Situs Inversus Totalis - A Rare Case Report

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ABSTRACT

BACKGROUND
A 60 years old previously asymptomatic female was admitted with accelerated hypertension and she was incidentally found to have mirror images of normal anatomical organs in chest and abdomen called situs inversus totalis. Situs inversus totalis is a congenital anomaly characterized by transposition of abdominal viscera associated with dextrocardia. Complete situs inversus is a rare syndrome of autosomal recessive inheritance. The anatomic, pathologic, embryologic and aetiology of complete situs inversus and related abnormalities are presented in this case with special emphasis to genetic correlation.

KEYWORDS
Accelerated Hypertension, Complete Situs Inversus, Congenital Anomaly, Dextrocardia, Transposition of Abdominal Viscera.


BACKGROUND
Situs inversus totalis is a congenital condition, in which the major visceral organs are reversed from their normal position. The situs inversus is present in 0.01% of the population. It is an autosomal recessive genetic condition, sometimes it can be X-linked and also found in identical twins.1,2 The persons with situs inversus totalis are usually asymptomatic and have a normal life expectancy. The situs inversus totalis has been estimated to occur once in about 6000 - 8000 live births. Situs inversus with levocardiA3 is another rare condition occurring in 1 in 22,000 of general population, in which the heart is found on normal side of thorax.

CASE REPORT
A 60 years old female patient was admitted with a history of headache and giddiness of 2 days’ duration. She did not give any history of significant illness in the past. There was no significant illness in the family. On examination, she was conscious, oriented, her pulse rate was 80 beats per minute, BP was 180/100 mmHg in supine and upright position. She did not have any signs of focal neurodeficit.

Her investigations revealed a haemoglobin of 12.4 gm%, total leucocyte count of 10100/mm3 with polymorphs of 86%, lymphocytes 12%, PCV 40.6%. Blood sugar, renal function tests, liver function tests and sr. electrolytes were within normal limits. She was not immunosuppressed. Chest radiogram showed Dextrocardia. Ultrasound abdomen revealed liver in left hypochondrium, spleen in right hypochondrium. Inferior vena cava noted left to the aorta, that is features of situs inversus totalis. Her ECG revealed inversion of P waves in leads I and aVL, dominantly negative QRS waves in leads I, V1-V6, extreme QRS axis, flattened T waves in aVR and inverted T waves in lead I and aVL. The features suggesting dextrocardia. Her 2D Echo revealed Dextrocardia with normal sized cardiac chambers and normal LV function. With the clinical and investigation results, a diagnosis of situs inversus totalis was made. She was started on calcium channel blockers for her hypertension. She was completely asymptomatic till date.

Left Sided Leads

Right Sided Leads
Chest Radiogram

USG Abdomen and Pelvis
- Liver in left hypochondrium.
- Spleen in right hypochondrium.
- Features s/o situs inversus totalis.

CT Abdomen and Pelvis
situs inversus totalis because of the cardiac position as well as the atrial
chambers and reversed abdominal viscera.

The terms levocardia and dextrocardia indicate the
direction of the cardiac apex at birth
In levocardia the base to apex axis is
towards the left and reverses in case of dextrocardia. Isolated
dextrocardia is situs solitus with dextrocardia. The cardiac
 apex points to the right, but the viscera are otherwise in their
usual positions. Situs inversus dextrocardia is situs inversus
totals because of the cardiac position as well as the atrial
chambers and reversed abdominal viscera.


totalis

1. D-loop or right sidedness, that is right ventricle is anterior
and to the right of the left ventricle (normal).
2. L-loop or left sidedness, right ventricle is posterior and to
the left of left ventricle (inversion of ventricles).

Dextrocardia is cardiac malposition in which the heart is in
the right hemithorax with the base to apex axis pointing to
right[11]; 3% of people with situs inversus totalis have
some form of congenital heart disease.[12,13] About 25% of
individuals with situs inversus totalis have an underlying
condition known as primary ciliary dyskinesia.

PCD is the dysfunction of the cilia that manifests itself
during the development of foetus. Situs inversus with PCD
together known as Kartagener syndrome characterised by
trip of situs inversus, chronic sinusitis and bronchiectasis.[13,14,15,16,17]

CONCLUSION
Situs inversus totalis is mostly an incidental finding. Most
individuals are unaware of having situs inversus totalis in
them and they come to know during screening investigations
for unrelated illness. Though Kartagener syndrome can be
associated with situs inversus totalis, so physician must be
vigilant in suspected cases and should take help of radiological
investigations to confirm the diagnosis. Situs inversus also
complicates organ transplantations as donor organs will
almost certainly come from situs solitus donors.

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