Abstract: **P611**

**A case of arrhythmogenic right ventricular cardiomyopathy**

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Introduction

Arrhythmogenic right ventricular cardiomyopathy (ARVC), also known as arrhythmogenic right ventricular dysplasia, is a heritable heart-muscle disorder that predominantly affects the right ventricle. Progressive loss of right ventricular myocardium and its replacement by fibrofatty tissue is the pathological hallmark of the disease. ARVC is one of the leading causes of arrhythmic cardiac arrest in young people and athletes. We present a clinical case of Arrhythmogenic right ventricular cardiomyopathy

Case Presentation

A 25-year-old female presented with a one year dyspnea after being diagnosed as systemic lupus erythematosus by a hospital in our city one year ago. She was diagnosed with brain tumor 17 years ago at the age of 8. Her treatments for brain tumor were surgery, chemotherapy and radiation therapy And then her family history decided for her to stop taking medicine and did not to follow treatment plan. Since then she has noticed she has no period. Her symptoms include fatigue, weakness, inability to lose weight (or weight gain), puffiness, constipation, Physical examination may, periorbital puffiness, brittle hair and eyebrow loss. Other findings were normal.

Results

Echocardiogram: EF 53%, normal size and wall thickness of the left ventricle, slightly dilated left atrium, enlargement of the right ventricular 50mm and RVOT 47mm with reduced contractile function TAPSE 11mm, apical, aneurysm-akinesia and endocardial ventricular hypertrabeculation especially at apex. Mild mitral regurgitation and severe tricuspid regurgitation. Medium pericardial effusion without right ventricular depression. Cardiac MRI showed normal volume and function of the left ventricle (EF 55%), right ventricle with 114 ml/m2 indexed end diastolic volume, reduced right ventricular function (EF 29.6%), areas of dyskinesia of the free wall at the mid and apex. Late gadolinium enhancement of the free wall right ventricular at the mid and apex. Hypertrabeculation of septal wall at the apex. Medium pericardial effusion, no cardiac tamponade.

Discussion

Diagnosis of ARVC relies on a scoring system, formulated in 2010 by the revisited Task Force, with two major or one major and two minor criteria or four minor criteria based on the demonstration of a combination of defects in right ventricular morphology and function, characteristic depolarization/repolarization electrocardiogram abnormalities (negative T waves and/or "epsilon" waves in right precordial leads), characteristic tissue pathology, typical arrhythmias, family history, and the results of genetic testing.

Conclusion

In ARVC symptoms usually appear between the ages of 30–50. Especially in young patients the most common clinical presentation of ARVC are palpitations and syncope due to ventricular tachycardia with left bundle
branch morphology. Cardiac magnetic resonance (MRI) is considered the best imaging modality in evaluating the RV in ARVC.