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Case Report

Agenesis of right lobe of liver; a rare anomaly: a case report

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ABSTRACT

Agenesis of right lobe of liver is a rare finding and was defined as the absence of liver tissue on the right side with preservation of the middle hepatic vein without previous disease or surgery. Congenital agenesis of right hepatic lobe is a rare anomaly which is found incidentally in radiologic examination. Here we present a case of 22 year old female who came with abdominal distension suspecting liver cirrhosis she was investigated and on imaging studies incidentally it was revealed that there is absence of right lobe of liver.

Keywords: Liver, Right lobe agenesis, Computed tomography

INTRODUCTION

The liver is wedge shaped, largest abdominal viscera occupying a substantial portion of the upper abdominal cavity. It occupies most of the right hypochondrium and epigastrium and extends into left hypochondrium as far as left lateral line and under the right dome of diaphragm.[1-4]

Anatomically liver is divided into right and left lobes by the line of reflection of falciform ligament anteriorly, the fissure for ligamentum venosum posteriorly and fissure for ligamentum teres inferiorly. The anatomical right lobe is larger than that of left lobe.[1,3-5]

Agenesis if right lobe of liver is defined as the absence of liver tissue on the right side with preservation of middle hepatic vein without previous disease or surgery.[6] Agenesis of right lobe of liver is an extremely rare congenital anomaly.[6-8] Till now only 29 cases have been reported in the literature.[9,10] Generally it is accompanied by other anomalies of biliary tree such as ectopic gall bladder, retro or suprahepatic gall bladder.[7-9,11,12] and compensatory hypertrophy of left hepatic lobe.[11,13,14] Agenesis of right lobe of liver is an incidental finding revealed by ultrasonography, computed tomography or magnetic resonance because the condition is asymptomatic.[15,16]

Here we report a case of agenesis of right lobe of liver, discovered as an incidental finding during the diagnostic imaging studies for cirrhotic liver disease and s/o portal hypertension. The diagnosis of this anomaly is documented with cross sectional imaging including ultrasonography and computed tomography.

CASE REPORT

A 22 year old female patient who was examined by physician and was admitted to the private hospital for abdominal distension. She had similar symptoms for last 8 years; suspecting portal hypertension and cirrhotic liver disease she was referred for imaging investigations.

Imaging studies such as ultrasonography, computed tomography and Doppler study were done which revealed the absence of right hepatic lobe and hypertrophied left hepatic lobe.

Ultrasonography was done (Figure 1) which revealed the enlarged left hepatic lobe and absence of right hepatic

lobe. Further computed tomography was done to evaluate the absence of right lobe of liver and position of gall bladder (Figure 2). The position of gall bladder was on the right side of liver in a vertical position between the right costal arch and right dome of diaphragm. E/o gross clear ascites seen with varices and splenomegaly.

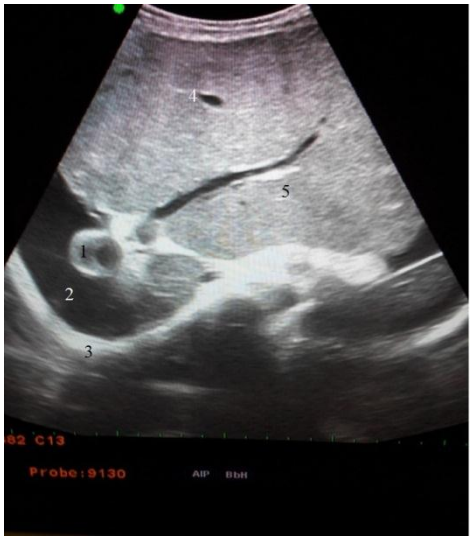

Figure 1: Ultrasonography.
1- Gall bladder, 2- Ascites, 3- Diaphragm, 4- Middle hepatic vein, 5- Left lobe of liver

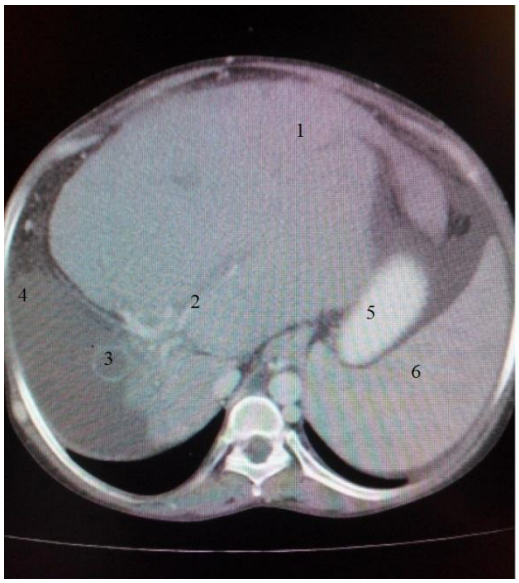

Figure 2: Computed tomography.
1- Left lobe of liver, 2- Left portal vein, 3- Gall bladder, 4- Ascites, 5- Stomach, 6- Spleen

Further Doppler study was done (Figure 3) which revealed the absence of right hepatic vein and right hepatic artery. The middle and left hepatic vein were normal and both formed a common stump which drained into inferior vena cava. The study also revealed that there was large left hepatic artery with elevated flow velocity in the left lobe suggestive of cirrhotic liver disease in the left lobe. The left portal vein showed low velocity flow and the most important finding was that the right portal vein was not identified.

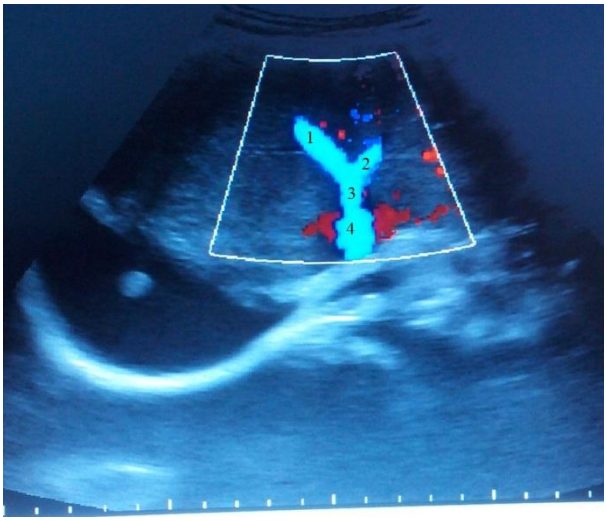

Figure 3: Doppler study.
1- Middle hepatic vein, 2- Left hepatic vein, 3- Common stump, 4- Inferior vena cava

Embryological basis

The embryological development of liver begins in the 2nd week of embryonic life with the formation of endodermal hepatic diverticulum at the distal part of foregut. The diverticulum grows within the septum transversum.[1,11,16] It has been known that this anomaly is caused by the developmental failure of right portal vein or an error of mutual induction between the primitive diaphragm and the endodermal diverticulum representing the primitive liver.[9,12]

DISCUSSION

Developmental anomalies of right lobe of liver was first reported in 1870 by Heller.[6,16] Anatomical variations of liver are common and occur during normal development of the organ and correspond to the variation in the distribution of liver territories.[6,16]

Lobar agenesis is defined as congenital absence of or incomplete development of liver tissue of the right or left hepatic lobe without prior surgery or hepatic disease[11,16] but the agenesis should be differentiated from a lobar atrophy of right hepatic lobe caused by cirrhosis, choangiocarcinoma, idiopathic portal hypertension and anomalies of other biliary tracts.[9,18,19]

Agenesis of right hepatic lobe is a rare congenital anomaly.[7,8] Advances and the increased use of non-invasive imaging techniques such as ultrasonography, computed tomography and magnetic resonance imaging led to more frequent detection of such variant anomaly. Several reports have described this anomaly on computed

tomography and ultrasonography[7] and found the absence of right hepatic lobe and compensatory enlargement of the left hepatic lobe. These same finding were present in our case where enlargement if left lobe of liver was present.

Agenesis of right hepatic lobe is always associated with absence of right hepatic artery, right hepatic vein and right portal vein. In our case the imaging studies revealed the absence of right hepatic vein, right hepatic artery and right portal vein. Agenesis of right lobe of liver may predispose the patient to the development of portal hypertension and oesophageal varices especially when the left lobe is enlarged.[20] In our case the patient had developed the portal hypertension along with ascites and varices were present.

The probable etiology of portal hypertension appears to be due to reduction in number of intrahepatic branches of portal vein that are not compensated for by increased density of the left lobe vasculature.[20]

In case of such congenital liver anomaly the gall bladder is often placed on the right side of liver against the diaphragm in a vertical position.[16] In our case the position of gall bladder is also on the right side of liver in a vertical position between the right costal arch and right dome of diaphragm.

Previous studies have revealed that the congenital agenesis of liver lobe affects the left lobe more than the right lobe and the agenesis of right lobe occurs slightly more often in men.[17] But in our case this rare anomaly was an incidental finding noticed in a female patient.

Also the studies reveals that agenesis of right lobe of liver is presented by absence of right hepatic vein and right portal vein.[11,17] Such similar finding was present in our case also.

Thus surgical knowledge of such anatomical agenesis is necessary for surgical planning, appropriate identification and intraoperative findings of postoperative approach to the therapy.[6]

Many of the patients with this anomaly remain asymptomatic clinically for many days as in our case where the patient came only when the ascitic fluid was collected in the right upper quadrant of abdomen. It is therefore important to consider agenesis of right lobe of liver in differential diagnosis when imaging studies revealing abnormal hepatic morphological characteristics associated with abdominal distension and portal hypertension.

CONCLUSION

We conclude that the knowledge of this rare congenital anatomical anomaly should be kept in mind of the surgeon. However we want to highlight the importance of understanding this rare condition. Surgeons must recognize the entity in order to deal appropriately with the findings. The patient needs to be educated regarding the benign evolution of the condition and future clinical implications.

Thus the knowledge of variation in liver may be of paramount importance to clinicians to diagnose hepatic disease, surgeons for carrying out liver related diseases, imaging personals for avoiding misinterpretation of images and anatomists and morphologists for new variants.

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