OLGU SUNUMU / CASE REPORT

İleri yaşta akut inferiyor miyokart enfarktüsü ile başvuran ve atriyal septal defekt anomalisinin eşlik ettiği doğumsal düzeltilmiş büyük arter transpozisyonu

Congenitally corrected transposition of the great arteries in a patient with atrial septal defect and acute inferior segment myocardial infarction

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Özet– Doğumsal düzeltilmiş büyük arter transpozisyonu, doğumsal kalp hastalıklarının yaklaşık olarak %1'ini oluşturan nadir bir anomalidir. Tanı konulduğu yaş ve yaşama oranları değişkendir ve pulmoner darlık, ventriküler septal defekt, atriyal septal defekt, atriyoventriküler bloklar, atriyoventriküler kapak yetersizlikleri gibi eşlik eden anomalilerle ilişkilidir. Literatürde tek koroner ostiyumunun ve atriyal septal defektin eşlik ettiği düzeltilmiş büyük arter transpozisyonu olguları nadirdir. Bu yazıda, akut inferiyor ST segment yükselmeli miyokart enfarktüsü ile başvuran ve anjiyografisinde sağ sinüs Valsalvadan köken alan tek çıkımlı koroner ostiyumunun eşlik ettiği düzeltilmiş büyük arter transpozisyonlu 55 yaşında erkek hasta sunuldu. Hastanın sirkümfeks arterine başarılı perkütan anjiyoplasti ve stent implantasyonu yapıldı. Ekokardiyograf, düzeltilmiş büyük arter transpozisyonuna eşlik eden atriyal boşluklar arasındaki negatif kontrastlanmayı gösterdi. Hasta tıbbi tedavi ile ameliyat sonrası sekizinci günde taburcu edildi. Bizim bilgilerimize göre, bu olgu, düzeltilmiş büyük arter transpozisyonu, atriyal septal defekt, tek ostiyumdan köken alan koroner arterler ve akut miyokart enfarktüsünün birlikte görüldüğü ilk olgudur.

Summary– Congenitally corrected transposition of the great arteries is a rare anomaly accounting for approximately 1% of clinically apparent congenital heart disease. Age at time of diagnosis and survival rates vary and depend on associated anomalies, including pulmonary stenosis, ventricular septal defect, atrial septal defect, atrioventricular block, and atrioventricular valve regurgitation. Reported cases of corrected transposition of the great arteries with single coronary ostium anomaly and atrial septal defect are very few. In this article a 55-year-old male who presented with acute inferior ST-segment elevation myocardial infarction and coincidental single coronary ostium arising from the right sinus of Valsal-va, as observed on coronary angiography is presented. Successful balloon angioplasty and stenting of the circumflex artery were performed. Echocardiography demonstrated the corrected transposition of the great arteries with negative-contrast enhancement between the atrial chambers. The patient was discharged with medical therapy on the eighth postoperative day. To our knowledge, this case is the frst report to describe corrected transposition of the great arteries, atrial septal defect, single coronary ostium, and acute myocardial infarction as comorbidities.

Corrected transposition of great arteries (CTGA) is a rarely seen congenital heart disease in which atrioventricular (AV), and ventriculoarterial discordances were observed.[1] Its incidence is nearly 1 % among all congenital heart diseases.[2] Generally it is associated with pulmonary stenosis, ventricular septal defect (VSD), atrial septal defect (ASD), atrioventricular blocks, atrioventricular valve insufficiency, and heart failure. Patients without these concomitant conditions are rarely (5-10%) encountered, and they can live till advanced ages without any relevant symptoms.[3] The incidence of coronary artery anomalies has been 1-2 % in studies perfromed with coronary angiograms, and autopsies, and among them most frequently single coronary artery anomalies are seen.[4]

In the literature, patients with single coronary artery anomalies detected on coronary angiograms who had presented with acute myocardial infarction and undergone successful percutaneous coronary interventions (PCIs) have been reported.[5]

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However coronary arteries of some cases with CCTGA have also undergone PCI.[6] However in our literature reviews, we haven't encountered cases with myocardial infarction with acute ST-segment elevation in association with single coronary artery anomaly, and ASD who had undergone successful PCIs.

Kısaltmalar:

ASD	*Atrial septal defekc*
AV	*Atrio-ventricular*
Cx	*Circumflex*
CTGA	*Corrected transposition of the grtett arteries*
EKG	*Electrocardiography*
CAG	*Coronary angiography*
LMCA	*left main coronary artery*
PCI	*Percutaneous coronary intervention*
RCA	*Right coronary artery*
RV	*Right ventricle*
LSV	*Left ventricle*
VSD	*Ventricular Septal defect*

CASE PRESENTATION

A 55-year-old male patient applied to the emergency service with complaint of chest pain. His medical history was unremarkable except for 30 pack-years of smoking. His physical examination revealed the following characteristic features: blood pressure, 80/60 mm Hg; heart rate, 54 bpm, moderately well-poor general health state, normal orientation, and cooperation On respiratory system examination rales were heard bilaterally over both basal segments. Auscultation of the cardiovascular system revealed fixed splitting of the second heart sound, 1/6 systolic ejection murmur over pulmonic, and mesocardiac area, and 2/6 pansystolic murmur at apex, and tricuspid valve. His electrocardiograms (EKGs) revealed sinus rhythm, heart rate of 55 bpm, ST elevation in leads D2, D3,and aVF, ST-depression in leads V2 – V6. Pathologic Q wave was detected in leads. V1-V3 Septal Q wave was not seen in leads V5,and V6. In posterior EKGs obtained ST-elevation was seen in the leads D2,D3,aVF, V7, V8, and V9 (Figure 1). On telecardiograms, increase in cardiothoracic ratio, and bilateral bronchovascular branching, and flatenning of the left upper border of the heart were seen. Mass CK-MB was 32 ng/ml (normal range: 0.1–5 ng/ml), and high-sensitivity troponin T, 146 pg/ml (normal range: 0–14 pg/ml). Acetylsalicylic acid (300 mg p.o.) , clopidogrel (600mg. p.o.) and 7500 IU IV heparin were given to the patient diagnosed as ST –segment elevation myocardial infarction. Infusion of dopamin in physiologic saline (0.9 % NaCl) solution was iniiated for the patient whose blood pressure persisted at lower levels, and he was urgently brought into coronary angiography laboratory (CAG) with the intention of performing emerrgency revascularization. Coronary angiography demonstrated that coronary arteries arised from the right sinus of Valsalva, and complete occlusion of the proximal part of the cicumflex coronary artery (Cx).

At the same session drug-eluting stent was implanted into Cx through percutaneous angioplasty (Figure 2). After the procedure the patient was brought into coronary intensive care unit. During their follow-up the patient maintained a stable hemodynamic state, and on his transthoracic echocardiogram a trabeculated systemic ventricle, a moderator band were observed. Besides a morphologic right ventricle (RV) was seen in normal anatomical location of the left ventricle) and vice versa).

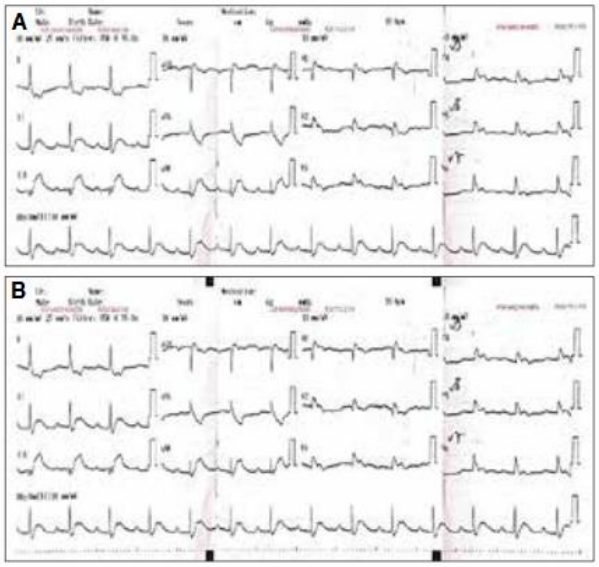

Figure 1. Electrocardiography. Sinus rhythm HR, 55/bpm ST-elevation in leads D2, D3,and aVF; ST-depression in leads, V2-V6; since septal electrical activation is from right to left, Q wave is observed in leads V1-V3. In leads V5-V6 **(A)**, posterior elektrokardiography: Sinüs rhythm HR, 59/bpm ST-ellevation; in leads, D2-D3-AVF, and V7-V8-V9 **(B)**.

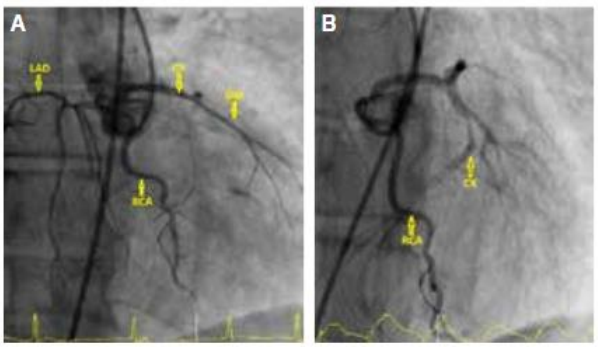

Figure 2. On left anterior oblique view of coronary angiography, complete occlusion of proximal segment of the circumflex artery is seen **(A)**, following percutaneous transluminal coronary angioplasty, and implantation of a drug-eluting stent **(B)**.

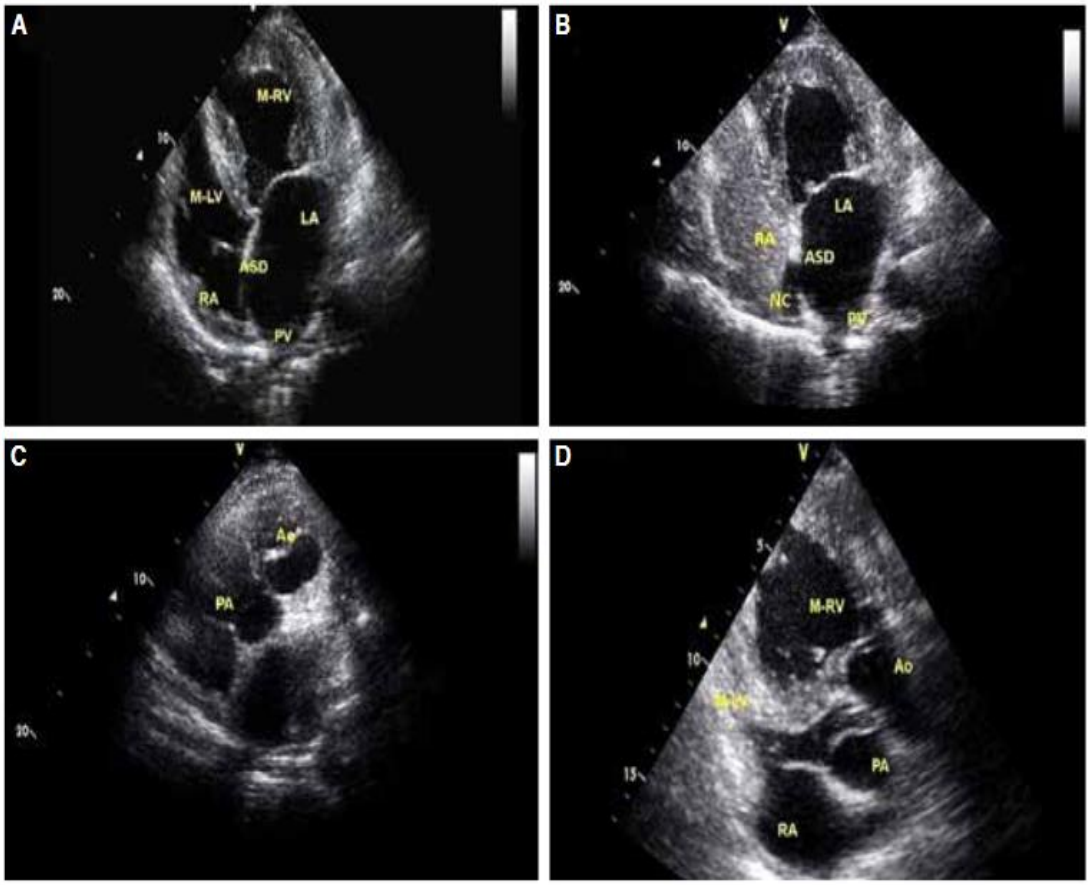

Figure 3. Right ventricle was observed in normal anatomic location of the left ventricle. (hypertrophic). Morphologic left ventricle was seen in the normal anatomic location of the right ventricle. Pulmonary vein drains into the left atrium. **(A)**. Defect was observed in the interatrial septum secundum. On contrast-enhanced echocardiogram negative contrast enhancement was noted. **(B)**. Aorte was localized anteriorly and displaced to the left, while pulmonary artery was situated posteriorly and displaced to the right **(C)**. Aorta arises from morphologic right ventricle. Pulmonary artery stems from the morphologic left ventricle **(D)**.

Second-degree insufficiency was detected in AV valve opening into morphologic RV, while in AV valve opening into morphologic LV first-degree insufficiency was observed. Aorta was positioned anteriorly and displaced to the left, while pulmonary artery was situated posteriorly and displaced to the right side. According to biplane Simpson method RV ejection fraction was measured as 40 percent. In contrast-enhanced echocardiography performed for the identification of the defect in interatrial septum secundum, negative contrast enhancement was seen on electrocardiograms (Figure 3). The patient diagnosed as congenitally CTGA, and ASD was discharged on 8. day of his hospitalization with prescription of medical treatment

DISCUSSION

Corrected transposition of great arteries is a rarely seen condition among congenital heart diseases with an incidence of about 1 percent.[2] In this anomaly, atrial chambers retain their normal position, while ventricles are transposed as their mirror-image symmetry (ventricular inversion, AV discordance). Besides pulmonary artery arises from morphologic LV, and aorta from morphologic RV (ventriculoarterial discordance).[7] Aorta is situated anterior to the pulmonary artery.[8]

Most frequently seen anatomic abnormalities include atrial, and visceral situs solitus, and L-loop ventricle.[9]

Completely normal cases with congenitally corrected transposition of great arteries comprise 5-10 % of all cases Patients with CCTAG without concomitant prevalently seen anomalies as wide ASD, VSD, pulmonary stenosis, heart failure, can remain asymptomatic till advanced ages.[10] However the patients with associated anomalies are treated during childhood are diagnosed,and treated with usual symptoms, and signs of easy fatiguability, cyanosis, inability to gain weight, and growth retardation. Nometheless, our patient remained asymptomatic till advanced age.

In patients with corrected transposition of great arteries, distribution of coronary arteries does not usually change, and follow ventricles.[11] Coronary artery arising from the right sinus of Valsalva generally behaves as morphologic left main coronary artery (LMCA) courses within the right AV groove, and branches into interventricular artery, and Cx, and moves around the mitral valve. The artery arising from the left sinus of Valsalva behaves like a morphologic right coronary artery (RCA), courses in the left AV groove, gives infundibular, and marginal branches, and moves around the tricuspid valve. Noncoronary sinus is usually assumes an anterior location.[12]

In invasive, and non-invasive imaging modalities, the incidence of detected coronary anomalies ranges between 0.6, and 5.6 percent. Various incidence rates have been reported in patients who had undergone conventional CAG (0.6-1.3 %) or coronary angiography with multi-slice computed tomography (0.7-18.4 %), and in autopsy studies (0.3 %).[13] In patients with corrected transposition of great arteries most frequently seen coronary artery anomaly is a coronary artery with a single ostium which crosses over RV outlet.[14] In the literature, coronary anomalies with a single ostium originating from the right sinus of Valsalva have been cited, besides the cases where the left main descending artery,and RCA arising from the right, and Cx from the left sinus of Valsalva have been also reported..[15] In our case right, and left major coronary arteries arised from a single coronary ostium in the right sinus of Valsalva, and LMCA course along the right AV sulcus, and branched into anterior interventricular, and Cx arteries. However morphologic RCA progressed along the left AV sulcus, and divided into infundibular, and marginal branches.

However in our literature reviews, we haven't encountered cases with myocardial infarction with acute ST-segment elevation in association with single coronary artery anomaly, and ASD who had undergone successful PCIs. Echocardiographic examination has a crucial importance in coronary artery patients with single coronary ostium detected during coronary angiography. Even these patients undergo coronary angiography and revascularization one should not forget that without performing echocardiographic examinations diagnosis of cardiac anomalies may be overlooked. It should be also considered that in the determination of the systolic function of the morphologic RV which is the systemic ventricle, along with 2D echocardiographic evaluation, RV function should be assessed with sophisticated echocardiographic methods as 3D echocardiography, tissue Doppler, and strain imaging to obtain valuable information about systemic ventricular functions

Conflict of Interest: None declared

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Anahtar sözcükler: Akut miyokart enfarktüsü; atriyal septal defekt; düzeltilmiş büyük arter transpozisyonu.

Keywords: Acute myocardial infarction; atrial septal defect; corrected transposition of great arteries.