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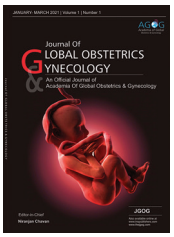
Case Report



Antenatal Journey and Outcome in a Patient with "Situs Inversus Totalis"

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ABSTRACT

Complete situs inversus is a rare condition, inherited in an autosomal recessive pattern, caused by the inversion of cardiac tube rotation during embryonic development. It affects about 1 in 10,000 births. We hereby report an interesting case where situs inversus was identified during the antenatal period and had an uneventful outcome. A 28-year-old woman, G2P1L1, with 32.3 weeks pregnancy following a spontaneous conception presented to the outpatient department of a tertiary care hospital for antenatal care registration with a recent diagnosis of situs inversus totalis. With regular follow-up delivered a full-term baby by cesarean section at 39.1 weeks gestation. Despite the rarity of this condition, it resulted in an uneventful pregnancy course.

Key words: Complete situs inversus totalis, C-section, Pregnancy

INTRODUCTION

Aristotle first identified this phenomenon in animals, and Fabricius later observed it in humans. Typically, thoracic and abdominal organs are situated on one side of the midline. For example, the heart, aortic arch, stomach, and spleen are on the left, while the liver and gallbladder are on the right. This typical left-right positioning of asymmetrical organs is known as situs solitus. When this left-right symmetry is disrupted, it leads to situs inversus. Situs inversus can be either total or partial. In total situs inversus, also called mirror-image dextrocardia, the heart is on the right side of the midline, while the liver and gallbladder are located on the left. The precise cause of this condition remains unclear, but it is thought to follow an autosomal recessive inheritance pattern.[1]

CASE REPORT

A 28-year-old woman, G2P1L1 with 32.3 weeks of gestation with spontaneous conception with previous Fagerström test for nicotine dependence presented to the outpatient department for antenatal care registration at Sion Hospital in June 2024, with a recently diagnosed case of situs inversus totalis. At 24 weeks of gestation, the patient was admitted to Shatabdi Hospital i/v/o acute febrile illness and moderates anemia. Ultrasound abdomen was done s/o liver and gallbladder situated on the left side, spleen and heart situated on the right. Chest X-ray with abdominal shield was done s/o dextrocardia. The patient was referred from there for further evaluation and management at a tertiary care hospital.

On physical examination, pulse rate of 88/min with a blood pressure of 120/70 mmHg. On auscultation, normal heart sounds were heard in the right 5th intercostal space. Electrocardiogram (ECG) showed predominantly negative P wave, QRS complex and T wave in lead 1, positive QRS in lead a VR, right axis deviation, and reverse R wave progression in pre-cordial leads. The echocardiogram revealed situs inversus dextrocardia, normal chambers, valves, normal left ventricular (LV) function, no pulmonary arterial hypertension, with LV ejection fraction of 60%. Fetal echocardiogram was normal. The patient consistently attended her follow-up visits, during which all antenatal investigations were within normal limits. The patient was asymptomatic throughout the pregnancy. At 39.1 weeks of gestation, a patient came to the emergency department with complaints of pain in abdomen. An emergency lower segment cesarean section was performed under spinal anesthesia due to fetal distress during labor. A full-term female baby weighing 2.7 kg was delivered. The intraoperative and post-operative course was uneventful, and the mother was discharged on the fifth post-operative day. Initial assessments of the newborn, including ultrasonography and echocardiography, were conducted and showed no abnormalities. Her 6-week post-partum follow-up was normal.

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DISCUSSION

The word "Situs" describes the arrangement of the cardiac atria and internal organs with relationship to each other. In situs solitus, the organs are in their typical positions. In situs inversus, however, the arrangement is reversed; the morphologic right atrium is situated on the left side, and the left atrium is on the right. This reversal extends to the lungs as well, where the left lung may have three lobes and the right lung may have two which is normally not so. In addition, we see that in situs inversus, the liver and gallbladder are on the left side, while the stomach and spleen are on the right.[2]

Situs inversus is a rare condition that is encountered in approximately 1 in 10,000 live births.[3] Complete situs inversus is even less common and is inherited in an autosomal recessive manner. It affects both sexes equally and shows no variation among different races. Recent research indicates that defects in left-right asymmetry are likely due to genetic abnormalities.[3]

Situs inversus can be categorized as either partial or total. It is further subdivided into two types: Situs inversus with levocardia and situs inversus with dextrocardia. In the latter (also known as situs inversus totalis), the heart, atrial chambers, and abdominal organs are arranged in a mirror image of the normal anatomy. Most individuals with situs inversus present with dextrocardia as their sole cardiac anomaly.[4]

The incidence of congenital heart diseases, such as atrial or ventricular septal defects, tetralogy of Fallot, and pulmonary artery hypoplasia, in patients with situs inversus and dextrocardia, is relatively low, ranging between 3 and 5%. Among these patients, 80% have a right-sided aortic arch.[2] In contrast, situs inversus with levocardia is a rare entity and is almost always associated with congenital heart disease. Conditions, such as Kartagener's syndrome, asplenia, and polysplenia are often observed in association with situs inversus with levocardia. Kartagener's syndrome, which affects 20% of individuals with situs inversus, is characterized by bronchiectasis, sinusitis, and situs inversus. However, only 50% of those with Kartagener's syndrome have situs inversus.[2] In our case, the patient did not exhibit any symptoms that were suggestive of Kartagener's syndrome.

Diagnosis relies on clinical examination and various diagnostic methods, such as electrocardiograms, echocardiography, radiography, ultrasonography, computed tomography scans, magnetic resonance imaging, and barium studies.[1] In our study, we diagnosed the condition using ultrasonography and echocardiography.

The prognosis for isolated dextrocardia depends on any structural or functional defects, as it is often linked to severe congenital anomalies. However, life expectancy in patients with situs inversus totalis with dextrocardia is similar to the general population.[3] Our patient had no associated congenital abnormalities and was asymptomatic during pregnancy.

Situs inversus often goes undiagnosed unless discovered incidentally during investigations for other conditions.[3] Our patient, evaluated for acute febrile illness and anemia, was diagnosed with situs inversus through a chest X-ray and abdominal ultrasound. Her pregnancy and post-partum period were uneventful.

CONCLUSION

In cases of situs inversus totalis, pregnancy typically proceeds without complications, provided there are no associated congenital anomalies. With careful monitoring, most women with this condition can expect a normal pregnancy and post-partum experience.

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