Echo-Electrocardiographic Imaging of Arrhythmogenic Right Ventricular Cardiomyopathy: Original Image

Aritmojenik Sağ Ventrikül Kardiyomiyopatisinin Eko-Elektrokardiyografik Görüntüsü

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Arrhythmogenic right ventricular cardiomyopathy (ARVC) is a familial cardiac disease characterized by ventricular arrhythmias and sudden cardiac death (SCD). It is most frequently inherited as an autosomal dominant trait with incomplete and age-related penetrance.[1] Its diagnosis is challenging because most patients are asymptomatic, and SCD is often the first manifestation of the disease. A critical component of the screening for and diagnosis of this disease is noninvasive imaging of the right ventricle.[2] A 23 years old asymptomatic patient was evaluated for possible ARVC in the context of family screening. His father was known to be survivor of SCD due to ARVC at age 48 and had undergone an implantable cardiac defibrillator (ICD) implantation, and his uncle was a victim of SCD due to unknown cause at age 45. On electrocardiography, a pronounced epsilon wave, T-wave inversion and localised QRS prolongation (>110 ms) in right precordial leads were detected (Figure 1). On echocardiography (Figure 2), a focal right ventricular apical aneurysm with excessive trabecular derangement and sacculations (thin arrow in 2A, B and C), and a hyper-reflective moderator band (arrow in Figure 2A) were seen. Severe right ventricle and right ventricle outflow tract enlargement (parasternal long-axis dimension of 48 mm, asterix in Figure 2C) and severe right ventricle systolic dysfunction were also detected. An ICD was implanted for primer prevention and he was listed for heart transplantation.

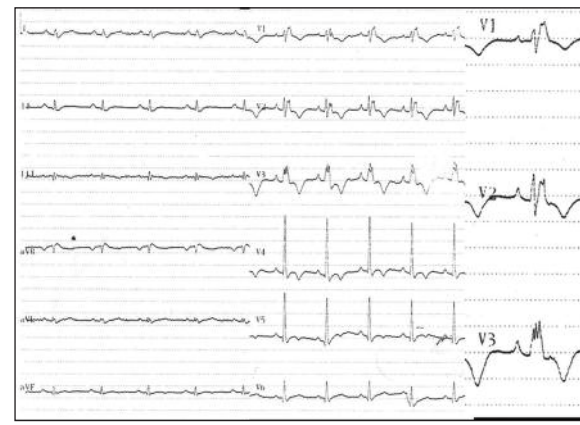

FIGURE 1: Electrocardiogram showing a pronounced epsilon wave, T-wave inversion and localised QRS prolongation (>110 ms) in right precordial leads.

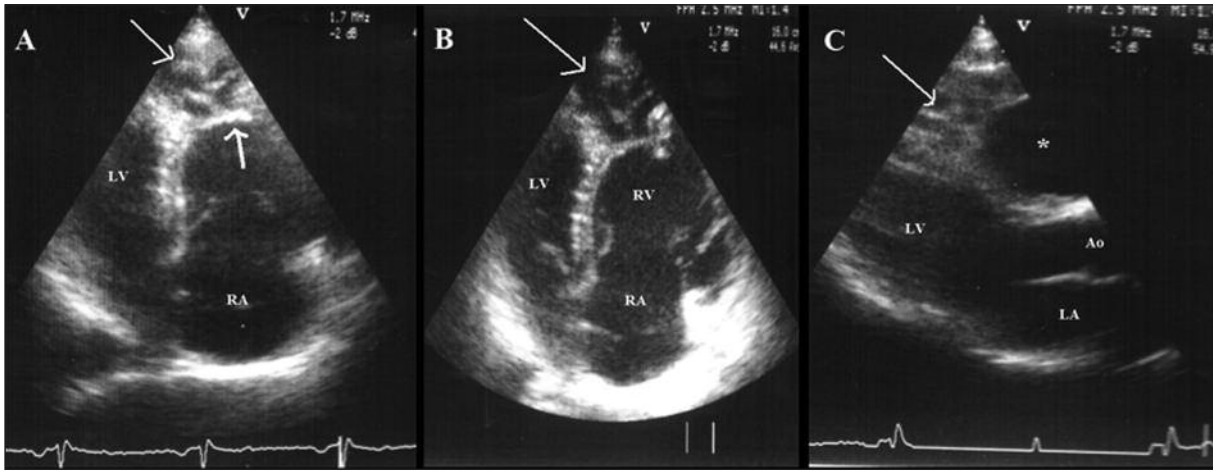

FIGURE 2: Echocardiogram showing typical morphologic (hyper-reflective moderator band, thick arrow in A; trabecular derangement and sacculations, thin arrow in A, B and C) and functional (severe RV and RV outflow tract enlargement and RV systolic dysfunction) abnormalities.
RA, right atrium; RV, right ventricle; LA, left atrium; LV, left ventricle; Ao, Aort.

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