Isolated situs ambiguus hepatitis: a rare case report

Introduction
Situs ambiguous 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, liver was found to be occupying a central position with a gallbladder on the right side of ligamentum teres hepatitis. No variation was seen in other abdominal viscera, vessels of abdomen or thoracic viscera and vessels. This isolated ambiguous 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.

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
Isolated situs ambiguous hepatitis

References


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.

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).