Myopericytoma of the Neck Originating From the Middle Scalene: A Case Report

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Abstract
We report the case of a myopericytoma of the neck. A 23-year-old female noticed a small, nontender mass in her left supraclavicular fossa. The mass grew over a period of 5 months, prompting the patient to seek evaluation. There were no motor or sensory deficits. Imaging suggested a mass originating from the middle scalene muscle. Computed tomography–guided core needle biopsy demonstrated a spindle cell neoplasm with smooth muscle differentiation. Complete surgical excision was performed. Histopathological and immunohistochemical evaluation of the tissue sample suggested myopericytoma. Myopericytoma is an extremely rare tumor of the head and neck. To our knowledge, this is the first reported case of a myopericytoma originating from a scalene muscle.

Keywords
myopericytoma, pathology, scalene, tumor, imaging

Introduction
First described in 1998, myopericytomas are rare, benign, and soft tissue neoplasms.¹ They are recognized by the World Health Organization as a subset of perivascular tumors characterized by oval-to-spindle cell proliferation that show myoid/myopericytic differentiation.¹ Myopericytomas reportedly occur more commonly in males than females (3:2) and present at a median age of 47 but have been described in teenagers and octogenarians alike.² A 10% to 20% recurrence rate is often cited.³ Myopericytomas characteristically originate from cutaneous and soft tissues but have been described originating from skeletal muscle, bone, and most recently, visceral organs.² Although most commonly found in the distal extremities, they have infrequently been identified at other sites including the head and neck and trunk.²⁻⁴ However, to our knowledge, no case presents a myopericytoma originating from a scalene muscle. We report the case of a myopericytoma of the neck originating from the middle scalene of a 23-year-old female.

Case Report
A 23-year-old otherwise healthy female presented to clinic for evaluation of a nontender mass in her left supraclavicular region. The mass had grown during the preceding 5 months. She denied pain, swelling, numbness, tingling, limitation of neck and upper extremity range of motion, recent illness, recent weight change, fevers, or night sweats. On physical examination, her neck was soft and supple with no evidence of lymphadenopathy. Her left upper extremity was warm and well perfused without evidence of edema, without sensory or range of motion deficits, 5/5 strength, and a 3+ radial pulse.

Magnetic resonance imaging demonstrated a relatively well-circumscribed mass in the left supraclavicular fossa measuring 2.5 cm in greatest transverse dimension. The mass was isointense to muscles on T1- and hyperintense on T2-weighted sequences and demonstrated avid but heterogeneous, predominantly peripheral, enhancement. It was attached to the lateral aspect of the middle scalene muscle and was separate from the brachial plexus, which extended anteriorly and inferiorly to the mass (Figure 1).

A computed tomography–guided core needle biopsy demonstrated bland spindle cell neoplasm with smooth muscle differentiation. Excision of the mass was performed. Intraoperatively, the tumor was found to be densely adhered to the periosteum of the transverse processes of the spine. No direct involvement of the brachial plexus was appreciated.

The lesion was excised. Histopathologically, the lesion was nodular and well circumscribed but unencapsulated. The stroma was predominantly collagenous but a few myxoid areas were present. There was no significant necrosis. Narrow, thin-
walled branching vessels were found in some areas (Figure 2). Overall the lesion was moderately cellular and composed of eosinophilic elongated cells with plump, smoothly demarcated, and elongated nuclei (Figure 3). Mitotic figures are present but at a rate of less than 1 per 10 high-power fields.

Immunohistochemical stains (Dako Envision method, Dako North America, Carpinteria, California) demonstrated immunoreactivity to smooth muscle actin (clone 1A4). Staining for β-catenin (clone β-catenin-1) was cytoplasmic only. The tissue was negative for cytokeratin (AE1/AE3), S100 (polyclonal), CD34 (clone QBend 10), CD68 (clone PG-M1), CD117 (polyclonal), and desmin (clone V9).

The diagnosis was myopericytoma of the neck originating from the middle scalene muscle. The patient is doing well without evidence of motor or sensory deficit 5 months postoperatively.

**Discussion**

First described by Zimmerman in 1923, pericytes are pluripotent stem cells that are capable of differentiating into modified smooth muscle cells normally occurring around capillaries.⁵ Myopericytomas refer to neoplasms of partially differentiated...
pericytic stem cells. In the differential diagnosis, myofibroma, angioleiomyoma, glomus tumor, perivascular epithelioid cell neoplasm (PEComa), and hemangiopericytoma should be considered. Hemangiopericytoma should be given particular consideration on account of its histologically similar vascular growth pattern to myopericytoma and due to the fact that this diagnosis is more commonly seen in the head and neck.

The most widely accepted method of diagnosis for these rare perivascular tumors is excisional biopsy followed by histological and immunohistochemical evaluation. Myopericytomas are identified by concentric, onion-skin pattern of round-to-ovoid eosinophilic spindle cells around vascular lumina. Immunohistochemical staining of these cells identifies them as smooth muscle actin and h-caldesmon positive with myoid differentiation. A 2011 case report suggests an ultrasound-guided core needle biopsy as an alternative method of diagnosis. However, the vascular nature of this tumor may limit this approach to superficial tumors such that local pressure would provide sufficient hemostasis.

Although widely considered benign, a small number of malignant myopericytoma cases have been described. The malignant nature of these can often be identified by the presence of active mitoses, necrosis, and nuclear atypia on histologic evaluation. Further, low Ki-67 labeling would indicate a benign clinical course. In the more common scenario of benign myopericytoma, there is a reported 10% to 20% recurrence rate. This is most likely attributed to either the extension of cords of tumor beyond the main lesion, as it is circumscribed but not encapsulated, or multifocal disease and development of a new primary lesion secondary to field change. Additional risks of recurrence may include the inability to perform a wide excision due to the proximity of critical structures.

Due to the small number of diagnosed cases, the management of myopericytoma is not well described. Although benign in nature, definitive diagnosis is not frequently made until postexcisional laboratory evaluation. Although myopericytomas are often asymptomatic, they can present symptomatically with pain, neurologic deficits, or motor deficits. Along with physical deformity, these symptoms are indications for complete surgical excision.

Initial postoperative management should include routine follow-up to assess wound healing and neurologic and motor function. Due to the risk of recurrence, surveillance examinations and imaging should be strongly considered. The typical slow growth of these lesions may require longer term follow-up.

Declaration of Conflicting Interests
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