Arrhythmogenic Right Ventricular Cardiomyopathy

Guy Fontaine MD

University Pierre et Marie Curie, La Salpêtrière Hospital, Paris, France

KEY WORDS: ventricular tachycardia (VT), arrhythmogenic right ventricular cardiomyopathy (ARVC), arrhythmogenic right ventricular dysplasia (ARVD), myocarditis

IMAJ 2014; 16: 375–376

In this issue of *IMAJ*, Belhassen et al. [1] describe an interesting case of a 78 year old patient who experienced multiple episodes of cardiac arrhythmias during a 40 year period. This arrhythmia was finally identified correctly as ventricular tachycardia and not supraventricular tachycardia. The most frequent cause of VT in the elderly is surely related to coronary artery disease. However, after elimination of coronary artery disease, the remaining possible diagnoses were arrhythmogenic right ventricular cardiomyopathy or a healed myocarditis, which had to have been considered before in order for this case to be classified as "idiopathic" VT.

A positive diagnosis of ARVD was suggested by the left bundle branch block electrocardiographic pattern, indicating that VT originated in the right ventricle. However, it is known that VT originating in the left ventricular septum may also exhibit a LBBB pattern [2]. The absence of disease progression during a 40 year period has been observed in my experience but this is not a common feature of ARVD [3].

A surprising positive diagnosis of ARVD was provided by magnetic resonance imaging but was not confirmed by repeated echocardiographic examination. However, MRI did not include gadolinium testing, which would have shown the presence of fibrosis in healed myocarditis or a possible different pattern in the event of ARVD [4].

Myocarditis is a disease with a polymorphic pattern ranging from a transient acute episode with complete healing to the fulminant form leading to hyper-acute heart failure and death in a few days or weeks. The histologic sequel of myocarditis is clusters of adipocytes and fibrosis [4]. In addition, it was recently confirmed that signs of inflammation and possible myocarditis occur more frequently at the time of ventricular arrhythmias in two right ventricular cardiomyopathies, e.g., arrhythmogenic right ventricular dysplasia and the Brugada syndrome [5,6].

In attempting to identify the disease mechanism, genotyping should have been the first step [7]. If positive for a mutation already known in ARVD in which PKP2 is the most frequent, then the title of the authors' article should be arrhythmogenic right ventricular dysplasia and not arrhythmogenic right ventricular cardiomyopathy, reserving the term ARVC for the wide spectrum of these diseases. Some of them are already known under a different term or will be discovered with the progress of genetics and molecular biology [8,9].

Apart from this discussion on the physiopathogenesis and subsequent terminology of the disease, it is important to emphasize the wise clinical approach of Dr. Belhassen who proposed the two treatment approaches: namely, implantation of a cardioverter defibrillator or anti-arrhythmic drug therapy provided that an electrophysiologic study is performed to ascertain its effectiveness. The latter approach, which was indicated as his preference, was also acknowledged by the patient and eventually proved its effectiveness. The other therapeutic approaches could have been VT ablation using three-dimensional electroanatomic mapping systems [10]. The newest approach could also rely on the interesting dechanneling technique, which requires further investigation [11].

The direction in the future will be to develop new techniques to block progression of the disease, such as the induced Programmed Stem Cells (iPSC) technique, which is able to reproduce the disease-in-the-dish from fibrocytes containing the genetic specific genetic material of each patient [9].

VT = ventricular tachycardia
ARVD =arrhythmogenic right ventricular dysplasia
LBBB = left bundle branch block

ARVC = arrhythmogenic right ventricular cardiomyopathy

Correspondence:
Dr. G. Fontaine
email: guy.fontaine2@numericable.fr

References

1. Belhassen B, Aviram G, Viskin S. Arrhythmogenic right ventricular cardiomyopathy: an unusual possible cause of arrhythmia in a 78 year old man with a 40 year history of palpitations. *IMAJ* 2014; 16: 385-7.
2. Josephson ME, Horowitz LN, Farshidi A, Spear JF, Kastor JA, Moore EN. Recurrent sustained ventricular tachycardia. 2. Endocardial mapping. *Circulation* 1978; 57: 440-7.
3. Hulot JS, Jouven X, Empana JP, Frank R, Fontaine G. Natural history and risk stratification of arrhythmogenic right ventricular dysplasia/cardiomyopathy. *Circulation* 2004; 110: 1879-84. Epub 2004 Sep 27.
4. Cihakova D, ed. Myocarditis. Rijeka, Croatia: Intech Publishers, 2011.
5. Bonny A, Lellouche N, Ditah I, et al. C-reactive protein in arrhythmogenic right ventricular dysplasia/cardiomyopathy and relationship with ventricular tachycardia. *Cardiol Res Pract* 2010; doi: 10.4061/2010/919783
6. Bonny A, Tonet J, Márquez MF, et al. C-reactive protein levels in the Brugada syndrome. *Cardiol Res Pract* 2011; doi: 10.4061/2011/341521.
7. Fressart V, Duthoit G, Donal E, et al. Desmosomal gene analysis in arrhythmogenic right ventricular dysplasia/cardiomyopathy: spectrum of mutations and clinical impact in practice. *Europace* 2010; 12: 861-8.

8. Fontaine GH. The multiple facets of right ventricular cardiomyopathies. *Eur Heart J* 2011; 32: 1049-51.
9. Kim C, Wong J, Wen J, et al. Studying arrhythmogenic right ventricular dysplasia with patient-specific iPSCs. *Nature* 2013; 494: 105-10.
10. Arbelo E, Josephson ME. Ablation of ventricular arrhythmias in arrhythmogenic right ventricular dysplasia. *J Cardiovasc Electrophysiol* 2010; 21: 473-86.
11. Berruezo A, Fernández-Armenta J, Mont L, et al. Combined endocardial and epicardial catheter ablation in arrhythmogenic right ventricular dysplasia incorporating scar dechanneling technique. *Circ Arrhythm Electrophysiol* 2012; 5: 111-21.