

Agnesis of Left Hepatic Lobe: A Rare Congenital Anomaly. Case Report

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Abstract

Case Report

Morphologic anomalies of the liver, as opposed to many other visceral organs, are very rare. Left lobe liver agnesis is defined as the absence of liver tissue on the left side of the liver in the absence of previous disease or surgery. It is commonly an accidental finding on imaging investigations or at the time of abdominal surgery. We report the case of left liver lobe agnesis diagnosed in a 68-year-old man with a history of urothelial carcinoma of the bladder, and the diagnosis of his liver congenital abnormality was made accidentally on a CT of chest-abdomen-pelvis (initially done to look for secondary localisations of his urothelial carcinoma). The main imaging findings of this condition are reviewed briefly, with associations and the most common differential diagnoses.

Keywords: Agnesis, Lobe, Liver, Imaging.

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INTRODUCTION

Liver lobe agnesis is defined as the absence of liver tissue on the right or left side of the gallbladder fossa, without evidence of previous surgery, trauma or any disease leading to hepatocellular dysfunction and atrophy. It remains a rare condition. Only some cases have been reported in the literature [1].

This condition is generally asymptomatic with normal liver function parameters, and it is generally a coincidental discovery, uncovered by autopsy or imaging examination [2].

CASE REPORT

A 68-year-old male with a medical history of urothelial carcinoma in the bladder, with no liver disease nor ablative surgery or abdominal trauma history. Whose physical examination was normal. Routine laboratory tests were normal. A contrast enhanced computed tomography (CT) was performed looking for metastasis, CT images revealed enlargement of the right hepatic lobe, especially the lateral segment. The left hepatic lobe was absent as well as left hepatic artery, left portal vein, and left hepatic biliary system. This appearance pleaded for left liver lobe agnesis associated to compensatory hypertrophy of right hepatic lobe.

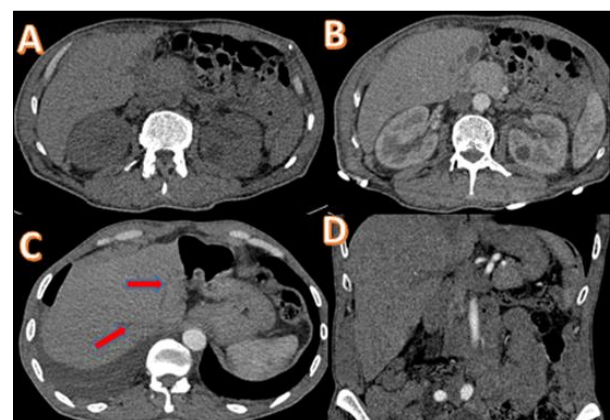


Figure 1: Abdominal contrast enhanced CT scan in axial section before IV contrast injection (A) and after (B) and coronal section (D) showing absence of the left hepatic lobe, left hepatic artery, left portal vein, and left hepatic biliary system. The left hepatic vein is also absent, only the right and middle ones are present (red arrows) (C). A compensatory hypertrophy of right hepatic lobe is noted.

DISCUSSION

Hepatic lobe agnesis is a rare developmental anomaly. Only some cases have been reported in the literature. It is commonly noted incidentally at autopsy, surgery or imaging investigations. Its knowledge is important especially for adequate planning of surgical procedures. This condition was first reported by Arnold and Ashley-Montagu in 1932. The incidence of lobar agnesis has been reported to be 0.005% in 19,000

autopsy cases. The left lobe more affected than the right one [3].

The pathogenesis of hepatic lobe agenesis is owed to defective and interrupted processes during embryonic development of the liver [3].

Clinically most patients are generally asymptomatic with no consequences in prognosis. However, it is important to know whether it is acquired (atrophy) or congenital (agenesis). Some of the affected patients may have additional anomalies and anatomical changes like partial or complete absence of the right part of the diaphragm, bowel malrotation, choledochal cyst or altered localization of the gallbladder. In addition, a tendency to develop portal hypertension [4].

Pages *et al.*, categorized developmental abnormalities of liver morphology as follows: Agenesis (absence of a lobe that is substituted by fibrous tissue); aplasia (one of the lobes is small and its structure is abnormal, with few liver trabeculae, numerous bile ducts and abnormal blood vessels); or hypoplasia (one of the lobes is small but its structure is normal) [4]. According to this classification, our case could be classified as agenesis. The radiologic findings can be detected in US, CT or MRI. The absence of liver tissue to the left side of the gallbladder fossa is the most significant finding for the diagnosis. The absence of the falciform ligament, blood vessels and biliary ducts is also strong evidence of left liver lobe agenesis [5, 6].

In addition to congenital agenesis, trauma, diseases such as liver cirrhosis or previous liver segment resections must also be taken into account in the diagnosis. After excluding the differential diagnoses mentioned above, the rare diagnosis of left lobe agenesis of the liver can be given. In this situation, no further specific therapy is required. However, because of the

presence of concomitant hepatic disease (NASH), annual sonographic and laboratory monitoring is recommended [6].

CONCLUSION

Left hepatic lobe agenesis is a rare congenital anomaly. Awareness about this condition is crucial for clinicians and radiologists. The imaging characteristics should be considered when evaluating a patient with hepatic lobe agenesis.

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