


## CASE REPORT

# Thoracic myopericytoma in an older adult, rare but possible: A case report

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## Abstract

Myopericytoma is a rare tumor generally arising from skin and soft tissues of extremities, trunk, head, and neck regions, rarely from visceral sites. An intrathoracic visceral localization may carry a broad differential diagnosis including primary lung, pleura and chest wall lesions, or metastatic lesions. To date, any radiological features have been recognized and diagnosis of myopericytoma with intrathoracic localization remains still challenging. Here, we describe the case of a subpleural lesion incidentally diagnosed in an older adult affected by gastric cancer. Radiological features did not allow a differential diagnosis between a benign lesion, a primary tumor, or a metastasis. After resection, the histological examination showed histopathological features congruent with the diagnosis of myopericytoma. This unusual presentation reflects the need to share clinical, radiological, and histopathological data about this uncommon but frequently misdiagnosed disease.

## KEYWORDS

myopericytoma, perivascular tumor, sub-pleural lesion, thoracic myopericytoma

## INTRODUCTION

Myopericytoma is a rare tumor generally characterized by cells of different shapes arising from perivascular myoid cells, called myopericytes.<sup>1</sup> It represents a quite recently delineated entity previously classified as a variant of hemangiopericytoma and currently classified by the World Health Organization (WHO) as soft tissue tumor, belonging to the group of peripheral blood cell/vascular cell tumors.<sup>2</sup>

The etiology is currently unknown, even if a co-relation with trauma or viral infections, particularly Epstein–Barr virus (EBV) in patients with acquired immunodeficiency syndrome, has been suggested.<sup>3,4</sup>

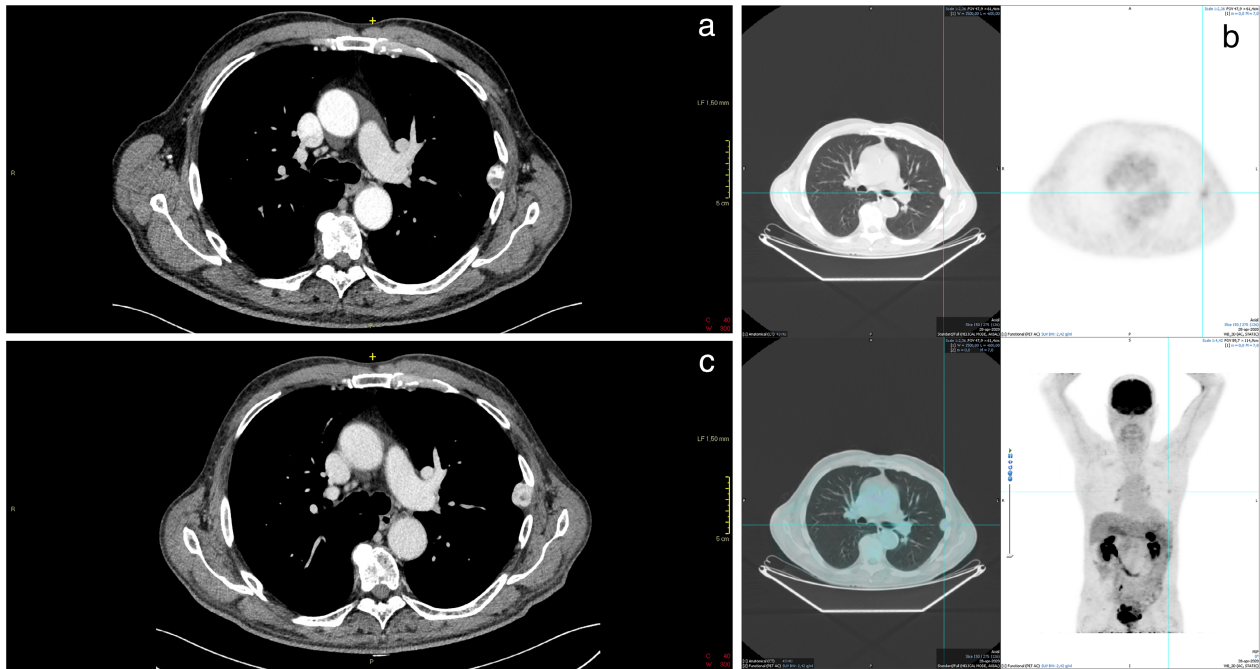
It is generally a tumor of childhood age, whereas it is uncommon in adults. It typically arises from distal extremities, occurring as single or multiple cutaneous or subcutaneous nodes, but it may also originate from other districts,

such as the trunk, head, and neck, and occasionally from the visceral organs.<sup>5–16</sup>

Histologically, myopericytomas are characterized by the presence of several blood vessels and a series of ovoid, plump, spiny, and/or round perivascular myoid cells, with eosinophilic cytoplasm.<sup>1,17</sup> Generally, a positive staining for h-caldesmon, smooth muscle actin, and myosin heavy chain are frequently described, whereas immunostaining for desmin is often negative.<sup>1,17</sup>

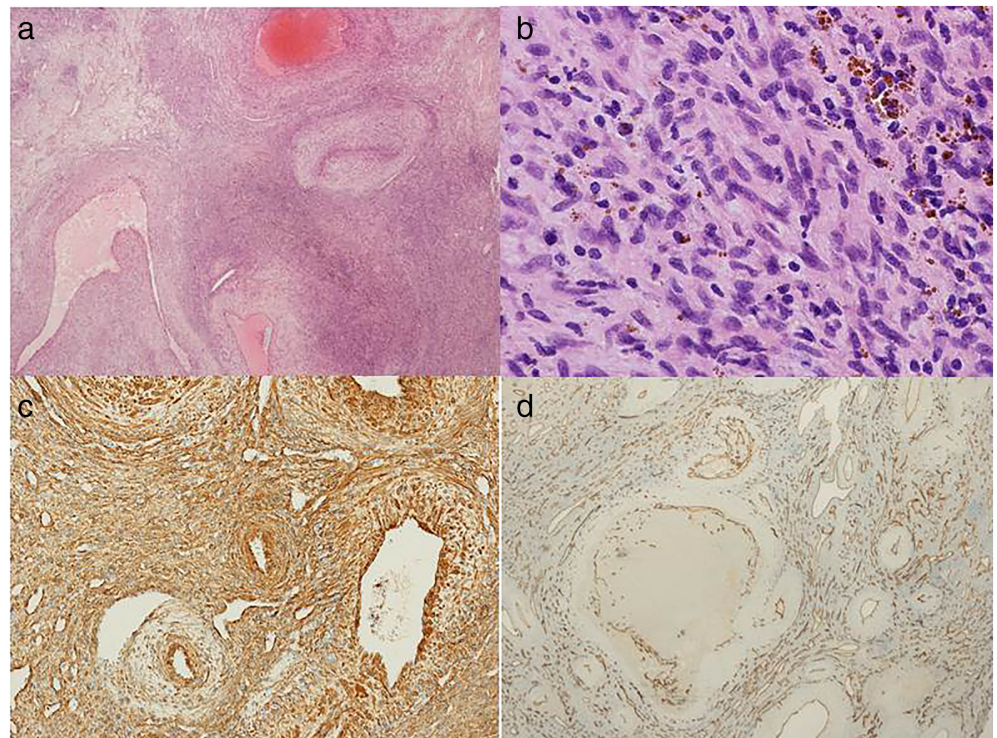
Most myopericytomas present as benign in nature with an indolent clinical course, even if a malignant behavior has been described in a few cases, usually occurring in deeper locations and showing alarming morphological and immunophenotypic features such as the presence of an infiltrative growth, high mitotic activity, atypical cells, and necrosis.<sup>10,17,18</sup>

Here, we report a rare presentation of thoracic myopericytoma, diagnosed in an older male affected by gastric cancer.



**FIGURE 1** (a) Axial CT scan showing subpleural lesion of  $\sim 19$  mm in size with regular margins and high contrast enhancement, arising from the thorax wall. (b)  $^{18}\text{F}$ -fluorodeoxyglucose (FDG) positron emission tomography (PET) demonstrating a mild FDG uptake (SUVmax 2.4) of the thoracic lesion. (c) One-year-after axial CT scan showing the known subpleural lesion increased in size ( $24 \times 22$  mm), highly vascularized without signs of infiltration

**FIGURE 2** Histological findings. (a) The tumor was composed of blanching thick-walled blood vessels with cellular stroma (Hematoxylin and Eosin stain (H&E)  $10\times$ ). (b) The intermixed cellular stroma was composed of small spindled and ovoid cells with limited amounts of pale eosinophilic cytoplasm without atypia or pleomorphism. Deposits of hemosiderin were also present (H&E  $40\times$ ). (c) The tumor cells were extensively positive for h-caldesmon. (d) Staining for CD31 revealed a diffuse proliferation of variably sized blood vessels



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In a 77-year-old male, an axial computed tomographic (CT) scan was performed during the radiological staging for

a gastric cancer and showed a subpleural lesion of  $\sim 19$  mm in size, with regular margins and high contrast enhancement, arising from the thorax wall (Figure 1(a)). At the subsequent  $^{18}\text{F}$ -fluorodeoxyglucose (FDG) positron emission

**TABLE 1** Summary of studies on primary intrathoracic myopericytomas

Reference	Localization	No. of patients	Age of patients (y)	Metastases at the diagnosis	Treatment approach	Recurrence after treatment	Outcome
Cao et al. <sup>6</sup>	Lower lobe of right lung	1	52	No	Lobectomy	No	Alive at 3 y
Edgecombe et al. <sup>7</sup>	Upper lobe of right lung	1	58	No	Wedge resection	No	Alive at 3 y
Song et al. <sup>8</sup>	Right and left lungs	1	26	No	Multiple pulmonary wedge resection	No	Alive at 3 y
Hodges et al. <sup>10</sup>	Pleura	1	57	No	Robotically assisted thoracic resection	NR	NR
Mun et al. <sup>11</sup>	Upper and lower lobe of left lung	1	63	No	VATS left lower lobectomy and upper wedge resection	No	Alive at 34 mo
Lombardi et al. <sup>12</sup>	Case 1. Lower lobe of left lung (subpleural site)	2	68	No	Partial resection	NR	NR
	Case 2. Right lobar bronchus (endobronchial site)		63	No	Right thoracotomy and lower lobectomy	NR	NR

Abbreviations: NR, not reported; No, number (of patients); Y, years; Mo, months; VATS, video-assisted thoracoscopic surgery.

tomography (PET), the lesion presented a mild FDG uptake with uncertain significance (SUVmax 2.4) (Figure 1(b)).

One year later, the subpleural lesion has increased in size (24 × 22 mm), always highly vascularized, but without signs of infiltration (Figure 1(c)).

After multidisciplinary discussion, given the fact that primary or metastatic cancer could not be excluded, the patient underwent elective surgery. Complete resection of the lesion was performed by video-assisted thoracoscopy. The postoperative course was uneventful and the patient was discharged 2 days after the intervention.

Pathologic examination revealed a tumor consisting of, for the most part, small spindled or ovoid neoplastic cells with limited amounts of pale eosinophilic cytoplasm arranged around numerous delicate thin-walled vascular channels, whereas, focally, there were larger myoid nodules with a whorled architecture and a hyaline stroma (Figure 2 (a)). The larger nodules bulged into vascular lumina beneath an intact layer of endothelium. No significant atypia or pleomorphism was found (Figure 2(b)). Tumor cells were diffusely positive for h-caldesmon, whereas desmin and STAT6 were negative (Figure 2(c)). Staining for CD31 highlights the delicate small vessels (Figure 2(d)). All these features fitted very well with a diagnosis of myopericytoma.

According to the uncertain malignant potential of the lesion because of the lack of significant atypia or pleomorphism, the patient continued with the periodic surveillance already ongoing for the gastric cancer.

## DISCUSSION

Myopericytoma represents a rare neoplastic entity with hemangiopericytoma-like vascular pattern, mainly occurring

in childhood to mid-adult years, with a predilection for skin and soft tissues of the distal extremities. Less commonly, it can arise at other sites, including proximal extremities, head, neck, and trunk.<sup>1</sup> Myopericytoma with an intrathoracic localization have been rarely reported. Clinic-pathological features are summarized in Table 1.<sup>6–8,10–12</sup>

Limited data are available about radiological presentation of this type of neoplasm. In the ultrasound examination, the lesions appear homogeneously hypoechoic with well-defined margins and clear blood flow.<sup>19</sup> Although at conventional CT scans the lesions may have a wide spectrum of presentation (they can be hypodense or isodense, homogeneous or not, occasionally calcified, and with clear boundaries), at contrast-enhanced CT evaluation myopericytoma appears as a homogeneous mass with complete uptake or just with peripheral enhancement that is typical of larger lesions with central necrosis.<sup>6,8,15,19</sup> At contrast-enhanced MRI, myopericytoma presents intense enhancement. At conventional MRI, lesions generally appear hypointense in T1W1 scans, whereas in T2W1 scans they show a heterogeneous hyperintensity.<sup>20</sup> This type of tumor presents a low-moderate uptake of FDG at PET examination.<sup>20</sup>

Even if the high vascularization pattern, together with the low FDG uptake, may be pathognomonic findings, the radiological diagnosis of myopericytoma with intrathoracic localization remains still challenging. In fact, the visceral localization is very unusual, especially in older adults, as in this clinical case, thus entering into differential diagnosis with the more frequent epithelial neoplastic lesions of the lung, pleura, and chest wall or with other rare entities such as solitary fibrous tumor, glomangiosarcoma, and perivascular and mesenchymal tumors.

Therefore, an adequate histological and immunohistochemical characterization is mandatory for a definitive

diagnosis and for identifying those rare cases with more aggressive features.

In summary, we report a rare case of intrathoracic myopericytoma made further unusual by the age of onset. Because radiological features appear to be not pathognomonic, it is important to spread awareness about this rare tumor among radiologists and clinicians, especially in the case of unusual presentations or clinical behavior.

#### AUTHOR CONTRIBUTION

M.C.N. and M.N. collected, analyzed data, and wrote the case report. M.G.P. contributed to data curation (histological images). M.M. contributed to data curation (radiological images). A.M. contributed to data curation (radiological images). E.G., M.A.P., P.S., and A.A. reviewed and edited the manuscript. All authors read and approved the final manuscript.

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#### CONFLICT OF INTEREST

The authors declare that they have no conflicts of interests.

#### DATA AVAILABILITY STATEMENT

All datasets generated for this study are included in the article.

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#### REFERENCES

1. 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(5):513–25. <https://doi.org/10.1097/00000478-199805000-00001>
2. Fletcher CD, Bridge JA, Hogendoom PC, et al. World Health Organization classification of Tumours of soft tissue and bone. Lyon: IARC Press; 2013.
3. Laga AC, Tajirian AL, Islam MN, Bhattacharyya I, Cohen DM, Plamondon CJ, et al. Myopericytoma: report of two cases associated with trauma. *J Cutan Pathol*. 2008;35(9):866–70. <https://doi.org/10.1111/j.1600-0560.2007.00910.x>
4. Calderaro J, Polivka M, Gallien S, Bertheau P, Thiebault JB, Molina JM, et al. Multifocal Epstein Barr virus (EBV)-associated myopericytoma in a patient with AIDS. *Neuropathol Appl Neurobiol*. 2008;34:115–7. <https://doi.org/10.1111/j.1365-2990.2006.00865.x>
5. Weng X, Zhang M, Guan H, Sun Y, Wu R, Xu M, et al. Myopericytoma of the stomach: report of one case and review of literature. *Int J Clin Exp Pathol*. 2020;13(7):1771–8.
6. Cao JH, Xu JP, Li YC, Lai J, Li Q. Pulmonary myopericytoma: a case report and review of the literatures. *Chin Med J (Engl)*. 2009;122(6): 755–7.
7. Edgecombe A, Peterson RA, Shamji FM, Commons S, Sekhon H, Gomes MM. Myopericytoma: a pleural-based spindle cell neoplasm off the beaten path. *Int J Surg Pathol*. 2011;19(2):247–51. <https://doi.org/10.1177/1066896910381897>
8. Song XL, Sun XY, Zhang GL, Yu YW, Wang CH. Myopericytoma presenting as multiple pulmonary nodules. *Intern Med*. 2012;51(6): 639–42. <https://doi.org/10.2169/internalmedicine.51.6531>
9. Mainville GN, Satoskar AA, Iwenofu OH. Primary malignant myopericytoma of the left atrium—a tumor of aggressive biological behavior: report of the first case and review of literature. *Appl Immunohistochem Mol Morphol*. 2015;23(6):464–9. <https://doi.org/10.1097/PAI.0b013e3182598554>
10. Hodges K, Zou Y, Everett A, Murthy S, Ahmad U. Pleural myopericytoma: a rare neoplasm in a difficult location. *J Thorac Cardiovasc Surg*. 2018;156:e129–31. <https://doi.org/10.1016/j.jtcvs.2018.01.072>
11. Mun JM, Kim JI, Ko TY, Park SD. Surgical treatment of primary pulmonary Myopericytoma. *Korean J Thorac Cardiovasc Surg*. 2018; 51(3):220–2. <https://doi.org/10.5090/kjtc.2018.51.3.220>
12. Lombardi M, Ibrahim M, Ruco L. Myopericytoma-like tumors of the lung: report of two cases. *Respir Med Case Rep*. 2021;1(32):101355. <https://doi.org/10.1016/j.rmcr.2021.101355>
13. Dhingra S, Ayala A, Chai H, Moreno V, Zhao B. Renal myopericytoma: case report and review of literature. *Arch Pathol Lab Med*. 2012;136(5):563–6. <https://doi.org/10.5858/arpa.2011-0387-CR>
14. Lau SK, Klein R, Jiang Z, Weiss LM, Chu PG. Myopericytoma of the kidney. *Hum Pathol*. 2010;41(10):1500–4. <https://doi.org/10.1016/j.humpath.2010.02.014>
15. Zhang Z, Yu D, Shi H, Xie D. Renal myopericytoma: a case report with a literature review. *Oncol Lett*. 2014;7(1):285–7. <https://doi.org/10.3892/ol.2013.1678>
16. Borella F, Lucchino F, Bertero L, Ribotta M, Castellano I, Carosso A, et al. Clinico-pathological features of gynecological myopericytoma: a challenging diagnosis in an exceptional location. *Virchows Arch*. 2019;475(6):763–70. <https://doi.org/10.1007/s00428-019-02645-2>
17. Mentzel T, Dei Tos AP, Sapi Z, et al. Myopericytoma of skin and soft tissues: clinicopathologic and immunohistochemical study of 54 cases. *Am J Surg Pathol*. 2006;30(1):104–13. <https://doi.org/10.1097/01.pas.0000178091.54147.b1>
18. McMenamin ME, Fletcher CD. Malignant myopericytoma: expanding the spectrum of tumours with myopericytic differentiation. *Histopathology*. 2002;41(5):450–60. <https://doi.org/10.1046/j.1365-2559.2002.01537.x>
19. Rho BH, Lee SK, Kwon SY. Myopericytoma of the neck: sonographic appearance and sonographically guided needle biopsy. *J Clin Ultrasound*. 2011;39:469–72. <https://doi.org/10.1002/jcu.20827>
20. Shaikh F, Huang B, Buchman C, et al. Multimodality molecular imaging correlation of a GLUT1-positive myopericytic lesion with a typical features—a case study. *OMICS*. 2013;2:1000111.

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