

Arrhythmogenic right ventricular dysplasia /cardiomyopathy with coronary artery fistula: an uncommon occurrence

Aritmojenik sağ ventrikül displazili koroner arteriyel fistül: Nadir bir oluşum

A 20-year-old man presented at our cardiology clinic for evaluation of progressive dyspnea and fatigue. His past family history included sudden cardiac death in two siblings at 3 and 14 years age. Physical examination showed blood pressure of 80/50 mmHg, pulse rate of 95 beats/min. Chest X-ray revealed cardiomegaly with a cardiothoracic ratio of 70%.

An electrocardiogram revealed sinus rhythm, right bundle branch block and frequent, premature ventricular beats (Fig 1). The echocardiogram showed right ventricular dilatation and hypocontractile areas. Left ventricular functions and dimensions were within normal limits (ejection fraction: 54%). Selective angiography of the left coronary system demonstrated a fistula arising from the proximal left anterior descending coronary artery. The fistula appeared to empty pulmonary artery (Fig 2). Other coronary arteries were normal. Pressures in left ventricle, pulmonary artery, right ventricle and right atrium were 100/12 mmHg, 20/12 mmHg, 20/15 mmHg and 15 mmHg, respectively. No left-to-right shunt was detected; specifically, there was no evidence of an atrial

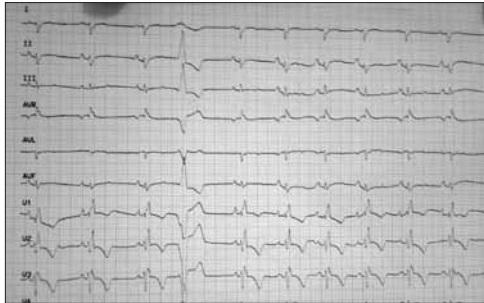


Figure 1. Electrocardiogram showing premature ventricular ectopic beats



Figure 2. Fistula arising from the proximal left anterior descending artery (above white arrow) and possibly emptying into the pulmonary artery (bottom arrow)

septal defect or of an anomalous pulmonary venous return to explain the right-sided chamber enlargement. Right ventriculography revealed right ventricular dilatation and slow evacuation of contrast medium due to severely depressed systolic function (Fig 3).

Computed tomography (CT) was performed with a 16-slice scanner after an intravenous injection of contrast medium. Images demonstrated low-density areas (-108 to -77 HU) indicative of focal fatty infiltration particularly in the anterior and inferior walls and along the right ventricular side of interventricular septum (Fig. 4). Dilatation of the right ventricle; fatty tissue in conspicuous trabeculae of the right ventricle, especially in the anterior wall and inferior (diaphragmatic) wall; and a scalloped appearance (bulging) of the right ventricular wall were characteristic findings at CT to diagnose arrhythmogenic right ventricular cardiomyopathy (ARVC) (1-3). Fatty tissue in the left ventricle and ventricular septum is seen relatively frequently in ARVC, and fat in the ventricular septum is another useful finding for diagnosis of ARVC with CT

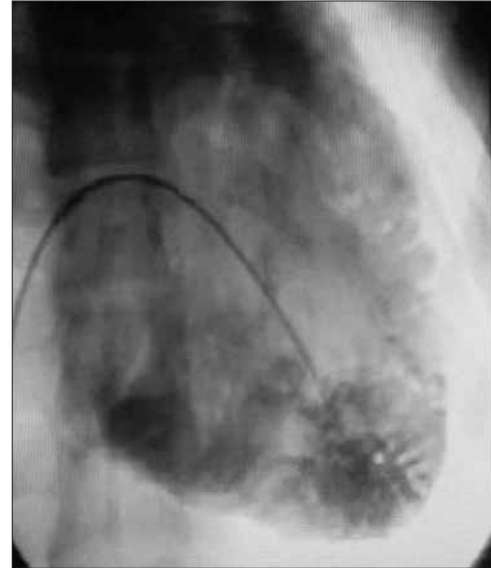


Figure 3. Right ventriculography revealing right ventricular dilatation, deep fissures and enlarged myocardial trabeculae



Figure 4. Computed tomography of the right ventricle. Fatty tissue is observed in the in the conspicuous trabeculae of the anterior wall of the right ventricle (arrows) with low attenuation (-108 to -77 HU)

(1). Multislice computed tomography (CT) may be useful for detecting myocardial fat infiltration and diagnosing ARVC (1-3). Because of its excellent spatial and temporal resolution, CT has received much attention in diagnosing of ARVC. It has been reported that CT findings of ARVC are (a) a dilated right ventricle, b) abundant epicardial adipose tissue, (c) conspicuous trabeculations with low attenuation, (d) a scalloped appearance of the right ventricular free wall, and (e) intramyocardial fat deposits (1, 2).

Coronary artery fistula is an uncommon clinical entity with an incidence in selected series ranging from 0.26% to 0.40% of congenital cardiac anomalies. Many adults are asymptomatic if the fistulae are small. Symptoms of fatigue, dyspnea, angina (due to "steal" phenomenon), atrial arrhythmia, signs of congestive heart failure, pulmonary hypertension or infective endocarditis are seen. In one report, patients older than 20 years had dyspnea on exertion (35%), fatigue (8%) or angina (22%). Conversely, only 9% of those <20 years of age had had such symptoms (4, 5).

As a consequence, we postulated that the small fistula did not contribute to the our patient's right heart dilatation producing a steal phenomenon.

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Hyperthyroidism as a rare cause of complete AV block

Tam AV blokun nadir bir nedeni olarak hipertiroidizm

Hyperthyroidism commonly causes cardiovascular manifestations such as, sinus tachycardia, atrial fibrillation and atrial or ventricular premature complexes. However, complete atrioventricular (AV) block or other AV conduction defects, although being a rare entity in thyrotoxicosis, is an important condition which should be recognized (1).

A 28-year old woman was admitted to emergency department with nausea, vomiting and then sudden loss of consciousness. In the emer-

gency department, on physical examination, blood pressure was 120/75 mmHg, heart rate was 52 beat/min. She had a systolic murmur of 1/6 degree on the mesocardiac area, and she had a 2-3 cm solitary palpable thyroid nodule. Electrocardiogram (ECG) revealed complete AV block (Fig. 1) with heart rate 52 beat/min. Chest X-ray was considered to be normal. Echocardiography revealed only moderate mitral valve insufficiency. We did not consider temporary transvenous pacemaker because of patient's hemodynamic stability. In her medical history, she had been diagnosed as hyperthyroidism, and did not get any medical treatment within the two last years.

Complete blood count revealed a hemoglobin level of 10.8 gr/dl and hematocrit-32.5 g/dl. Thyroid function tests confirmed hyperthyroidism with a serum free triiodothyronine (FT3) level of 13,4 pg/ml (1,7-4,9), serum free thyroxine (FT4) level of 5,4 ng/dl (0,7-2,0) and thyrotropine (TSH) level of <0.01 (0,4-4,1) IU/dl. The other biochemical parameters including auto-antibodies belong to connective tissue disorders, angiotensin converting enzyme (ACE) activity for sarcoidosis, were normal. Anti-thyroid treatment (propylthiouracil, PTU) was started. Her rhythm resolved from complete AV block to second degree AV block (Mobitz type 2) on her fourth day of hospitalization (Fig. 2), to first degree AV block on the seventh day (Fig. 3) and to normal rhythm on the eighth day of hospitalization. Thyroid ultrasonography was performed and a 3 cm solid nodule on the right lobe and a 1 cm solitary nodule on the left lobe were observed.

Treatment of the AV block in hyperthyroidism is based on the treatment of the hyperthyroidic condition. Because, electrocardiographic findings returned to normal in the short time after antithyroid treatment in these patients. The palpitation in patient with hyperthyroidism is commonly due to tachyarrhythmia, however, it may also be due to bradyarrhythmia or complete AV block. β -blocker treatment of palpitation due to bradyarrhythmia or complete AV block may be dangerous in patients with hyperthyroidism. Recognition that complete AV block can complicate thyrotoxicosis is important. That is why, electrocardiography should be taken before treatment of palpitation.

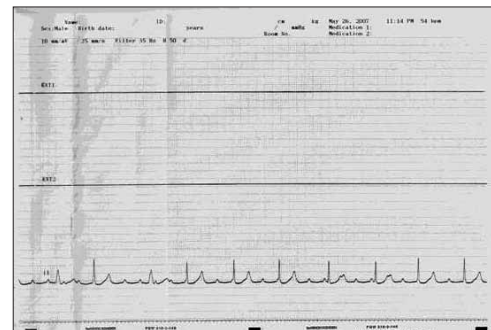


Figure 1. Complete AV block on the ECG performed at the time of submission

AV-atrioventricular, ECG-electrocardiogram

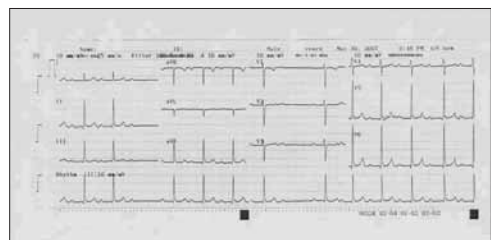


Figure 2. Mobitz type 2 on the ECG performed on the fourth day of hospitalization

ECG - electrocardiogram