

SITUS INVERSUS TOTALIS AND CORRECTED TRANSPOSITION OF THE GREAT VESSELS IN A 50 YEAR OLD WOMAN

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Situs inversus totalis'de iç organlar normal pozisyonda karşı tarafta bulunur. Büyük arterlerin düzeltilmiş pozisyonu ise atrioventriküler ve ventriculoarterial uyumsuzluk ile karakterizedir. Situs inversus totalis ile beraber görülen büyük arterlerin düzeltilmiş pozisyonu çok nadirdir. Bu olguda 50 yaşında hem situs inversus totalis'i hem de büyük arterlerin düzeltilmiş pozisyonu olan bir kadın tartışıldı. Situs inversus totalis'de büyük arterlerin düzeltilmiş

pozisyonunda cerrahide problem olacağı için bu anomalilerin bilinmesi çok önemlidir.

Bu nedenle bu hastaların anomalilerinin tanınması kalp cerrahları için faydalıdır.

Anahtar kelimeler:Situs inversus totalis, Büyük arterlerin düzeltilmiş pozisyonu

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GİRİŞ

Situs inversus totalis (SIT), which refers to a mirror-image reversal of the normal position of the internal organs, is a rare and confusing situation. It occurs in association with other anomalies and syndromes, which may disturb the surgical procedure for diseases coexistent with SIT. Congenitally corrected transposition of the great arteries (CCTGA) is a rare form of congenital heart disease. Life expectancy of patient with CCTGA is limited by the onset of systemic (morphological right) ventricular failure in their 40s or 50s[1]. Fewer than 30 patients older than 40 years have been reported in the literature[2-5].

Reports of CCTGA are rare[2-5] and the combination with SIT is even more scarce[1,6] In this case, 50 year-old woman with SIT and CCTGA has been discussed.

CASE REPORT

A woman with syncope was admitted to the Sani Konukoğlu Medical Centre five years ago. Her ECG arrhythmia and complete AV heart block. Echocardiography showed that her heart was located

on the left side, the aorta arose from the morphological right ventricle, which was identified by the three leaflet tricuspid valve and the pulmonary trunk that arose from the morphological left ventricle. No intracardiac shunts were found. In coronary angiography, it was observed that left main coronary artery arose from right hand sinus of Valsalva and then bifurcated to anterior descending and circumflex branches and the right coronary artery arose from left hand sinus of Valsalva. The aortic arch and the thoracic aorta were located on the right side of vertebral column (Fig. 1). The aorta arising from the right side and the pulmonary artery arising from left side of the heart was observed again. The upper abdominal ultrasonography demonstrated that she had situs inversus viscerum; as liver was located on the left side (Fig. 2) and the spleen was located on the right side of the abdomen but her heart was located on the left side.

She had a history of pacemaker insertion because of complete heart block. The patient had been alive for 5 years after the insertion of a pacemaker.

DISCUSSION

SIT refers to a mirror-image reversal of the normal position of the internal organs. The first representation regarding situs inversus was described by Aristotle in subhuman forms, but transposition of the viscera had not been recognized until Fabricius

Fig. 1 Ventriculography. The aortic arch and the thoracic aorta are located on the right side of vertebral column.

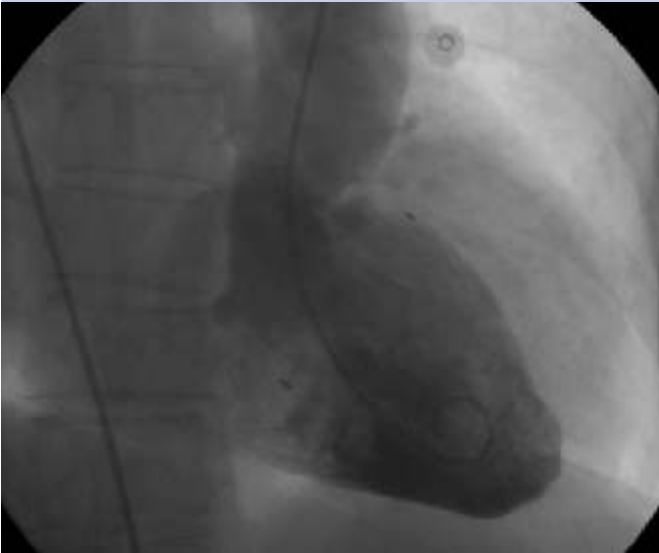
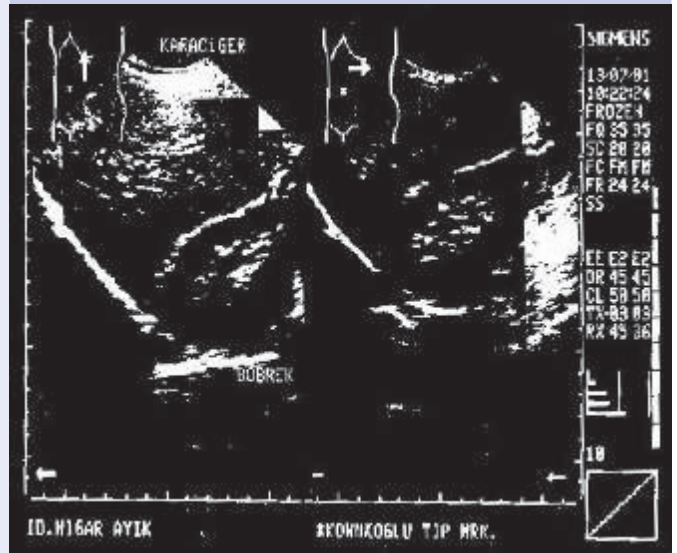


Fig. 2 Ultrasound showed left-sided liver.



reported a case of reversed liver and spleen in a human in 1600[7]. The incidence of SIT is 1 in every 8,000-25,000 births[8]. SIT occurs in association with other anomalies and syndromes.

CCTGA is a rare form of congenital heart disease first described by Von Rokitansky in 1875 and characterized by atrioventricular and ventriculoarterial discordance[9]. The atrioventricular discordance implies that the morphological right atrium drains into the morphological left ventricle, and the morphological left atrium into the morphological right ventricle. Thus, the left ventricle supplies the pulmonary circulation, and the right ventricle supports the systemic circulation. CCTGA accounts for less than 1% of congenital heart disease cases, and is usually associated with a severely reduced life expectancy owing to ventricular septal defects (74%), pulmonary valve stenosis (74%), systemic (tricuspid) valve abnormalities (38%), and complete heart block (5%)[2]. Only 1-10% of individuals with CCTGA have no associated defects[2-5]. Their life expectancy is limited by the onset of systemic (morphological right) ventricular failure in their 40s or 50s[1].

Patients with CCTGA rarely survive to old age. In the literature, there is a few case report. Only three 72 or 80 years old patients were reported[10-11]. Fewer than 30 patients older than 40 years have been reported in the literature[1-5]. Our patient was 50 years old and she is living now.

Much controversy exists regarding the ability of the morphological RV to support the systemic circulation. In patients with CCTGA, who present as a naturally occurring model of this adaptation, systemic ventricular failure is the cause of death in more than 50%[2]. A progressive deterioration of the systolic

function of the systemic ventricle has also been described in patients without associated cardiac defects. Complete heart block and increasing left (tricuspid) atrioventricular valve regurgitation are known contributing factors[1]. Our patient had a history of pacemaker implantation due to complete AV block.

CCTGA without associated intracardiac defect is very rare. CCTGA usually presents with one or more associated intracardiac anomalies. The most common associated anomalies are ventricular septal defect, pulmonary outflow tract obstruction and abnormalities of the systemic atrioventricular valve[2,5,12]. Our patients had no intracardiac shunts.

CCTGA is very singular if it is associated with situs inversus totalis[1,6]. Allwork et al examined 32 patients with CCTGA and out of these patients, 29 patients (90.6 percent) had in situs solitus and only 3 patients (9.4 percent) had SIT[1]. Our patient had SIT; the liver located on the left side and the spleen located on the right side of the abdomen.

The patients with CCTGA and SIT usually undergo surgical repair. The presence of anatomical anomalies significantly increases operative risk. Therefore, recognition of anomalies with this patient may be useful for a cardiac surgeon.

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