Pleural myopericytoma: A rare neoplasm in a difficult location

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Coronal CT and intraoperative photograph demonstrating pleura-based myopericytoma.

Central Message

A 57-year-old woman had primary intrathoracic myopericytoma, which was discovered incidentally on a chest radiograph. The patient successfully underwent robotically assisted resection.

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Video clip is available online.

The spectrum of pleural tumors is broad and comprises entities with varying degrees of malignant potential. Generally, solitary pleura-based tumors are treated with resection to alleviate symptoms, establish diagnosis, and exclude malignancy. Myopericytoma, a tumor that is rarely identified as a pleura-based lesion, may share radiographic and pathologic characteristics with solitary fibrous tumors.¹ We present the case of a patient who underwent robotically assisted resection of a primary intrathoracic myopericytoma.

CLINICAL SUMMARY

A 57-year-old woman presented for evaluation of a pleura-based mass identified incidentally on a chest radiograph. Her medical history was remarkable only for uterine fibroids, for which she had undergone a hysterectomy. At

the time of presentation, she had a 2-month history of dry cough and persistent dyspnea. Axial computed tomographic imaging showed a 3 × 4 × 3-cm apical, pleura-based mass with smooth margins (Figure 1, A and B) that did not demonstrate fludeoxyglucose F 18 (INN fludeoxyglucose [¹⁸F]) uptake. A core-needle biopsy sample was consistent with solitary fibrous tumor. Magnetic resonance imaging was performed to assess neurovascular involvement and demonstrated abutment of the brachial plexus and close proximity to the subclavian vessels without neurovascular encasement (Figure 1, C). Because of the risk of malignancy or malignant transformation, robotically assisted thoracoscopic resection, was offered with the plan to convert to

FIGURE 1. A and B, Computed tomography demonstrates a heterogeneous, pleura-based mass at the apex of the left side of the chest that lies in close proximity to the left axillary artery. C, Postcontrast T1 magnetic resonance imaging demonstrates an enhancing lesion that abuts the brachial plexus without neurovascular encasement.

FIGURE 2. A, Robotic port placement. B, Intraoperative photograph of the mass, which was well encapsulated and rich in vascular component. C, Representative histologic section with hematoxylin and eosin stain, demonstrating a tumor composed of uniform cells with a prominent hemangiopericytoma-like vascular pattern.

left hemiclamshell in case of significant subclavian artery involvement.

The patient underwent elective robotically assisted resection of the tumor. She was positioned in the right lateral decubitus position and securely taped to the operating table, with padding on both sides such that table could be rotated significantly without the risk of the patient moving off the table. In the event of emergency conversion to left hemiclamshell, the plan was to rotate the operating table steeply to the left to facilitate exposure. A camera port was placed in the seventh intercostal space, robotic instrument ports in the sixth space, and an assistant port in the eighth intercostal space. The tumor was encapsulated and adherent to the adventitia of the subclavian artery as it traversed the apex of the left side of the chest (Figure 2). There was a network of collateral vessels arising from the pleura and chest wall, which were controlled with a series of clips. The internal thoracic artery was identified and preserved. The tumor

was dissected from the subclavian artery adventitia with bipolar electrocautery. A plane was developed between the tumor capsule and the subclavian artery adventitia, and both structures were successfully preserved. The dissection was intentionally kept in the periadventitial plane of the subclavian artery, and the brachial plexus was not encountered during dissection. Several small feeding vessels were ligated, and the tumor was completely excised (Video 1). Postoperative recovery was uneventful, and the patient was discharged home on postoperative day 2.

Pathologic evaluation demonstrated a tumor composed of uniform cells with a prominent hemangiopericytoma-like vascular pattern. Lesional cells were negative for S100 protein, desmin, AE1/AE3, and CD31. CD34 highlighted the vessels with weak and patchy positivity in the tumor cells. The tumor cells were strongly positive for smooth muscle actin (SMA) and negative for STAT6, supporting a diagnosis of myopericytoma.

DISCUSSION

Myopericytoma represents a rare entity with hemangiopericytoma-like vascular pattern, which primarily affects the skin and soft tissues of the extremities. Less commonly, these tumors are identified affecting the head, neck and trunk.¹ The lesional cells are related to myopericytes, contractile cells that surround capillaries and small vessels.² They are defined by specific histopathologic criteria, including smooth muscle actin positivity.³ To date, there have been only a small number of case reports of primary intrathoracic myopericytomas (Table 1).⁴⁻⁷ There are no data on the clinical characteristics of intrathoracic myopericytoma, but extrapolation from tumors of the extremities and head and neck suggest that these lesions tend to follow an indolent course. In a series of 46 patients with median follow up of 48 months (range 7-168), Mentzel and colleagues³ found that despite marginal or incomplete excision in 23 of 46 cases, only two myopericytomas recurred locally. Nevertheless, there have been reports of

VIDEO 1. Intraoperative video demonstrating robotically assisted resection of an apical myopericytoma at the thoracic inlet. Video available at: [http://www.jtcvsonline.org/article/S0022-5223\(18\)30337-4/fulltext](http://www.jtcvsonline.org/article/S0022-5223(18)30337-4/fulltext).

TABLE 1. Case reports of primary intrathoracic myopericytomas, excluding tumors of the thoracic spine, in English-language journals

Reference	Clinical and anatomic details	Treatment approach	Outcome
Cao et al, ⁴ 2009	Right lower lobe lung nodule, asymptomatic	Right lower lobectomy for suspected primary lung cancer	Alive with no recurrence at 3 y
Edgecombe et al, ⁵ 2011	Pleura-based right upper lobe nodule, asymptomatic, enlarging during 5 y of follow-up, core biopsy suggested myopericytoma	Right upper lobe wedge resection	Alive with no recurrence at 3 y
Song et al, ⁶ 2012	Multiple bilateral lung nodules, symptoms of dry cough without dyspnea	Bronchoscopy followed by multiple pulmonary wedge resections for diagnosis and treatment	Alive with no recurrence at 3 y
Mainvill et al, ⁷ 2015	Malignant left atrial tumor with brain, thoracic spine, and liver metastases, presented with CNS symptoms	Staged resection of primary tumor, occipital lobe metastasis, and thoracic spinal metastasis	Alive at 8 mo with extensive metastatic disease

A PubMed search was conducted for articles containing “myopericytoma” in the title; all results were reviewed for inclusion. CNS, Central nervous system.

malignant myopericytoma. McMenamin and Fletcher⁸ reported a series of 5 patients with malignant myopericytoma; 4 patients had metastases develop, and 3 died within 1 year. Malignant myopericytoma was defined by high mitotic rate and myoid-appearing ovoid to spindle cells showing at least focally striking perivascular orientation as in benign myopericytoma.⁵

Myopericytoma represents a rare intrathoracic neoplasm, which may easily be mistaken for solitary fibrous tumor based on imaging characteristics. The clinical implications of this entity have not been defined. Myopericytomas seem to have some degree of malignant potential, however, so complete resection should be considered.

Multiple published series have demonstrated that pleura-based tumors can be safely resected with minimally-invasive techniques. In such reports, resection margin and oncologic outcome have not been shown to vary by surgical approach. Data on robotically assisted techniques for the resection of chest wall tumors are sparse, but extrapolation from the thoracoscopic surgery literature would suggest that robotic resections can be performed with similar outcomes. Dense tumor adhesions can make resection challenging in

proximity to neurovascular structures. In case of minimally invasive resections, it is important to be prepared to achieve open vascular exposure.

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