Incidental Pleural-Based Pulmonary Lymphangioma

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Pulmonary lymphatic disorders are rare and are often mistaken for serious pulmonary diseases. Among such disorders are pulmonary lymphangiectasis, which is often fatal in children; lymphangiomatosis, which comprises multiple lymphangiomas and typically has multiorgan involvement; and lymphatic dysplasia syndrome, which results in peripheral lymphedema and pleural effusions. Pulmonary lymphangiomas, perhaps the most common of the four main manifestations of pulmonary lymphatic disorders, are focal congenital malformations that consist of atretic nonfunctional lymphatic tissue separated from the lymphatic drainage system.

 Mediastinal lymphangiomas comprise 10% while intrapulmonary lymphangiomas make up less than 1% of all lymphangiomas. More than 90% of thoracic lymphangiomas occur in children younger than 2 years and may be more prevalent in those with superior vena cava syndrome. Secondary forms develop in adults as a result of lymphatic channel obstruction caused by radiation, surgery, or infection.

Patients with lymphangiomas can be asymptomatic for many years and often have symptoms only after vital structures are compressed by the lesion. As such, asymptomatic lymphangiomas are typically found incidentally on chest radiographs or computed tomography (CT) scans without any unique visible characteristics. Although biopsies can identify malignant or benign lesions, empiric resection is often performed as a precautionary measure. If surgical means are pursued, however, it is important to remove the entire lesion to avoid tumor regrowth.

In the present report, we describe a woman who had chronic pain in her right upper arm and shoulder. A pleural-based lesion was found incidentally and was initially suspected to be the cause of her pain. However, further examination suggested otherwise. The present report is, to our knowledge, the first published case of isolated pleural-based pulmonary lymphangioma as well as a fluorodeoxyglucose (FDG)-avid pleural lesion with this pathology.

Report of Case

A 38-year-old woman presented to the outpatient Pulmonary Clinic at the Penn State College of Medicine in Hershey, Pa, complaining of chronic pain in her upper arm and shoulder on the right side.

The patient reported that while she was at work 3 years earlier, a forklift trapped her right upper arm and shoulder against a wall, resulting in immediate and persistent numbness and tingling in the right hand. Initial surgical treatment included decompression of the right radial nerve and cervical spinal fusion of vertebrae C4 through C7. However, chronic, unrelenting pain persisted. The patient therefore sought neurosurgical consultation for possible brachial plexus surgery. The neurosurgeon ordered a thoracic CT scan, the results of which revealed a right apical pleural-based lesion.

The neurosurgeon referred the patient to a community pulmonary specialist, who monitored the lesion. In 1 year, the lesion grew from 1.5 cm × 1.2 cm to 1.6 cm × 1.4 cm. A positron emission tomography (PET) scan confirmed the presence of a right apical pleural-based nodule. The pulmonologist suspected a malignant primary lung mass and referred the patient to an osteopathic physician (M.G.B.) at the Penn State College of Medicine.
The patient denied fever, chills, night sweats, and weight loss. She also stated that she did not have chest pain, shortness of breath, cough, wheezing, or hemoptysis. She had no history of seasonal or perennial postnasal drip, rhinitis, sinusitis, asthma, bronchitis, or pneumonia. She had intermittent headaches but no history of seizures or skin lesions. The patient had no history of alcohol or drug abuse, and though she never smoked cigarettes, she had exposure to secondhand smoke from both parents throughout childhood. Before the work injury, which left her disabled, she had worked as a laborer in a battery factory for 6 years, and before that, in a fabric factory. The patient also reported that she had consistently received age-appropriate preventive health screening.

On physical examination, the patient’s blood pressure was 120/84 mm Hg; heart rate, 72 beats per minute; respiratory rate, 16 breaths per minute; and body mass index, 30. She appeared healthy and in no distress, with neither cervical nor axillary adenopathy. Cardiovascular examination revealed a regular heart rate without murmurs. Her lungs were clear to auscultation with normal percussion notes and no point tenderness with chest wall compression. Chest excursion and diaphragm descent were normal. The patient’s fingernails were normal, her fingers were not clubbed, and she had no peripheral edema or skin or joint lesions. The results of her neurologic examination were normal except for 4/5 muscle strength in right shoulder abduction and diminished sensation on the palmar aspect of the medial two fingers on her right hand.

The initial CT scan showed a focal area of nodular thickening measuring 60 Hounsfield units—similar to the density of muscle—located in the lateral right apical lung pleura. There were no other lung or pleural-based masses, no mediastinal, hilar, or axillary adenopathy, and no pleural effusion. The heart and great vessels were normal, as were the postsurgical changes from the spinal fusion.

A second CT scan 3 months later revealed an increase in lesion size. A PET scan taken immediately afterward showed a single 1 cm × 2 cm FDG-avid lesion at the same location. Three core biopsies under CT guidance yielded benign, CD31-positive tissue, which is consistent with lymphangioma. A CT scan with intravenous contrast administered to the right antecubital fossa showed no evidence of structural compression to right apical lymphatic flow. The patient returned for a third CT scan 3 months later, at which point the lymphangioma measured 1.3 cm × 2.2 cm between the lateral aspect of the second and third ribs on the right side. Minimal dependent bibasilar atelectasis was present. The bony structures were otherwise unremarkable, and no right shoulder mass was seen on the margin of the images.

The patient’s condition was presented to the multidisciplinary thoracic tumor conference group at the Penn State College of Medicine. The lymphangioma was extra-pulmonary and was determined to be an unlikely source of the patient’s chronic pain. Because no vital structures were obstructed, the lesion was not excised. However, as consensus dictated at the conference meeting, the patient continues to receive follow-up CT scans for serial observation of the lesion. Gabapentin, prescribed by the patient’s family physician, alleviated her chronic arm and shoulder pain, though the source of that pain was not found.

Discussion
Cases of lymphangioma have been reported in the form of isolated parenchymal lesions, chest wall lesions, and multiple cystic lesions throughout the thorax. However, lymphangiomas presenting as solitary pulmonary lesions are rare.

Likewise, lymphatic abnormalities are typically found in any region of the body where lymphatic drainage exists—most commonly, the head and neck, axilla, and abdomen. In the chest, such lesions are most frequently found in the mediastinum, accounting for up to 4.5% of mediastinal tumors. Of the adult patients who have solitary pulmonary lesions, most are asymptomatic. Other clinical features, such as cough and dyspnea, may be present if the patient’s vital structures are compromised.

Lymphatic dysplasia syndrome and lymphangioma are the two most common diseases in their class, with 90% of lymphangiomas occurring in children younger than 2 years. In the present case, the absence of radiographic pleural effusion excluded the presence of a chylothorax, therefore eliminating the possibility that the patient had lymphatic dysplasia syndrome. With lymphangiomas, CT scans indicate the location, size, and density of lesions, but they cannot establish the diagnosis. While the lesion in the present report measured 60 Hounsfield units, lymphangiomas are typically ~4 to 34 Hounsfield units and are smooth cystic masses. Spiculated lesions can also occur. The high density of the mass described in the present report was unlike any previous lymphangiomas found in the medical literature.

Magnetic resonance imaging (MRI) is considered the most precise modality for characterizing lesion tissue and for determining tumor extension, particularly in the case of lymphangiomas. Because the lesion in the present report was sufficiently delineated using a contrast medium with CT scans, an MRI was not ordered. However, a PET scan was ordered because the lesion appeared to be noncystic. The scan revealed that the lesion was FDG-avid, yet, to our knowledge, FDG avidity has not been exposed previously on the PET scans of lymphangiomas. While PET scans have been used to identify malignant lesions, malignant degeneration has not been reported. To identify areas of increased glucose metabolism, F-18 FDG PET scans are ideal. Nonmalignant FDG-avid lesions can occur in granulomatous diseases as well as in normal cardiac, renal, and gastrointestinal structures. However, the cellular source of the FDG-avid signal in the present report is unknown.
Because lymphangiomas may be difficult to differentiate radiographically from primary lung cancer, surgical wedge resection is often considered. In the present case, the lesion was not excised because the patient’s pain was unrelated to the presence of the lymphangioma and no vital structures were obstructed. However, in such cases, continued observation is necessary to monitor tumor growth. Sclerotherapy has also been reported as an alternative treatment option to alleviate vital structure compression.17

**Conclusion**

Lymphatic anomalies frequently mimic other pathologic processes, particularly neoplastic processes. The rare case presented in the current report illustrates the necessity of a broad differential diagnosis for focal pleural-based lesions as well as the potential for diagnosis through core biopsy to avoid

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In the past, lymphangiomas were distinguished from hemangiomas by the lack of intraluminal blood. However, core biopsies of lesion tissue using immunohistochemical and elastin staining (Figure 1) are more useful in identifying lymphangiomas from other types of lesions. Unlike other lesions and tumors, lymphangiomas are dilated, closely apposed, endothelium-lined vascular spaces that have adventitial connective tissue but lack a media.15 Congenital lymphangiomas are embryologic remnants of lymphatic tissue that failed to connect to efferent channels, while secondary lymphangiomas result from mechanical obstruction to existing lymphatics. Lymphatic-specific vascular endothelial growth factors C and D and molecular cell surface markers—such as the vascular endothelial growth factor 3 receptor—allow molecular characterization and exploration of trophic factors in the occurrence of these lesions.16

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**Figure 1.** Core biopsy of a pulmonary pleural-based lesion revealed benign tissue consistent with lymphangioma. The CD31 antibody immunohistochemical stains (A, magnification ×200, and B, ×400) reveal the specificity for platelet endothelial cell adhesion molecule, confirming the endothelial nature of this lesion. The elastin stain (C, ×200) highlights elastin protein in the walls of the vessels, suggesting the presence of a vascular structure with elastic tissue. The standard hematoxylin and eosin stain (D) is shown at ×100.
surgical resection. A thorough patient history and physical examination, along with proper imaging studies, tissue sampling, and expert consultation, are paramount in guiding the treatment of a patient with an unusual finding.

References

CASE REPORT

Figure 2. Computed tomography scan with intravenous contrast of a solitary pleural-based pulmonary lymphangioma (arrow) suggests that lymphatic flow was not impeded.

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