Dilated Right Ventricle with Impaired Systolic Function

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CASE REPORT

We describe a 50-year old female admitted to our cardiac department with an episode of palpitations and associated dyspnea. She had never had any previous medical history. Physical examination revealed an irregular heart rate of 130 beats per minute, blood pressure of 122/70 mmHg and no pyrexia. Twelve–lead ECG demonstrated atrial fibrillation. Transthoracic echocardiogram showed a dilated right ventricle. The right ventricular outflow tract was measured at 40 mm and the maximum dimension of the middle third of the right ventricle was measured at 43 mm. Right ventricular (RV) systolic function was impaired; the RV fractional area change was 32%, tricuspid annular plane systolic excursion (TAPSE) was measured at 14 cm and the systolic wave of the pulsed tissue Doppler from the lateral tricuspid annulus was 9 cm/sec. There was mild tricuspid regurgitation with no pulmonary hypertension. An abnormal shape and function of the interventricular septum was detected. Left ventricle and valves were normal.

The transesophageal echocardiogram revealed a secundum atrial septum defect (ASD) of about 0.6 cm in diameter. The ratio of pulmonary to systemic blood flow (Qp/Qs) was measure at 1.3, which is not consisted with a significant left to right shunting and the RV dilation. Atrial fibrillation was converted to sinus rhythm with amiodarone and the patient was discharged home, while she was advised to return for re-evaluation but unfortunately she did not return to clinic.

Two years later the patient was admitted again with a sustained ventricular tachycardia of left bundle – branch block morphology which required defibrillation therapy. Magnetic resonance imaging (MRI) showed RV dyskinesia and impaired systolic function (RV ejection fraction of 33%). A new echocardiogram uncovered an aneurysm at the anterior wall of the right ventricle. She met two major criteria according to the International Task Force for the diagnosis of arrhythmogenic right ventricular cardiomyopathy (ARVC).

COMMENT

Arrhythmogenic RV cardiomyopathy is a familiar myocardial disease characterized by progressive fibrofatty replacement of the RV myocardium which constitutes a substrate for electrical instability. The diagnosis of ARVC relies on the presence of structural and functional abnormalities of the right ventricle, electrophysiological abnormalities and a family history. Echocardiography plays an important role in the evaluation of patients with ARVC. However, assessment of the right ventricle is difficult, requiring a combination of different echocardiographic modalities and a high clinical suspicion.
REFERENCES

