Diagnosis and Management of Primary Pulmonary Leiomyosarcoma

Laura M. Arnold, OMS III
Sudeep D. Burman, DO
Albert H. O-Yurvati, DO

Leiomyosarcomas are cancers of smooth muscle cells that can arise from any location but occur most often in the uterus, retroperitoneum, or intraabdominal region. Primary leiomyosarcomas of the lung are extremely rare and are often diagnosed as a mass on routine chest radiography. Metastasis is uncommon and typically occurs late in the disease process, indicating the importance of early detection. Surgical excision is generally curative, offering patients optimal prognoses. The authors describe a 56-year-old asymptomatic man who was found to have a pulmonary mass on routine chest radiography. Further testing revealed a primary pulmonary leiomyosarcoma. The tumor was excised and postoperative osteopathic manipulative treatment was applied to help facilitate patient recovery. A brief discussion of the typical clinical presentation, prevalence, and medical management of these tumors is included.

From the Departments of Surgery at University of North Texas Health Science Center and at Plaza Medical Center in Fort Worth, Texas. Address correspondence to Albert H. O-Yurvati, DO, University of North Texas Health Science Center, Department of Surgery, 855 Montgomery St, Fort Worth, TX 76107-2553.

E-mail: albert.yurvati@unthsc.edu

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Leiomyosarcomas are neoplasms of smooth muscles that most commonly arise from the uterus, gastrointestinal tract, or soft tissue. Primary pulmonary leiomyosarcomas are rare, accounting for less than 0.5% of all malignant lung tumors.1 The earliest case was described in 1903 by Davidsohn.2 In 1999, Cordes et al3 reported that 127 cases had been reported in the literature. When seen in the lung, it is important to establish whether the tumor originated in the lung or if it is secondary to metastases from a separate location, the latter being much more common. To definitively establish the lung as the primary site for a leiomyosarcoma, all other possible sites of origin within the body must be excluded.

A primary leiomyosarcoma of the lung may originate from the smooth muscle of the pulmonary parenchyma, bronchi, or pulmonary arteries, in order of decreasing frequency.4 Most leiomyosarcomas appear to arise from around the hilum and presumably originate from peribronchial smooth muscle fibers, most frequently the larger bronchi of the left lower lobe.1,2,5

Patients may present with symptoms similar to those seen in other primary pulmonary tumors, including cough, dyspnea, hemoptysis, sputum, chest or back pain, and weight loss. Alternately, patients may be asymptomatic on presentation, as seen in the present case report. Because these tumors grow and metastasize slowly, they are virtually curable with early detection and treatment.

Report of Case

A 56-year-old white male aircraft mechanic presented to his primary care physician for his annual physical examination. A routine radiographic image of the chest exposed a 4- to 5-cm mass in the lower lobe of his left lung. The primary care physician ordered a computed tomography (CT) scan of the chest, abdomen, and pelvis to evaluate for evidence of metastases. On the CT scan, a mass was noted in the left lung base with no abnormalities present elsewhere. The patient’s primary care physician referred him to a consulting cardiothoracic surgeon. The patient presented to the cardiothoracic surgeon (A.H.O.) 2 weeks after being seen by his primary care physician. He had a 35-year history of tobacco use, smoking one pack of cigarettes per day for 30 years, as well as a history of squamous cell carcinoma on his hand and basal cell carcinoma on his shoulder. His family history was significant for cancers of the bladder, pancreas, liver, and neck. Additional patient risk factors included workplace chemical and fuel exposure. The patient denied any upper respiratory symptoms, exercise limitations, or contact with individuals who had respiratory symptoms.

On physical examination, vital signs were stable. The patient’s lungs were clear to auscultation with slightly diminished breath sounds bilaterally. Oxygen saturation was 96% on room air. Pulmonary function tests showed evidence of air trapping and bronchodilator response most likely secondary to reactive airway disease. Forced vital capacity (FVC) was 5.2 L (86% predicted); forced expiratory volume in the first second of expiration (FEV₁) was 3.2 L (71% predicted); and FEV₁/FVC was 63%.

The CT scan revealed a 3.5 × 2.2-cm bilobulated mass in...
the anterior basal portion of the left lung lobe abutting the major fissure. Some thickening and fluid in the oblique fissure were also noted. An interventional radiologist performed a CT-guided biopsy, which revealed a mesenchymal lesion consistent with a smooth muscle cell–type lesion. Bundles of spindle-shaped cells with small nuclei and a few large hyperchromatic nuclei were seen. A light scattering of lymphocytes and focal necrosis were evident. Minimal mitotic activity and some nonneoplastic lung tissue were present.

A positron emission tomographic/CT whole body scan was taken to rule out disease involvement in other areas of the body. A 3.7-cm lobulated mass with moderate increased activity was seen in the inferior lingula. There was no evidence of metastatic disease. A repeated chest radiograph, taken 2 months after the initial radiograph, showed a poorly marginated nodular opacity measuring 4.5 cm in the basilar left lower lobe. There was no evidence of infiltrate, mass, or effusion.

The patient underwent a left thoracotomy with a left lower lobectomy. Mediastinal lymphadenectomy of the left upper paraatracheal, left lower paraatracheal, para-aortic, and subcarinal lymph nodes was negative for metastatic disease. The sarcoma was 4 cm and did not involve the lymphovascular bed or the overlying visceral pleura. The surgical margins of resection were negative for malignancy. The sarcoma was 5.2 cm from the bronchial margin of resection. The remaining lung parenchyma demonstrated focal areas of benign pneumocyte hyperplasia and mild emphysematous-type change.

The surgical pathology report revealed a well-circumscribed malignant spindle cell tumor characterized by spindle cells with blunt nuclear ends and markedly increased areas of mitotic activity. The average mitotic rate was 18 mitotic figures per 10 high-powered fields. Punctuate areas of necrosis were also identified. The presence of hypocellularity areas along with hyalinized fibrosis were noted centrally. The malignant spindle cells tested positive for S100 antigen, smooth muscle actin, and vimentin. Malignant spindle cells tested negative for anticytokeratin 5.2, caldesmon, cytokeratin 5/6, cytokeratin 7, cytokeratin 20, CD34, C-Kit, desmin, epithelial membrane antigen, myogenin, pancytokeratin, and thyroid transcription factor-1. The spindle cell tumor had no epithelial differentiation. Overall morphologic features favored a high-grade sarcoma with evidence of smooth muscle differentiation, making this a leiomyosarcoma.

The lung expanded on the left side postoperatively, and the patient had an uneventful recovery. Osteopathic manipulative treatment (OMT) comprising soft tissue and myofascial release to the upper thoracic and chest wall regions and gentle rib raising techniques was applied postoperatively twice daily. Pedal lymphatic pump technique was also used to augment systemic lymphatic flow.

He was discharged in stable condition on postoperative day 3 with a final diagnosis of high-grade spindle cell sarcoma consistent with leiomyosarcoma. He was instructed to follow up with an oncologist for his leiomyosarcoma diagnosis and with his primary care physician for smoking cessation.

Comment

Leiomyosarcomas primary to the lung occur mainly in adults and have a greater prevalence among males, with a 2.5:1 male to female ratio. However, cases have been reported in patients aged 4 years to 83 years. Primary leiomyosarcomas of the lung are extremely rare. Known risk factors include radiation therapy, chemotherapy (eg, cyclophosphamide, melphalan, nitrosoureas), and environmental and occupational exposures (eg, arsenic, dioxin, phenoxy herbicides, vinyl chloride). Many patients with primary pulmonary leiomyosarcomas present similarly to those with other primary tumors of the lung (eg, bronchogenic carcinoma), with symptoms including cough, dyspnea, hemoptysis, sputum, chest or back pain, and weight loss. However, other patients may be asymptomatic on presentation, as in the present case. Primary leiomyosarcomas of the lung arise most commonly in the left lower lobe, as seen in the patient in the present report. Initially, a mass is detected on routine chest radiography. Radiologically, sarcomas appear as nonspecific, solitary, well-defined nodules with smooth margins. To differentiate a primary pulmonary leiomyosarcoma from bronchogenic carcinoma, an excisional biopsy is needed. Diagnosis of primary pulmonary leiomyosarcomas should only be considered when there is no evidence of an occult primary tumor elsewhere in the body. In women, it is important to closely examine the uterus for any evidence of tumor. After tissue biopsy confirms sarcoma, preoperative staging generally includes CT scans of the lungs and magnetic resonance imaging of the primary lesion to determine whether metastases have occurred. Metastasis is uncommon in leiomyosarcomas and typically occurs late in the disease process.

Histologically, leiomyosarcomas are characterized grossly by a grey or white firm surface. Microscopically, malignant spindle cells with cigar-shaped nuclei arranged in interweaving fascicles are noted. Mitotic figures, multinucleation, nuclear atypia, prominent vascularity, scanty cytoplasm, and zonal necrosis are common, while calcification, cavitation, pleural effusions, and pneumothorax are not common. Immunohistochemically, such tumors stain with antibodies to actin, smooth muscle actin, desmin, and vimentin. Leiomyosarcomas are generally negative for carcinoembryonic antigen, cytokeratin, leukocyte common antigen, neuroendocrine filament, and S100 protein.

The goals of treatment are to obtain local and systemic control of the sarcoma while preserving function and quality of life. If preoperative evaluation reveals no evidence of metastases, then treatment is surgical. Patients with large primary tumors may receive preoperative radiation treatment in hopes of decreasing the size of the mass before surgical resection. Depending on the size and grade of the tumor, radiation...
therapy, chemotheraphy, or both may be indicated as an adjuvant therapy. If radiation therapy is not planned, then wide-margin excisions of 3 to 6 cm are generally recommended.8,10

Lymph node resections are generally not necessary because primary leiomyosarcomas of the lung rarely show lymph node involvement. In the present case report, the tumor was resected with a 5.2-cm margin, and no lymph node metastases were evident. An exploratory thoracotomy should be done in all cases where there are no demonstrable metastases.5

Low-grade lesions and all lesions smaller than 5 cm are generally considered “cured” if adequately resected. Low-grade tumors do not tend to recur and complete resection is therefore adequate.9 By contrast, the median survival for patients with unresectable sarcoma or metastatic disease is about 12 months. Primary leiomyosarcomas, if lobulated and not affixed to the chest wall, have a much better prognosis for surgical cure than do bronchogenic carcinomas of the lung.11 Tumor cell necrosis, high cellularity, mitotic counts above 4 mitotic figures per 10 high-powered fields, pleomorphism, and nuclear atypia are all predictive of more aggressive behavior and therefore increased chance of recurrence.11

Osteopathic Manipulative Treatment

The use of OMT as an adjuvant treatment in postoperative recovery of surgical patients can be beneficial. O-Yurvati et al12 examined the effects of OMT immediately following coronary artery bypass graft (CABG) surgery to improve respiratory function and alleviate the anatomic deformation caused by the operation. In patients receiving OMT, cardiac index, saturated venous oxygen, and thoracic impedance were measured before CABG surgery, immediately after surgery, and 5 to 10 minutes after postoperative OMT.12 The study12 showed significant hemodynamic benefits in CABG patients who had received OMT versus those who did not receive OMT. Patients had improved cardiac function (measured by the cardiac index) and improved perfusion (measured by the thoracic impedance and saturated venous oxygen).12 This physiologic study12 suggests that OMT improves fluid homeostasis and hastens recovery following a surgical procedure involving a median sternotomy.

In 1998, Radjieski et al13 described using OMT as adjuvant therapy for acute pancreatitis and found a decreased length of hospital stay (mean, ~3.5 days). Goldstein et al14 reported in 2005 that the combination of postoperative OMT and preemptive morphine in patients undergoing an elective total abdominal hysterectomy had improved analgesic efficacy and comfort in the immediate 48-hours postoperation. As suggested in these studies,12,14 OMT in postoperative patients has the potential to reduce adverse pulmonary complications, hasten recovery, and decrease hospital stay, therefore potentially lowering hospital costs. As more research is performed on the effects of OMT, osteopathic physicians will have opportunities to educate patients and colleagues about the benefits of OMT.

Conclusion

The present case emphasizes the important role of primary care physicians in detecting malignant pathologies of the lungs. If these lesions are detected in the early phase, resection is essentially curative. Primary care physicians also play a vital role in patient follow-up because they can be pivotal in identifying recurrence of this rare tumor. Osteopathic physicians are uniquely situated to use osteopathic manipulative medicine to directly improve the patient’s initial recovery from surgery and reduce the long-term musculoskeletal dysfunctions associated with a major thoracotomy.

References