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Case Report

Incidental finding of intrathoracic caudate lobe of the liver associated with an arterovenous malformation ☆,☆☆

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ABSTRACT

In this case report we describe an occasional finding of intrathoracic caudate lobe protruding through the diaphragm foramen in a 73-year-old woman who came to our attention for a screening for coronary artery disease, due to the presence of cardiovascular risk. The patient had no symptoms. The computed tomography showed, a circumscribed homogeneous soft tissue mass that protruding through the aortic diaphragmatic foramen that was as isodense as the liver. Moreover was revealed an abnormal artery emerging from the celiac tripod which, through the diaphragmatic foramen, ends in an inferior pulmonary vein creating an arteriovenous malformation.

Introduction

The finding of an ectopic supradiaphragmatic liver is an extremely rare anomaly and is part of the differential diagnosis of a mediastinal mass, often diagnosed as an incidental finding during examinations for other clinical reasons. The first supradiaphragmatic ectopic liver was descripted by Hansborough and Lipin in 1957 [1] and, from then, only 30 cases have been reported in the literature [2–5].

Abnormal location of liver tissue is typically found in abdominal organs such as the gallbladder, spleen, adrenal gland and pancreas. Intrathoracic liver tissue is rare and may occur due to congenital abnormalities or after trauma or a surgical procedure [3].

In most cases it is asymptomatic and, if present, symptoms are variable and non-specific, such as cough, difficulty breathing, chest or abdominal pain. An extensive diagnostic workup usually follows this incidental finding, starting with chest X-ray, chest CT, CT angiography, magnetic resonance imaging

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(MRI) and possibly invasive procedures (usually video-assisted thoracic surgery for biopsy) [4].

Differential diagnoses often include esophageal cysts, lung tumors or mediastinal lesions. A correlation between this finding and pulmonary sequestration is described in the literature, although the etiology of an intrathoracic liver coinciding with pulmonary sequestration is not well known [2].

Adin et al. developed a classification of ectopic liver formations based on morphology, location and relationship to the liver assessed by imaging, recognizing 3 classes. Class I: herniated liver tissue through a diaphragmatic defect; Class II: supradiaphragmatic liver mass connected to the liver through a vasculobiliary pedicle; Class III: ectopic liver tissue separated from the liver, generally more distant [6].

Here we describe an asymptomatic case of an intrathoracic caudate lobe protruding through the diaphragm, incidentally detected during a coronary computed tomography angiography (CCTA) for screening purposes. The patient then underwent a total body CT scan on suspicion of a mediastinal mass.

Case report

A 73-year-old woman presented to our department for coronary angiography with computed tomography. The patient had no symptoms and had the CT scan as a screening for coronary artery disease (CAD), due to the presence of cardiovascular risk factors such as family history, hypertension and hypercholesterolemia.

In her remote pathological history, the patient reported that she had undergone transcatheter foam sclerotherapy surgery for pelvic varicocele at our department in 2007 [7,8].

She also reported suffering from Raynaud's syndrome since childhood, for which she does not receive any specific therapy [9]. The patient denied any previous history of trauma or other abdominal surgery. She performed an abdominal ultrasound in the past in which the only pathological finding was the presence of uncomplicated cholelithiasis.

A coronary computed tomography (CT) angiography with contrast medium was performed, which revealed the presence of some calcific and mixed coronary plaques that do not lead to significant CAD [10], but showed, as a collateral finding, a circumscribed homogeneous soft tissue mass in the right para-mediastinal area in the lower middle mediastinum (Fig. 1).

Consequently, with the patient's consent, a total-body CT scan was performed to characterize the finding.

A non-enhanced scan and a series of dynamic scans during arterial phase, portal phase and a late phase showing enhancement pattern of the lesion, which revealed a 3,5 cm solid tissue formation, protruding through the aortic diaphragmatic foramen, connected to the abdominal liver by a sub-centimetric peduncle and the absence of a recognizable caudate lobe. This intrapulmonary mass was as isodense as the liver (Fig. 2).

In addition, CT showed an abnormal artery emerging from the celiac tripod which runs through the diaphragmatic foramen and ends in an inferior pulmonary vein, thus creating an arteriovenous malformation (Fig. 3).

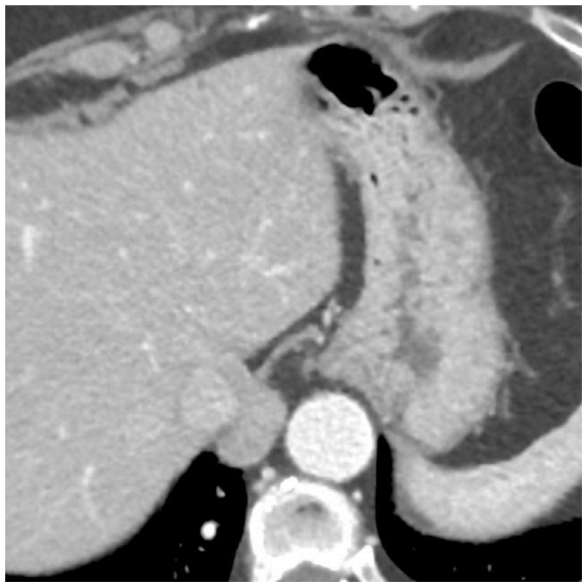

Fig. 1 – A circumscribed homogeneous soft tissue mass in the right para-mediastinal area in the lower middle mediastinum accidentally finding during a coronary computed tomography angiography (CCTA).

Discussion

The occurrence of ectopic liver tissue is extremely rare (incidence less than 0.5 per cent), especially in supradiaphragmatic locations [5].

Most frequently described within the abdominal cavity, ectopic liver tissue usually occurs as accessory lobes or nodules adherent to intra-abdominal organs and structures, such as the gallbladder, spleen, pancreas, adrenals, peritoneum, esophagus, and ligaments, usually connected to the liver by connective tissue [11,12].

In contrast, intrathoracic presentation of liver tissue is exceptionally rare and is mostly found in the right hemithorax [2].

Only 30 cases of intrathoracic liver tissue have been reported in the literature since 1957, 2 of which were associated with pulmonary sequestration [2,5].

The etiology of intrathoracic ectopic liver tissue is uncertain. Two main mechanisms have been hypothesized: an embryogenetic origin, due to abnormal migration of liver tissue into the thoracic cavity before complete closure of the diaphragmatic membranes, or an iatrogenic or post-traumatic origin, after surgery on the diaphragm or high-impact trauma [3,4].

The mass may present as a separate formation from the remaining liver, taking the name 'ectopic liver nodule', or with a pedicle connected to the orthotopic liver that crosses the diaphragm or passes through a hiatus [2,13].

Adin et al. developed a classification of ectopic liver formations based on morphology, location and relationship to the liver assessed by imaging, recognizing 3 classes.

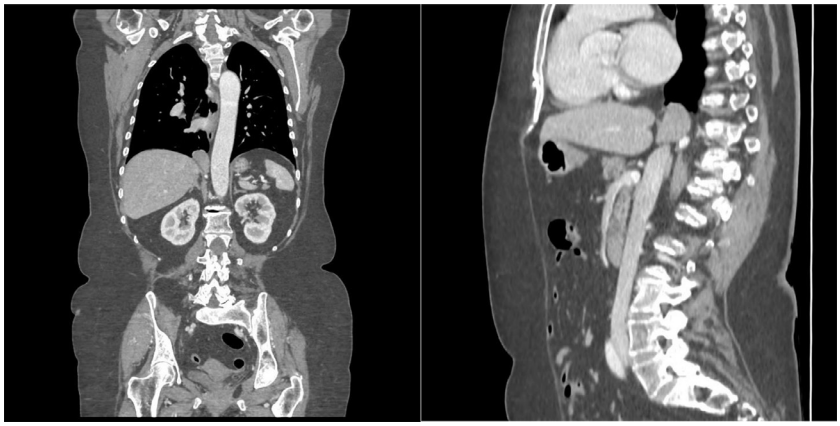

Fig. 2 – (A, B) An intrapulmonary mass as isodense as the liver protruding through the aortic diaphragmatic foramen, connected to the abdominal liver by a sub-centimetric peduncle.

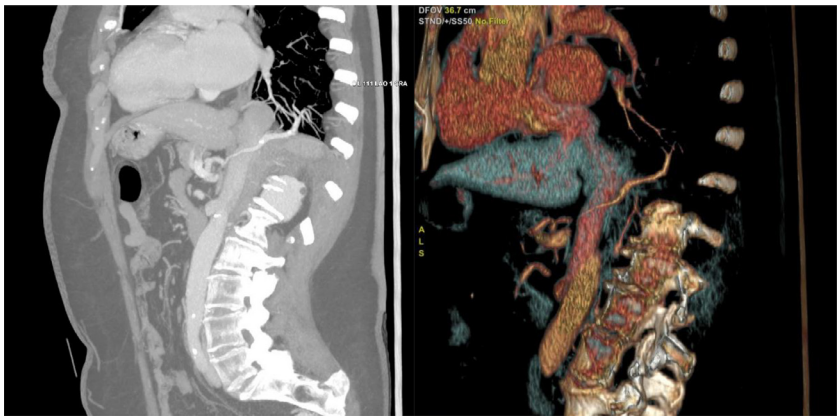

Fig. 3 – An arteriovenous malformation between an abnormal artery emerging from the celiac tripod and inferior pulmonary vein.

Class I: herniated liver tissue through a diaphragmatic defect; Class II: supradiaphragmatic liver mass connected to the liver through a vasculobiliary pedicle; Class III: ectopic liver tissue separated from the liver, generally more distant [6].

The most frequent symptoms are cough, dyspnea, chest pain, dyspepsia, digestive difficulties and hemoptysis, although in most cases the patient is asymptomatic [2,14].

Consequently, diagnosis is often occasional and relies on advanced imaging.

Ultrasonography is often negative, as in our case, while chest X-ray may show the presence of a radio-opaque mass, better evident in the lateral projection. A differential diagnosis should then be made with posterior pleural, pulmonary and mediastinal masses such as lymphomas, neurogenic tumors, benign fibrous tumors, pericardial, or esophageal neoplasms [15,16].

In most cases, 3D CT with contrast of the thorax and abdomen allows adequate characterization of the finding, defining its densitometric features and relationships with the diaphragm and adjacent structures such as the pleura, lung parenchyma and mediastinum. It is also necessary to study the vascular relationships with the portal, systemic and pulmonary circulation [4].

In addition, contrast-enhanced MRI can be a useful tool to diagnose liver tissue in doubtful cases, due to the possibility of administering gadolinium-based liver-specific contrast agents (HBAs) [17].

In our case, the diagnosis was incidental, thanks to the performance of a CCTA in an asymptomatic patient. The diagnostic procedure was continued with a thoraco-abdominal triphasic CT scan, which allowed us to determine the normal hepatic nature of the supradiaphragmatic mass, recognizing it as an ectopic caudate lobe connected to the liver by a tissue and vasculobiliary pedicle running through the aortic diaphragmatic foramen, adjacent to the right diaphragmatic pillar (class II from Adin et al. [6]). CT showed the absence of compressive effects of the mass on the aorta and other mediastinal structures and the absence of pulmonary sequestration or adjacent atelectatic lung parenchyma. These details, in combination with the absence of symptoms and a negative history of trauma or previous surgery, suggested a congenital origin of the abnormality and therefore, in agreement with the patient,

no further diagnostic or interventional procedures were performed, but a distant follow-up was opted for.

CT images also showed a peculiar vascular abnormality associated with the finding, not previously described in cases of supradiaphragmatic liver tissue, namely the presence of an accessory hepatic artery from the celiac tripod, which runs adjacent to the pedicle of the ectopic caudate lobe through the diaphragm. The vessel provides some arterial branches to the caudate lobe and has an abnormal drainage into the ectasic pulmonary venous branch for the postero-basal segment of the right lower lobe, resulting in a small arteriovenous malformation (AVM) at that level, which is not clinically significant. Follow-up will also be useful to assess the evolution of this finding when it is first encountered.

In conclusion, for the diagnosis of supradiaphragmatic hepatic ectopic tissue, a correct diagnostic framing with imaging methods, particularly CT with contrast medium, is essential to avoid further invasive procedures, especially in cases of asymptomatic patients who do not require surgery [3,18].

Furthermore, CT is also essential in the follow-up of such findings to exclude evolution towards liver disease, such as cirrhosis or neoplasms, or mechanical events such as pedicle stenosis or torsion [19,20].

Ethics human rights

The authors declare that the work described has been carried out following the Declaration of Helsinki of the World Medical Association revised in 2013 for experiments involving humans.

Author contributions

All authors attest that they meet the current International Committee of Medical Journal Editors (ICMJE) criteria for Authorship.

Availability of data and material

Not applicable.

Patient consent

The authors declare that this report does not contain any personal information that could lead to the identification of the patient. Informed consent was obtained from the patient.

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