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Case Report

Congenital Corrected Transposition of the Great Arteries (ccTGA): An Incidental Case Report

Konjenital DüzeltilmişBüyük Arterlerin Transpozisyonu (KD-BAT): Raslantısal bir vaka sunumu

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ABSTRACT

Congenital corrected transposition of the great arteries (ccTGA) is a rare disorder with variable cardiac malformations that can result in a wide range of clinical outcomes, from asymptomatic to mortality. In ccTGA cases, the right atrium is connected to the left ventricle by the mitral valve, and this structure is supported by the pulmonary artery. After opening into the left atrium, the pulmonary veins are connected to the right ventricle by the tricuspid valve, and this structure continues with the aorta. As a result, systemic venous blood with low oxygenity goes to the lungs through the pulmonary artery and blood with high oxygenity from the pulmonary veins goes to the systemic circulation through the aorta and cyanosis is not seen. Although most cases are clinically asymptomatic in childhood, findings are more common in late adolescence and adulthood in the presence of additional lesions, such as ventricular septal defect or Ebstein anomaly. Bradycardia (with or without heart failure), complete AV block, tachyarrhythmia and congestive heart failure may be present. In addition, in later years, heart failure may develop and patients may die because the structure that functions as a pump to the systemic circulation is in the right ventricular musculature.

Keywords: *echocardiography, congenital corrected transposition of the great arteries (ccTGA), children*

ÖZET

Konjenital düzeltilmiş büyük arter transpozisyonu (KD-BAT), oluşturduğu değişken kalp malformasyonları neticesinde asemptomatik durumdan ölüme kadar geniş bir yelpazede klinik sonuçlarla karşımıza çıkabilen nadir görülen bir hastalıktır. KD-BAT olgularında; sağ atrium sol ventriküle mitral kapak ile bağlanır ve bu yapı pulmoner arter ile desteklenir. Pulmoner venler sol atriuma açıldıktan sonra triküspid kapak ile sağ ventriküle bağlanır ve bu yapı aorta ile devam eder. Bu durumun sonucunda; oksijenitesi düşük olan sistemik venöz kan pulmoner arter aracılığıyla akciğerlere, pulmoner venlerden gelen oksijenitesi yüksek olan kan ise aort aracılığıyla sistemik dolaşıma gider ve siyanoz görülmez. Çoğu olgu klinik olarak çocukluk döneminde asemptomatik olmakla beraber bulgular daha çok geç adolesan ve erişkin dönemlerinde eşlik eden ventriküler septal defekt veya Ebstein anomalisi gibi ek lezyonların varlığında bulgular ortaya çıkmaktadır. Bradikardi (kalp yetmezliğinin eşlik ettiği veya etmediği), komplet AV blok, taşiaritmi, konjestif kalp yetmezliği ile bulgu verebilmektedir. Ayrıca ileriki yıllarda sistemik dolaşıma pompa işlevi gören yapı sağ ventrikül kas yapısında olduğu için kalp yetmezliği gelişmekte ve hastalar bu nedenle hayatlarını kaybedebilmektedir.

Keywords: *ekokardiyografi, büyük damarların konjenital düzeltilmiş transpozisyonu, çocuk*

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INTRODUCTION

Congenitally corrected transposition of the great arteries (ccTGA) is a rare congenital heart malformation. Its prevalence is 0.03 per 1,000 live births, accounting for approximately 0.05% of congenital heart malformations.(1) Normally, the systemic venous return morphologically joins the right atrium. This atrium is morphologically connected to the right ventricle by the tricuspid valve and is supported by the pulmonary artery. After opening into the left atrium, the pulmonary veins are connected to the left ventricle by the mitral valve, and this structure is supported by the aorta. This is referred to as atrioventricular and ventriculoarterial concordance.(2,3)

In patients with ccTGA, systemic venous return joins the right atrium with normal atrial placement. This atrium is connected to the left ventricle by the mitral valve and is supported by the pulmonary artery. After opening into the left atrium, the pulmonary veins connect to the right ventricle through the tricuspid valve and this structure continues with the aorta.(2,3)In infants with ccTGA, although the right and left ventricles are displaced, systemic venous blood with low oxygenation goes to the lungs using the pulmonary artery and blood with high oxygenation from the pulmonary veins goes to the systemic circulation via the aorta; therefore, cyanosis is not seen.(4) In this study, we report a 17-year-old male with an incidental diagnosis of ccTGA who presented after a suicide attempt.

CASE REPORT

A 17-year-old male patient was admitted to the emergency department of our hospital after taking 60 pieces of Quetiapine 25 mg tablets for suicidal purposes. Blood pressure was 125/70 mmHg, and pulse rate was rhythmic at 75 beats/minute. Physical examination revealed no pathologic findings except for mild stupor. Glasgow's coma score was 13. Cardiovascular system examination revealed a grade 1-2 murmur in the mesocardiac focus. Laboratory tests reportedhemoglobin as 15.8 g/dl, leukocytes as 6620/mm3, ck as 384 IU/l, CK-MB as 0.91 ng/ml, and Troponin-T as 24.6 ng/ml. Both hepatic and renal function tests and coagulation parameters were normal. The patient's electrocardiogram (ECG) revealed the presence of Q waves in the right precordial deviations (V1-V2) and the absence of Q waves in the left precordial deviations (V5-V6) (Figure 1). The patient was monitored, and hydration therapy was initiated.

The patient underwent a cardiological examination due to possible side effects of quetiapine. Echocardiography (ECHO) revealed systemic venous flow-right atrium-morphologic left ventricle-pulmonary artery outflow on the right side, pulmonary veins-morphologic right ventricle-aortic outflow on the left side, and mild tricuspid valve insufficiency (Figure 2). The echocardiographic examination showed that the patient experienced exertional fatigue during the last few years. There was no rhythm disturbance on Holter ECG. The patient was discharged uneventfully on the third day of hospitalization. The consent of the patient's parents was obtained for this study.

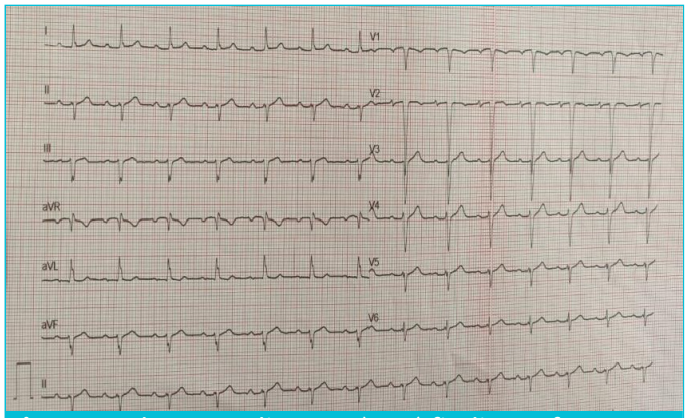

Figure 1. Electrocardiogram (ECG) findings of our patient showed the presence of Q waves in the left axis, right precordial deviations (V1-V2) and absence of Q waves in the left precordial deviations (V5-V6). Atrioventricular (AV) block was not detected.

DISCUSSION

It has been well established that ccTGA is a rare form of congenital heart disease with a broad spectrum of associated cardiac pathologies and postnatal clinical outcomes.(5) In this case study, we report a case of ccTGA incidentally detected in a 17-year-old asymptomatic patient who presented to the emergency room due to a suicide attempt. Examination of the cardiovascular system was unremarkable except for a low-intensity systolic murmur in the mesocardiac focus. ECG showed the presence of Q waves in right precordial deviations and the absence of Q waves in left precordial deviations, which should normally be present, and the diagnosis of ccTGA was verified by transthoracicechocardiography.

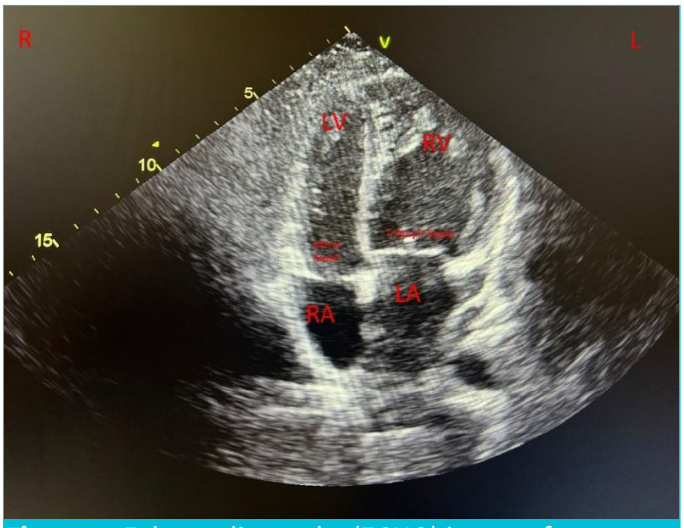

Figure 2. Echocardiography (ECHO) image of our patient; pulmonary veins-left atrium-tricuspid valve-right atrium-aorta-systemic circulation is observed on the left. On the right, systemic venous flow-right atrium-mitral valve-left ventricle-pulmonary artery-lungs are observed.

Patients with ccTGA are clinically asymptomatic during childhood, but symptoms become more frequent in late adolescence and adulthood.(4) In early childhood, findings occur only in the presence of additional lesions, such as concomitant ventricular septal defect (VSD) or Ebstein anomaly.(3) In this period, patients are diagnosed mainly after cardiological evaluation, which is performed due to the incidental detection of ECG abnormalities and/or auscultation of tricuspid regurgitation murmur. In adolescence and adulthood, bradycardia (with or without heart failure), complete atrioventricular (AV) block, tachyarrhythmia and congestive heart failure may be present. Additionally, heart failure develops in later years of life and patients may die because the structure that functions as a pump to the systemic circulation is situated in the right ventricular musculature.(3-6)

Jain et al.(7) reported that a four-year-old boy who presented for circumcision was incidentally diagnosed with ccTGA (with no other valvular or septal defects) on ECHO performed after suspicious findings were observed on the ECG during the preparation for surgery. Obongonyinge et al.(8) incidentally detected ccTGA in five African children aged between one and 13 years and observed VSD in four of these patients and septal wall abnormalities and severe right ventricular dysfunction in one of them. Hsu et al.(9) shared their results in 56 pediatric patients with ccTGA who were followed surgically for 17 years. They reported that better results were obtained in long-term follow-up, especially in children with single ventricle palliation.

In conclusion, each patient may present with a different combination of cardiac defects. Therefore, the significance of each defect should be analyzed. The observed combination of defects may also not always explain stability. The present case report aims to remind us that the diagnosis of ccTGA can be made incidentally during late adolescence and early adulthood based on the coincidental ECG findings.

Patient Consent Form / Hasta Onam Formu

The parents' of this patient consent was obtained for this study.

Conflict of Interest / Çıkar Çatışması

The authors declared no conflicts of interest with respect to authorship and/or publication of the article.

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REFERENCES

1. Šamánek M, Voříšková M. Congenital heart disease among 815,569 children born between 1980 and 1990 and their 15-year survival: A prospective Bohemia survival study. Pediatr Cardiol 1999; 20(6): 411-7.
2. Graham TP, Markham L, Parra DA, et al. Congenitally corrected transposition of the great arteries: An update. Curr Treat Options Cardio 2007 ;9: 407-13.
3. Kutty S, Danford DA, Diller GP, et al. Contemporary management and outcomes in congenitally corrected transposition of the great arteries. Heart 2018; 104: 1148-55.
4. Cohen J, Arya B, Caplan R, et al. Congenitally Corrected Transposition of the Great Arteries: Fetal Diagnosis, Associations, and Postnatal Outcome: A Fetal Heart Society Research Collaborative Study. J Am Heart Assoc 2023;12:e029706.
5. Murtuza B, Barron DJ, Stumper O, et al. Anatomic repair for congenitally corrected transposition of the great arteries: a single-institution 19-year experience. J Thorac Cardiovasc Surg 2011; 142: 1348-57.
6. Baruteau AE, Abrams DJ, Ho SY, et al. Cardiac Conduction System in Congenitally Corrected Transposition of the Great Arteries and Its Clinical Relevance. J Am Heart Assoc 2017; 6(12): e007759.
7. Jain G, Joshi A, Sathe V, et al. A Case Report on the Anaesthetic Management for a 4-Year-Old Child Undergoing Cardiac Catheterization to Confirm the Diagnosis of Congenitally Corrected Transposition of Great Arteries. International Journal 2020; 3(5): 237-41.
8. Obongonyinge B, Namuyonga J, Tumwebaze H, et al. Congenitally corrected transposition of great arteries: a case series of five unoperated African children. Journal of Congenital Cardiology 2020; *4*(1): 8.
9. Hsu KH, Chang CI, Huang SC, Chen YS, Chiu IS. 17-year experience in surgical management of congenitally corrected trans–

position of the great arteries: a single-center's experience. Eur J Cardiothorac Surg 2016; 49: 522-27.