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Case Report

A unique case of fetal situs inversus totalis with associated malformations

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ABSTRACT

Situs inversus totalis is a congenital positional anomaly characterized by transposition of abdominal and thoracic viscera. It refers to inverted position of internal organs. It can be either total or partial. Situs inversus totalis is also termed as Situs inversus with dextrocardia. Here, we are reporting autopsy findings of an anomalous male fetus of 17 weeks gestation born to a 27 years old woman who was gravida 3, para 1 and abortion 1. Prenatal ultrasound was done and was diagnosed as Situs inversus totalis associated with cleft lip and palate, short corpus callosum. After termination of pregnancy in Dept of Obstetrics and Gynecology AIMS Hospital fetus was sent to Anatomy department for autopsy. The detailed information was noted down from the case sheet of mother. The formalin was injected to cranial, thoracic and abdominal cavity. After fixation, the autopsy was done following the standard protocol. It was noted that thorax viscera were trans positioned, left lung was trilobed, right lung was bilobed and cardia on right side of thoracic cavity. The abdominal viscera were trans positioned with liver, gall bladder, caecum, appendix, ascending colon on left side and stomach, spleen, pancreas, descending and sigmoid colon on right side. Midline malformations, such as cleft lip and cleft palate were noted. Brain was not fixed well and corpus callosum could not be visualized. Situs inversus totalis is the rarest congenital anomaly. Exact etiology is unknown. Its incidence is 0.01%. Various modalities such as Ultrasonography, computed tomography can be used to diagnose situs inversus.

Keywords: Fetal autopsy, Formalin, Dextrocardia, Left lung trilobed, Cleft lip, Cleft palate

INTRODUCTION

Situs inversus is a congenital condition in which the major organs of the thorax and abdomen are reversed or mirrored from their normal positions. Situs inversus is a short form of the Latin phrase “Situs inversus Viscerum” meaning “inverted position of the internal organs, as first described by Marco Severino in 1643.[1]

It is generally classified into three: Situs solitus in which the organs are in normal position. Situs inversus is a congenital anomaly characterized by transposition of abdominal and thoracic viscera. It can be either complete - Situs inversus totalis (dextrocardia) or partial (levocardia)- Situs abdominis and Situs ambiguous in which there is visceral malposition. Situs inversus was first described by Aristotle in animals.[2] Matthew Baillie described the complete mirror-image reversal of the thoracic and abdominal organs in situs inversus. The incidence has been reported to vary widely between 1 in 4,000 to 20,000 live births, with a male: female ratio - 3:2.[2] Situs inversus can occur alone without any other abnormalities or it can be a part of a syndrome with various other defects such as Kartagener Syndrome or Primary Ciliary Dyskinesia. It is characterized by bronchiectasis, sinusitis, and situs inversus and affects 20% of patients with situs inversus.[1,2] However, only 50% of patients with Kartagener syndrome have situs inversus.[2] The exact etiology is not proven but it has been inherited in different ways in different families. Autosomal recessive and X-linked inheritance have been

reported. Prenatal diagnosis is done by ultrasonography and in adults by X-ray of chest and computed tomography of abdomen.[3]

CASE REPORT

We hereby report autopsy findings of an anomalous fetus born to a woman aged 27 yrs with the obstetric history of G3P1L1A1 reported with a to the obstetric department at Adichunchanagiri Institute of Medical Sciences, B.G Nagara, on 11thJuly 2024. The detailed history was taken. She gave h/o amenorrhea 6 months. she had taken two doses of tetanus toxoid. She was on folic acid, iron and calcium supplementation. She gave h/o one abortion of anomalous fetus at 16 weeks and details were not available. There was no h/o of consanguineous marriage, no h/o diabetes or hypertension. There was no history of smoking, alcohol consumption and exposure to teratogenic drugs. The obstetric scan report showed single live intrauterine gestation of 17 weeks 3 days and the anomaly scan showed fetus with cleft lip and cleft palate, short corpus callosum and Situs Inversus Totalis. The concerned obstetrician decided for medical termination and induction of labor was done. She aborted male fetus of 340 grams on 13th July 2024.

Autopsy report

The baby was sent to department of anatomy for autopsy after a written consent from the woman. The prenatal ultrasonographic reports and other investigation reports were collected. The formalin was injected into the cranial, thoracic and abdominal cavity. After fixation, autopsy was done according to the standard protocol.

External features

Male fetus weighed about 340 grams with placenta, 265.4 grams without placenta, height was 21 cm, head circumference was about 15 cm, mid arm circumference was 3 cm, abdomen and chest circumference measured about 13 cm and 13.5 cm respectively.

The autopsy findings are as follows.

Head and neck

Cleft lip and cleft palate were noted. Brain was not fixed well, so corpus callosum could not be appreciated.

Thorax

Heart was on the right side of thoracic cavity the left lung was trilobed and right lung was bi lobed.

Abdomen

Right side of abdominal cavity showed following organs, fundus of stomach with greater curvature, duodenum & pancreas and spleen, and descending colon, sigmoid colon. Left side of abdomen showed liver, gallbladder, caecum & appendix and ascending colon.

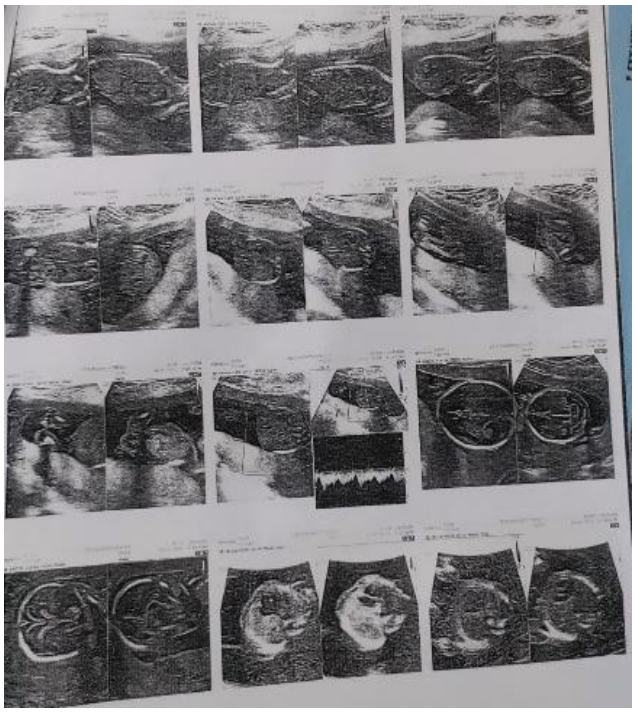

Figure 1: The prenatal ultrasound scan report.

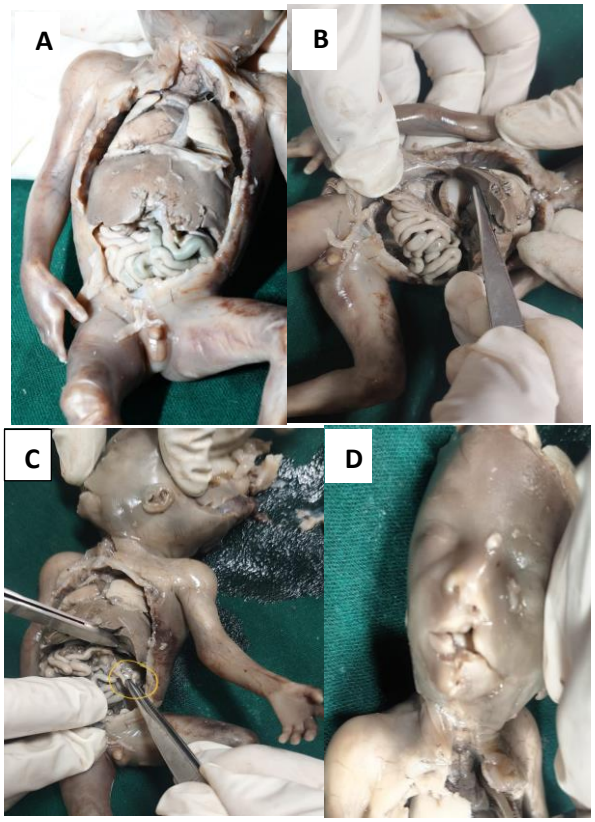

Figure 2: A) Situs inversus totalis. B) Dextrocardia and liver on left side. showing fundus of stomach on right side. C) Appendix on left side. D) Cleft lip.

DISCUSSION

Situs inversus totalis is the rarest congenital anomaly in which the organs of the chest and abdomen are arranged in a perfect mirror image reversal of the normal positioning. This condition was uncovered in cadavers in 1600 by Fabricus where he described the reversal of the liver and spleen in a patient.[3] It is common in age group of 19 to 45 years (28.8 %) and 46 to 65 years (29.9%).[4] In 1897, Vehsemeyer demonstrated the transposition of the viscera using X-rays.[3] The clinical significance of Situs inversus was not grasped until the advent of X-ray imaging which made diagnosis in humans easy and reliable.[1]

Most people with situs inversus can live a normal life without symptoms or disability. However, there are lot of comorbidities of varying frequency associated with situs inversus. Several cardiovascular, gastrointestinal as well multisystemic comorbidities like duodenal atresia, biliary atresia, gastroschisis with malrotation, congenital coronary abnormalities, ventricular septal defects etc. have been shown to be associated with this condition.[4]

Table 1: Comparison of autopsy reports with other studies.

S. no	Author year	Period of gestation	Sex of baby	Condition of fetus/ neonate	Investigation reports	Autopsy
1	Julie et al[5] (2018)	33 Weeks	Male	Died after birth	X-ray chest	Situs inversus totalis
2	Sharada Sharma[1] (2021)	27-30 Weeks	Female	Dead fetus	X-ray and Abdominal USG	Situs inversus totalis
3	Jordan Devera[6] (2021)	Preterm	-	Shifted to NICU due to wet lung	X-ray USG	-
4	Abnish Kumar[7] (2014)	7 Months	Female	Respiratory distress	Chest X-ray	Situs inversus totalis
5	Nagraj Jawali[8] (2015)	Full Term	Female	Admitted to NICU due to respiratory distress and shock	Chest X Ray and Abdominal Scan	Situs inversus totalis
6	Present study (2024)	17 Weeks 04 days	Male	Aborted fetus	USG Anomaly scan	Situs inverus totalis cleft lip and palate

The present case autopsy findings are comparable with the case reported by Sharada S et al.[1] Where as in other studies, there were preterm babies admitted with respiratory distress and later died and autopsy findings are reported. Jordan D et al, reported a preterm baby admitted to NICU with respiratory distress, situs inversus totalis was diagnosed after X-ray chest, and later referred to higher centres.[6] Abnish K et al, reported a 7 months old female baby with severe respiratory distress.[7] Investigations done and situs inversus totalis was confirmed. Baby later died and autopsy was done.

Abdullah Al Saleh et al, reported a boy aged 5 years, who was healthy with no medical complaints, but was diagnosed with situs inversus totalis.[9] John Osaretin Osaren Khoae et al, reported a review of 191 published cases of situs inversus stating that situs inversus totalis comprises 82.7%, situs inversus ambiguous up to 13.6% and situs inversus abdominis with levocardia up to 9.4%.[4]

Embryological basis

The embryological basis for situs inversus totalis can be "Laterality disorders" caused due to multifactorial, genetic and environmental factors. It is associated with ciliary abnormalities as well as the genes NODAL and PITX 2.[10] Embryonic rotation of midgut in opposite direction leads to situs inversus.

Failure of bulboventricular loop

A bulboventricular loop to migrate into left hemithorax may result in dextrocardia. The absolute cause of situs inversus totalis is unknown.

CONCLUSION

In our study, we observed an aborted fetus showing dextrocardia with transposition of both thoracic and abdominal viscera. The patient with situs inversus totalis may be associated with extra cardiac anomalies. The exclusive part of our study was midline malformations like short corpus callosum, cleft lip and cleft palate were noted along with situs inversus totalis. The etiological cause of this case is unknown.

Surgeons and radiologists need to be aware of this anomaly during the preoperative and surgical management. Routine premedical examination helps the patient to be aware of his condition, thereby preventing wrong diagnosis possibly death due to delay in surgical management

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