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Case Report

Situs Inversus Totalis: A Rare Case Report with Review of Literature

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Received: June 11, 2021; **Published:** October 28, 2021

Abstract

Situs inversus is a rare congenital condition in which the major visceral organs are reversed or mirrored from their normal positions. It involves complete transposition (right to left reversal) of the thoracic and abdominal organs. The association of neonatal intestinal obstruction with situs inversus totalis is extremely rare with only a few cases reported in the literature to this date. Associated other congenital anomalies may lead to a poor survival rate. We describe a neonate with an antenatally detected duodenal obstruction who also had situs inversus totalis and other intra-cardiac defects.

Keywords: *Situs Inversus; Neonate; Congenital; Intestinal; Mortality*

Abbreviations

SI: Situs inversus; SIT: Situs Inversus Totalis; USG: Ultrasonography; DBS: 'Double-Bubble Sign; DO: Duodenal Obstruction; SNCU: Sick Newborn Care Unit; OT: Operation Theatre; APGAR: Color (Appearance), Pulse (Heart Rate), Grimace (Reflexes), Activity (Muscle Tone) and Respiration; PFO: Patent Foramen Ovale; IPPV: Intermittent Positive-Pressure Ventilation; DA: Duodenal Atresia; POD: Postoperative Day; CHD: Congenital Heart Disease; TR: Tricuspid Regurgitation

Introduction

Situs inversus (SI) occurs in 1 to 2 in 10,000 live births. Situs inversus totalis (SIT) involves complete transposition (right to left reversal) of the thoracic and abdominal organs [1]. The association of congenital anomalies with neonatal intestinal obstruction is extremely rare with only a few cases reported in the literature to this date. We describe a neonate whose antenatal USG showed a 'double-bubble sign' (DBS), suggestive of duodenal obstruction (DO), and got incidentally diagnosed as SIT on postnatal echocardiography.

Case Presentation

A day 1 male neonate of gestational age 38 weeks and body weight 2450 gram admitted to the SNCU, directly from the Labour room OT due to antenatal USG showed DBS with distended stomach (Figure 1). His mode of delivery was an elective cesarean section. The baby cried immediately after birth and had APGAR scores of 8 and 9 at one and five minutes respectively. There was no history of maternal diabetes, drug abuse, polyhydramnios, consanguinity, or family history of congenital anomalies. Initially, the neonate was active, euther-

mic, euglycemic, and hemodynamically stable. Chest auscultation revealed cardiac apex to be on the right 4th intercostal space along the midclavicular line. 2D echocardiography showed dextrocardia, patent Foramen Ovale (PFO) measuring 6 mm in size with a left to right shunt and tricuspid regurgitation gradient of 54 mm Hg suggestive of persistent pulmonary hypertension of newborn. The baby passed meconium within 24 hours of life. From day three, the neonate developed abdominal distension and bilious orogastric aspirates. Subsequently, he developed tachypnoea with labile oxygen saturation. The babygram revealed dextrocardia with stomach bubble on the right side, liver opacity on the left side with few distended bowel loops in the upper abdomen, and paucity of gas shadows in the lower abdomen (Figure 2). USG whole abdomen revealed the liver and gall bladder situated in the left hypochondrium and stomach and spleen in the right hypochondrium with both kidneys being normal. After pediatric surgery consultation, preoperative preparations were done and informed consent was taken from the parents for surgery. The operation was done under general anesthesia with IPPV and rapid sequence induction was done. On exploration, we found the liver and duodenum on the left side, while the stomach and spleen on the right side (Figure 3). There was duodenal atresia (DA), annular pancreas, and malrotation of the gut. Ladd's procedure and duodenoduodenostomy (diamond anastomosis) were done. The baby was put on ventilatory support due to hemodynamic instability and was administered vasopressor infusion. On 2nd POD, the baby was passing greenish mucoid stool. But gradually the clinical condition deteriorated and the baby developed fulminant septicemia with a subsequent downhill course and unfortunately expired on the 4th POD.

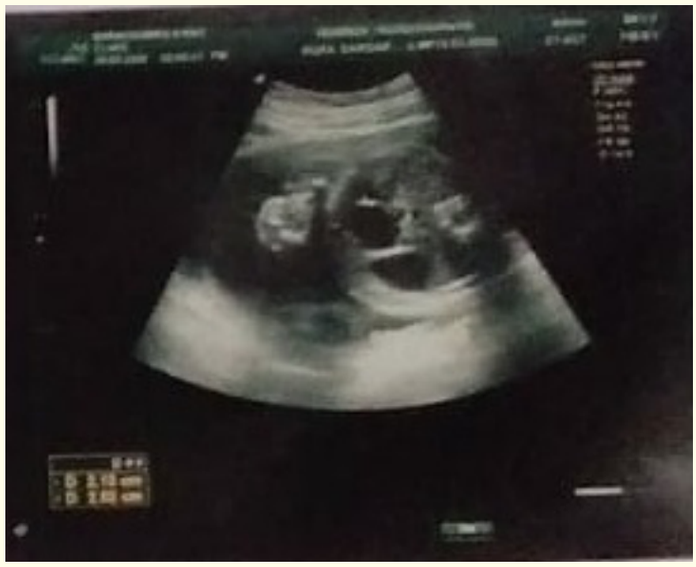

Figure 1: *An antenatal ultrasonography, at 20th weeks of gestation showing "Double Bauble Sign" with a distended stomach.*

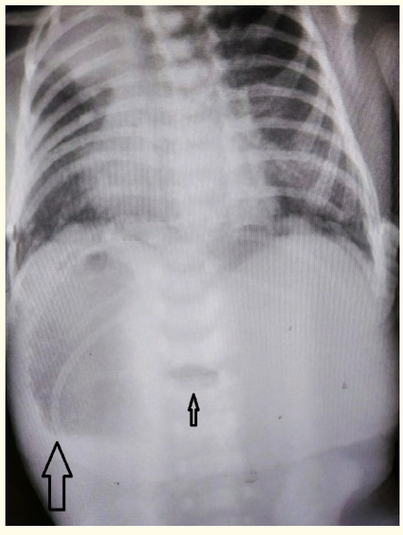

Figure 2: *A baby-gram on 2nd day of life showing dextrocardia, Double Bauble Sign with stomach bubble on the right side, liver opacity on the left side and absent gas shadows in the lower abdomen.*

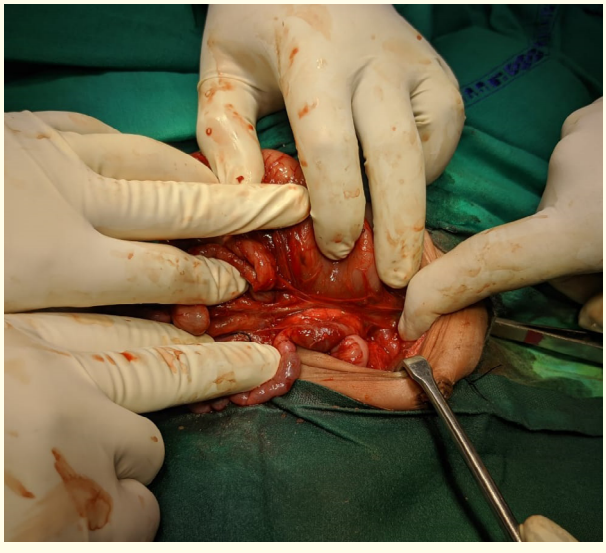

Figure 3: *Per-operative picture showing the duodenal C-loop on the left side, the stomach and spleen on the right side and an annular pancreas in a 7-day old neonate with Situs inversus.*

Discussion

Fabricius first described the SI in humans, while Vehemeyer provided radiological evidence. Three types of situs have been reported in the literature. These are- (i) situs solitus, (ii) Situs inversus, and (iii) situs ambiguous [2]. In SIT, there is a complete right to left reversal of all of the viscera including dextrocardia; the morphologic right atrium is on the left and the left atrium is on the right. The normal pulmonary anatomy is reversed such that the left lung has three lobes and the right lung has two. The liver and gallbladder are located on the left, and the spleen and stomach are on the right side.

Multiple factors are responsible for these anomalies; gestational diabetes, maternal drug abuse, retinoic acid exposure, conjoined twinning, consanguinity, and familial inheritance [3]. Eroschenko reported that SI is generally an autosomal recessive genetic condition, although it can be X-linked [4]. In our case, there was no history of consanguinity, family history of congenital anomalies, gestational diabetes, or maternal drug exposure.

A patient with SI may remain asymptomatic and unaware until he/she undergoes laparotomy for a surgical cause. A SI may be associated with duodenal atresia, annular pancreas, biliary atresia, preduodenal portal vein, diaphragmatic hernia, lung cyst, genitourinary anomalies, ear, eye, and vertebral defects [5]. In our case, the baby had duodenal atresia with an annular pancreas and malrotation of the gut. The diagnosis can be made by radiographs, upper gastrointestinal contrast studies, abdominal ultrasound, and laparotomy for a specific indication, as we see in our case. The prevalence of association of CHD with SIT is about 5 - 10% of cases and thus, echocardiography is important and recommended [3,4]. In our case, the baby had a 6 mm PFO with a Left to Right shunt and a TR gradient of > 40 mmHg suggestive of persistent pulmonary hypertension of the newborn.

The incidence of DA is 1 in 4000 to 1 in 15000 live births, and about half of them are associated with other organ anomalies. The association of DO and SIT is extremely rare, with only 20 cases reported in the literature to date [6]. The management of DA/ malrotation with or without associated SI follows the same principles [7]. The classic treatment for malrotation is Ladd's procedure. If the duodenal obstruction is caused by an intra-luminal web, excision with duodenoplasty is recommended. Duodenoduodenostomy is preferred to avoid iatrogenic biliopancreatic injury because of the proximity of the ampulla of Vater to the site of the duodenal web [8].

The survival rate is low when SI is associated with other congenital anomalies, so prenatal diagnosis is important [9]. Doctors should be aware of this anomaly, as preoperative diagnosis helps to prevent surgical mishaps that may occur due to failure to recognize the reversed anatomy or atypical history [10]. In our situation, the baby had an unfavorable course because of a slight delay in the diagnosis, gangrenous area in the gut due to atresia, and post-operative septicemia.

Conclusion

The surgical importance of these anomalies is that it poses a problem in the interpretation of the clinical signs and symptoms. Hence, it causes a dilemma in diagnosis. Incisions of approach to organs to be operated upon have to be planned according to the mirror image transposition of organs and viscera in dextrocardia with SI.

Acknowledgement

Dr. Tapas Kumar Sabui [MD, DCH, MNAMS] Professor and Head, Department of Pediatric Medicine, R G Kar Medical College, Kolkata, India.

Conflicts of Interest

Nil.

Sources of Support/Funding Support

Nil.

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Volume 10 Issue 11 November 2021