

Dextrocardia and *Situs Inversus Totalis* in a Turkish Subject: A Case Report

Dextrocardia y *Situs Inversus Totalis* en un Sujeto Turco: Reporte de un Caso

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SUMMARY: Dextrocardia with *situs inversus* is an uncommon anomaly affecting about 1 to 2 per 10,000 in the general population. This report describes an adult male patient with dextrocardia and in a Turkish subject. The photographic illustrations revealed transposition of some of the visceral organs such as the spleen was located right and the liver and gall bladder on the left. The heart was flattened and flipped to the right. Many people with *situs inversus totalis* are unaware of their unusual anatomy until they seek medical attention for an unrelated condition. So, early detection may lead to a successful surgical management and consequently offer a safer chance of survival. This report showed that dextrocardia and *situs inversus* can be seen amongst Turkish subjects.

KEY WORDS: Dextrocardia; *Situs inversus totalis*; Rare anomaly.

INTRODUCTION

'Situs' means the position of heart i.e. the cardiac atria and viscera. When there is occurrence of mirror image it is termed as '*Situs inversus*' i.e. mirror image of as that suppos on the normal position. Dextrocardia is a term used exclusively for defining the positioning of heart i.e. the tip of heart points to the right side. A very few cases of *situs inversus totalis* has been described in literature i.e. Dextrocardia with *Situs inversus* (Jasrotia *et al.*, 2016).

Dextrocardia with *situs inversus* is a condition that is characterized by abnormal positioning of the heart and other internal organs. *Situs inversus* refers to the mirror-image reversal of the organs in the chest and abdominal cavity. Some affected people have no obvious signs or symptoms). In people affected by dextrocardia, the tip of the heart points towards the right side of the chest instead of the left side. However a small percentage of people also have congenital heart defects, usually transposition of the great vessels (National Organization for Rare Disorders, 2007; MedlinePlus, 2014; Wilhelm, 2018).

Situs inversus totalis is a very rare autosomal recessive entity (Piryani *et al.*, 2007). normally does not

cause any organ dysfunction, few patients with *situs inversus totalis* have been previously reported in the literature. This report showed that dextrocardia and *situs inversus* can be seen amongst Turkish subjects.

CASE REPORT

An 87-year-old male patient was admitted to Erciyes University urology clinic with a complaint of blood in the urine. The patient continues to consume 1 pack (20 cigarettes) per day for 40 years. Examination of the cardiovascular system revealed a normal pulse rate of 86 minute and blood pressure was 130/80 mmHg. Ultrasonography showed a 4 cm solid mass lesion in the anterior wall of the bladder (Figs. 1 and 2). The patient underwent cystourethroscopy and transurethral resection (TUR-CT) was performed on the bladder tumor. As a result of pathology, high-grade interchangeable epithelial carcinoma and tumor invasion were detected in the bladder muscles. The patient underwent thoracic and abdominal computed tomography (CT) for staging of bladder cancer.

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CT images revealed. **T2N0M0** was defined as clinical stage and radical cystoprostatectomy was performed. The pathological staging was consistent with preoperative clinical staging and the patient was followed up with routine uro-biological follow-up.



Fig. 1. Computerized tomography image of patient (Heart is on the right side).

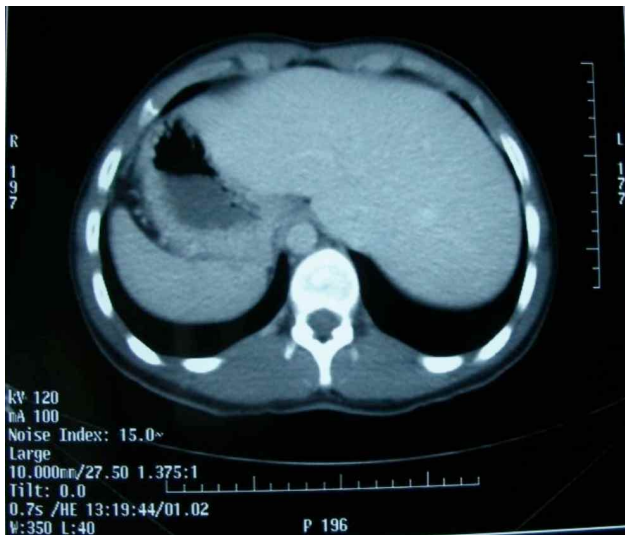


Fig. 2. Computerized tomography image of patient (Liver is on the left side).

DISCUSSION

Situs inversus totalis was first described by Aristotle in animal and by Fabricius in humans. *Situs inversus totalis* is defined as a complete mirror-image transposition of the thoracic and abdominal viscera (Piryani *et al.*). With the increasing use of surgical modalities in the treatment, various

clinical manifestations are faced by surgeons, one of which is *situs inversus totalis*. Dextrocardia with *situs inversus* is a rare anomaly occurring in about one of 10,000 people. This anomaly may not be diagnosed until later in life, in some cases *situs inversus* is commonly associated with serious primary ciliary dyskinesia and splenic malformations (Deuse & Reitz, 2009). Its frequency is between 1 in 8,000 to 1 in 20,000. It may be *situs inversus totalis* or incomplete in less than 10 % of cases (Chaturvedi & Thomas, 2003).

Dextrocardia with *situs inversus* is asymptomatic. Many people with *situs inversus totalis* are unaware of their unusual anatomy until they seek medical attention for an unrelated condition (Chaturvedi & Thomas).

Situs inversus with dextrocardia also is termed as *situs inversus totalis* because the cardiac position as well as atrial chambers and abdominal viscera, is a mirror image of normal anatomy (Van Praagh, 1977). There are some case reports on *situs inversus totalis* with various types of cancers, almost all of the reports describe that the surgical procedures are difficult because of the anatomic abnormality, and there are no reports of the etiological relationship between *situs inversus totalis* and cancer (Özben *et al.*, 2010). *Situs inversus* may be associated with other congenital anomalies such as duodenal atresia, asplenism, multiple spleens, ectopic kidney, horseshoe kidney and various pulmonary and vascular abnormalities. *Situs inversus totalis* that is associated with primary ciliary dyskinesia is known as Kartagener syndrome (Bohun *et al.*, 2007; Ortega *et al.*, 2007).

David A. *et al.* in their case report found that the stomach and spleen were located on the right side, while the liver was on the left side. The gallbladder was located in the epigastric area toward the left side (Ofusori *et al.*, 2009; Özben *et al.*).

Typically, persons having *situs inversus* with dextrocardia without other congenital anomaly have a normal life expectancy and have a similar risk of getting acquired disease as that of other person of same age and sex group. In the rare instances of cardiac anomalies, life expectancy is reduced, depending on the severity of the defect (Bohun *et al.*).

Detection of bladder cancer in our patient also confirms the increase in the rate of cancer in *situs inversus*; *situs inversus* is asymptomatic and does not cause any long term problems, it is very dangerous if the condition is not diagnosed prior to surgery. *Situs inversus* is present in 0.01 % of the population of United States (Wilhelm) but its incidence in Turkey is unknown.

Nawaz H. *et al.* (2005) in their report described a case of two newborns with *situs inversus* in association with congenital duodenal obstruction.

Surgeons and radiologists should beware of this anomaly during the presurgical and surgical management and also encourage a routine medical examination which could give a signal to the existence of this condition if present (Ofusori *et al.*). This will help the patient when afflicted with certain clinical conditions such as appendicitis; where the referred pain will be on the left side rather than the right thus leading to wrong diagnosis and possibly death due to delay in surgical management (Chaturvedi & Thomas; Ofusori *et al.*).

CONCLUSION

The condition is rare and with limited literature. We therefore, in conclusion, sensitize the surgeons and radiologists to beware of this anomaly during the presurgical and surgical management and also encourage a routine medical examination which could give a signal to the existence of this condition if present. Additionally, we believe that the sharing of experience of this rare type of patient is important and will be able to guide future surgical procedures.

Informed consent: Written informed consent was obtained from patient who participate in this study.

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RESUMEN: La dextrocardia con *situs inversus* es una anomalía poco frecuente que afecta aproximadamente de 1 a 2 personas por 10.000 en la población general. Este informe describe un paciente masculino adulto con dextrocardia. Las figuras revelaron que la transposición de algunos de los órganos viscerales, como el bazo, se ubicada a la derecha y el hígado y la vesícula biliar a la izquierda. El corazón fue aplastado y girado hacia la derecha. Muchas personas con *situs inversus totalis* desconocen su anatomía inusual hasta que buscan atención médica por una afección no relacionada. Por lo tanto, la detección temprana puede llevar a un manejo quirúrgico exitoso y, en consecuencia, ofrecer una posibilidad más segura de supervivencia. Este informe mostró que la dextrocardia y el *situs inversus* se pueden encontrar entre los sujetos turcos.

PALABRAS CLAVE: Dextrocardia; *Situs inversus totalis*; Anomalía rara.

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