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Case Report

Heterotopic supradiaphragmatic liver tissue mimicking a thoracic tumor: a diagnostic dilemma and VATS management

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ABSTRACT

Heterotopic supradiaphragmatic liver tissue (HSLT) is an extremely rare condition, with about 30 cases documented in the literature since its first description. Because it often shows no symptoms and looks similar to pleural or parenchymal tumors on imaging, HSLT poses a significant diagnostic challenge. Although the cause remains unknown, a definitive diagnosis usually requires surgery. While conservative treatment is suitable for clinically confirmed cases, surgical exploration becomes necessary when malignancy cannot be ruled out. In such cases, the videothoracoscopic approach (VATS) is considered the gold standard for both diagnosis and treatment. Here, we report a case of HSLT discovered incidentally during imaging for acute appendicitis. We review the diagnostic difficulties and recommend VATS as the primary surgical option for unclear lesions above the diaphragm.

Keywords: choristoma, liver, thoracic surgery, video-assisted, diaphragm, incidental finding, diagnosis, differential

Introduction

The incidence of ectopic liver tissue is less than 0.5%, with the majority of cases detected in the abdomen [1]. Supradiaphragmatic presentation is exceptionally rare; since the first description by Hansbrough and Lipin [2], approximately 30 cases have been reported across various age groups. Supradiaphragmatic liver tissue is classified as an accessory liver lobe if connected to the liver parenchyma, or as ectopic (heterotopic) liver if no such connection exists [3]. Diagnosis is challenging due to the asymptomatic nature of the condition, leading to frequent incidental detection [4]. Herein, we present a case of heterotopic supradiaphragmatic liver tissue (HSLT) unconnected to the liver, identified incidentally during surgery for acute appendicitis.

Case Report

A 33-year-old non-smoker male presented with right flank pain. His medical history was significant for an appendectomy performed eight months prior, during which a thoracic lesion was incidentally detected but not surgically addressed. He also had a history of multinodular goiter and thyroiditis and was not on any active medication. Physical examination and laboratory findings were unremarkable.

Follow-up thoracic computed tomography (CT) confirmed a 12 mm nodular lesion in the lateral basal segment of the right lower lobe (Figures 1a,b). To further characterize the lesion, Positron Emission Tomography/Computed Tomography (PET/CT) was performed, which revealed a diaphragmatic pleural-based, smoothly circumscribed soft tissue nodule measuring 14 x 12 mm with a SUVmax of 2.2. Due to the suspicion of a pleural or parenchymal tumor, video-assisted thoracoscopic surgery (VATS) was planned. Surgical exploration revealed a purple-blue nodular lesion, approximately 15 mm in length, located on the diaphragm with a broad base and distinct from the lung parenchyma. The lesion was completely excised from the diaphragmatic pleura using an ultrasonic energy device. The postoperative course was uneventful, and the histopathological examination confirmed the diagnosis of heterotopic supradiaphragmatic liver tissue (HSLT) (Figures 2a-d). Written informed consent was obtained from the patient for publication of this case report and any accompanying images.

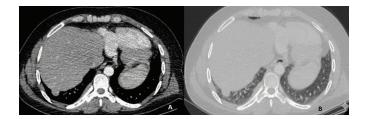


Figure 1. Axial chest CT scans demonstrating a 12mm nodular lesion located in the lateral basal segment of the right lower lobe. Mediastinal window view (**A**), lung parenchymal window view (**B**).

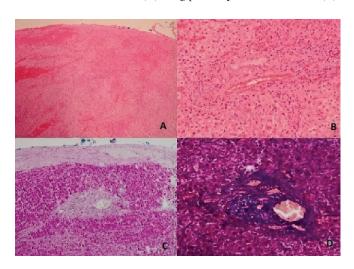


Figure 2. Histopathological evaluation of the resected specimen. Liver parenchyma exhibiting hemorrhagic changes encapsulated by fibrous tissue (H&E, x4) (**A**), portal areas showing minimal inflammatory infiltration with accompanying hydropic degeneration in the adjacent parenchyma (H&E, x20) (**B**), Periodic Acid-Schiff staining demonstrating positive hepatocellular glycogen accumulation (x10) (**C**), Masson's Trichrome staining revealing mild fibrous expansion in select portal areas, corresponding to an ISHAK fibrosis score of 1 (x20) (**D**).

Discussion

Heterotopic supradiaphragmatic liver tissue (HSLT) is an exceptionally rare entity with an obscure etiology. While often attributed to congenital abnormalities, instances secondary to trauma or surgical intervention have also been reported [5]. Clinically, its presentation varies by localization, frequently mimicking intrathoracic pathologies such as pleural or parenchymal tumors, pulmonary sequestration, or neurogenic tumors [6]. In the present case, the patient lacked any history of congenital disease, trauma, or relevant thoracic surgery. Although an appendectomy was noted in the patient's history, it was performed subsequent to the lesion's detection and was therefore unrelated. While HSLT can

be diagnostically challenging to distinguish from liver herniation, we definitively excluded herniation upon thoracoscopic exploration, which revealed a fully intact diaphragm and no evidence of traumatic sequelae.

The diagnosis of HSLT is complicated by the distinct absence of specific clinical symptoms. While frequently detected incidentally, the principal diagnostic challenge lies in establishing an accurate preoperative identification [7]. Radiological imaging remains the cornerstone of diagnosis. Although CT and PET/CT are instrumental in differentiating benign from malignant tumoral lesions, Magnetic Resonance Imaging (MRI) serves as a valuable adjunctive modality specifically for confirming the hepatic nature of the tissue [8]. In the present case, the patient was asymptomatic with no significant etiological history. The lesion was identified incidentally during imaging for acute appendicitis and was subsequently evaluated via PET/CT to rule out malignancy.

While existing literature documents the potential for HSLT to undergo malignant transformation to hepatocellular carcinoma or degenerate into cirrhosis, conservative management is generally preferred for asymptomatic patients with a definitive preoperative diagnosis. However, the radiological mimicry of HSLT to benign intrathoracic neoplasms often precipitates unwarranted surgical interventions [9]. Conversely, when preoperative diagnostic uncertainty persists, surgical exploration becomes imperative. In the present case, we proceeded with videothoracoscopic exploration and total excision, as the supradiaphragmatic lesion could not be definitively distinguished from potential diaphragmatic or parenchymal malignancies.

When surgical intervention is indicated, the videothoracoscopic approach (VATS) represents the gold standard. Despite the significant diagnostic challenges posed by the rarity and asymptomatic nature of HSLT, it must be considered in the differential diagnosis of supradiaphragmatic lesions. Therefore, rather than avoiding intervention, we advocate for the utilization of minimally invasive videothoracoscopic exploration to ensure accurate diagnosis and definitive management when uncertainty remains.

Declaration of conflicting interests

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Authors' contribution

HY; organized the article and wrote the paper, HY,FY; contributed to the data collection, HY,FY,FH; revised the article. All authors revised the manuscript. The authors read and approved the final manuscript.

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