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A Case Report Of Complete Situs Inversus

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ABSTRACT

A 45 year male while being dissected during undergraduate teaching was discovered as a case of complete situs inversus, a rare congenital abnormality. The thorax and the abdomen dissection revealed a complete mirror image of the normal visceral arrangement. The heart possessed an atrial septal defect. The abdomen revealed enlarged spleen, liver and gall bladder .The gall bladder was full of stones, with stones present in the common bile duct, too. The left kidney had an accessory renal artery supplying it. Our aim is to describe the anatomic relations of the viscera in the situs inversus, so as to decrease the incidence of the mishappenings in the clinical practice and to correlate the anomaly embryologically, which in this case seems to be due to the defect in the lateralization .

Keywords: Atrial septal defect, Kartagener's syndrome, mirror-image organs, complete situs inversus, situs solitus.

Situs solitus is the usual arrangement of organs and vessels within the body. The systemic atrium is on the right with a right-sided trilobed lung, liver, gallbladder, and inferior vena cava. The pulmonary atrium is on the left with a left-sided bilobed lung, stomach, single spleen, and aorta. The cardiac apex is on the left.

Situs inversus is a rare congenital abnormality. The incidence of complete situs inversus is estimated as 1/8000 in the general population, (Douard et al.,2000) . It indicates mirror-image location of the viscera relative to situs solitus. There are two major subcategories of situs inversus: situs inversus with dextrocardia and situs inversus with levocardia. Situs inversus with dextrocardia is characterized by mirror-image location of the heart and viscera relative to situs solitus, with the cardiac apex, spleen, stomach, and aorta located on the right ; the liver and inferior vena cava (IVC) located on the left.

Situs inversus are often asymptomatic and discovered accidentally. It may be associated with anomaly of the other organs like lungs, heart, spleen , liver and intestine, which usually present in the childhood or adulthood with respiratory distress or cyanosis, obstruction and other complications. It further complicates diagnosis and management of acute abdominal pain, (Ann et al,2002). Fifty percent of people suffering from Kartagener's syndrome have situs inversus, (McManus et al., 2004). As such it is important, that its recognition during the emergency settings to minimize the mishappenings at surgery or other interventions.

This rare variety of situs inversus is due to defect in lateralization but the exact mechanism of alteration of the rotation is still unknown. The defect may be either due to genetic or environmental factors such as teratogens.

A 45 year old man, farmer, residing in West Bengal was admitted to the hospital with complain of severe abdominal pain and features of obstructive jaundice. He soon expired and donated his body to the Department of Anatomy, Calcutta National Medical College, and Kolkata. The man was dissected and an assessment was made for the location of the solid and hollow organs of the thorax and the abdomen; the location of the cardiac apex, aortic arch, abdominal aorta, and IVC; the presence of spleen; the pancreas; the presence of vascular anomalies ; and the presence of underlying disease processes.

Thorax

The thorax examination revealed that the case had a heart with its apex to the right. The heart externally possessed four chambers. The atria were placed on the left and possessed

auricles. The right ventricle was anterior to the left ventricle and led to the pulmonary trunk. Pulmonary artery was on the right of ascending aorta. While the ascending aorta is on the left side, arising out of the left ventricle (Fig. 1B). It lead to the arch of the aorta with its branches which was directed backwards and to the right, (Fig. 1,B).The arch of the aorta is crossed by right phrenic nerve ,which pierced the dome of the right diaphragm, and right vagus nerve from before backwards. The right recurrent laryngeal nerve hooked arch of the aorta below against ligamentum arteriosum, (Fig. 1,D) .

The two atria communicated through an atrial septal defect, (Fig.1 ,D and E).The ventricles revealed normal architecture, with normal bicuspid , tricuspid valves and interventricular septum.

The superior vena cava present on the left side received the arch of the azygos vein, on the left . The Superior hemizygos vein present on the right side drained into the azygos at the level of thoracic seven vertebra (T7) while the inferior hemiazygos drained into the same at the level of thoracic eight vertebra (T8) . Both the hemiazygos had an intercommunication through a venous channel at their termination (Fig.1,C).The IVC drained into the left atrium through an opening in the diaphragm left of the midline , accompanied with the left phrenic nerve .

The esophagus was on the right and behind the trachea. Trachea had divided into two bronchi .Left being more vertical shorter and wider dividing into three subdivision, while the right one dividing into two (Fig. 1 E). Right tracheo esophageal groove had right recurrent laryngeal nerve .The thoracic duct was on the left to the aorta and after piercing the diaphragm crossed the midline at the level of T 4-T5 to the right,(Fig.1,B).

The left lung had three lobes while the right had two lobes .

Abdomen

The examination of the abdomen also revealed that the viscera were the total mirror image of situs solitus the spleen, stomach and aorta were located on the right and the liver, gall bladder and inferior vena-cava on the left side (Fig. 2,A).The present case had mirror-image location of the bowel and mesenteric vessels, too. The stomach, jejunum, and descending colon were located on the right, and the ligament of Treitz, ileum, and ascending colon are located on the left. The jejunum was shown to lie primarily to the right of midline and the ileum to the left. Inversion of the superior mesenteric artery was seen.

The gall bladder on the left was enlarged, full of multiple large stones (Fig . 2, B and C). The common bile duct was dilated full of small stones, adhered to the gall bladder. The porta-hepatis relation was reversed along the left free margin of the lesser omentum (Fig.2,B).

The liver was present on the left side, enlarged and nodular. The liver had two lobes the left larger, to the left of the midline while the right smaller lobe, right on the right side of the midline , (Fig 2,A (L)). The spleen was present on the right side and exhibited splenomegaly (Fig. 2,A). The splenic artery was observed which divided into multiple segmental arteries long before entering the hilum .The splenic vein was present posterior to the arteries and possessed large caliber. The consistent relationship between the spleen and the stomach was seen.

Pancreas head was located right to the midline, within the C loop of the duodenum .The body, triangular, demonstrated a groove at its superior border created by the splenic artery. The tail was present in the lienorenal ligament at the hilum of the spleen.

The aorta was located to the right of midline and the IVC on the left (Fig.2, D).The renal veins drained in the IVC while the testicular vein on the right side drained into the right renal and the left testicular vein drained into the IVC directly. The renal arteries as well as the gonadal arteries arose from the abdominal aorta and supplied the respective organs i.e., the kidney and the testis. An extra branch was observed from the abdominal aorta supplying the left kidney at its lower pole (Fig2, D).

The left kidney was placed at a lower

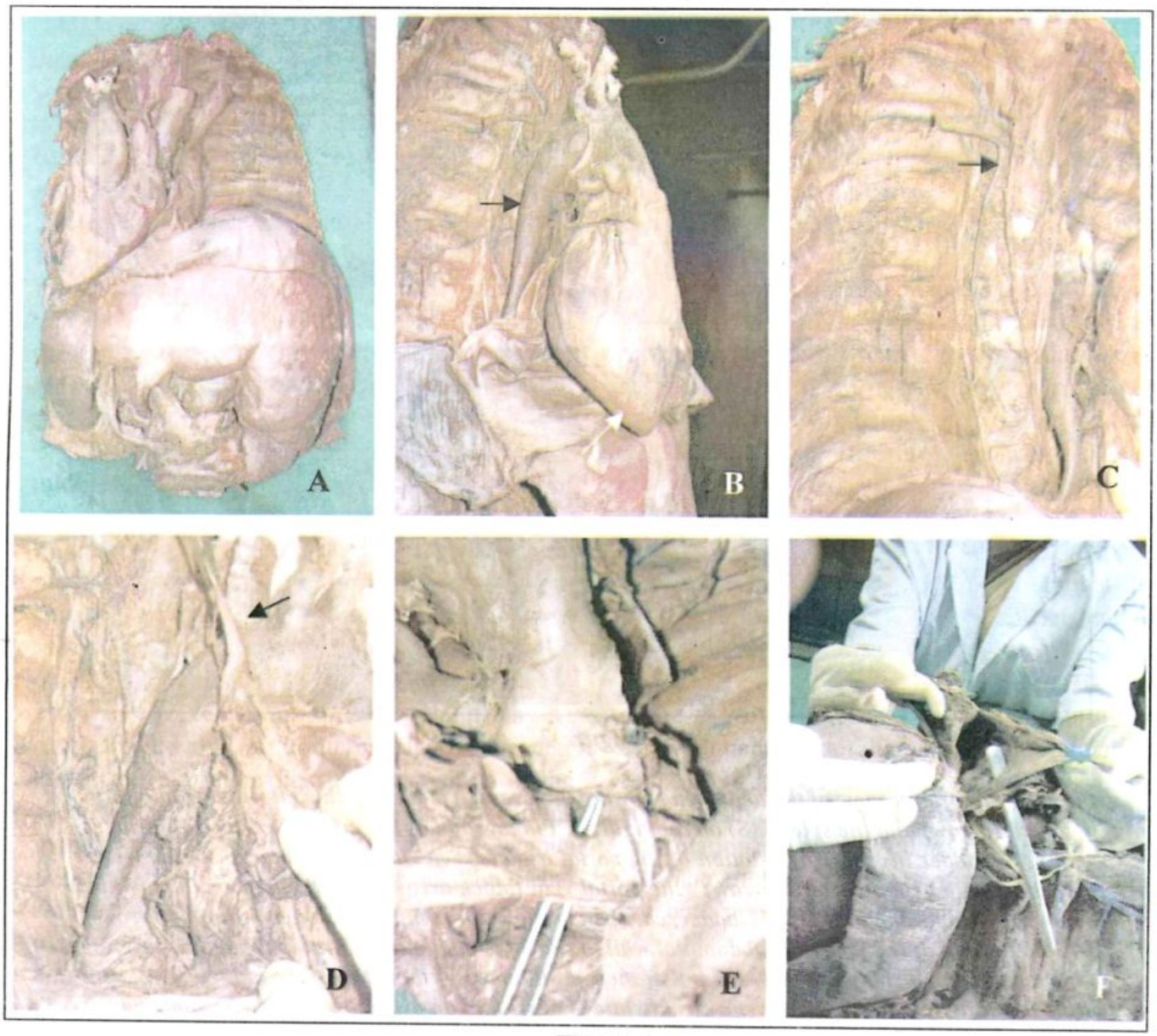

Figure 1

Fig. 1. Complete situs inversus

A. Panoramic view of the thorax and the abdomen showing complete situs inversus. B. Photograph of the right side view of the thorax showing the apex of the heart at the right (white arrow), the arch of aorta with its branches ,the descending aorta (arrow) , the thoracic duct, and the superior hemiazygos, the inferior hemiazygos, and the sympathetic chain on the right. C. Photograph of the posterior thoracic wall showing the superior hemiazygos (arrow) and the inferior hemiazygos draining into the azygos vein, and the thoracic duct traversing from left to right side at the level of T4 (white arrow). D. Photograph to show the right recurrent laryngeal nerve (arrow) hooking the arch of aorta against the ligamentum arteriosum. E and F .Photograph of the right atrium of the heart with a forcep placed in the atrial septal defect emerging out of the left atrium pulmonary vein openings .

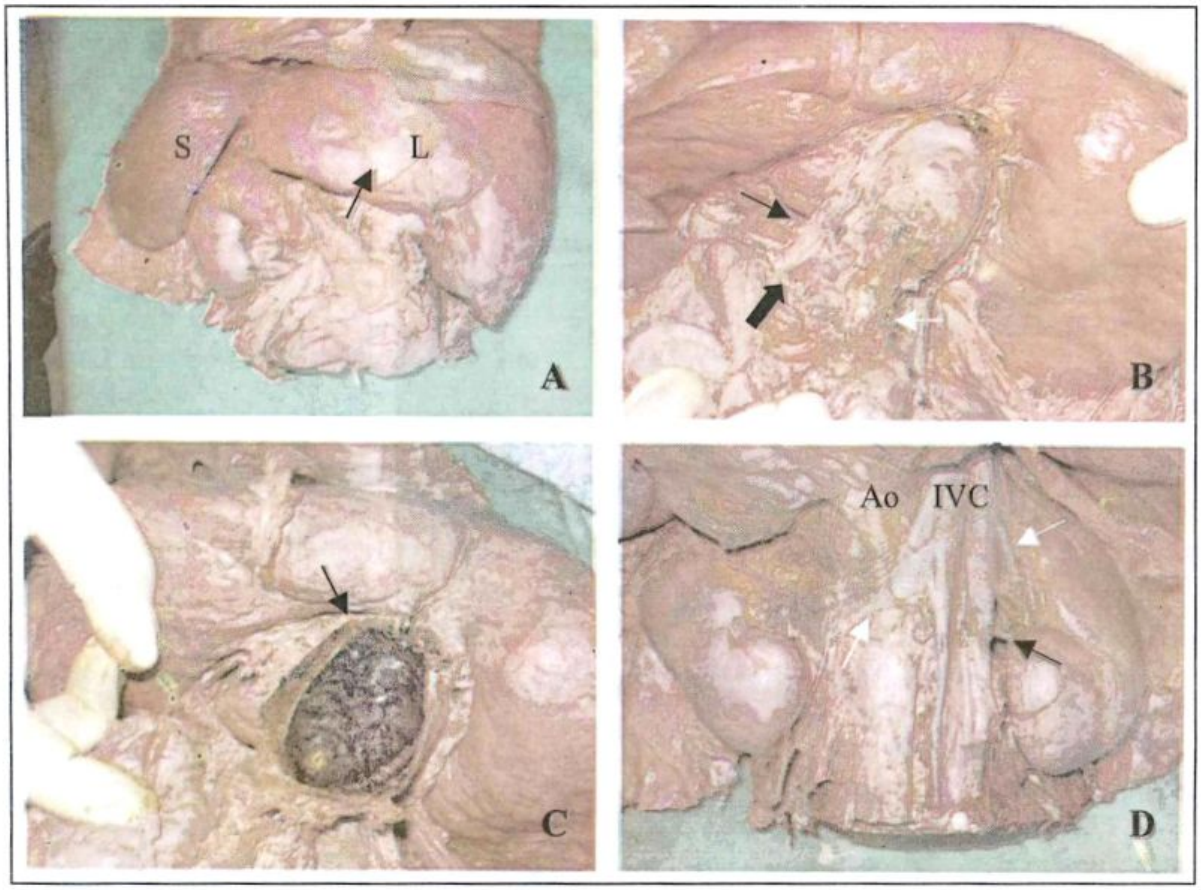

Figure 2

Fig 2 A . Panoramic view of the abdomen showing the liver (L) placed on the left with the falciform ligament (arrow) and the spleen (S) on the right. B. Photograph of the inferior surface of the liver with gall bladder in situ. The relation of the portal triad is seen at the free margin of the lesser omentum , the hepatic artery (arrow) , the portal vein (block arrow) and the hepatic duct (white arrow). C Photograph of the gall bladder, cut open to show large multiple stones, within it (arrow). D. Photograph of the posterior abdominal wall to show the abdominal aorta (Ao) and the inferior vena cava (IVC) displaying total reversal of their positions as well as that of the kidney . The renal veins draining into the inferior vena cava (white arrows) and an extra artery is seen supplying the lower part of the left kidney, (arrow).

position than that of the right kidney. At the hilum the vein artery and the ureter were placed from before backwards.

Disease Correlation

Autopsy finding suggests that he had cholecystitis ,cholelithiasis, choledocholithiasis; and might be suffering from portal hypertension, as suggested by hepato-slpenomegaly.

DISCUSSION

In the present case the orientation and the position of the various organs like heart venous system, lung ,liver, spleen and the stomach with the intestinal loop strongly suggest it to be mirror image of situs solitus and a case of complete sirtus inversus ; situs inversus with dextrocardia . According to Gray's (39th edition), embryos may be considered to be constructed with three orthogonal spatial axis . The cephalocaudal axis, the dorsoventral axis and the bilateral axis plus a further temporal axis . The bilateral axis is the consequence of the formation of the first two axes. Initially the right and left halves of the body are bilaterally symmetric with the temporal axis , modification of the embryo relative to its original axes can be seen .The originally midline ventral structure specially those derived from the

splanchnopleuric mesenchyme , such as the cardiovascular system and the gut are subject to extensive shifts .Changing from the bilateral symmetric arrangement to a whole body that is now chiral .It is puzzle that this symmetry is handed (chiral) rather than random, i.e., it is always asymmetric in the same direction .Defects in the lateralization may be responsible for the congenital anomalies of the heart , bronchial branching ,lung lobation , portal venous anomalies. The frequent association of these anomalies with one another strengthens the view that they are due to a common defect and that is this defect is due lateralization .

The heart present on the right side of the midline , presented with a defect in the interatrial septum at the upper part of the fossa ovalis suggesting a variation in the length of septum primum and the septum secundum, and consequently its non closure. In situs inversus the common cardiac malformations include; Atrial septal defect or Ventricular septal defect , reported by Trehan et al.,2003 ; or it may be associated with a common atrioventricular canal, univentricular heart, transposition of the great arteries, or anomalous venous connections including total anomalous pulmonary venous return (Saha et al., 2004).The association of the abonormality of the heart in relation to situs inversus could not be elicited , as these congenital abnormalities are associated with situs solitus also .

Patients with situs inversus demonstrate mirror-image location, not only of the solid organs and heart, but also of the bowel and mesenteric vessels (Ann et al., 2002; Le et al., 2003) . In the present case the stomach, jejunum, and descending colon are located on the right, and the ligament of Treitz, ileum, and ascending colon are located on the left. The orientation of the bowel is reversed rather than malrotated relative to situs solitus. The jejunum was shown to lie primarily to the right of midline and the ileum to the left. Inversion of the superior mesenteric artery was observed. The consistent relationship between the spleen and the stomach was seen and can be explained by the fact that splenic tissue develops in the dorsal mesogastrium and thus in this case present on the right side of the midline .The liver with the two lobes: the right smaller lobe and the left larger lobe ,was on the left of the midline . The gall bladder full of stone seems to correlate with the cholecystitis, and the enlarged spleen and liver with the portal hypertension

The present situs inversus had no anomaly of the lung and no complain of respiratory diseases or infertility and therefore we can rule out its association with kartagener's syndrome or primary ciliary dyskinesia . The cause for situs inversus holds a different explanation , as abnormal situs in 50% are associated with conditions like primary ciliary dyskinesia (McMannus et al., 2004) or renal hepatic pancreatic dysplasia , the view holds that the situs inversus seem to be independent of positional information .Brown et al., 1991 depending on primary ciliary dyskinisia had suggested 'dynein , as the component in the establishment in lateralization .While Rautiainen et al., 1990 proposed that a common mechanism is responsible for both the orientation of laterally information and also for ciliary orientation.

The etiology of the abnormal lateralization are both genetic and environmental. Arnold et al., 1983 suggested it to be autosomal recessive ,while Niikawa et al., 1983 suggested autosomal dominant transmission and X-linked predisposition was proposed by Mathias et al., 1987. Casey (1998), suggested that this defect may be due to genetic abnormality , e.g. HAND, ZIC3, Shh, ACVR2B and Pitxz genes. Environmental factors like the teratogens were held responsible for abnormal lateralization.

Based on the case report , the most probable cause seems to be the abnormal lateralization but we cannot rule its association with the genetic factors . Thus by correlating the relations and the position of the various organs found in complete situs inversus the surgeons can reduce the mishappenings in the in emergency and other interventions.

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