KLİNİKİ MÜŞAHİDƏLƏR **AÇIQ GİRİŞ (OPEN ACCESS)**

Full recovered sudden cardiac arrest as a result of tachycardia induced CMP related to miocarditis in a patient with Marfan syndrome.

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Description –A 22-year-old male with Marfan syndrome presented with dyspnea andpalpitations. He has been following by cardiologist for 4-5 years. During the last examination, the patient had supraventricular tachycardia and was referred to a specific center. In admission, the patient had a decrease in the ejection fraction and was hospitalized due to symptoms of heart failure.
Learning outcomes – Atrial arrhythmias also occur with increased frequency in patients with Marfan's syndrome. Atrial arrhythmias do not carry the same risk of sudden death as ventricular arrhythmias but are still clinically significant.

Patient presentation - A 22-year-old boy, 194 cm tall, weighting 94 kg, with Marfan syndrome, came to our clinic with complaints of palpitations, dyspnea, quick fatigue (NYHA Class II). Two days before hospitalization, progressive mitral valve insufficiency and one or two non-sustained VT episodes and frequent episodes of atrial tachycardia were revealed on 24 Hour Holtermonitoring. When patient has applied to our clinic 2: 1 atrial tachycardia was recorded (pulse 152) on ECG. In ECHO LVEF was 40-45% (global hypokinesia), pear-shaped aortic root dilatation (annulus 32mm, sinus valsalva 43mm, sinotubular junction 36mm), moderate mitral regurgitation, mild tricuspid regurgitation, left ventricular and atrial enlargement (LV 58/38, LA 44mm), SPAP 40 mmHg was recorded. On physical examination, blood pressure was 90/70 mmHg (left arm), 100/70 (right arm), heart rate - 140 bpm. The thorax is the pectusexcavatum. On auscultation, heart sounds are weakened, systolic murmur grade 4/6 is heard at the apical point of hearing, breathing sounds are clear.

The most common type of arrhythmia in patients with Marfan syndrome are premature atrial or ventricular beats. This was not related to aortic root dilatation, left atrial dilatation, LV dilatation or function. It was presumed that deficiency of fibrillin-1 in

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patients with MFS, which causes microfibril abnormality in the matrix of the myocardium, affects conduction of impulses.

Initial work up -The patient was hospitalized with a diagnosis of atrial tachycardia and heart failure. During the first hour of hospitalization, sudden cardiac arrest occurred and ventricular fibrillation was seen on monitore. CPR were initiated immediately in accordance with the recommendations, cardioversion was performed (with 220J), and in a short time (5 minutes) the patient's vital signs and sinus rhythm were restored. Bedside Echocardiography showed more reduced ejection fraction (LVEF 30%) after CPR. Cordaron infusion was started at ICU.Laboratory parameters were Hemoglobin 14.4 g/dl, WBC 8.93, Troponin I- 0.014 ng/ml, Potassium 4.2 mmol/l, Calcium 9.4 mg/dl, creatinine 1.1 mg/dl, CRP 1.71 mg/l. Vital signs were A/T 100/70mmHg, HR 120bpm. Medical treatment, Spironolactone 25mg, Cordarone tab 200mg, Ramipril 2.5mg, Metoprolol 12.5mg, Enoxaparin 0.6ml were started. After 7 days, the patient was discharged home hemodynamically stable and without any neurological problems.

The first choice for arrhythmia treatment in a patient with Marfan's syndrome are Metoprolol. Beta-blockers, a group of drugs used to decrease blood pressure and heart rate, have been recommended by guidelines as the first line medical treatment of Marfan syndrome. And also

Diagnosis and management - Once the patient's condition stabilized, an MRI of the heart was performed to identify myocardial disease that could cause heart failure and arrhythmias. LVEF was recorded 31% on MRI, EDV 195 ml, ESV 134 ml. İnitially, left ventricular damage is more similar to ARVD, with severe diastolic dysfunction. Myocarditis-related tachycardia and tachycardia-induced cardiomyopathy were thought to be present and followed by medical treatment.

Follow up - At the follow-up examination after 6 months, the ECG is sinus rhythm, there are no specific changes. The patient has no complaints (NYHA Class I). On repeated MRI, the systolic function of the left ventricle is completely normal,LVEF 55%, EDV 159ml, ESV 69ml, the left ventricular damage is more reminiscent of myocarditis. ECHO showed normalization of the ejection fraction (LVEF 55% asynchronous movement), reduction of left atrium size (LA 36mm), decreased SPAP - 35mmHg, mild MR, mild TR. Treatment is Ramipril 2.5 mg, Metoprolol 25 mg, Spironolactone 25 mg.

Conclusions -Mild but significant myocardial dysfunction and arrhythmias are more common in patients with Marfan syndrome than in other patients. Therefore, we recommend monitoring myocardial function and arrhythmias in all patients with Marfan syndrome. Patients with left ventricular enlargement, decreased function, palpitations and additional cardiovascular risk factors are at higher risk populations, so we recommend more frequent follow-up examinations, such as a routine 12-lead ECG, cardiac ultrasound, 24-hour ambulatory ECG, and if necessary CMRİ.

Published guidelines on the cardiovascular management of patient's with Marfan disease have been based on case reports rather than randomized controlled trials.

References:

1. Myocardial disease and ventricular arrhythmia in Marfan syndrome: a prospective study

Laura Muiño-Mosquera, Hans De Wilde, Daniel Devos, Danilo Babin, Luc Jordaens,

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Əlavə məlumatlar.

Müəlliflərin töhfələri.
Konsepsiya və dizayn, Məlumatların əldə edilməsi, təhlili və ya təfsir, Əlyazmanın tərtibi, Əlyazmanın mühüm intellektual məzmun üçün tənqidi təftişi, Statistik təhlil, Məlumatların idarəedilməsi, Araşdırma, Əldə edilmiş dəstək, maliyyə və nəzarət: bütün müəlliflər bərabər qaydada. Müəlliflər yekun əlyazmanı oxuyub və təsdiq edib.

Maliyyələşdirmə.
Məqalənin hazırlanması məqsədilə aparılan təhlil və araşdırmalar üçün heç bir kənar maliyyə əldə edilməmişdir. Heç bir digər qurum və ya sponsor təşkilatlararaşdırmanın və ya tədqiqatın və ya təhlilin dizaynı və aparılmasında; məlumatların toplanması, idarə edilməsi, təhlili, məlumatların təfsirində, habelə əlyazmanın hazırlanması, nəzərdən keçirilməsi və ya təsdiqində heç bir rola malik olmayıb; əlyazmanın nəşrə təqdim edilməsi haqqında qərarların verilməsində iştirak etməmişdir.

Məlumat və materialların əlçatanlığı.
Təhlil zamanı istifadə olunan və/yaxud təhlil edilən məlumatlar (datalar) müəlliflərə və ya jurnalın redaksiyasına müraciət etməklə əldə edilə bilər.

Bəyannamələr.
Etik Komitənin icazəsi və məlumatlı razılıq.
Hər bir iştirakçıdan yazılı və ya uyğun olduqda şifahi məlumatlı razılıq alınıb. Etik Komitə (AKC, Azərbaycan) bu təhlili təsdiq edib.

Maraqların toqquşması.
Müəllif(lər) hər hansı maraqların toqquşmasını bəyan etməyiblər.

Müəlliflərə dair təfərrüatlar.
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