An Incidental Discovery of the Intrathoracic Accessory Liver Lobe in a 72-Year-Old Man: Case Report and Literature Review

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Abstract: Accessory liver lobe is a rare finding, with the most common cases being accessory liver tissue on the gallbladder wall separate from the orthotopic liver. As the incidence of the ectopic liver is low there are only several case reports in published literature that describe similar presentations. We report a case of intrathoracic liver lobe that was connected to the main liver by a thick pedicle. Due to benign presentation, the patient was discharged without any surgical intervention. This case highlights the importance of understanding anatomical variability of internal organs, understanding the risks of torsion and malignant transformation of the accessory liver tissue. The literature review provides an excellent overview of published case series and reports, and outlines current recommendations on imaging, diagnosis, and management.

Keywords: accessory liver lobe; intrathoracic liver; liver/abnormalities; liver/surgery; HCC; liver malignancy

1. Introduction

Intrathoracic accessory liver tissue is a rare anatomical variation characterized by the presence of ectopic liver tissue within the thoracic cavity. Normally, the liver is in the upper right quadrant of the abdominal cavity, but in some cases, small fragments of liver tissue can develop in unusual locations, such as the chest. This condition can be congenital or acquired, and its precise cause is still not fully understood. Due to the rarity of the condition, the clinical significance of intrathoracic accessory liver tissue is poorly understood. However, in some cases, the presence of ectopic liver tissue in the thoracic cavity can cause symptoms due to the compression on nearby organs. In these cases, the presenting symptoms can be cough, shortness of breath, chest or back pain, indigestion, or dysphagia. Occasionally, accessory liver can transform into hepatocellular carcinoma that requires prompt treatment, therefore, the involvement of the multidisciplinary team is paramount. Diagnosing intrathoracic accessory liver tissue usually requires a combination of imaging techniques, such as X-rays, computed tomography (CT) scans, or magnetic resonance imaging (MRI). Additionally, a biopsy may be necessary to confirm the presence of liver tissue and rule out other conditions such as lung cancer, mesothelioma, neurogenic tumor, thymoma, or hydatid cyst.

The treatment for intrathoracic accessory liver tissue depends on the symptoms and underlying causes. In asymptomatic cases, no intervention may be required, and regular monitoring may be sufficient. However, if symptoms such as pain, respiratory distress, or gastrointestinal issues occur, surgical removal of the ectopic liver tissue may be recommended. Special care needs to be taken should malignancy be suspected.

While intrathoracic accessory liver tissue is an uncommon condition, this case report highlights the remarkable complexity and variability of human anatomy. Further research...
is needed to better understand the underlying mechanisms of accessory liver lobe and ectopic liver tissue formation and clinical implications of this condition to enhance medical care and patient outcomes.

2. Detailed Case Description

An otherwise healthy 72-year-old man presented to the hospital after sustaining a fall. He had no other comorbidities; his past medical history was significant for tonsillectomy at the age of 12. He was unable to bear weight on his left ankle and was admitted under orthopedics for further assessment. He became febrile to 38.4 on day three of admission. The chest X-ray revealed a right lower lobe chest mass. Laboratory studies revealed white blood cells of $7.4 \times 10^9/L$, platelets of $647 \times 10^9/L$ and hemoglobin of 105 g/L, his biochemistry was unremarkable. C-reactive protein was 30 mg/L. Physical examination did not demonstrate any abnormality, there was no palpable mass in the right upper quadrant, and chest examination was normal with equal air entry bilaterally. The neurological exam was normal.

Computer tomography of the chest showed a well circumscribed lesion that appears to arise from the liver through a defect in the diaphragm, Figure 1a,b. The lesion is supplied by the branch of middle hepatic artery and a draining branch of middle hepatic vein. Based on these imaging findings; the patient was diagnosed with an accessory transthoracic liver lobe. As imaging findings did not suggest underlying malignancy and the alpha-fetoprotein level was 1 kIU/L a conservative course of action was deemed appropriate with regular cross-sectional imaging and specialist follow-up.

![Figure 1. (a) Coronal view of CT chest. There is a circumscribed lesion measuring 41 mm × 19 mm in the right hemithorax that extends through a defect in the diaphragm measuring 36 mm. The lesion arises from the liver and is supplied by the branch of the middle hepatic artery and a draining branch of middle hepatic vein. (b) Axial view chest CT demonstrating the lesion.](image)

The patient remained stable throughout admission. The source of the fever was not apparent, he was prescribed a short course of oral antibiotics to ensure coverage for hospital acquired pneumonia. His condition stabilized; he was discharged after physiotherapy clearance on day five of admission. He was advised to follow up with an upper gastrointestinal surgeon in six months and with his family doctor. To the best of our knowledge, the patient did not present to any other health facility and remained in good health.

3. Discussion

Both intrathoracic and intrabdominal accessory liver lobes (ALLs) have been described in the literature. ALLs are often undiagnosed and frequently discovered incidentally during
surgery or autopsy. Patients with an intrabdominal ALL occasionally complain of sudden abdominal pain, chest pain, nausea, or vomiting. The symptoms are frequently due to the compression of nearby structures such as biliary ducts, the stomach or duodenum [1]. More severe symptoms are caused by complications such as torsion, traumatic rupture, or infarction [2,3]. Torsion is the most common and serious complication [1,3]. It has been extensively described in several case reports [1,2]. Patients with torsion of an ALL typically experience severe stomach pain due to hemadostenosis, vascular obstruction, ischemia, putrescence, or even rupture and bleeding. ALLs can also be associated with congenital biliary atresia, congenital diaphragmatic defects, and angiocavernoma [4,5].

There are two ways in which liver tissue can be abnormally located in the thoracic cavity. When these lesions are connected by a pedicle to the orthotopic liver they are classified as ALL. Alternatively, when the tissue is located in the thoracic cavity independently it is referred to as ectopic liver tissue. While these conditions typically do not cause any symptoms, in some cases they may lead to chest pain, coughing, hemoptysis, dyspnea, or back pain [6]. As the tumor can compress nearby organs or interfere with their normal function it can cause gastrointestinal symptoms, for example reflux and dysphagia [7].

Commonly paravertebral lesions are discovered alongside ALLs. Depending on its location, an ALL may be mistaken for a pulmonary tumor, pleural tumor, pulmonary sequestration, neurogenic tumor, or hydatid cyst [8].

In the autopsy case series, the incidence of accessory liver tissue (ALT) was reported as 0.23%; however with development of laparoscopy and more advanced cross sectional imaging the incidence rose to 0.7% [8–10].

Adin et al. described three main types of intrathoracic ALT. [11], Figure 2. In Type I the accessory liver is connected to the orthotopic liver by a thick stock and represents a direct extension of the liver into the thoracic cavity. The type II: the ALT is in the thoracic cavity and attached to the orthotopic liver by a thin vascular pedicle; it can be located within the liver parenchyma or in the mediastinum. In Type III, the intrathoracic ALT has no connection to the orthotopic liver. Same as in Type II, it can be in the lung parenchyma or in the mediastinum.

Both congenital and acquired mechanisms of ALT have been described [12,13]. During embryological development human liver arises as a ventral outgrowth of the caudal part of the foregut adjacent to the diaphragm. At 4 weeks, the septum transversum does not separate the thoracic and abdominal cavities entirely, it has openings on each side. It has been speculated that a portion of the liver may grow cranially, preventing the complete closure of the septum transversum. Once fully developed, the septum transversum defect may be fully sealed off, thus separating the liver tissue within the thoracic cavity resulting, therefore, in Type III ALT. Alternatively, intrathoracic liver develops from a separate intrathoracic hepatic diverticulum or from hepatic cells that migrated into the pleural cavity via patent pleuro-peritoneal canals (Type III). Anatomical studies involving the necropsies of 172 rats have supported a genetic explanation, as they indicate that ALL is
associated with an autosomal recessive gene [5]. Currently, two hypotheses exist regarding the mechanism of ALL formation: (i) the embryonic liver protrudes outward and creates an accessory lobe during embryonic development, or (ii) an accessory lobe develops due to intra-abdominal pressure caused by the growth of the tunica muscularis recti and the enlargement of the liver [14,15]. Congenital diaphragmatic defects may contribute to the formation of an intrathoracic ALT. This should not be confused with a normally developed liver that prolapsed into the chest cavity [16]. Several authors indicate a correlation between congenital abnormalities in other organs and accessory liver lobes. Ito et al., Elmasalme et al., Grunz et al., and Sanguessa et al. have reported on the association of omphalocele and an accessory liver lobe in pediatric patients [14,17–29]. The diagnosis was made in children aged from one day to 14 years, possibly due to increased intra-abdominal pressure.

Acquired cases of ALT usually appear post thoracic trauma where diaphragmatic and liver integrity has been disturbed [2]. These are usually present as Type I and are easily diagnosed on cross sectional imaging. Fixed herniation of liver tissue into the thoracic cavity due to diaphragmatic hernia can be congenital and acquired with trauma being the most common etiology. Traumatic diaphragmatic hernia is a condition in which the abdominal contents herniate through a tear in the diaphragm. It usually occurs in young men who have been involved in motor vehicle accidents, and up to 8% of patients with major blunt trauma may experience it [24]. However, patients typically do not experience any significant symptoms related to the diaphragmatic tear, and clinical findings are often obscured by other major trauma. Strangulation of the herniated viscera is typically a late manifestation [24,25]. Chest radiographs can reveal specific findings, such as the intrathoracic herniation of the stomach and visualization of the nasogastric tube above the left hemidiaphragm. These findings, however, are not specific for liver herniation but rather for traumatic diaphragmatic injury [25]. Other findings may include apparent elevation, distortion, or obliteration of the diaphragmatic outline, abnormal lucencies adjacent to the diaphragm, and contralateral shift of the mediastinum.

A separate consideration should be given to the blood supply and bile drainage of the accessory liver lesion. A case report by Han et al., illustrates a case of type III intrathoracic ALT with arterial and venous supply originating from the abdomen, i.e., from hepatic artery and portal vein. Unfortunately, the case report does not specify if there was a separate bile duct [26]. Chapman and colleagues presented a case of a 12-year-old patient with Arnold–Chiari malformation who was unable to walk. The patient had deep venous thrombosis, and during an echocardiography, an intracardiac mass was found that partially blocked the right atrial-inferior vena caval junction. The mass, which consisted of a benign hepatic parenchyma, was removed along with a part of the inferior vena cava. Histological examination confirmed that the mass was benign. The blood supply originated from cardiac vessels and IVC [27]. Chen and Huang reported a case of an accessory liver lobe that protruded through the IVC foramen into the mediastinum. The ALT was supplied by two aberrant vessels from the right hemidiaphragm [28]. These case reports illustrate that blood supply to the intrathoracic ALT may originate from abdomen as well as from the thoracic cavity and is usually provided by nearby organs, we were unable to identify pattern of the blood supply.

Like any intrathoracic lesions, ALLs are frequently managed by a multidisciplinary team (MDT) including specialists from oncology, interventional radiology, thoracic surgery, hepatobiliary surgery, pathology, imaging and anesthesiology departments. Multidisciplinary
Surgical team meetings have a major influence on the management strategies for patients with thoracic neoplasms [29].

As this is frequently an incidental finding in the chest X-ray, the next investigation of choice is frequently computed tomography (CT). Diagnosis is straightforward in cases where an ALL is connected to the orthotopic liver by a pedicle. When located in the thorax, the ALL needs to be distinguished from a tumor of the pleura, lungs, chest wall, or diaphragm whereas an ALL in the pelvic cavity should be distinguished from a benign or malignant tumor of the pelvic organs. An ALL located on the surface of the liver or abdominal organs should be distinguished from pathological alterations in the liver, gall bladder, pancreas, spleen, or adrenal glands [6]. A 3D contrast-enhanced CT of the chest and upper abdomen is frequently valuable for any intrathoracic masses that are protruding from the abdominal cavity. An MRI scan can also be employed to specifically assess the association of the mass with the lung tissue and liver [6]. Hepatic angiography, which reveals the vascular connection of the mass with the liver, is another diagnostic approach used in cases of accessory liver tissue. When the diagnosis is not obvious, and especially in the cases of ectopic liver tissue (Type III per Adin classification) a transthoracic needle biopsy or transbronchial biopsy would be recommended [30]. The biopsy approach heavily depends on the available resources and the proximity of the lesion to the chest wall or to the bronchus.

Several case reports describe the malignant transformation of ALL [28]. As far as the carcinogenic process is concerned, ineffective bile reabsorption and altered blood outflow may contribute to the malignant transformation [28]. Currently, there is no evidence to suggest that ALLs with malignant transformations are more prevalent in patients with liver cirrhosis. There are limited reports of ALLs associated with benign lesions such as focal nodular hyperplasia [31]. To our knowledge, there is only one case report describing the interventional embolization of the feeding artery of ALLs with HCC followed by laparoscopic surgery [32]. Huo et al. stated that while FDG PET/CT is effective for identifying numerous liver conditions, its sensitivity in the early detection of hepatocellular carcinoma is suboptimal [33]. Additionally, Arakawa et al. observed that serum alpha-fetoprotein (AFP) levels were higher than 1000 ng/mL in 12 out of 19 cases of ALLs or ectopic liver with HCC (63%). When the serum AFP level continues to rise with no significant abnormality in the native liver, although rare, the possibility of ALL or ectopic liver with HCC should be considered [34].

The surgical removal of intrathoracic accessory liver tissue may be considered in cases when the patient is symptomatic or where there are concerns about malignant transformation. Surgery may be offered in cases where the accessory liver lobe may become twisted, leading to ischemia. Several cases were published on the torsion of the accessory liver lobe; however, none in the intrathoracic liver [21,35]. Minimally invasive techniques such as thoracoscopic surgery can be utilized, providing a less invasive approach with reduced postoperative complications. However, the decision to proceed with surgery should be carefully evaluated and individualized based on the potential risks and benefits for each patient. Several authors suggest the resection of the ALT as a prevention strategy [36–38]. Similarly, surveillance without surgical intervention has been described in cases with no concerns for malignant transformation. Ultrasonography is reported as a good surveillance imaging modality, this patient, however, would benefit from CT given the intrathoracic location of the accessory lobe [11].

4. Conclusions

The study of intrathoracic accessory liver tissue sheds light on the remarkable complexity and variations of human anatomy. While it is a rare condition, understanding its characteristics and clinical implications can contribute to improved diagnosis and management for affected individuals.

Further research is needed to deepen our understanding of the underlying mechanisms of the embryological origins of the accessory liver tissue as well as the genetic
predispositions and long-term outcomes associated with intrathoracic accessory liver tissue. This will enhance our ability to provide appropriate care and support to those affected by this condition.

Additionally, it is important to note that intrathoracic accessory liver tissue is often found incidentally during investigations for unrelated medical conditions. This emphasizes the significance of thorough diagnostic evaluations to identify any anatomical variations or abnormalities within the thoracic and abdominal cavities. Several imaging modalities can be used for primary investigation and for subsequent surveillance with cross-sectional studies being the investigation of choice. In this case, the greatest concern was for the malignant transformation of the ALL. As no malignant features were detected during the examination and the patient did not show any symptoms for over seventy years, the option of yearly surveillance through CT imaging of the chest and abdomen, liver function tests, and specialist follow-up was presented. There is no scientific evidence to suggest that yearly follow-up is superior to six-month follow-up. Therefore, the decision was made based on clinical judgment.

Due to its rarity, the clinical significance of intrathoracic accessory liver tissue is not well-documented. However, in some cases, the presence of ectopic liver tissue in the thoracic cavity can lead to respiratory symptoms such as cough, shortness of breath, chest, or back pain. It can also result in gastrointestinal symptoms if the ectopic liver tissue compresses nearby organs or interferes with their normal functioning. In some cases, intrathoracic accessory liver tissue has malignant potential, but the mechanism of that is, sadly poorly understood.

The management of intrathoracic accessory liver tissue typically involves a multidisciplinary approach, with input from radiologists, thoracic surgeons, upper gastrointestinal surgeons, hepatologists and medical oncologists. The treatment decision is based on the specific situation of the individual, considering factors such as the presence of symptoms, the size and location of the ectopic liver tissue, the type of connecting pedicle, and any associated complications.

In conclusion, the described case report is a rare presentation of asymptomatic intrathoracic liver tissue that most likely represents a case of congenital diaphragmatic malformation with an accessory intrathoracic liver lobe. The management depends on the symptoms, the anatomy of the accessory lobe, and on clinical and biochemical suspicion of the underlying malignancy. The understanding of the anatomy of the accessory liver lobe is pivotal for preoperative planning and for the stratification of malignant potential.

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