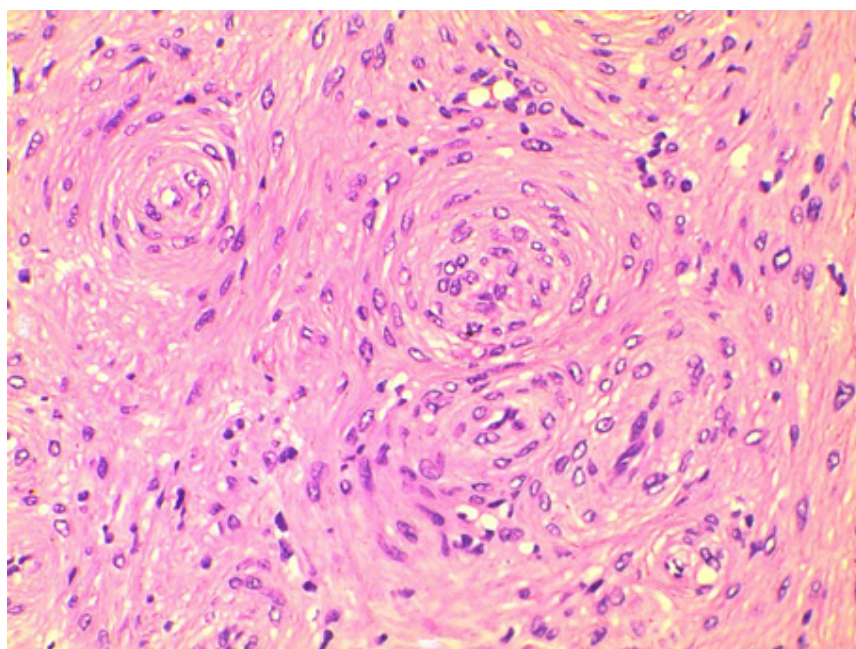


FIG. 2: Plump eosinophilic cells arranged around blood vessels in a concentric pattern. (H&E x200)

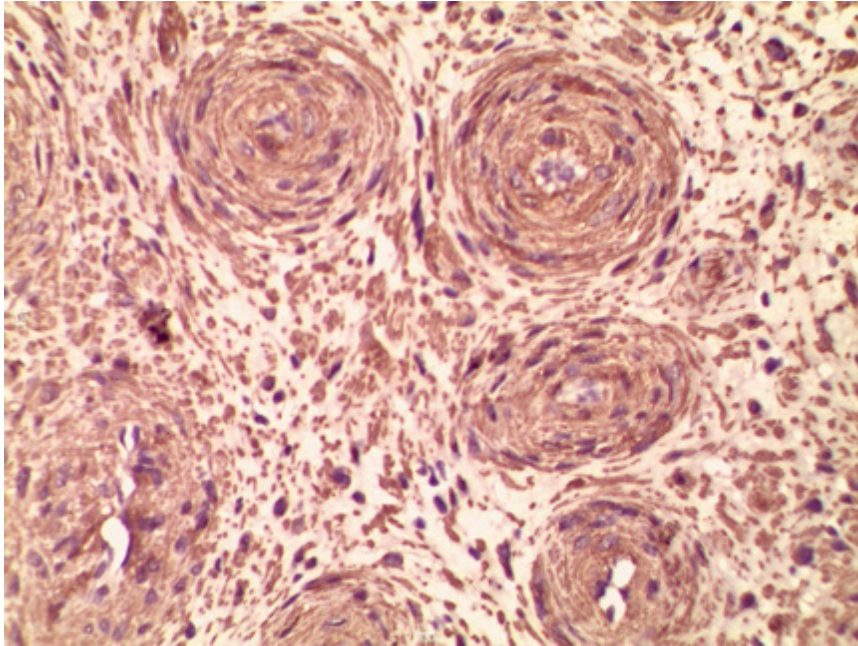


FIG. 3: The tumour cells show diffuse positivity for smooth muscle actin (IHC, x200)

myoid appearing cells, oval- to-spindle shaped with eosinophilic cytoplasm, nuclear enlargement and occasional multinucleation.⁵ Characteristic concentric perivascular growth of cells in varying proportions with vascular extension has been reported in malignant pericytomas. This is in addition to evidence of SMA positivity

suggesting myopericytic differentiation. Areas of necrosis may also be evident.⁵

MPC is a relatively new disease entity, first reported by Granter *et al* in 1998⁶ as belonging to a group of neoplasms showing perivascular myoid differentiation. Later, the World Health Organization (WHO) in their 2002 classification

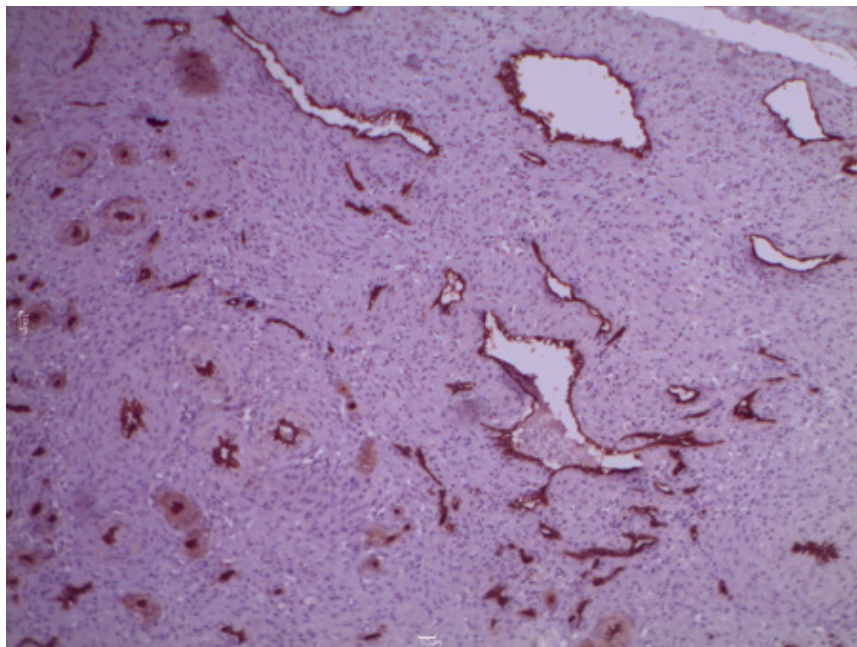


FIG. 4: The tumour cells negative for CD34 while normal endothelial cells are positive. (IHC, x100)

recognized MPC as separate entity with glomangiopericytoma as a subtype of MPC while myofibroma is grouped separately from MPC.¹

MPC has a wide histological spectrum varying from lesions similar to myofibromatosis to those resembling glomus tumours and angiomyoma. A mixture of compact cellular areas and variable numbers of vascular channels are seen with prominent vascular branching imitating a hemangiopericytoma-like appearance.⁷ The term MPC is proposed to include lesions such as myofibromatosis, adult myofibroma, glomangiopericytoma and infantile haemangiopericytoma.⁶ All these entities have overlapping histological features and show differentiation towards myopericytes.⁷ It shows features between vascular smooth muscle cells and pericytes hence representing a transitional form in between the two types of cells. Microscopical evaluation of a benign form of myopericytoma reveals a fibrocollagenous capsule along with islands of concentric arrangement of cells. Characteristic perivascular arrangement of neoplastic cells around numerous small to medium-sized vessels is a typical finding,⁴ along with many thin-walled branching staghorn vessels. Morphological features of MPC are shared with hemangiopericytoma, glomus tumors, myofibroma (MF), and solitary fibrous tumour, hence it becomes important to differentiate them. Immunohistochemical analyses appear to be imperative in reaching the correct diagnosis. Tumour cells in MPC show positive staining for smooth muscle actin and vimentin while a negative staining for CD34 and desmin. Our case also was negative for CD34 and desmin. Glomus tumors are morphologically different as they do not show the characteristic concentric arrangement of neoplastic cells. A solitary fibrous tumour on the other hand can be differentiated from myopericytoma, on the basis of immunoreactivity for vimentin, CD34, Bcl-2 and CD99.⁴ Differentiating from myofibroma is difficult as they share an overlapping spectrum of growth patterns. Categorization of tumours as MPC or MF depends on the predominant growth pattern. Perivascular arrangement of plump spindle cells in a concentric fashion favours the diagnosis of MPC while a typical biphasic pattern of central primitive spindle cells and peripheral myoid/myofibroblastic cells is characteristic of MF. The primitive spindle cells have scant cytoplasm and are arranged around branching blood vessels while the myoid cells, located peripherally, are spindle cells with

abundant pale eosinophilic cytoplasm with a fascicular or whorled appearance embedded in a myxoid matrix.^{2,5,8} These areas tend to bulge and invaginate into the intralesional blood vessels.⁹

The overlapping features of the different lesions led the authors to include them in a comprehensive spectrum under the term MPC, which include myopericytoma, myofibroma and glomangiopericytoma (GPC).⁶ The presumed cell of origin, the myoid cell, is located perivascularly and possesses features of both smooth muscle cells and glomus cells which explains the spectrum of growth patterns and the overlapping histological features seen in these uncommon tumors.

Conclusion

Myopericytoma is an uncommon benign spindle cell tumor, typified by a vascular architectural pattern similar to hemangiopericytoma and showing features of perivascular myoid (myopericytic) differentiation. It is a newly proposed term included in the WHO classification. The overlapping histological features of perivascular arrangement of tumour cells seen in a number of tumors have led to a varied differential diagnosis. Immunohistochemical evaluation is a useful aid to arrive at the correct diagnosis. In view of the recent delineation of the lesion both clinicians and pathologists need to be aware of myopericytoma.

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REFERENCES

1. Folpe AL, McMenamin ME. Pericytic (perivascular) tumours. In: Fletcher CDM, Unni KK, Mertens F, editors. Pathology and genetics of tumours of soft tissue and bone. Lyon, France: IARC Press; 2002: 135–9.
2. Dray MS, McCarthy SW, Palmer AA, *et al.* Myopericytoma: a unifying term for a spectrum of tumours that show overlapping features with myofibroma. A review of 14 cases. J Clin Pathol. 2006; 59: 67–73.
3. Ide F, Obara K, Yamada H, Mishima K, Saito I. Intravascular myopericytoma of the oral mucosa: a rare histologic variant in an uncommon location. Virchows Arch. 2007; 450: 475–7.
4. Lau SK, Klein R, Jiang Z, Weiss LM, Chu PG.

- Myopericytoma of the kidney. *Hum Pathol.* 2010; 41: 1500-4.
5. McMenamin ME, Fletcher CD. Malignant myopericytoma: expanding the spectrum of tumours with myopericytic differentiation. *Histopathology.* 2002; 41: 450-60.
 6. Granter SR, Badizadegan K, Fletcher CD. Myofibromatosis in adults, glomangiopericytoma, and myopericytoma: a spectrum of tumors showing perivascular myoid differentiation. *Am J Surg Pathol.* 1998; 22: 513-25.
 7. 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: 104-13.
 8. Bansal S, Desai RS, Shetty S. Myopericytoma of oral cavity: a case report and review of literature. *Int J Oral Maxillofac Pathol.* 2013; 4: 31-5.
 9. McMenamin ME, Calonje E. Intravascular myopericytoma. *J Cutan Pathol.* 2002; 29: 557-61.