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A Pathologically Benign, but Clinically Pathologic Neck and Mediastinal Lipoma

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Abstract

Lipomas are benign mesenchymal tumors that are generally found in the subcutaneous tissue but have been reported in virtually all organs throughout the body. We present a rare fibrolipoma of a 78-year-old female involving the neck and the mediastinum causing airway and cardiovascular compression. This patient underwent a staged resection, first addressing the neck component to relieve the airway compression, then addressing the intrathoracic portion via a right thoracotomy.

Introduction

Lipomas are benign, well-circumscribed, mesenchymal-derived tumors that are generally found in the subcutaneous tissue but have been reported in virtually all organs throughout the body. Only 13% are found in the head and neck, and even less in the mediastinum or chest. Lipomas are reported to represent only 1.6-2.3% of all mediastinal tumors. Although often slow growing and asymptomatic, at larger sizes they may contribute to symptoms of dyspnea, dysphagia, cardiac arrythmias, or even cause death from mass effect.

Clinical Summary

The patient is a 78-year-old female with a history of mediastinal tumor status post resection 20 years ago via a median sternotomy, who presented to the ED with dyspnea and cough. She was noted to have

a large left neck mass and within an hour of presentation was intubated for acute hypoxic respiratory failure. A CT scan (Figure 1, Figure 2) demonstrated 21x29 cm mass extending from the neck, through the thoracic inlet and displacing several mediastinal structures with extension to the diaphragm. There was tracheal and esophageal deviation to the right associated with partial airway obstruction. Although clinical and radiological findings were suspicious for a sarcoma, two biopsies confirmed fibrolipoma.

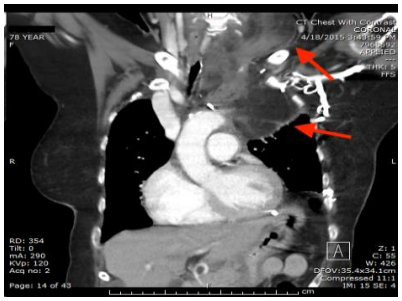

Figure 1: Mass extending from the neck, through the thoracic inlet (red arrow).

Figure 2: Mass within the thoracic cavity displacing several mediastinal structures with extension to the diaphragm (red arrow).

The patient was taken to the OR following resuscitation. She underwent a staged resection, first via a cervical approach, followed by transthoracic mediastinal debulking. Resection of the mass was performed via a left neck incision from the sternocleidomastoid to the thoracic inlet. The tumor measured 10x15cm and was noted to be firm and well-circumscribed. She tolerated the procedure well, was extubated on the fifth post-operative day and discharged on the ninth post-operative day.

The following month, she was taken for a mediastinal tumor debulking via a right thoracotomy. Due to her age, the lipoma's slow growth, the involvement of the aortic arch and its branches, we decided to limit the debulking to the right chest. The mass was well-circumscribed, and was dissected free from the esophagus, the azygous vein, the pericardium, and the diaphragm (Figure 3). She tolerated the procedure and was extubated on the second post-operative day. Her postoperative course was complicated by a chylothorax which improved with a low-fat diet. She was subsequently discharged home. The patient's six-month follow-up CT scan demonstrated a stable mass in the left chest measuring 13.7x13.2x11.4 cm. Now, six years later at the age of eighty-five, she continues to deny any dysphagia or dyspnea. The mass has since grown to 17.5x11.2x14 cm but the patient remains asymptomatic.

Figure 3: Resected mediastinal fibrolipoma.

Discussion

Lipomas are benign, well-circumscribed, mesenchymally-derived tumors. They are most commonly found in the subcutaneous soft tissue; however, they can be found almost anywhere throughout the body. Only 13% are found in the head and neck, and even less in the mediastinum. Lipomas are reported to represent only 1.6-2.3% of all mediastinal tumors [1]. Within the thorax, several types have been described, originating from the mediastinum, diaphragm, pleura and even within the bronchi. We present a unique case of mediastinal fibrolipoma. Histologically, lipomas are characterized by encapsulated, mature adipocytes varying slightly in size. However, there are multiple lipoma variants described, including fibrolipoma. Like lipomas, fibrolipomas histologically demonstrate mature adipocytes and a lack of atypical cells, but must have focally increased fibrous tissue [2]. Clinically, the lipoma and fibrolipoma expand and displace tissue, rather than infiltrate it, as is seen with liposarcomas [3].

There is but one fibrolipoma described in the English literature that is not primarily associated with the esophagus [4]. Our case report represents the second. Based on a classification system described as early as the 1930s, thoracic lipomas can be: (a) hourglass or dumbbell lipomas that pass through the intercostal space or thoracic inlet, or (b) purely intrathoracic lipomas [5-7]. We present a case of the former.

Lipomas are often slow growing and asymptomatic, however at larger sizes they may cause dyspnea, dysphagia, and cardiac arrhythmia, or even cardiac arrest [1,6-9]. Our patient presented with a neck mass and respiratory distress, requiring endotracheal intubation.

Chen et al. reported a patient with dyspnea secondary to right lung collapse as a result from mass effect due to a giant pleural lipoma [9]. Also, Jack et al. reported a case of a pericardial lipoma causing severe left ventricular dysfunction [8]. The patient refused surgery and subsequently suffered cardiac arrest due to the tumor's compression of the heart.

On CT imaging, lipomas are well-defined masses composed of homogenous fat (-50 to -150 HU) and demonstrate an obtuse angle with the chest wall,often displacing adjacent pulmonary parenchyma and vasculature. Infiltration of surrounding structures rather than displacement, a heterogenous architecture, or attenuation values greater than -50 HU should raise suspicion for liposarcoma [10].

However, computed tomography alone is insufficient in differentiating between lipoma and liposarcoma [1].

Fine-needle aspiration can aid in the diagnosis of an intrathoracic mass; however, it may be difficult to differentiate between lipomas and well-differentiated liposarcoma based on such a sample [11]. In our case, the more aggressive liposarcoma was ruled out on fine-needle aspiration analysis using fluorescence in-situ hybridization (FISH) analysis of MDM-2, which plays a role as a negative regulator of the p53 tumor suppressor. This patient's FISH analysis was negative for MDM-2 gene amplification.

A management algorithm of intrathoracic lipomas has not been well-established. Some authors recommend observation of asymptomatic intrathoracic lipomas with serial imaging [12,13]. However, others recommend surgical excision because of symptoms or risk for mass effect on intrathoracic structures, as well as because preoperative differentiation between lipomas and liposarcomas is very difficult and not well established [14,15]. Due to the uncertainty regarding tumor histology and the risk for recurrence, formal resection of large intrathoracic lipomas should be attempted, but when unable, tumor excision should be performed to the greatest extent possible without causing undue morbidity and mortality.

Outcomes are typically good following resection of intrathoracic lipomas. They may recur locally at a rate of <5%; however, deep-seated intra-thoracic lipomas tend to have a higher recurrence rate due to the difficulty associated with performing a formal resection of the tumor in its entirety [11]. As with our patient, the clinical scenario may preclude a formal resection of the tumor. Our patient's advanced age and the tumor-vessel relationship with the innominate and left common carotid artery increased the risk of obtaining a formal resection outweighing its benefits, for this benign, slow-growing tumor. The goal of her operation was primarily to reduce the mass effect of the fibrolipoma in order to allow for improved pulmonary physiology to facilitate extubation.

There are multiple case reports describing partial lipoma excisions when formal resection would have resulted in undue morbidity or mortality, such as a case with tumor involving the brachial plexus [11]. Interestingly, there are several cases reported by Wurlitzer et al. in which the tumor ceased further growth following a partial excision [16].

Conclusion

Our case is only the second in the English literature to describe an intrathoracic fibrolipoma not associated with the esophagus. While intrathoracic lipomas and fibrolipomas are benign tumors, surgery may be indicated due to compressive symptoms and potential airway or hemodynamic compromise. While we were able to obtain a tissue diagnosis of fibrolipoma pre-operatively, the inability to distinguish a lipoma or lipoma subtype from a liposarcoma in other cases may be an indication for surgery. Formal resection is preferable to reduce risk of recurrence, however partial excision can be considered based on the clinical scenario to reduce risk of undue long-term morbidity or mortality.

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