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An Anatomical Study Of Situs Inversus Totalis

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Abstract
Situs inversus is a short form of Latin phrase "situsinversusviscerum" which means transposition of internal organs from its normal anatomical position. Usually the anomaly results from mechanical disturbance during the rotation of the gut during its development and when it is associated with dextrocardia, it is termed as Situs InversusTotalis.
The routine dissection revealed heart in the right side (dextrocardia), stomach was on right side, the liver in the left side .left lobe of liver was bigger than the right. The gall bladder was present on the visceral surface of the left lobe of liver, duodenum on left side, pancreas and spleen were on the right side. We hereby report a rare case of situsinversustotalis as clinicians may be aware and give due importance of any case of right sided chest pain and conduct proper investigations to diagnose such rare anomalies.

Key words: Situs inversus, dextrocardia

INTRODUCTION:

Situs inversus is a congenital genetic abnormality in which organs in the thorax and the abdomen are mirror image to their normal positions. Situs inversus was characterized by the transposition of the thoracic and abdominal visceral organs. Usually the anomaly results from mechanical disturbance during the rotation of the gut during its development and when it is associated with dextrocardia, it is termed as Situs Inversus Totalis. It occurs once in about 8-10,000 births. There are types depending on the position or arrangements of the organs. If it is mirror image of normal positioning means it is referred as Situs inversus totalis. If only lungs and heart are transposition it is Situs inversus thoracalis. If the liver, stomach and spleen are in transposition it is Situs inversus abdominalis.

MATERIALS AND METHOD

During our routine dissection in the Institute of Anatomy, Madras Medical College, Chennai, we came across an adult male cadaver, aged about 50 year identified as a rare condition Situs inversus totalis.

Observation

The male cadaver aged about 50 years was dissected and their observations were as follows:

Thorax cavity:

The Right Lung had 1fissure and 2 lobes and the left lung had 2 fissure 3 lobes (fig.1). The hilar structures appeared normal. Apex of the heart was on right side (fig 2) and chambers were reversed. The great vessels like superior vena cava, inferior vena cava were normal but seen on left side. The Arch of aorta, brachiocephalic trunk was also on the right side.

Diaphragm:

The Crus, central tendon, dome of diaphragm were normal; oesophageal opening was on the left side.

Abdomen:

In the abdomen the liver was on the left hypochondrium (fig. 3). The left lobe of the liver was larger than the right lobe of liver. Gall bladder was lodged inferiorly. The head of pancreas, uncinate process and neck were curved by the duodenum on the left side but the body and tail of pancreas, spleen was on the right side. Spleen was on the right hypochondrium. The spleen seems to be enlarged and bifurcated (fig.4). Right kidney was located at higher level than Left kidney (fig.5). External and internal genital organ were well developed. Coils of small intestine were in the midline, colon, caecum and appendix were seen in the normal anatomical position.

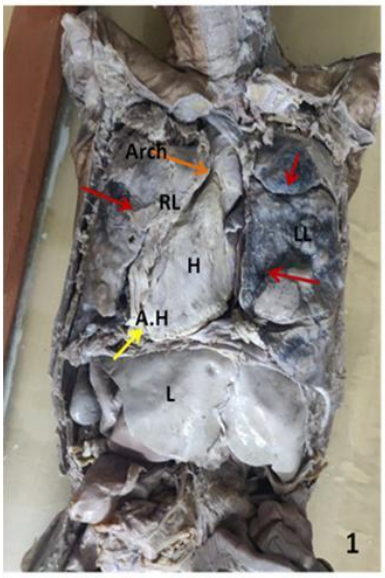

Fig. 1 Situs inversus – The transposition of the thoracic and abdominal visceral organs. The fissures of the lungs are showed in red arrow. RL-Right lung; LL-Left lung; H- Heart; L-Liver; A.H- Apex of heart; Arch – Arch of aorta.

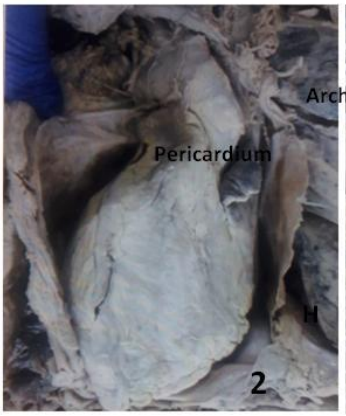

Fig. 2 Showing the apex of the heart (A.H)on the right side after cutting the pericardium. Arch of aorta is also visible. H- heart; Arch- Arch of aorta.

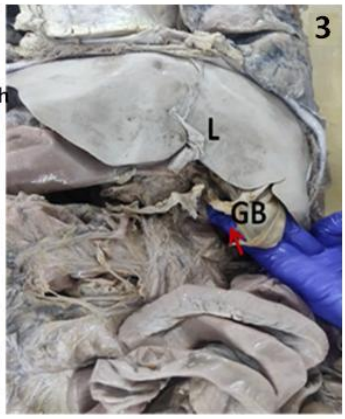

Fig. 3 Presence of liver and gall bladder (red arrow) on the left side

Fig. 4 Enlarged bilobed (red circle) spleen on the right side

Fig. 5. Right & Left kidney (red arrow). The right kidney is little higher in the level than the left kidney.

DISCUSSION:

The transposition of internal organs from its normal anatomical position in the present case may be explained embryologically. In the 3rd week of gestation primitive streak(the primary organizer), decide the right-left axis of the embryo. Primitive node appears on the cranial part of the primitive streak. The primitive node induces the nodal expression lefty-2, and beats the cilia present on the primitive node towards the left of embryonic disc up to the lateral plate mesoderm[1].

Some Authors also suggest that the transposition of the thoraco abdominal viscera in the present case may be related to malrotation of the heart tube and gastrointestinal system during the embryonic development.[1].

Development Of The Heart

The malposition of the heart in situs inversus can be explained with reference to its development. There are two rotational movements in the development of the heart occurs as heart tube over grows pericardium. According to researchers, in the first phase of embryonic development, there is 180° rotation around a transverse body axis, which brings the heart-anlage to the future thoracic region. During the second rotation around the sagittal and longitudinal axis, this results in the heart being properly positioned in the mediastinum[2].

Development Of Abdominal Viscera

In Situs solitusienormally the rotation of the midgut occurs at 270 degree anticlockwise rotation around the axis of superior mesenteric artery . During the rotation of midgut elongation of intestinal loop and coiling of jejunum and ileum take place. Large intestine shows elongation without coiling[2].

In Situs Inversus instead of normal rotation, midgut may have rotated to270 degree clockwise direction. This may result in the developing thoraco-abdominal organs to be the mirror image positionof the abdominal and thoracic viscera[3] The transposition of the abdominal viscera in situs inversus may be related to malrotation of the gastrointestinal system during the embryonic development[4]. Interestingly, some researchers defined a gut rotation determining factor is usually located in the left side of the body.The disturbance of this gut rotation determining factor during ontogenesis may be solely responsible for situs inversus. An earlier study described the positive role of homeobox gene Pitx2 in the looping mechanism of the heart and the gut[5]. Logan et al[l6] described “The transcription factor Pitx2 mediates situs-specific morphogenesis in response to left-right asymmetric signals. It has been thought that the disturbance of this gut rotation-determining factor during ontogenesis may be solely responsible for Situs inversus.

CONCLUSION

Situs inversus totalis may also be associated with numerous cardiac anomalies such as atrial septal defect, ventricular septal defect, transposition of great vessels, abnormal atrioventricular valves, absent coronary sinus, conduction abnormalities, double-outlet right ventricle, total pulmonary venous defect and pulmonary valve stenosis. The most prevalent gastrointestinal abnormalities associated with situs inversus are hepatic dysfunction, splenic dysfunction, respiratory abnormalities, and associated midline defects. The present case highlighted though it may be an incidental finding at routine dissection, the clinicians and anaesthesiologist may be aware and give due importance of any case of right sided chest pain and conduct proper investigations to diagnose such rare anomalies. This cadaver has been preserved in our museum.

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