Indian Journal of Clinical Anatomy and Physiology 2019;6(3):373–375

Content available at: iponlinejournal.com

Indian Journal of Clinical Anatomy and Physiology

Journal homepage: www.innovativepublication.com

Case Report

Embryological and clinical significance of situs inversus totalis: Rare case presenting with cholelithiasis and choledocholithesasis

William[1,*], S K Goyal[2]

[1]*Dept. of Anatomy, Government Medical College, Kota, Rajasthan, India*
[2]*Goyal Endoscopy & Research Center, Kota, Rajasthan, India*

ARTICLE INFO

Article history:
Received 17-10-2018
Accepted 12-12-2018
Available online 12-10-2019

Keywords:
Situs inversus totalis (sit)
Bile duct stone
Gut rotation. Choledocholithiasis
ERCP
Situs inversus

ABSTRACT

Situs inversus is a rare, autosomal recessive condition that is incidentally discovered. It may occur with dextrocardia, levocardia, or situs ambiguous, the latter with either right or left isomerism, along with asplenia or polysplenia. Incidence of situs inversus varies from 0.002% to 1%. Mostly it occurs in males. [1–3] Situs inversus totalis is the mirror image of the situs solitus, which describe the normal position of the thoracic and abdominal viscera. It may be associated with other genetic diseases such as Kartagener's syndrome. [4] This condition does not affect the life expectancy or quality of life. [1,3] Individuals with situs inversus anomaly exhibit a variety of associated gastrointestinal abnormalities. These anomalies involve liver, biliary tract, stomach, spleen and the intestines. There are few cases in which the anomaly was never detected during lifetime because of the absence of any presenting symptom. Such anomalies may be incidental findings during autopsies or cadaveric dissections. Any mechanical disturbance which occurs in any of the two rotational movements may give rise to abnormal positioning of the heart or dextrocardia. Some researchers defined a gut rotationdetermining factor which is usually located in the left side of the body. [5] The transposition of the abdominal viscera in the present case may be related to malrotation of the gastrointestinal system during the embryonic development. It has been thought that the disturbance of this gut rotation determining factor during ontogenesis may be solely responsible for situs inversus. [5]
Aristotle discovered this alternation in animals while in humans it was described by Fabricius in1600. Vehemeyer first identified situs inverses in 1967 with x-rays. [4,6] First case of SIT was by Venu et al (1985) [3] in America and latest by Kamani L et al (2014) [7] in Pakistan. The exact etiologies remains obscure but attempts have been made to explain it on the basis of a complex gene with variable expression. [6] More than one genetic mutation was implicated in etiopathogenesis. [8]
Mohamed S et al 2013 reported a male case of situs inversus which remained undiagnosed until the age of 50 years. He presented as chest pain and collapsed even before starting treatment and was diagnosed SITS on autopsy. [9]
Clinical manifestation depends on the location of the compromised organ. It does demand greater attention from surgeons and endoscopist because diagnosis can be c.

1. Case Report

A male of 72 years old was brought to Goyal hospital and endoscopic center, Kota on 14th may 2015 with complaint of pain in left upper abdomen with nausea, vomiting. Jaundice was present. On Ultrasonography he was diagnosed to have Cholelithiasis and Choledocholithasis. Liver, gall bladder and CBD duct were present on left side of the body. All the other abdominal viscera like caecum, appendix, ascending colon, duodenum, and pancreas were also on left side of body, while the spleen, splenic flexure, descending colon, sigmoid colon were on right side. So the case of situs inversus totalis was diagnosed which was confirmed with MRCP, echocardiography and CT scan.

ERCP was planned to remove the CBD stones. For this procedure the setup has to change because it is designed for normal anatomy of body. All the instruments and other supporting machines was adjusted for this patient.

* *Corresponding author.*
E-mail address: dr.williammasih@yahoo.com (William).

https://doi.org/10.18231/j.ijcap.2019.081
2394-2118/© 2019 Published by Innovative Publication.

Usually the left lateral position is given to patient but here procedure was done in right lateral position, endoscopic surgeon also stood on right side and c-arm on left side. CBD duct cannulation attempted. Pancreatic duct cannulated and stenting, precut splitting and CBD duct cannulation done. CBD dilated upto 10mm with 10mm canula and cannulated. Stone removed with balloon sweeping. Plastic stent number 7 Fr deployed and kept in situ.

Post-operative was uneventful

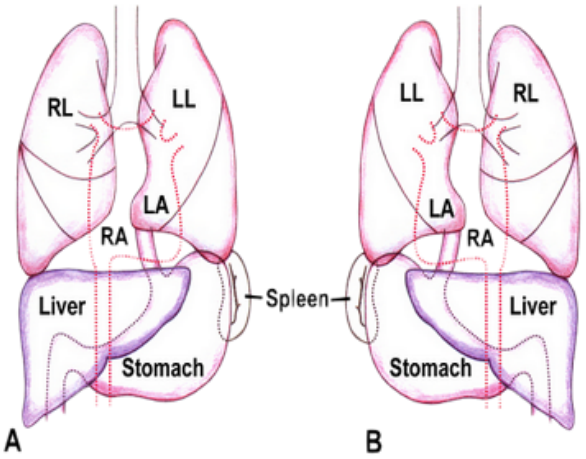

Fig. 1: Situs inverses totalis (SIT)

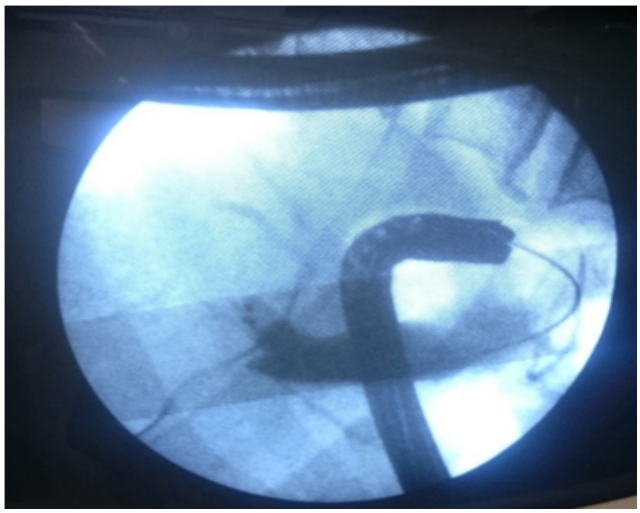

Fig. 2: X-Ray (C-Arm) view checking the position of guide wire in CBD duct

2. Discussion

75 year old male patient having pain in left hypochondrium and signs of biliary obstruction which was confirmed with sonography, MRCP and Echography

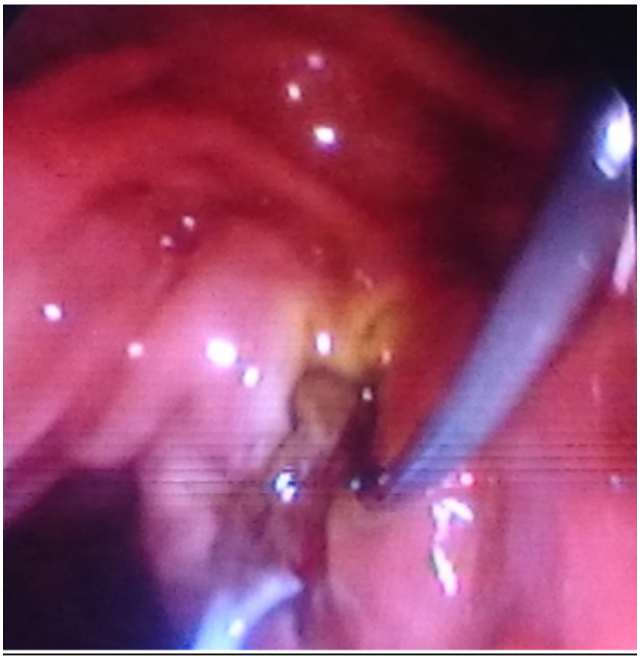

Fig. 3: Endoscope in situ

According to past researchers, situs anomalies exhibit a wide spectrum of disorders in which many of the organs fail to develop in proper symmetry and these are accompanied with malrotation of the intestines.[1]

In 1949 Blegen[10] reported diagnostic error in 45% of all patients with situs inversus and where incorrect incision in 31% patient was given for operation for gall stones. Vehemeyer first identified situs inversus in 1967 with x-rays[4,10] Currently this type of error is minimized because of noninvasive imaging method to arrive at correct preoperative diagnosis as in this case.

Twenty per cent of the cases may be associated with bronchiecstasis and chronic sinusitis due to primary ciliary dyskinesia and this is termed as Kartegenar syndrome.[4] No proper history of any respiratory disease was obtained in the present case.

Nawaz et al reported two extremely rare cases in which the situs inversus abdominus was associated with congenital partial duodenal obstruction secondary to a duodenal diaphragm with a central aperture in one case, whereas the other child had completed duodenal atresia as well as Fallot's tetralogy.[11]

The malposition of the heart in situs inversus can be explained with reference to its development. There are two rotational movements which are involved during the development of the heart. In first phase of embryonic development, 180° rotation occurs around a transverse body axis, which brings the heart-anlage to the future thoracic region.[5] Second phase rotation occurs around the sagittal and longitudinal axis and finally heart is properly positioned

in the mediastinum.[5] The exact cause of situs inversus is not known. It has been suggested that the rotations of the cardiac tube brings about all the other changes that follow. However the fundamental underlying mechanism is still not known. Any mechanical disturbance which occurs in any of the two rotational movements may give rise to abnormal positioning of the heart or dextrocardia.[1,5] The present case can be explained embryo logically. The transposition of the abdominal viscera in the present case may be related to malrotation of the gastrointestinal system during the embryonic development. Interestingly, some researchers defined a gut rotation determining factor which is usually located in the left side of the body[5]). It has been thought that the disturbance of this gut rotation determining factor during ontogenesis may be solely responsible for situs inversus.[5] An earlier study described the positive role of homeobox gene Pitx 2 in the looping mechanism of the heart and the gut.[12]

It may be possible, perhaps, to determine the genetic basis for the concerned genes associated with the rotation in any case of situs inversus. Cockayne (1938) first proposed an autosomal recessive gene which involved in complete transposition of the viscera[13] However, Campbell in 1963 highlighted that there was insufficient evidence to support such a conclusion.[14]

Anatomic anomaly of the patient makes ERCP and operation more challenging and demanding technical skill. Endoscopic procedures such as sphintertomy, stent placement, and stone extraction are more difficult than in normal patient. Crucial factor for ERCP in situs inverses patient is to put the patient in different position during procedure.

The instrumentation, side view endoscope is built for right side position of liver with papilla on medial side of duodenum.

Endoscopic sphincterotomy and stone extraction are standard procedures for the removal of bile duct stones. Therapeutic ERCP for bile duct st ones has a success rate of 96-100%.[15]

SIT increases the technical difficulties and the possibilities of complications of therapeutic interventions. Pathak et al[8] reported a case of ERCP performed in SIT. Keeping the patient in the prone position and the endoscopist at the left side, but that procedure was done with rotation of the instrument. In this case, we succeeded to pass the stomach and to reach the duodenum with 180 degrees turn and complete the sphinterotomy and stone extraction.

3. Conclusion

In the present case, situs inversus totalis was explained embryologically. Endoscopic sphincterotomy and stone extraction are standard procedure for the removal of bile duct stones. The instrument for the side view endoscope is built for the right position of the liver with the papilla at the medial side of the duodenum. While this report shows that an experienced endoscopic surgeon can achieve the same results in the conventional way as it is possible when anatomical changes occur as in this case.

4. Source of funding

None.

5. Conflict of interest

None.

References

1. Sadler TW. Langman's medical embryology. London: Lippincott Williams and Wilkins ; 2010,. 8th edn.
2. Melchor JM, Perez R, Torres M, Rodiguez V. Situs inversus: Reporte de dos casos. *Cir Ciruj*. 2000;68:72–75.
3. Venu RP, Geenen JE, Hogen W, Johnson GK, Tayler AJ, Stewart ET. ERCP and endoscopic sphincterotomy in patients with situs inversus. *Gastointestinal Endosc*. 1985;31:338–340.
4. Kinney TB, Deluca SA. Kartageners syndrome. *Am Farm Physician*. 1991;44:133–139.
5. Considerations. *Int Med J*. 2013;20(2):264–265.
6. Embryologie DS, Auflage N, Stuttgart. Thieme ; 1965,. .
7. Garca-Fernndez FJ, Infantes JM, Torres Y, Mendoza FJ, Alcazar FJ. ERCP In complete situs inversus viscerum using a mirror images technique. *Endoscopy*. 2010;42(2):316–317. suppl.
8. Pathak KA, Khanna R, Khanna N. Situs inversus with cholelithiasis. *J Postgrad Med*. 1995;41:45–46.
9. Cockayne EA. The genetics of transposition of the viscera. *Quarterly Med*. 1938;(7):479–479.
10. Fiocca F, G D, Ceci V, Cereatti F, Simonelli L. ERCP in total situs inversus viscerum. *Case Rep Gasroenterol*. 2008;2:116–120.
11. Kamani L, Kumar R, Mahmood S, Jafri S, Siddiqui E. Therapeutic ERCP in patient with situs inversus total is and ampullary diverticulum. *J Coll Physcian Surg Pak*. 2014;24:365–366.
12. Nawaz A, Matta H, Hamchou M. Situs inversus abdominus in association with congenital duodenal obstruction: a report of two cases and review of the literature. *Pediatr Surg Int*. 2005;21(7):589–592.
13. Campione M, Steinbeisser H, Schweickert A, Deissler K, Bebber FV, Lowe LA. The homeobox gene Pitx 2: mediator of asymmetric left-right signaling in vertebrate heart and gut looping. *Dev*. 1999;126:1225–1234.
14. Mohamed S, Srijit D, Ohnmar H. A Case of Situs Inversus Totalis: Embryological and Clinical A Case of Situs Inversus Totalis: Embryological and Clinical Considerations. *Int Med J*. 2013;20(2):264–265.
15. M AJ, Hoche F. Laparoscopic cholecystectomy in situs inversus totalis: is it safe? *J Laparoendosc Adv Surg Tech A*. 2001;11:229–231.

Author biography

William Professor

S K Goyal Senior Surgeon

Cite this article: William , K Goyal S. Embryological and clinical significance of situs inversus totalis: Rare case presenting with cholelithiasis and choledocholithesasis. *Indian J Clin Anat Physiol* 2019;6(3):373-375.