DOI: 10.7860/IJARS/2016/23272:2197

Case Report

Anatomy Section

Situs InversusTotalis with Malposition of Caecum and Ascending colon

VIDYA GUNASEKARAN, GLADWIN VICTOR RAJ, PARKASH CHAND

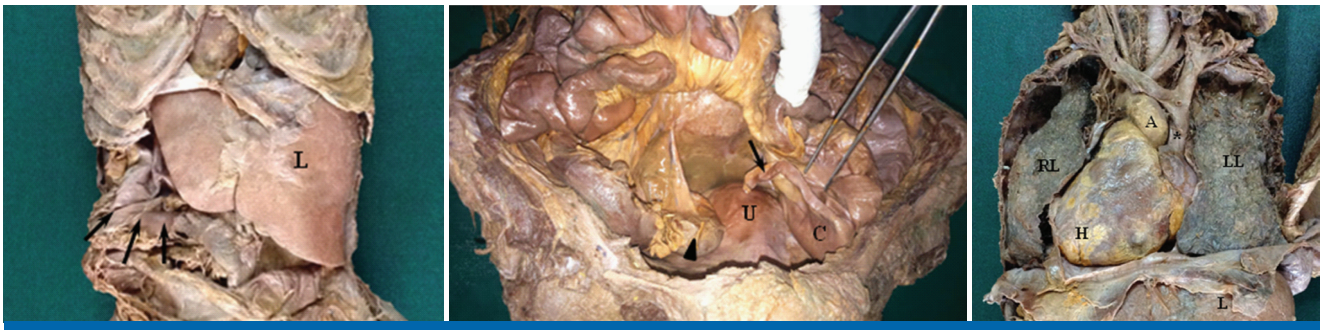
ABSTRACT

Situs Inversus Totalis (SIT) is an inversion anomaly involving the position of abdominal and thoracic viscera with respect to the midline. The medical and surgical management of these patients are technically more challenging because of the mirror image location of the viscera. The objective of the present case report is to document a rare case of SIT associated with malposition of caecum and ascending colon. A routine gross anatomy dissection was carried out for demonstration for the undergraduate students, in a formalin fixed female cadaver. Abdominal and thoracic region dissection was carried out following standard procedures. An inversion anomaly associated with malposition of colon was encountered, which was documented appropriately. Inversion anomaly of abdominal organs was observed with the liver on the left and stomach and spleen situated on the right side of the body. The position of large intestine per se was lower and it was found that the caecum and appendix was located deep down, in the left pelvic region with a short segment of ascending colon. In the pelvis, uterus was identified and was retroverted in position. Further, dissection of the thoracic region confirmed dextrocardia in the same cadaver. The position of great vessels in the thorax and the abdomen were also inverted with regards to the midline. These anatomical variations should be kept in mind during clinical and radiological examination in patients presenting with unusual signs and symptoms of abdominal region. Further, inversion of technical procedures is mandatory for effective surgical management of such SIT cases.

Keywords: Dextrocardia, Heterotaxy, Inversion anomaly, Kartagener's syndrome, Lateralization defect

CASE REPORT

During routine dissection for the undergraduate medical students in our Institute with an annual intake of 150, we observed an inversion anomaly of abdominal organs, in a female cadaver of nearly 45 years of age. There were no significant findings in the external abdominal wall. Dissection of the abdominal region following the routine method, revealed inversion anomaly of the abdominal organs with the liver located on the left whereas, the stomach and single notched spleen were situated on the right side of the upper abdomen. The liver was vertically enlarged extending upto the left lumbar region. Gall bladder occupied the inferior surface of the liver with the direction of the fundus towards the left side [Table/Fig-1]. Pancreas was vertically inclined occupying the umbilical region and extending towards the right hypochondriac region, but not truncated. No malrotation of gut was noted in the cadaver, the small intestines occupied mostly the lesser pelvic region. The position of the larger intestine varied significantly. The caecum was found located lower in the left pelvic region in close relation with that of uterus and the appendix was projecting into the true pelvis [Table/Fig-2]. The ascending colon was a very short segment measuring approximately 14cm occupying the left iliac region and the transverse colon was obliquely placed extending towards the right hypochondriac region. The uterus was retroverted in position. The great vessels of abdomen, inferior vena cava and abdominal aorta also showed a complete inversion anomaly. Further, dissection of the thoracic region confirmed dextrocardia with the apex of the heart pointing towards the right side and inversion of major blood vessels with their branches and tributaries in this region viz., formation of superior vena cava with the right and left brachiocephalic veins, branches of arch of aorta and pulmonary trunk [Table/Fig-3]. The fissures of both the lungs were ill defined and demarcation of lobes of lungs in both the sides couldn't be ascertained. The right lung was narrow and long with the cardiac notch. Histopathological examination was inconclusive to rule out Kartagener syndrome of the lungs due of partial putrefaction of the lung tissue.



[Table/Fig-1]: Showing vertically enlarged liver extending upto left lumber region. *L – Liver, Black arrows – Stomach. **[Table/Fig-2]:** Caecum located lower in the left pelvic region in close relation with that of uterus and the appendix. *U-Uterus, C- Caecum, Black Arrow – Appendix, Black arrow head – Sigmoid colon **[Table/Fig-3]:** Showing dextro cardia and inversion of major blood vessels. *RL – Right Lung, LL- Left Lung, H- Heart, A- Aorta, L – Liver, * - SVC

DISCUSSION

Situs Inversus Totalis (SIT) is an inversion anomaly involving the position of abdominal and thoracic viscera with respect to the midline. Situs inversus should be differentiated from other situs anomalies such as situs inversus with levocardia and situs ambiguous or heterotaxy, since the percentage of associated congenital heart anomalies varies greatly between the individuals with different types of situs anomalies [1]. Situs Inversus Totalis can be a part of other associated syndromes such as Kartagener, in which the primary ciliary dyskinesia is pathognomonic [2]. SIT impose a greater challenge to the clinician in the diagnosis as well as the management of medical and surgical conditions. The surgical procedures done in SIT patients pose difficulty in the orientation and technique because of the mirror image location of the viscera in comparison to the normal individuals [1]. The problems in these patients are magnified, when SIT is associated with other rotational anomalies of gastro intestinal tract [3]. A rare case of SIT associated with malposition of caecum, ascending and transverse colon is reported in the present study to enlighten the knowledge of the surgeons and radiologists.

SIT is a rare but significant congenital anomaly with the mirror image location of the thoraco-abdominal organs. SIT can be associated with other clinical conditions such as Kartagener's syndrome, polysplenia, rotational abnormalities of gut and cardiac anomalies. The incidence of SIT is about 0.01% [4,5].

Though, the etiology of situs anomalies remains elusive, it has been suggested that the abnormalities in the primary looping of heart and lateralization defects may be the key factors.The occurrence of situs anomalies is usually sporadic, however familial cases of autosomal recessive transmission have also been reported. In addition, situs anomalies may be due to the influence of environmental factors as evidenced by its close association with diabetes mellitus. Recent human studies have revealed that mutations in the Activin receptor IIB gene, Connexin 43 gene and LEFTY A gene are involved in lateralization defects. Further, chromosomal abnormalities which includes translocations (such as between chromosomes 12 and 13 or chromosomes 11 and 20), paracentric inversions in chromosome 11, deletions (involving chromosomes 4, 10, 13, and 18), insertions (from chromosome 8q into 7q), placental trisomy 16 are also associated with lateralization defects [2,3,6].

The lateralization of human embryo is a complex process, in which the midline notochord cells play a key role in determining the left-right asymmetry. These midline notochord cells are the major signaling source for lateralization. The dynein arms of the nodal cilia maintain a gradient which acts as a barrier which regulates the flow of extra-embryonic fluid at the embryonic node towards the left which is necessary for the left-right determination.The defects in lateralization are related to the changes in the nodal fluid dynamics, secondary to the mutations in the genes coding for nodal ciliary motility. Due to this the midline barrier gets disrupted resulting in the separation of left from right which in-turn permits the mixing of molecules that are normally asymmetrically distributed in early embryos. This results in abnormal organ sidedness later in development [2,3,6].

Clinical presentation wise,there were various reports of duodenal perforation, cholecystitis, appendicitis and even sudden death in patients with SIT [7-9]. The diagnosis of patients with situs anomalies is usually incidental, but the greater task in these patients lies in complete work up to rule out other associated anomalies and also to differentiate SIT from isolated situs anomalies of heart and heterotaxy. Image studies of organs, spleen (asplenia, polysplenia); duodenum (malrotation); pancreas (truncated/normal) and heart (dextrocardia, levocardia, ASD, VSD) play a vital role in the diagnosis [7,9].

The surgical management of the situs patients pose a greater challenge because of reversal of surgical procedures due of

the mirror image location of the thoraco-abdominal organs [10-12]. Hence, preoperatively, a thorough assessment should be done to rule out associated thoraco-abdominal visceral, vertebral and cardiac anomalies. A contingency plan and a mock run for resuscitation CPR and defibrillation should be worked out with turn around of procedures, in these patients [13]. During surgery, necessary position changes of Laparoscopic monitors, ports, instruments and relative position of surgical team members are required for successful laparoscopic surgical procedures [14]. Interventional cardiac procedures require a skillful maneuvering of catheters and mirror image views, during transradial or transfemoral approaches in patients with dextrocardia [15]. Further, transplantation of viscera either as a donor or as a recipient of situs anomalies patients is complicated by the geometric rearrangement of structures [15].

CONCLUSION

Situs inversus anomalies should be kept in mind during clinical and radiological examination in patients presenting with unusual signs and symptoms of abdominal region. A thorough evaluation of associated anomalies reduces intraoperative challenges in patients requiring surgical management. Technical modifications intra-operatively should also be considered for successful surgical outcome in these patients.

REFERENCES

[1] Fulcher AS, Turner MA. Abdominal manifestations of situs anomalies in adults. *Radio Graphics*. 2002;22(6):1439–56.

[2] Swarhib M, Das S, Htwe O. A case of situs inversus totalis: embryological and clinical considerations. *Int Med J*. 2013;20(264):5.

[3] Bartram U, Wirbelauer J, Speer CP. Heterotaxy syndrome; asplenia and polysplenia as indicators of visceral malposition and complex congenital heart disease. *Biol Neonate*. 2005;88(4):278–90.

[4] Radhika D, Rekha NS, Mohan KVM. Dextrocardia with situs inversus–a case report. *Int J Anat Var*. 2011;4:88–89.

[5] Mallick IH, Iqbal R, Davies JB. Situs inversus abdominus and malrotation in an adult with Ladd's band formation leading to intestinal ischaemia. *World J Gastroenterol*. 2006;12(25):4093–95.

[6] Bartoloni L, Blouin J-L, Pan Y, Gehrig C, Maiti AK, Scamuffa N, et al. Mutations in the DNAH11 (axonemal heavy chain dynein type 11) gene cause one form of situs inversus totalis and most likely primary ciliary dyskinesia. *Proc Natl Acad Sci.* 2002;99(16):10282–86.

[7] Ortiz RO, Ali MJ, Lopez F. Clinical case of the month: a review of situs inversus and dextrocardia. *J State Med Soc*. 2015;167(2):102-04.

[8] Supriya G, Saritha S, Madan S. Situs inversus totalis–a case report. *IOSR Int J Appl Phys*. 2013;3(6):12–16.

[9] Talabi A, Sowande O, Tanimola A, Adejuyigbe O. Situs inversus in association with duodenal atresia. *Afr J Paediatr Surg*. 2013;10(3):275.

[10] Kyuno D, Kimura Y, Imamura M, Uchiyama M, Ishii M, Meguro M, et al. Pancreaticoduodenectomy for biliary tract carcinoma with situs inversus totalis: difficulties and technical notes based on two cases. *World J Surg Oncol*. 2013;11:312.

[11] Jindal V, Misra MC, Bansal VK, Choudhury N, Garg SK, Khan RN, et al. Technical challenges in laparoscopic cholecystectomy in situs inversus. *J Laparoendosc Adv Surg Tech A*. 2010;20(3):241–43.

[12] Machado NO, Chopra P. Laparoscopic cholecystectomy in a patient with situs inversus totalis: feasibility and technical difficulties. *JSLS*. 2006;10(3):386–91.

[13] Bajwa SJS, Kulshrestha A, Kaur J, et al.The challenging aspects and successful anaesthetic management in a case of situs inversus totalis. *Indian J Anaesth*. 2012;56:295–97.

[14] Arya SV, Das A, Singh S, et al. Technical difficulties and its remedies in laparoscopic cholecystectomy in situs inversus totalis: A rare case report. *Int J Surg Case Reports*. 2013;4:727–30.

[15] Potdar A, Sapkal G, Sharma S. Triple vessel percutaneous coronary intervention in a patient with situs inversus dextrocardia using a transradial approach. *Indian Heart J.* Published Online First: 2016; Mar 25.

AUTHOR(S):

1. Dr. Vidya Gunasekaran
2. Dr. Gladwin Victor Raj
3. Dr. Parkash Chand

PARTICULARS OF CONTRIBUTORS:

1. Senior Resident, Department of Anatomy, JIPMER, Puducherry, Puducherry, India.
2. Associate Professor, Department of Anatomy, JIPMER, Puducherry, Puducherry, India.
3. Professor, Department of Anatomy, JIPMER, Puducherry, Puducherry, India.

NAME, ADDRESS, E-MAIL ID OF THE CORRESPONDING AUTHOR:

Dr. Gladwin Victor Raj,
Associate Professor,
Department of Anatomy, Jawaharlal Institute of Postgraduate Medical Education and Research,
Puducherry-605006, India.
E-mail: drgladwin@gmail.com

FINANCIAL OR OTHER COMPETING INTERESTS: None.

Date of Publishing: **Oct 01, 2016**