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Case Report

Situs Inversus Totalis a Case Report

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ABSTRACT

Situs inversus totalis is a rare congenital autosomal recessively inherited condition in which there occurs transposition of all viscera with right sided heart (Dextrocardia) in a mirror image form. The term situs, inversus is a latin origin meaning inverted position of internal organs. Fabricus first described dextrocardia in 1600 AD, but severinus was the first to describe it with a complete situs inversus. Incidence lies between 1 in 10,000. This condition has linked with conditions like conjoined twinning, maternal diabetes, this condition may also associated with primary ciliary dyskinesia, congenital heart disorder and splenic abnormalities.

Keywords*: Situs Inversus Totalis, Dextrocardia, Autosomal Recessive Inheritance, Primary Ciliary Dyskinesia, Congenital Anomalies.*

CASE SUMMARY I REPORT:
A 27 year old female with $G_2P_2L_2$ of of obstetric score, admitted for safe confinement, delivered and ultrasound done to rule out Retained product of conception (RPOC).

On General examination:

- Pallor was present (Mild)
- Blood pressure recorded was 120/ 80 mm/Hg.
- On cardio vascular system examination – $S_1$and S_2 heart in right 5th intercortal space.
- On respiratory system examination – Bilateral air entry was present, no added rounds.
- On central nervous system examination - Patient is consious, alert and well oriented to time, place, person.
- On per abdominal examination: 1) Abdomen is soft. 2) No tenderness was present. 3) Stria albicans were present. 4) No scars present. 5)No organo megaly. 6) No dilated veins. 7) All quadrants moves equally with respiration.

Investigations:
Ultrasound abdomen – Urinary bladder empty folleys bulb in situ.
Uterus – Bulky, post-partum status.
Impression – Situs inversus (spleen on right side and liver on left hypochondria)

MRI Abdomen:
Liver is noted on left side of abdomen and spleen on right side. Dextrocardia noted with cardiac apex on right side.

CBC
Total WBC – 7000 cells / mm3
RBC count – 3.01millon / cumm
Platelet – 0.52 lakh / cumm (52,000)
Nuetrophils – 80%
Lymphocytes – 15%

Liver function test
Total Bilirubin – 1.8mg/ dl
Direct Bilirubin – 1.0mg/dl
Indirect Bilirubin – 0.8 mg/dl
SGOT – 67 Iu/L
SGPT – 203Iu/L
ALP – 172 Iu/L
Total protein – 5.0 g/dl
Serum albumin – 2.9 g/dl

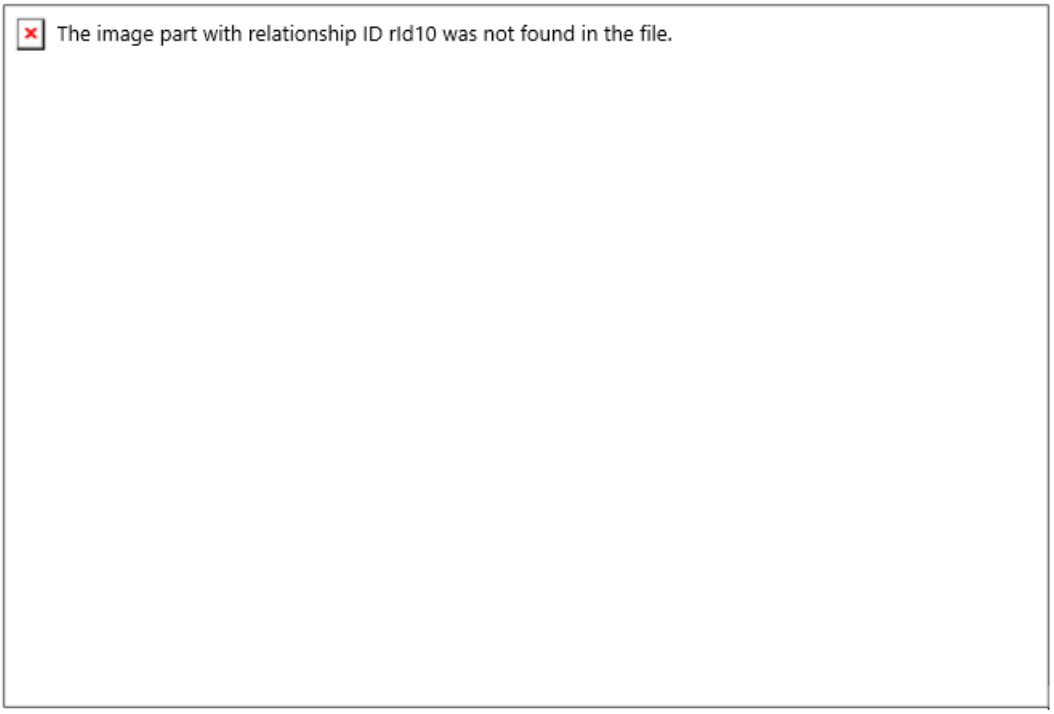
- Serum electrolytes – Within normal range
- Serum creatinine – 0.6 mg/dl
- Blood urea – 23 mg/dl
- C –Reactive protein – 2.5
- LDH – 405.75 U/L

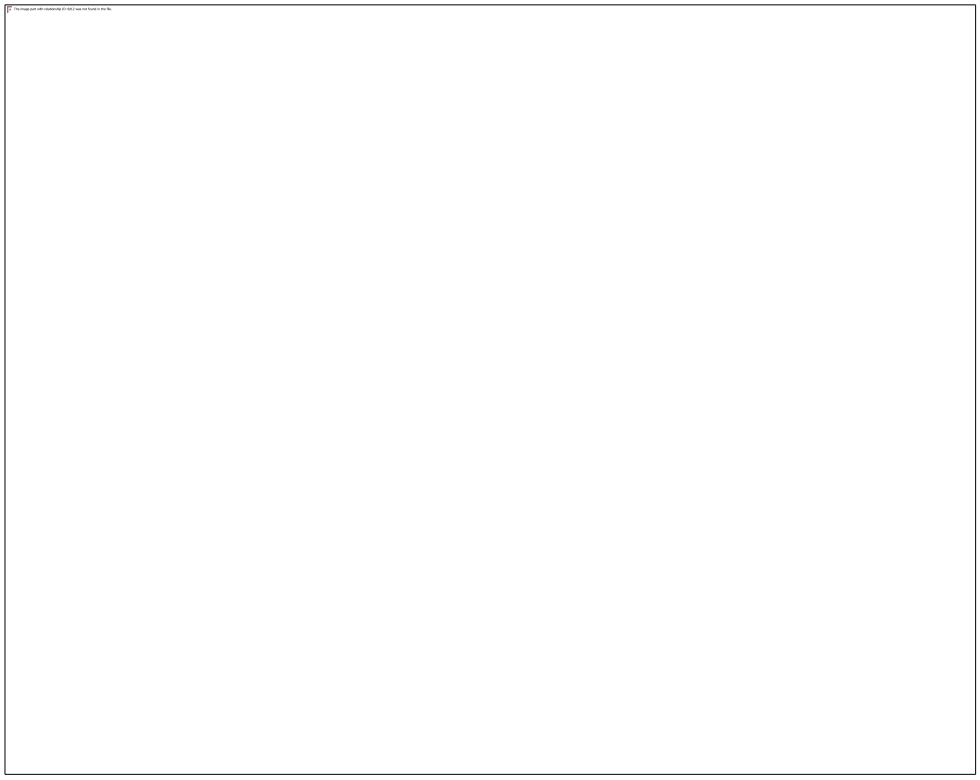
Thyroid profile
T3 - 0.3 ng/ml
T4 – 5.7 ug/dl
TSH – 1.12 uUl/ml

MRI FINDINGS

S

ECG FINDINGS





INTRODUCTION

Situs inversus totalis is a rare congenital condition characterized by a complete mirror – image reversal of the normal positioning of the internal organs.

Although the precise cause is unknown many familiar have experienced different inheritance patterns. Situs inversus may be the only anomaly present or it may be part of a syndrome that includes several other disorders.

Although, it's rare condition, understanding this anomaly is crucial due to it's unique challenges in clinical diagnosis, management and treatment due to its atypical presentation and potential for misinterpretation on imaging studies. Due to the uncommon occurrence of situs inversus totalis and limited understanding of the clinical implications of dextrocardia

with situs inversus, there is a critical need to document and analyse the individual cases to improve recognition, diagnostic accuracy and patient out comes.

DISCUSSION:

SIT is a rare condition characterized by complete mirror image reversal of the normal positioning of the internal organs. The typical or normal arrangement of the organs, where the stomach and spleen are on the left side of the abdomen, the liver and gallbladder is on the right and heart is on the left side of the thorax is reffered as situs solitus.Partial or complete situs inversus are both possible.

Situs inversus with dextro cardia is another name for SIT. It is distinguished by having the liver on left side of abdomen, the heart on the right side of the thorax.

The right lung has two lobes and the left lung has three, this condition is known as situs inversus incompletus or situs inversus with levocardia.

Situs inversus is a rare congenital abnormality occurring in approximately 0.001 to 0.01 of the population. It is inherited in an autosomal recessive manner.

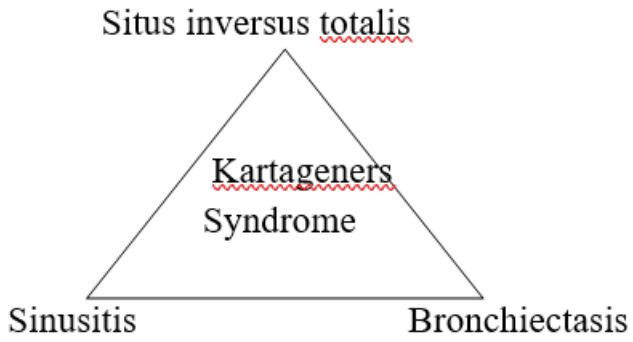
Situs inversus is the out come of the viscera and organs rotating in the other direction during the embroy's organogenesis. A series of signal molecules and genes co ordinate to establish laterality early in development.

A series of signal molecules and genes co- ordinate to establish laterality early development. Szenker – Ravi etal discovered a set of genes encode extra cellular proteins that directly contribute to the establishment of the left – right axis in animal species with cilia in left – right organisers. Defects in these processes generates heterotaxy, the aberrant development and arrangements of organs across the left – right axis, which can range from full inversion of the symmetry to selective organ miss arrangement.

Many persons who have situs inversus totalis are not aware of their unique anatomy until they go to the physician for anything unrelated.

While it may not cause any significant health problems on its own, it can have clinical importance in certain situations. This condition can also complicate surgical procedures as surgeons need to adopt their approach accordingly.

- When dextracardia is present in situs inversus there is a 3 – 5% chance of additional congenital cardiac disorders, typically involving the transposition of the great vessels.
- Kartageners syndrome, which compromises situs inversus totalis, sinusitis and Bronchiectasis is seen in patient with primary ciliary dyskinesia.



- No clinically significant cardio pulmonary symptoms are noted in our patient.
- There are also been reports of renal agenesis in some situs inversus cases.

Organ transplantation in SIT

It presents a unique challenge due to mirror image reversal of their organs. Surgeons must carefully assess the changed anatomy using modern imaging investigations and have particular surgical expertise to negotiate the challenges of transplant procedures in these patients.

Post transplant care and surveillance are critical for SIT people as they may experience a higher risk of complications that need rapid intervention.

Genetic Counselling for people with SIT
Situs inversus totalis is an autosomal recessive disorder, a child cannot have the condition unless both parents have the mutated genes for those with situs inversus totalis and their families, genetic counselling may be advised to help them comprehend the likelihood of passing the condition on to further generations. Genetic counselling for patients with SIT and their families can provide valuable information about the condition, assess the risk of inheritance, offer emotional support, assit with family planning decisions, help develop personalized medical management plans.

CONCLUSION:
SIT is uncommon condition in which the body's internal organs are mirrored from their normal locations. The exactcause is still unknown. Situs inversus is often undiagnosed unless it is incidentally discovered during investigations for other medical condition. To arrive at diagnosis it is crucial to thoroughly and systematically assess patients.

Physicians should be alerted to search for further associated abnormalities. When they find one congenital anamoly. correct diagnosis is crucial for interpretation of further symptoms and other diagnostic procedures, the clinical implications of SIT encompass challenges in diagnosis and procedures, potential congenital cardiac abnormalities, considerations for organ transplantation and clinical decision making.

Individuals with SIT should be informed about their condition so that they could inform health care provider they might encounter in their subsequent life...

Consent:
The patient gave written informed consent for this case report and any selected images to be published.

Conflict of interest:
The authors declare that they have no conflict of interest.

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