Atrial Arrhythmias in Arrhythmogenic Cardiomyopathy: At the Beginning or at the End of the Disease Story?

To the Editor:
Atrial arrhythmias in arrhythmogenic cardiomyopathy occur in 20–40% of cases. Recent analysis shows that atrial arrhythmias are associated with chronic disease, dilatation of the atria, left ventricular involvement, heart failure, death from cardiac causes, inappropriate intrathoracic defibrillator shocks and heart transplantation.1

Two cases of arrhythmogenic cardiomyopathy in which atrial arrhythmias were the first findings attracting interest are presented.

The first patient was a 47-year-old woman who suffered for several years from intermittent atrial fibrillation, was treated with cardioversion and was stable in sinus rhythm when ventricular premature beats and nonsustained ventricular tachycardias occurred. The ECG of the patient revealed sinus rhythm and localized right precordial QRS prolongation (QRS in [(V1+V2+V3)/(V4+V5+V6)] ≥1.2) as the only hint of arrhythmogenic cardiomyopathy. At echocardiography the right ventricle showed typical signs of arrhythmogenic cardiomyopathy, with dilatation of the right ventricular inflow and outflow tracts and sacculation of the apical region. Right ventricular angiography revealed a severe form of arrhythmogenic cardiomyopathy with trabecular disarray of the apical and inferior regions and trabecular hypertrophy of the right ventricular outflow tract (Figure).

In the second case, 55-year-old woman had atrial fibrillation documented as the first manifestation of arrhythmogenic cardiomyopathy. When admitted to hospital, her ECG was again in sinus rhythm and localized right precordial QRS prolongation and epsilon wave in lead V2 occurred. Echocardiography and right ventricular angiography presented typical signs of arrhythmogenic cardiomyopathy.

In summary, atrial arrhythmias are often the first arrhythmic manifestation in the development of arrhythmogenic cardiomyopathy.

As shown in the first case, ventricular arrhythmias appear many years later, when implantation of an ICD becomes necessary.

In the second case, atrial fibrillation appeared as the first arrhythmic event; ECG monitoring when in sinus rhythm under β-blockade could exclude neither supraventricular arrhythmias nor significant ventricular arrhythmias.

Atrial arrhythmias can appear as the first event in the clinical development of arrhythmogenic cardiomyopathy before ventricular arrhythmias appear. In the development of atrial arrhythmias typical risk factors are arterial hypertension and valvular diseases, but arrhythmogenic cardiomyopathy and Brugada syndrome are risk factors as well.2

On the other hand, atrial arrhythmias are typical arrhythmias in chronic arrhythmogenic cardiomyopathy, heart failure, left ventricular involvement and the need for heart transplantation as a therapeutic option.1

In arrhythmogenic cardiomyopathy, atrial arrhythmias do appear either at the beginning or at the end of the disease course and have an incidence of 20–40%.

References

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