İleri yașta akut inferiör miyokart enfarktüsü ile başvuran ve atrial 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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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 Valsalva, 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 first 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. Its incidence is nearly 1% among all congenital heart diseases. 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. The incidence of coronary artery anomalies has been 1-2% in studies performed with coronary angiograms, and autopsies, and among them most frequently single coronary artery anomalies are seen.

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.

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However coronary arteries of some cases with CCTGA have also undergone PCI. 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.

**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 flattening 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). Acetyl salicylic acid (300 mg p.o.), clopidogrel (600 mg. 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 initiated 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 emergency revascularization. Coronary angiography demonstrated that coronary arteries arised from the right sinus of Valsalva, and complete occlusion of the proximal part of the circumflex 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.

**Table**

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<th>Cardiac Anomalies</th>
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<tr>
<td>ASD Atrial septal defect</td>
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<tr>
<td>AV Atrio-ventricular</td>
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<tr>
<td>Cs Circumflex</td>
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<tr>
<td>CTGA Corrected transposition of the great arteries</td>
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<tr>
<td>EKG Electrocardiography</td>
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<tr>
<td>CAG Coronary angiography</td>
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<td>LMACa left main coronary artery</td>
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<tr>
<td>PCI Percutaneous coronary intervention</td>
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<td>RCA Right coronary artery</td>
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<td>RV Right ventricle</td>
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<tr>
<td>LSV Left ventricle</td>
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<td>VSD Ventricular Septal defect</td>
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**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 elektroardiography: Sinus 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).
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. It occurs because the 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). Aorta is situated anterior to the pulmonary artery.

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

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. 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. Nonetheless, 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, andCx, 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 arise 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

REFERENCES


Anatür sözlükler: Akut miyokart enfarktusu; atriyal septal defekt; düzeltilmiş büyük arter transpozyonu.

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