

A Case of Intravascular Myopericytoma Located in the Neck

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ABSTRACT

Background: Myopericytomas are rare tumors originating from perivascular myoid cells, with overlapping features of glomus tumors and myofibromas.

Case Report: We present a case of intravascular myopericytoma, a rare variant of myopericytomas, with a mass located in the neck. This case is accompanied by a review of the relevant literature discussing its histomorphological and immunohistochemical features. In histopathology, this tumor is characterized by spindle or epithelioid cells that surround the vessels in a concentric pattern.

Conclusion: While these tumors most commonly occur in extremities, they are rarely seen in the head, neck, and trunk. Even though they are generally considered benign, it's important to note that cases showing increased mitotic activity, recurrence, and metastasis have been reported.

Keywords: Myopericytoma, intravascular, angioleiomyoma.



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INTRODUCTION

Myopericytoma was first described by Granter in 1998 as a rare tumor typically found in the dermis and superficial soft tissues.¹ Most are slow-growing tumors with benign behavior, but a few have been noted to present recurrent disease and distant metastases. In 2002, it was classified as a pericytic tumors by the World Health Organization (WHO). Myopericytomas rarely occur in a blood vessel wall, with the intravascular form first identified by McMenamin et al.^{2–4} Here, we presented this rare form with histopathological features, alongside a literature review of a case involving a cervical mass.

CASE REPORT

A seventy-eight-year-old male patient was admitted to the otorhinolaryngology outpatient clinic complaining of neck swelling. Ultrasonography revealed a non-vascular, superficially located, oval-shaped, well-defined lesion, measuring 0.4x0.7 cm at level three on the right side of the neck. Suspecting lymphadenopathy, we performed a mass excision. Macroscopic examination of the mass revealed a cream-colored, uniformly encapsulated appearance, measuring 0.8x0.7x0.5 cm. The cross-section was cream-colored and solid in appearance. Microscopic examination revealed a well-defined, non-encapsulated tumor, displaying nodular growth towards the sur-

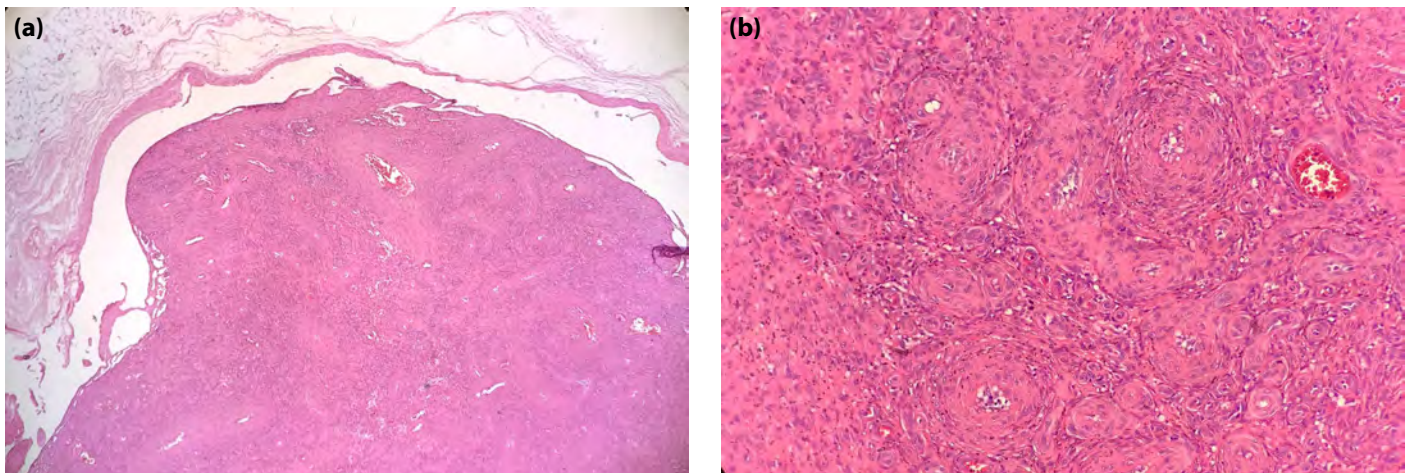


Figure 1. Tumor nodule located in vascular space (**1a**: H&E, 50X), and tumor cells demonstrating a concentric arrangement around the vessels (**1b**: H&E, 200X).

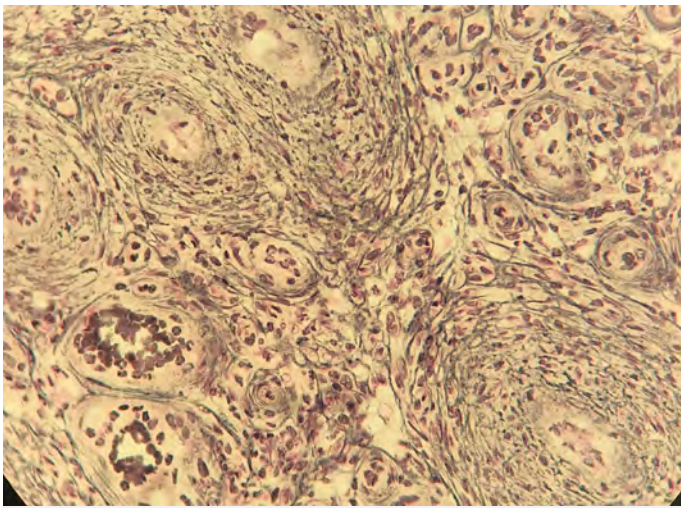


Figure 2. Tumor showing a concentric pattern around the vessels, revealed by Gomori reticulin staining (Gomori reticulin, 400x).

rounding vascular lumen (Fig. 1a). The tumor consisted of cells with oval shapes and eosinophilic cytoplasm that were arranged concentrically around the vessels in a myxoid and predominantly fibrous stroma where scattered lymphocytes were observed (Fig. 1a).

No necrosis or increased mitotic activity was noted. Histochemical studies revealed a concentric pattern around the vessels with reticulin fibers (Fig. 2). Immunohistochemical studies showed the tumor cells positively stained with Smooth Muscle Actin (SMA) and were negative for desmin, S100, CD34 (Fig. 3a, b).

DISCUSSION

Myopericytes have been described by Dictor et al. as cells with a transitional form between pericytes and vascular smooth muscle cells.^{3,5} Myopericytomas are characterized by the concentric arrangement of oval and spindle cells around numerous thin-walled vascular structures. These tumors most frequently occur in the thigh, hand, head, neck, trunk, foot, and heel.⁴ Predominantly found in males, they have been observed in individuals aged between 10-87 years (average 49 years).⁶ Although typically benign, reports of malignant cases with recurrence exist. Some suggest trauma may play a role in etiology. Lesions can be solitary or multiple. Myopericytomas affecting visceral organs have been associated with the Epstein-Barr Virus (EBV).³ However, the mechanism underlying EBV-associated soft tissue tumors (SMTs) remains unclear.⁷ In contrast to sporadic pericytic tumors, EBV associations have been found exclusively in Acquired Immunodeficiency Syndrome (AIDS) patients. Myopericytomas in AIDS patients show some differences from sporadic cases, such as rare sites of involvement and multifocal involvement.⁸ Reports of EBV-associated myopericytomas are extremely rare. EBV-related tumors located in the cranial, hepatic, spinal, bronchial, tongue, brain, vocal cords, and feet regions have been reported in patients with AIDS. Atypical localization and multifocality are usually noted in these patients.^{3,7,8}

Tumors are nodular, hard, gray-white, or hemorrhagic brown masses.⁹ They comprise mesenchymal proliferation, consisting of well-limited, non-encapsulated fusiform cell areas, growing concentrically around blood vessels.³ In addition to the classical solid nodular pattern, different morphologies may be encountered, including hemangiopericytoma-like, angioleiomyoma-like, cutaneous myofibroma-like, hypocellular

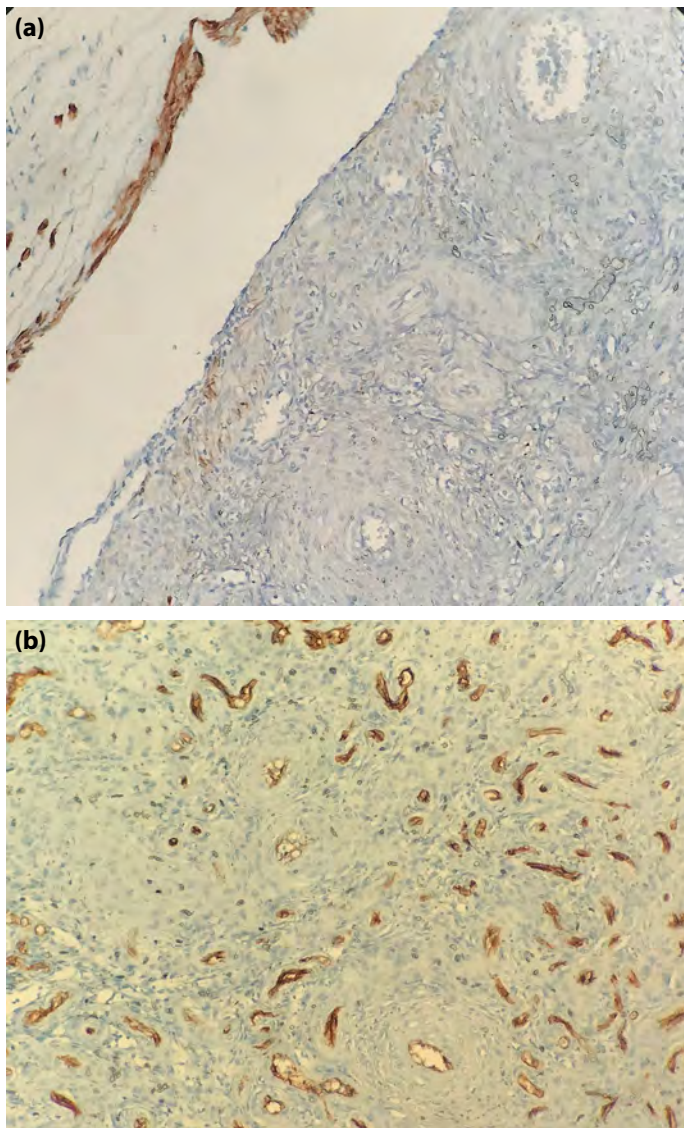


Figure 3. The vessel wall exhibits desmin staining (**3a**: DAB, 200x), whereas tumor cells are negative for both desmin and CD34 (**3b**: DAB, 400X).

fibroma-like, and glomoid myopericytomas.⁹ These tumors can develop within an existing vascular malformation. In such contexts, the differential diagnosis between proliferative and malformative vascular lesions is necessary to prevent inappropriate treatment.⁴ Necrosis is typically not observed, while rare mitosis may be seen.

Malignant cases are generally deeply located, with limited infiltration, and may exhibit mitosis, atypia, necrosis, and metastasis.¹⁰ Immunohistochemically, these cases are SMA and caldesmon positive; S100, CD34, pan-cytokeratin are negative. Desmin can be focally positive.

Table 1. Sex, age and site of intravascular myopericytoma cases in literature

Study	Sex and age	Site
McMenamin et al. ²	M/54	Thigh
Valero J et al. ³	M/48	Foot
Augusti J et al. ⁴	F/63	Foot
Manole et al. ⁶	M/36	Arm
Woollard AC et al. ¹¹	M/54	Hand
Ide F et al. ¹²	F/45	Oral
Xia CY et al. ¹³	M/50	Thigh
Park HJ et al. ¹⁴	F/79	Infraorbital
Ko JY et al. ¹⁵	M/67	Thigh
Mahapatra P et al. ¹⁶	F/59	Hand
Augusti J et al. ⁴	F/63	Foot
Kagoyama K et al. ¹⁷	M/78	Foot
Mohammed et al. ¹⁸	F/36	Foot

So far, a total of 12 cases have been reported in previous studies (Table 1).^{2-4,11-18} Pain is more common in intravascular myopericytoma cases, likely caused by thrombus formation due to the tumor. Unlike the focal positivity observed in myopericytomas, this type is desmin negative.³

In differential diagnosis, glomangioma, angioleiomyoma, and myofibroma should be considered. Desmin positivity and the presence of prominent smooth muscle cell fascicles are findings favoring angioleiomyoma. Glomangioma is characterized by eosinophilic cytoplasm, round and sharply bounded nuclei, absence of spindle cell component, and concentric arrangement similar to myopericytoma. Myofibromas are tumors with zonation and biphasic appearance, characterized by immature cells around the veins in a hemangiopericytomatous pattern, and between the myoid nodules in the central and peripheral myofibroblastic cell bands.² These findings may assist in the differential diagnosis with myopericytoma.

Local excision is typically sufficient. Magnetic resonance imaging and ultrasonography are useful for determining the tumor's location and planning the best surgical procedure.³ Rare aggressive tumors harboring Platelet-Derived Growth Factor Receptor Beta (PDGFRB) mutations or Neurotrophic Tyrosine Kinase, Receptor, Type 1 (NTRK1) fusions, or large lesions unsuitable for surgical resection, may benefit from treatment with imatinib, nilotinib, or similar tyrosine kinase inhibitors.¹⁹

CONCLUSION

Although most cases follow a benign course, this rare tumor can behave malignantly and there may be a small risk of recurrence. Different morphological subtypes should be considered in the differential diagnosis of cutaneous mesenchymal tumors.

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