eISSN 1308-4038

International Journal of Anatomical Variations (2012) 5: 16–17

Case Report

Isolated situs ambiguus hepatis: a rare case report

Published online June 11th, 2012 © http://www.ijav.org

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Received May 27th, 2011; accepted March 28th, 2012

Abstract

During routine dissection of abdomen of a 45-year-old male cadaver, liver was found to be occupying a central position with a gallbladder on the right side of ligamentum teres hepatis. No variation was seen in other abdominal viscera, vessels of abdomen or thoracic viscera and vessels. This isolated ambiguus liver makes this case unique and interesting. Recognition of this variation is useful for radiologist and surgeon for making of a correct diagnosis and surgical intervention plan.

Key words [situs] [situs ambiguus] [heterotaxy syndrome] [midline liver]

Introduction

Situs ambiguus or heterotaxy syndrome is a disturbance/arrangement in the usual left and right distribution of thoracic and abdominal organs, which does not entirely correspond to the complete or partial mirror image [1]. True incidence of heterotaxy is not known but is variably being described to be 0.004% to 0.0125% [1]. Heterotaxy is being classified into associated with asplenia (asplenic syndrome or right isomerism or bilateral right-sidedness) or associated with polysplenia (left isomerism or bilateral left-sidedness) [2].

Case Report

During routine dissection of abdomen of a 45-year-old male cadaver, for undergraduate training, we observed that liver was occupying a central position in the abdomen. It was occupying the epigastrium and left hypochondrium. The right surface of the liver was just right to the right midclavicular plane and the space between this surface and parietal peritoneum lining right costal margins and undersurface of thoraco-abdominal diaphragm was occupied by loosely arranged fibrous tissue. The apex and adjoining inferior surface of liver was partially covering the diaphragmatic surface of a spleen. The gallbladder was found to be on the inferior surface of liver on the right side of the interlobar fissure of liver and ligamentum teres hepatis; but it was occupying left hypochondrium.

The hepatic artery proper was arising from hepatic artery and divided into right and left hepatic arteries. The cystic artery was arising from right hepatic artery and was ascending to left side, crossing the common bile duct from its anterior aspect, to reach the gallbladder. Extrahepatic biliary ducts were found to be without any variation. All other abdominal and thoracic viscera were also found to be without any variation. Grossly, no anomaly was found on thorough examination of cardiac chambers. No other associated variation was observed in the large vessels of abdomen and thorax.

Discussion

During third week of human embryogenesis, with the appearance of primitive streak cranial-caudal, dorsal-ventral and left-right axis is established. First major break in left-right symmetry is seen with looping of heart tubes to right and clockwise 90 degrees rotation of stomach during fourth week of development. This left-right asymmetry is non-random and is highly conserved [3, 4].

Applegate et al. described a series of 21 heterotaxy cases, all were associated with intestinal malrotation and 18 were associated with cardiac anomalies [2]; but in the present case no cardiac malformation or intestinal malrotation was

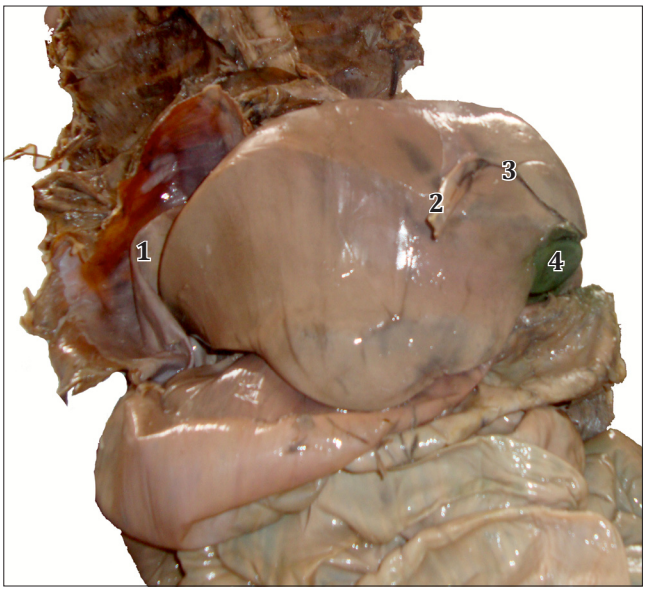

Figure 1. Photograph of left sided (midline) liver showing: empty space on right side of liver (1), falciform ligament (2), interlobar fissure (3) and gallbladder (4).

noted. They described a case with left/middle liver with a single spleen, as in our case, but having a midline gallbladder associated with left/middle stomach, intestinal malrotation, azygos/hemiazygos continuation and bilateral superior vena cava.

Similarly in a study of situs anomalies (10 situs inversus, 9 situs ambiguus) by Fulcher and Turner [5], only one case was reported having midline liver with a single spleen but associated with midline gallbladder and intestinal malrotation.

The presence of a heterotaxic liver without splenic variation (asplenia or polysplenia), intestinal, cardiovascular or pulmonary variation makes the case unique and interesting.

To conclude, a centrally situated liver without situs inversus may be present without numeric variation of spleen and other intestinal, cardiovascular and pulmonary variations/malformations. With increasing use of radiological imaging techniques, situs anomalies are detected with greater frequency. Therefore, the radiologist as well as the surgeon must be aware of the spectrum of the situs anomalies for a correct diagnosis and surgical intervention plan.

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