Kardiologia Polska 2013; 71, 10: 1079–1081; DOI: 10.5603/KP.2013.0265 ISSN 0022–9032

OPIS PRZYPADKU / *CASE REPORT*

From primary tricuspid regurgitation to arrhythmogenic right ventricular cardiomyopathy

Od pierwotnej niedomykalności zastawki trójdzielnej do arytmogennej kardiomiopatii prawej komory

Adrian Gwizdała, Hubert Popiak, Magdalena Janus, Stefan Grajek, Ewa Straburzyńska-Migaj

1st Department of Cardiology, Poznan University of Medical Sciences, Poznan, Poland

Abstract

Since arrhythmogenic right ventricular dysplasia is still an under-recognised clinical entity, its 'deceitful' course requires alertness of physicians, and — in particular — awareness of its less typical manifestations. Therefore, we present a case report of a 52-year-old male subject with signs and symptoms of right ventricular heart failure and marked tricuspid regurgitation.

Key words: primary tricuspid regurgitation, arrhythmogenic right ventricular dysplasia

Kardiol Pol 2013; 71, 10: 1079–1081

INTRODUCTION

Since arrhythmogenic right ventricular cardiomyopathy (ARVC) is still an under-recognised clinical entity [1], its 'deceitful' course requires awareness of less typical manifestations. Therefore, we present a case report of a 52-year-old male clarinetist who reported with signs and symptoms of right ventricular (RV) heart failure.

CASE REPORT

In 2006, the patient was first seen by his doctor due to worsening of physical capacity (NYHA class III), resting dyspnoea, peripheral oedemas and weight gain following pneumonia. His medical and family history was irrelevant. Physical examination revealed: loud systolic murmur in the left 4th intercostal space, pulmonary congestion and enlarged liver. The echocardiography showed significant RV and atrial enlargement (RVEDD 51 mm) with hypokinesis of the free wall, paradoxical interventricular septum movement, dilation of the tricuspid ring with lack of leaflet coaptation and major tricuspid regurgitation (TR) as well as right ventricular systolic pressure (RVSP) of 35 mm Hg. A 12-lead ECG showed atrial tachycardia 120–150 bpm with 2:1 atrioventricular conduction and rSr' complex in right precordial leads. A 24-h ECG revealed supraventricular ectopic beats (< 1,000/24 h) and single ventricular premature contractions of left bundle branch block morphology. An angio-computed tomography scan excluded pulmonary embolism as a cause of RV enlargement. Eventually, the consulting cardiologist made a diagnosis of severe primary TR and qualified the subject for surgical treatment. Prior to surgery, the patient underwent coronary angiography (January 2007, no significant stenosis) and another echocardiography (LVEDD 56 mm, RVDD 47 mm, LVEF 60%, TR IV, MR I/II, RVSP 20 mm Hg). Consequently, in February 2007, tricuspid annuloplasty was performed (Duran AnCore® Annuloplasty Ring, 620 R 31 mm, Medtronic).

The early post-surgical course was complicated with recurrent hydrothorax (treated with punctures and steroids), and atrial fibrillation (amiodarone). The echocardiography (August 2007) documented persistent RV dysfunction with progressive left ventricular (LV) involvement (LVEF 30%).

During the follow-up period, the patient's condition gradually improved (NYHA class II). However, RV failure shortly recurred and after three years the patient was referred to our tertiary reference Department of Cardiology for evaluation. At that time the clinical manifestation was typical of RV heart failure. However, the ECG revealed sinus rhythm with a distinct epsilon wave which had not been seen previously (Fig. 1). ARVC was suspected and then confirmed by

Address for correspondence:
Adrian Gwizdała, MD, PhD, 1st Department of Cardiology, Poznan University of Medical Sciences, ul. Długa 1/2, 61–848 Poznań, Poland, e-mail: agwizdala@ump.edu.pl
Received: 04.06.2012 **Accepted:** 21.06.2012

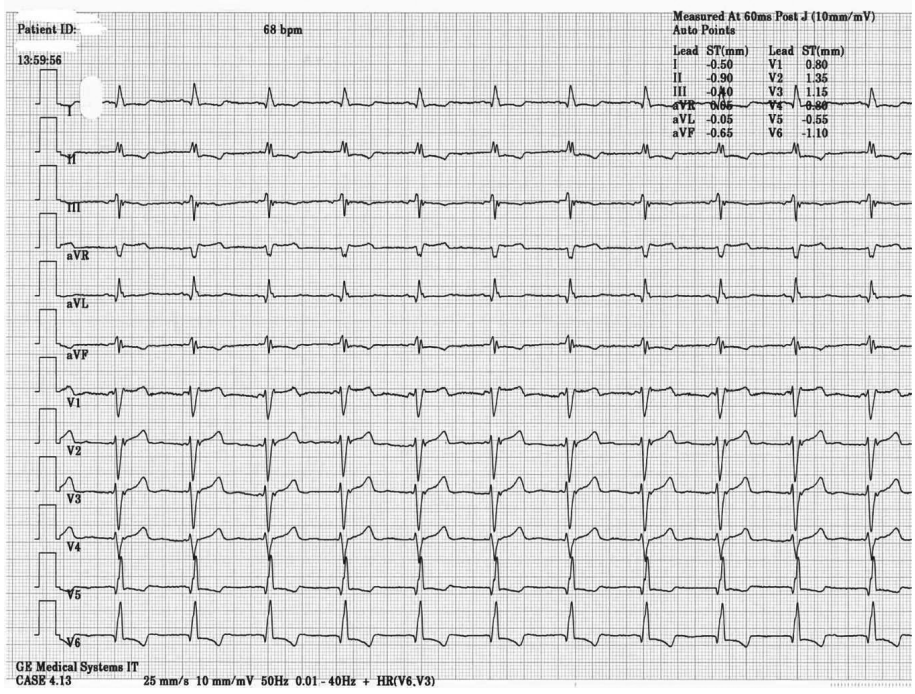

Figure 1. 12-lead ECG: epsilon wave in right precordial leads. QT dispersion is 40 ms

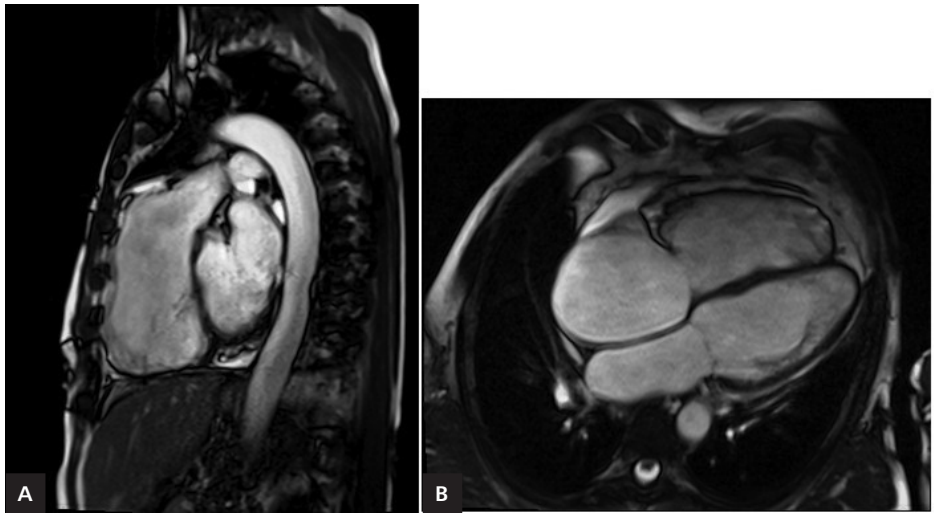

Figure 2. **A**, **B**. Cardiac magnetic resonance imaging showed impaired right ventricular systolic function with sacculations, dyskinesis and fibrosis of its free wall. Left ventricular systolic dysfunction plus paradoxical interventricular septum movement

both echocardiography and cardiac magnetic resonance imaging (Fig. 2).

Subsequent echocardiography (April 2010) showed typically dilated RV (51 mm) with dyskinetic and thin free wall and sacculations as well as secondary dilatation of the tricuspid annulus and large TR (LVEF of 50% with LVEDD 70 mm). Another 24-h ECG showed sinus rhythm and only 177 ectopic ventricular beats without ventricular tachycardia. Nevertheless, the patient met the ESC criteria of ARVC definite diagnosis (two major ones: RV dysfunction and structural alterations, and presence of epsilon wave) [2]. Beta-blocker therapy was initiated and implantable cardioverter-defibrillator (ICD) implantation was scheduled.

In August 2010, a 24-h ECG revealed predominating junctional rhythm mostly at 30–50 bpm and 1,850 single ventricular ectopic beats, and no ventricular tachycardia. ICD was implanted (Medtronic Maximo II DR).

Due to ongoing progression of biventricular heart failure, the subject was screened (November 2010) for heart trans-

plantation. The heart transplantation protocol-required tests documented mild pulmonary hypertension, peak oxygen consumption of 22 mL/kg/min, and elevated B-type natriuretic peptide level (1,214.5 pg/mL). Finally, the subject was referred for an outpatient follow-up. Regular ICD interrogation revealed no episodes of ventricular tachycardia or fibrillation thus far (March 2012).

DISCUSSION

ARVC is usually diagnosed secondarily to previous ventricular tachycardia or ventricular fibrillation. Typically, the clinical presentation includes previously asymptomatic young active individuals suffering from sudden cardiac death, especially during exertion [2, 3]. The presented subject is unique due to atypical manifestation that significantly delayed a correct diagnosis. There are at least a few causes of late diagnosis in this case. As mentioned above, natural manifestations of ARVC include a range of RV arrhythmias and/or progressive right or biventricular heart failure [4], but there are reports of less typical and confusing presentations [5]. Moreover, the diagnosis of primary TR seems to be very premature. Primary TR in the adult is rather a rare finding and results from e.g. Ebstein anomaly, rheumatic disease, infective endocarditis or degenerative lesions [6]. On the other hand, significant TR, apart from RV dysfunction, biventricular involvement and amiodarone treatment, has been found to be a strong predictor of poor prognosis in ARVC subjects [7].

The misleading lack of life-threatening ventricular arrhythmia probably hugely contributed to the misdiagnosis. Initially, the subject had a significant amount of neither single nor complex ventricular arrhythmias despite extensive ECG monitoring. Meanwhile, most papers report the predominance of ventricular arrhythmia in ARVC. Hulot et al. [8] revealed ventricular tachycardia of left bundle branch block morphology in all fatal cases, with premature ventricular contractions (> 1,000/24 h) and epsilon wave in 31% and 16% of subjects, respectively. Ventricular arrhythmias in routine ECGs were reported in almost all patients (27/28) analysed by Corrado et al. [9]. Although not being a rule, a lack of ventricular arrhythmia may be observed in a later phase when a diffuse disease involves both ventricles [10].

The ESC guidelines allow individuals to be free from ventricular arrhythmia while still meeting the criteria of a definite ARVC diagnosis. Even though in the majority ventricular arrhythmia may be the first, and often terminal, manifestation [9], clinicians need to be aware of less common presentations. More frequent evaluations may be required, including repeated ECG monitoring, magnetic resonance imaging, myocardial biopsy or genetic testing. As ARVC often affects young and active subjects, it requires early and accurate diagnosis to prevent sudden cardiac death (ICD implantation) and to avoid unnecessary procedures.

Conflict of interest: *none declared*

References

1. McKenna WJ, Thiene G, Nava A et al. Diagnosis of arrhythmogenic right ventricular dysplasia/cardiomyopathy. Task Force of the Working Group Myocardial and Pericardial Disease of the European Society of Cardiology and of the Scientific Council on Cardiomyopathies of the International Society and Federation of Cardiology. Br Heart J, 1994; 71: 215–218.
2. Thiene G, Nava A, Corrado D et al. Right ventricular cardiomyopathy and sudden death in young people. N Engl J Med, 1988; 318: 129–133.
3. Marcus FI, McKenna WJ, Sherrill D et al. Diagnosis of arrhythmogenic right ventricular cardiomyopathy/dysplasia Proposed Modification of the Task Force Criteria. Eur Heart J, 2010; 31: 806–814.
4. Marcus F, Fontaine G. Arrhythmogenic right ventricular dysplasia/cardiomyopathy: a review. Pace Pacing Clin Electrophysiol, 1995; 18: 1298–1314.
5. Kukla P, Biernacka EK, Jastrzębski M et al. Arrhythmogenic right ventricular cardiomyopathy with left ventricular involvement mimicking acute coronary syndrome: two case reports. Kardiol Pol, 2011; 69: 470–474.
6. Pravin MS, Aidan AR. Tricuspid valve disease. Curr Probl Cardiol, 2008; 33: 47–84.
7. Pinamonti B, Dragos AM, Pyxaras SA et al. Prognostic predictors in arrhythmogenic right ventricular cardiomyopathy: results from a 10-year registry. Eur Heart J, 2011; 32: 1105–1113.
8. Hulot JS, Jouven X, Empana JP et al. Natural history and risk stratification of arrhythmogenic right ventricular dysplasia/cardiomyopathy. Circulation, 2004; 110: 1879–1884
9. Corrado D, Basso C, Thiene G at al. Spectrum of clinicopathologic manifestations of arrhythmogenic right ventricular cardiomyopathy/dysplasia: a multicenter study. J Am Coll Cardiol, 1997; 30: 1512–1520.
10. Sen-Chowdhry S, Syrris P, Ward D et al. Clinical and genetic characterization of families with arrhythmogenic right ventricular dysplasia/cardiomyopathy provides novel insights into patterns of disease expression. Circulation, 2007; 115: 1710–1720.