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.1 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.1 Mediastinal lymphangiomas comprise 10% while intrapulmonary lymphangiomas make up less than 1% of all lymphangiomas.2 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.3 Secondary forms develop in adults as a result of lymphatic channel obstruction caused by radiation, surgery, or infection.1

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.
right shoulder mass was seen on the margin of the images. The bony structures were otherwise unremarkable, and no changes from the spinal fusion.

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.

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...
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


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