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Case Report

Ectopic Intrathoracic Liver Mimicking Pulmonary Nodules

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Abstract

A 48-year-old female with a remote history of pneumothorax and uterine fibroids was referred for further evaluation of pulmonary nodules suspicious for metastatic lesions. Abdominal computed tomography revealed an enlarged heterogeneous uterus and numerous right lower lobe pulmonary nodules raising suspicion for metastatic leiomyosarcoma. Right thoracoscopy with biopsy revealed the lungs to be normal, however ectopic hepatic tissue was found protruding through the diaphragm. Three months later the patient developed a recurrent pneumothorax requiring repeat thoracoscopy with pleurodesis and repair of multiple diaphragmatic defects. This case report discusses the diagnostic and treatment considerations of patients with ectopic intrathoracic tissue associated with diaphragmatic defects. In particular, patients who are found to have lower lung field nodules in association with history of pneumothorax should raise suspicion for diaphragmatic defects and ectopic liver.

Keywords: *Ectopic Liver; Accessory Liver; Thorax; Lung Nodule; Supradiaphragmatic Liver*

Introduction

Unlike catamenial pneumothorax in which ectopic endometrial tissue is associated with pneumothorax, intra-thoracic ectopic liver tissue has not previously been reported to be associated with pneumothorax. Ectopic liver tissue is a rare finding and in prior case reports of ectopic intrathoracic tissue, the abnormally positioned tissue is most commonly found attached to various intra-abdominal structures such as the spleen, omentum, and retroperitoneum by mesenteries or stalks [1]. In our case, ectopic hepatic tissue protruding through the diaphragm was seen in association with recurrent pneumothorax and multiple diaphragmatic pores. The course of our case, taken together with previous reports of recurrent pneumothorax not associated with ectopic tissue, suggests that diaphragmatic fenestration alone may be sufficient to contribute to the development of pneumothorax.

Case Report

A non-smoker, 48-year-old African American female with a history of uterine fibroids and anemia was initially evaluated for a large symptomatic fibroid. Abdominal CT scan revealed an enlarged heterogeneous uterus extending into the mid-abdomen as well as several pulmonary nodules clustered in the right lower lobe suspicious for metastatic lesions.

At the time of presentation, the patient denied pulmonary symptoms. She also reported a history of right-sided spontaneous pneumothorax one year prior to the current evaluation treated with chest tube placement. Past medical history was otherwise unremarkable.

Physical examination as well as laboratory results were unremarkable. CT scan of the chest revealed several pulmonary nodules clustered in the right lower lobe. The nodules were located fairly peripherally with the largest being 1.5 cm in size (Figure 1).

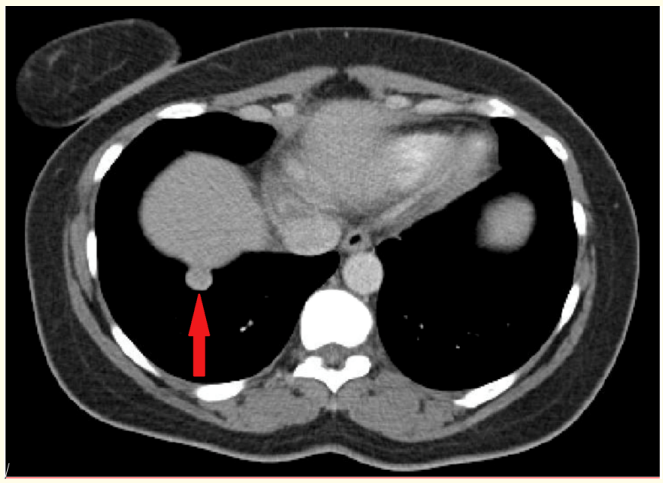

Figure 1: *CT scan of the chest demonstrating the appearance of a right lower lobe lung nodule (arrow).*

Based on the patient's radiological appearance and in the setting of her large fibroid uterus, the pulmonary nodules were felt to possibly represent benign metastasizing leiomyomas vs. metastatic leiomyosarcomas. She was presented in multidisciplinary tumor board and it was recommended she undergo diagnostic right thoracoscopy with biopsy.

Intraoperatively, her right hemidiaphragm was elevated. Additionally, there appeared to be implants along the diaphragmatic surface and a more subtle implant along the chest wall inferiorly. Upon further inspection it appeared that the implants on the diaphragmatic surface were ectopic hepatic tissue that had grown through Swiss cheese-like defects in the diaphragm in concordance with the CT findings of round nodules (Figure 2). Biopsy of the diaphragmatic implants revealed normal liver. A diagnostic wedge biopsy of the right lower lobe revealed normal lung tissue while the diaphragmatic biopsies were reported as partially encapsulated portions of liver tissue consistent with intrathoracic ectopic liver.

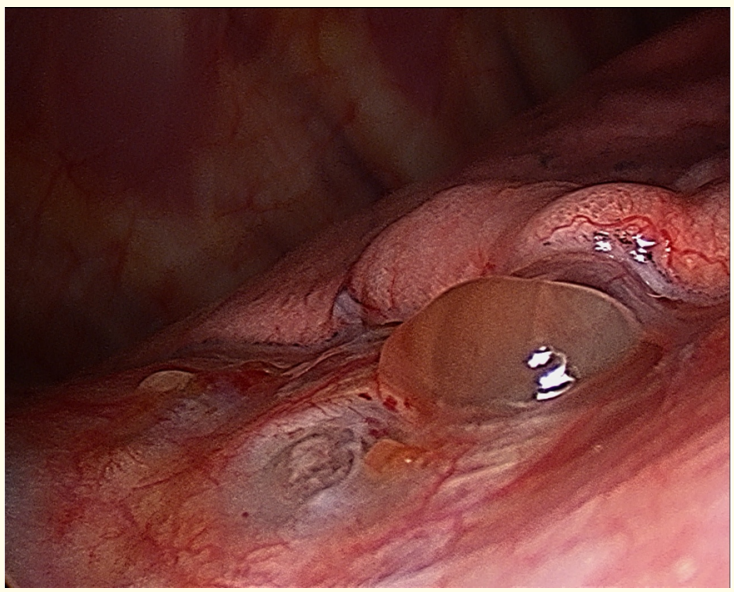

Figure 2: *Intraoperative findings of liver tissue piercing through defects in the diaphragm.*

The patient's postoperative course was uneventful; however, two months following surgery the patient presented with her second right pneumothorax. On this admission, given knowledge of her prior surgery findings, the patient was scheduled for right thoracoscopy, diaphragmatic plication, and talc pleurodesis. Intraoperatively, again numerous defects were noted in the diaphragm that were closed primarily together with diaphragmatic plication. The patient's postoperative course was uneventful, and she was discharged on postoperative day two. No pneumothorax was reported during 3 years follow-up.

Discussion

This case illustrates a rare presentation of ectopic intrathoracic hepatic tissue associated with recurrent spontaneous pneumothorax. Two aspects of the case are worthy of discussion: the CT appearance of the ectopic intra-thoracic liver mimicking pulmonary nodules, and the association of multiple diaphragmatic defects with spontaneous pneumothorax.

While in most patients the ectopic liver tissue has been present since birth there are few reports of intrathoracic liver due to iatrogenic causes such as diaphragmatic hernia repair [2] or following trauma [3]. In our patient's case, there was no prior history of trauma or lung infection; we therefore feel it is likely that the diaphragmatic defects were congenital rather than acquired. Collan., *et al.* [4] classify ectopic liver tissue into four main types: 1. A connecting stalk attached to an accessory liver lobe that reaches a considerable size, 2. A Small accessory liver lobe that is attached to the liver but is smaller than the first type, 3. Abnormally positioned liver, which is situated outside the liver without any connection to it, 4. Ectopic liver tissue, which is evident only microscopically. While the majority of case reports illustrating ectopic liver tissue are classified as Type 2 according to the Collan classification, our case is classified as Type 3 as the liver tissue was found on the thoracic aspect of the diaphragm without clear evidence of hepatic attachments. The abnormally positioned liver found in our case may be explained by the patient's large fibroid uterus and its pressure effect on other abdominal organs. It is possible that the uterus displaced the liver and in so doing allowed ectopic liver to protrude through the several diaphragmatic defects.

Another unique aspect of our case is the clinical presentation of recurrent pneumothorax. Our review of the literature indicates two possible explanations for pneumothorax in association with diaphragmatic defects and ectopic tissue. The first entity is catamenial pneumothorax, a condition in which pneumothorax occurs in conjunction with menstruation - this is thought to be a result of presence of endometriosis implants on the visceral pleura [5]. Among the several possible etiologies of catamenial pneumothorax, transdiaphragmatic passage of air through congenital or (more frequently) an acquired diaphragmatic defect is one of the most common causes. Although our patient did not have endometriosis, the small diaphragmatic defects seen in our case are similar to those described in the literature in patients with catamenial pneumothorax. Indeed, a report of recurrent pneumothorax in a woman without endometriosis but with multiple diaphragmatic fenestrations supports the hypothesis that diaphragmatic defects alone are sufficient to cause recurrent pneumothorax [6].

The second clinical syndrome that offers an explanation for pneumothorax associated with diaphragmatic defects is a group of symptoms known as porous diaphragm syndromes. Kirschner [7] attributes the trans-diaphragmatic passage of gases, fluids and other substances to small defects in the diaphragm, and he groups the resulting clinical presentations into porous diaphragm syndromes based on the substance that traverses the diaphragm. For instance, in cirrhotic patients, Kirschner writes that the diaphragmatic pores permit the egress of ascitic fluid from the abdomen into the ipsilateral hemithoracic pleural space, resulting in a cirrhotic hydrothorax [8]. According to this theory, the pneumothoraces in our patient could be explained by intraabdominal gases traversing defects in the diaphragm.

Conclusion

In patients with recurrent pneumothorax, occult diaphragmatic defects with or without associated ectopic liver tissue should be considered in the differential diagnosis. Abnormally positioned tissue at the lung base that has the appearance of lung nodules may in fact be ectopic intrathoracic liver. Therefore, during operative intervention for recurrent pneumothorax of unknown etiology, the surgical approach should consist of a diligent search for diaphragmatic fenestrations.

Conflict of Interest

There are no conflicts of interest.

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