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Case Report
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Dextrocardia with Situs Inversus in an Adult Turkish

Seher Yılmaz[1], Ayse Kristina Polat[2], Adem Tokpınar[1] and Özlem Bozkurt[3]*

[1]*Department of Anatomy, Yozgat Bozok University, Turkey*

[2]*Yozgat Bozok University, Turkey*

Abstract

Situs Inversus is a rare congenital anomaly caused by the displacement of the organs in the chest and abdomen. Situs inversus totalis known as the displacement of all organs according to the mirror image. Dextrocardia is used to describe the position of the heart which tip is pointing to the right instead of left side of the chest.

People having situs inversus totalis and dextrocardia live with normal life expectancy and are mostly asymptomatic. These anomalies may not be recognized for a long lifetime. Other congenital disorders such as heart diseases and lung infections may progress with this condition. Although situs inversus totalis with dextrocardia are rare, the lack of a comprehensive study of this anomaly in the literature indicates that further research is needed. In our report, we describe a case of dextrocardia with situs inversus totalis in a 21-year-old male.

Keywords: Situs inversus totalis, Dextrocardia; Rare anomaly

Introduction

Situs inversus totalis is a condition where the normal positions of the chest and abdominal organs are arranged in a mirror image. Situs inversus totalis is known to be a rare syndrome inherited by autosomal recessive genes. Situs inversus may be seen with levocardia or dextrocardia. In levocardia, the tip of the heart is pointing to the left and in dextrocardia it turns to the right. Dextrocardia is a developmental anomaly where a bulboventricular cycle is folded to the left of the primitive heart cycle in a reverse placement of normal. The anomaly causes the heart to be positioned on the right side of the chest along with the long right and downward orientation. The distribution of these abnormalities was found to be equal between genders as well as between races [1]. Situs inversus occurs in 1 in 4000 people in Japan, 1 in 5000 in Israel and 1 in 10,000 to 15,000 in Northern Europe and the United States. Dextrocardia occurs in about 1 in 10,000 people in the general population. The exact cause of this condition, although it is unknown, has been associated with several factors including incomplete penetration, maternal diabetes mellitus, and conjoined twin anomaly, as well as autosomal recessive genes.

People with situs inversus do not have a phenotypic difference and may maintain routine healthy life without any complication related to their medical condition. Most people with situs inversus totalis are unconscious until they are treated for a medical condition independent of this anomaly [2,3].

The diagnosis of dextrocardia is made by using a combination of clinical, radiological, electrocardiographic and echocardiographic findings. The anomaly is generally diagnosed on a routine prenatal sonogram, but not every radiologist can identify it, especially if there are no cardiac structural abnormalities. Diagnostic methods such as chest radiography and electrocardiogram are sufficient to diagnose dextrocardia, but current methods for instance echocardiography and magnetic resonance imaging are used for definitive diagnosis. Dextrocardia with situs inversus might be diagnosed incidentally during the evaluation of conditions not associated with the anomaly, in a routine medical examination or as indicated in this case report [4-7].

The purpose of this study is to present a case with Dextrocardia and Situs Inversus in our country.

Case Presentation

A 21-year-old male patient was admitted to the urology outpatient clinic of Erciyes University Hospital for urinary tract infection. His blood pressure was 130/80 mmHg and his pulse rate was 60 per min. No cardiovascular problems were observed. The patient stated that he had smoked 1 pack of cigarettes a day for 3 years. As a result of the ultrasonography, the organs of the patient

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Correspondence:
Seher Yilmaz, Department of Anatomy, Yozgat Bozok University, Yozgat, Turkey,
E-mail: sehery38@hotmail.com

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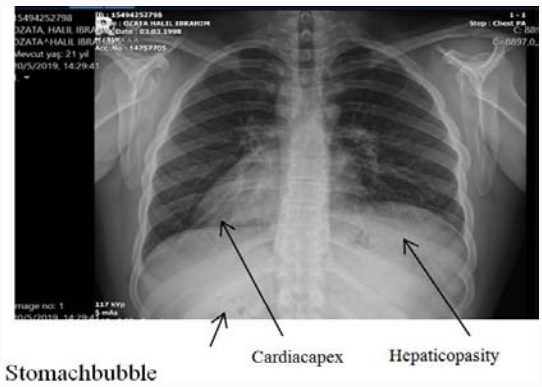

Figure 1: Posteroanterior (PA) chest X-Ray image of the patient. Position of the heart is changed according to the mirror image.

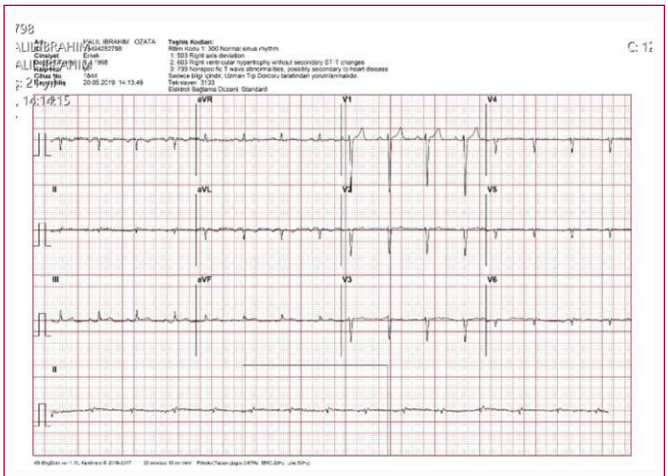

Figure 2: The ECG obtained from the patient revealed qRS positivity, which is the opposite of normal status in unipolar avR leads.

were displaced symmetrically. The diagnosis was as situs inversus totalis, accompanied by dextrocardia by chest X-Ray (Figure 1) and Electrocardiography (ECG) (Figure 2).

Discussion

Dextrocardia was first seen by Leonardo da Vinci in 1452 and later described by Marco Aurelio Severine in 1643 and more than a hundred years later was explained by Matthew Ballie [8]. "Situs" term is used to define the position of internal organs. "Solitus" refers to the normal position and "Inversus" means the opposite position [9]. Dextrocardia with situs inversus is a rare condition occurring in the general population is approximately one in every 10,000 people, but it is not yet known incidence in Turkey. There is no race-related difference for situs inversus and the male-female incidence is equal [10]. Situs inversus may be associated with other congenital problems such as asplenism, multiple spleens, duodenal atresia, horseshoe/ectopic kidney and various pulmonary/vascular abnormalities. Situs Inversus Totalis was also associated with Kartagener's Syndrome, a primary ciliary dyskinesia [11,12]. In our case, the cause of this anomaly was not found exactly. Follow-up of patients with situs inversus totalis and dextrocardia is only necessary if there are congenital heart defects or other associated syndromic features and this condition is not noticed in patients with a healthy life expectancy [13,14]. Recognition of situs inversus has a great importance for the prevention of surgical mishaps due to reversed anatomy or atypical history. For example, appendicitis causes left lower quadrant pain in a patient with situs inversus, while cholecystitis typically causes left upper quadrant pain. Cardiac situs is also determined by atrial location. In situs inversus, the morphological right atrium is on the left and the left atrium is on the right. However, the normal pulmonary anatomy is also reversed, so that the left lung contains three lobes and the right lung contains two lobes. In addition, the spleen and stomach are on the right and the liver and gallbladder are on the left. Situs inversus also may complicate organ transplantations because the donor organs are located in the situs solitus, which is the normal position. Therefore, problems arise from the geometric shapes of organs such as the heart and liver that need to be implanted [15,16]. Since this procedure also applies to the orientation of similar blood vessels, the correct steps need to be taken for the proper attachment of the blood vessels.

The diagnosis of situs inversus totalis associated with dextrocardia can be confirmed by electrocardiography, echocardiography, magnetic resonance imaging, abdominal ultrasonography, chest radiography and computed tomography scans [17]. ECG is an essential and interesting tool for diagnosis of dextrocardia. Echocardiography is required for precise analysis of cardiac position as well as for the recognition of associated congenital heart or vascular malformation. Sonographic and computed tomography studies are needed to confirm the positions of abdominal organs anatomy [7]. Computed tomography and magnetic resonance imaging are known to be the best method for demonstrating mirror anatomy of organs [18].

Surgeons and radiologists have to avoid this abnormality before or at the time of surgery, and should undergo a routine medical examination, that may indicate the presence of this condition. The medical examination will assist the patient when affected by clinical conditions such as appendicitis. If the pain is on the left side instead of the right side, it may lead to misdiagnosis and even death due to delay in surgical treatment [19].

Conclusion

Although situs inversus is a rare anomaly, studies dealing with this issue are limited. Increased awareness of this situation; surgeons and radiologists should be more careful to take the anomaly into consideration before and during surgery. It is also known that encouraging patients to undergo a routine medical examination that may indicate the presence of this condition will improve life quality of patients. In addition, sharing the experiences of rare patient type will provide guidance in future surgical procedures.

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