Situs inversus totalis - a case report

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Abstract

Situs inversus is a congenital positional anomaly characterized by transposition of abdominal viscera and when associated with right sided heart (Dextrocardia) is referred to as Situs inversus totalis. It is not so uncommon congenital positional anomaly but can be a diagnostic problem at times. The case was detected in the Department of Anatomy, Gauhati Medical College during the routine dissection. Situs inversus was first described by Aristotle in animals and by Fabricius in humans. Its incidence has been reported between 1 in 4000 to 20,000 live births. The exact etiology is unknown but Autosomal recessive and X-linked inheritance have been reported. It can also occur in association with syndromes such as Kartagener syndrome or Primary Ciliary Dyskinesia (PCD). Many people with situs inversus totalis are unaware of their unusual anatomy until they seek medical attention for any condition.

Keywords: dextrocardia, Kartagener syndrome, primary ciliary dyskinesia

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. Typically the heart lies on the left side of the body (levocardia), the liver lies on the right and spleen on the left. The lung on the left has two lobes while the lung on the right has three lobes. This normal arrangement is known as Situs solitus. The term Situs inversus is an abbreviated form of latin phrase "Situs inversus viscerum" meaning inverted position of the internal organs. Situs inversus can be either total or partial. Situs inversus totalis is also termed as Situs inversus with dextrocardia. It is characterized by the presence of the heart on the right side of the thorax, the stomach and spleen on the right side and liver on the left side of the abdomen. Here the left lung is trilobed and right lung is bilobed. If the heart remains on the normal left side of the thorax while the abdominal organs are placed exactly opposite to their normal site then it is termed as Situs inversus incompleitus or situs inversus with levocardia.

Situs inversus was first described by Aristotle in animals and by Fabricius in humans. Marco Severino first recognized dextrocardia in 1643. More than a century later, Matthew Baillie described the complete mirror-image reversal of the thoracic and abdominal organs in situs inversus. Its incidence has been reported to vary widely between 1 in 4,000 to 20,000 live births, with a male:female ratio-3:2. 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.

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 typified by bronchiectasis, sinusitis, and situs inversus and affects 20% of patients with situs inversus; however, only 50% of patients with Kartagener syndrome have situs inversus. Primary Ciliary Dyskinesia is caused by a defect in the cilia that impairs their normal movements and is thought to occur either at conception or within two weeks of embryonic life. It is suggested that the immobility of nodal cilia inhibits the flow of extra-embryonic fluid during the embryonic period, and this leads to the development of situs inversus. Its transmission mode is autosomal.
recessive inheritance, but its precise genetic mechanism is still unidentified. More than one genetic mutation including the gene mutations that cause ciliopathy and cystic renal diseases has been implicated in the etiopathogenesis.

Case report

The present case was observed while dissecting a 60 year old male cadaver during dissection teaching for medical undergraduates in the Anatomy department of Gauhati Medical College.

During routine dissection of thorax, the heart was found on the right side, and then the whole thoracic cavity and the abdominal cavity were dissected carefully and explored to see the abnormal positions of the thoracic and abdominal organs. After thorough exploration of the thoracic and abdominal cavities following observations were made:

- The heart was on the right side of the thoracic cavity (Fig. 1).
- Right turned Aortic arch (Fig. 2).
- Right lung was bilobed and cardiac impression was present on it with lingula (Fig. 3 & 4).
- Left lung was also bilobed (Fig. 5 & 6).
- Liver and Gallbladder were on left side while Spleen was on the right side of the abdominal cavity (Fig. 7 & 8).
- Fundus of Stomach was on the right and the first part of the duodenum was lying to the left of the midline in the left hypochondrium (Fig. 9).
- Caecum and Appendix was in the left iliac fossa while sigmoid colon was in the right iliac fossa (Fig. 10).
- Right testicular vein was draining into the right renal vein (Fig. 11).

Discussion

Situs inversus totalis is a rare anomaly with complete right to left transposition of the thoraco-abdominal organs. Incidence of situs inversus totalis is 0.01%.

Associated intra abdominal anomalies are polysplenia, asplenia, Kartagener’s syndrome, malrotation and duodenal obstruction are found to occur in such patients.

Total situs inversus also termed mirror image dextrocardia, is characterized by a heart on the right side of the midline while the liver and Gallbladder on the left side. Patients are usually asymptomatic and have a normal life.

Various modalities such as Echocardiogram, Radiographic Studies, Computed Tomography (CT) scans with oral and intravenous contrast, Ultrasonography (USG) and Barium Study can be used to diagnose situs inversus.

There have been isolated reports of situs inversus associated with peptic ulcer, ulcer perforation, amoebic liver abscess, acute cholecystitis, cholelithiasis, acute appendicitis and intestinal obstruction.

Gandhi et al, reported first case of perforated duodenal ulcer with situs inversus in 1986.

In 1983, Ruben et al reviewed 23 cases of abdominal situs inversus cases. They found 12 abdominal situs inversus with dextrocardia, 10 abdominal situs inversus with levocardia and one with partial heterotaxia. Major intraabdominal anomalies produced surgical emergencies in seven neonates in the first year. Five of the seven neonates had associated major congenital heart disease accounting for two of the three deaths in this series.

Situs inversus occurs more commonly with dextrocardia. A 3-5% incidence of congenital heart disease is observed in situs inversus with dextrocardia, usually with transposition of the great vessels. Of these patients, 80% have a right-sided aortic arch. Situs inversus with levocardia is rare.

Embryological Significance

When the structure or arrangement of one or more of the laterally asymmetric organs is abnormal it is thought that errors in the specification or interpretation of left-right information may have occurred in early embryogenesis. Defects of lateralization may be responsible for some congenital anomalies of the heart.
Fig. 1: The heart on the right side of the thoracic cavity

Fig. 2: Right turned aortic arch

Fig. 3: Right lung (medial aspect)

Fig. 4: Right lung (lateral aspect)

Fig. 5: Left lung (medial aspect)

Fig. 6: Left lung (lateral aspect)

Fig. 7: Contents of abdomen (liver & GB on left side)

Fig. 8: Pancreas (body & tail) & spleen (right side)

Fig. 9: Pancreas and Duodenum (left)

Situs inversus totalis - Malamoni & Joydev
bronchial branching, lung lobation, major vessels, portal venous anatomy, the spleen and gastrointestinal mesenteric attachments. Since these anomalies are frequently associated, they are due to a common defect and this defect is one of lateralization. In amniotes, the organs develop from a bilaterally symmetric epiblast. The asymmetry is first apparent in the heart. Abnormal lateralization of the cardiac segments has unquestionably serious consequences and abnormal situs may occur in more than one segment of the heart tube. For example, there can be mirror image dextrocardia in which all segments are inverted or transposition of the major vessels alone. Dextral looping of the embryonic heart tube is highly conserved in vertebrates, but the mechanism of looping and its consistent handedness are not understood. There are both genetic and environmental influences in the aetiology of abnormal lateralization.

Conclusion
Situs inversus is an uncommon congenital anomaly involving the thoracic and abdominal viscera. Exact aetiology is still unknown. Situs inversus usually remains undiagnosed unless it is diagnosed accidentally while investigating for another associated ailment. A diagnostic dilemma arises whenever thoracic and abdominal pathology occurs in patients with situs inversus. Although an uncommon anomaly, to choose a proper surgical incision site for thoracic and abdominal exploration pre-operative recognition of the condition is important.

References


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