A Case of Arrhythmogenic Right Ventricular Dysplasia Associated with FANA Positiveness:

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Arrhythmogenic right ventricular dysplasia (ARVD) is an unusual disease in which a predominantly right-sided cardiomyopathy is associated with ventricular arrhythmias. It is based on the presence of ventricular arrhythmia with left bundle branch block (LBBB) configuration and morphologic changes or motion abnormalities localized in the free wall of the right ventricle (RV). These abnormalities are usually associated with RV enlargement and diffuse hypokinesia with no identifiable etiology.

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We report a case presenting attacks of ventricular arrhythmia with LBBB in whom we found right ventricular abnormalities and unusual positiveness of fluorescent anti-nuclear antibody (FA-NA) for this disease.

Case report

A 45 year-old woman was admitted to the cardiology department for spontaneous tachycardia attacks of short duration for more than a year. She also complained of pain in her hands and knees, with no history of arthritis, and paleness of her hand during cold weather for a few years.

On her physical examination early systolic murmur graded as 2/6 and doubled second heart sound on the left sternal border of 2nd intercostal space were heard. Prominence of pulmonary arch was striking on her telecardiography. No pathological findings were seen in her hands, feet, cervical, lumbosacral, or sacroiliac X-ray films. ECG revealed incomplete RBBB, T negativeness between V1 and V6, and rare VPCs. During palpitation attack, ventricular tachycardia with LBBB conduction were observed (Fig. 1). Laboratory tests showed positiveness of fluorescent antinuclear antibody (FANA specled type), anti-DNA: 86.3u/ml; C3: 95.1 u/ml; ESR: 23 mm/h, CRP and RF (-); cryoglobulin (-); HBsAg (-). Raynaud's phenomenon was not observed on cold pressor test.

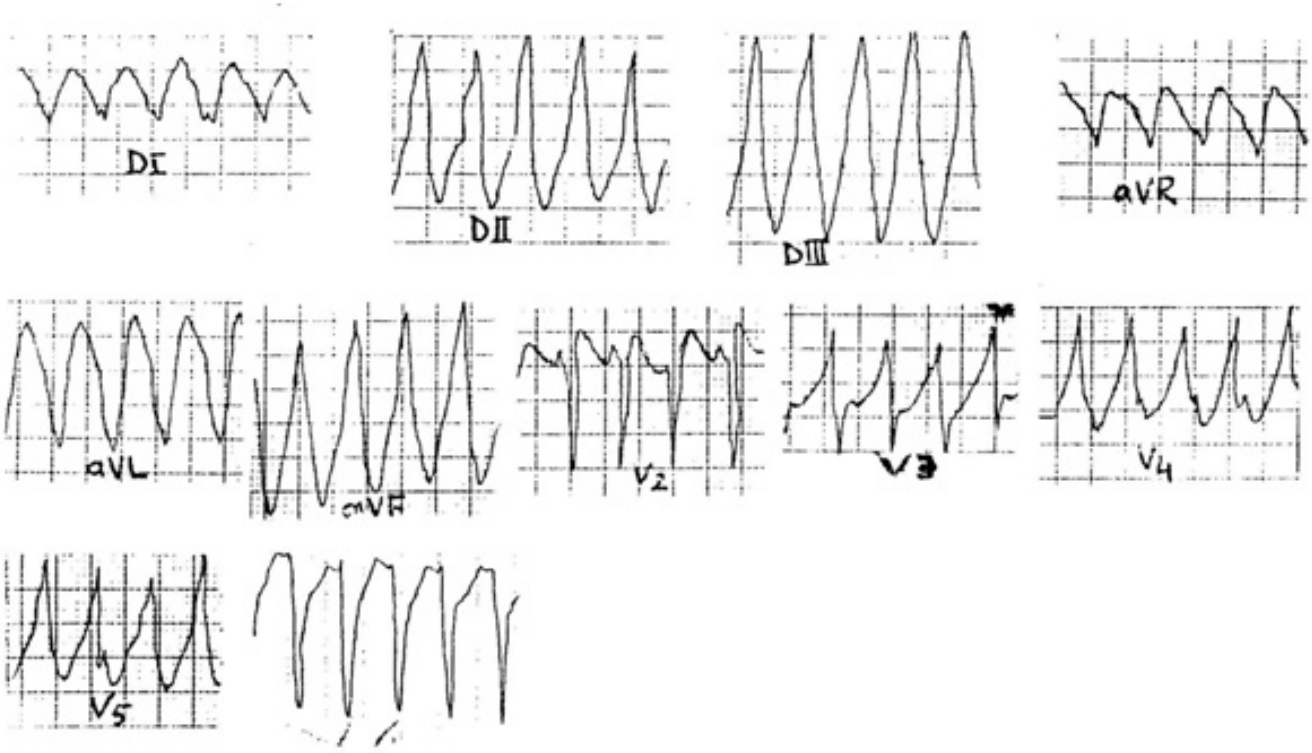


Fig. 1: ECG during tachycardia with LBBB conduction

Echocardiography showed marked enlargement of the right atrium, right ventricle, and pulmonary artery. Coronary angiography and left ventriculography were normal (Fig. 2,3). Right ventriculography revealed severe enlargement of the right ventricle and main pulmonary artery (Fig. 4). Attempts to reach to the pulmonary artery were not successful. Right ventricular pressure was 36/0/17 mmHg and left ventricular 135/0/8 mmHg.

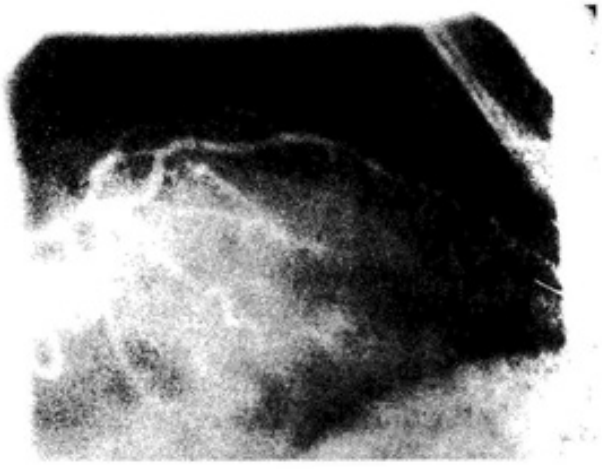


Fig. 2: View of right anterior oblique position of coronary angiography within normal limits.

Fig. 3: View of left ventriculography within normal limits

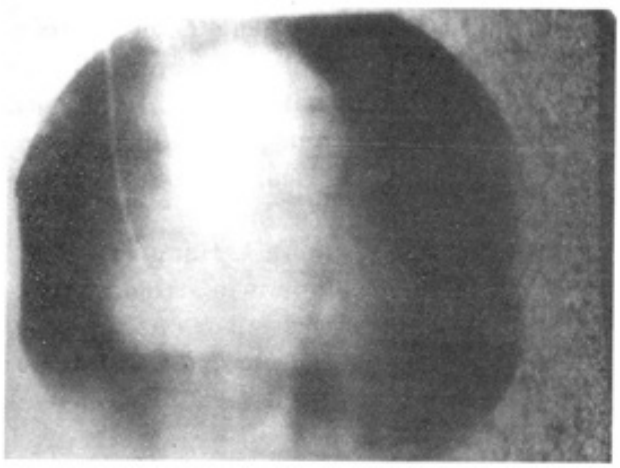


Fig. 4: Severely enlarged right ventricle and main pulmonary artery with right ventriculography

Endocardial biopsy from the right ventricle revealed marked myocardial muscular dege-neration (Fig.5,6,7,8).

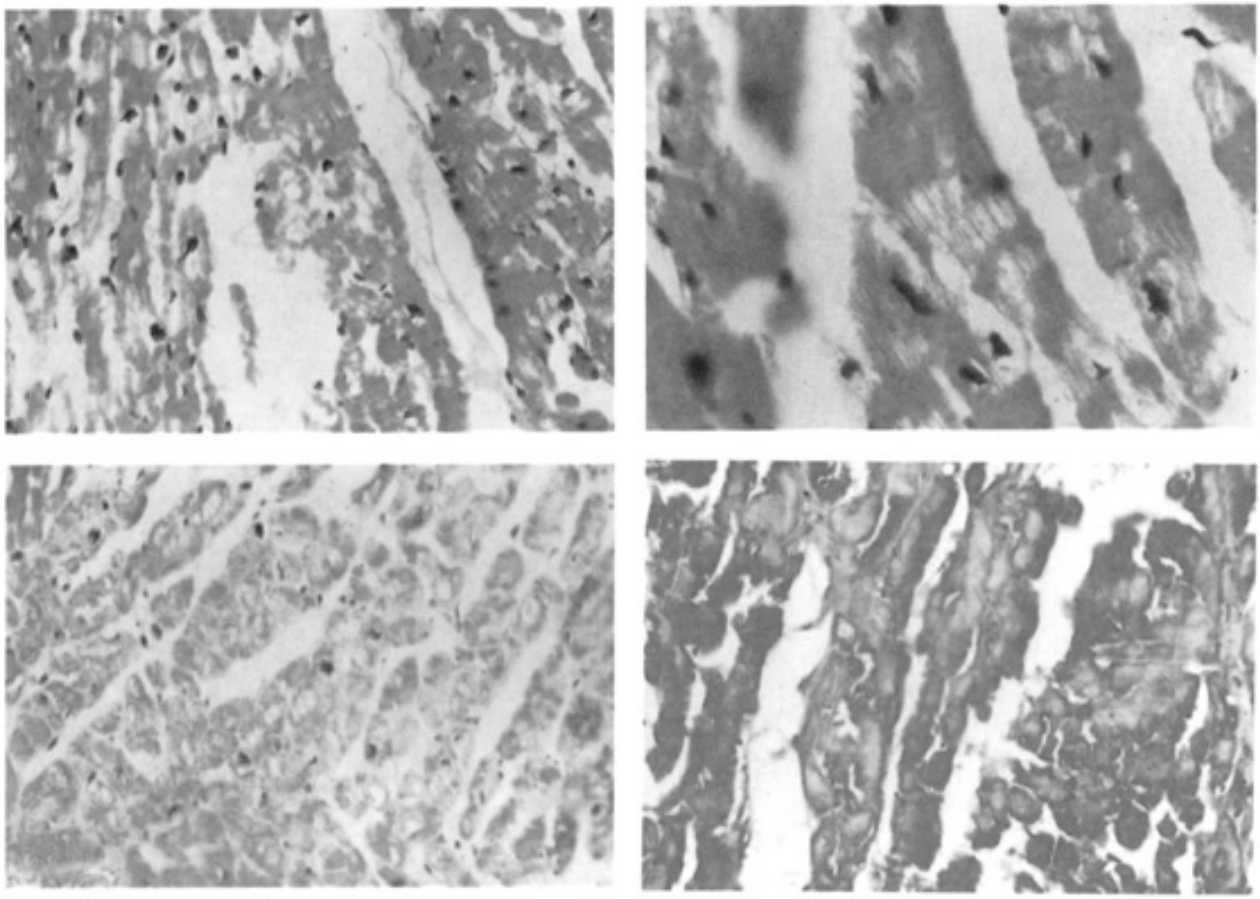
Follow up:

After unsuccessful treatment with Verapamil and Procainamid consequently, Amiodarone was begun and remained the patient symptom-free for more than a year.

Discussion

With these aforementioned findings "arrhythmogenic right ventricular dysplasia" was diagnosed. All except FANA positiveness were consistent with the findings stated in literature. Nothing was found in literature about the association of FANA positiveness and right ventricular dysplasia or about whether it is a coincidence or a rare feature of this disease.

Our patient has several features that are typical of ARVD[1,2,3,4,5,6]. In addition she showed some unusual manifestations of this cardiomyopathy. (eg. FANA positiveness). All findings except FANA positiveness were consistent with the findings stated in litera-



Figs. 5,6,7,8: Histopathological findings.

ture. The causes of FANA positiveness were all reviewed.

As far as we know this is the first report of FANA positiveness associated with ARVD. Because both are rare conditions their association may not have been fortuitous and may be a rare feature of this disease.

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