Sudden death due to arrhythmogenic right ventricular cardiomyopathy associated with hemodynamically significant myocardial bridging

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Abstract: Arrhythmogenic right ventricular cardiomyopathy is a genetically transmitted cardiac disorder, characterized by increased myocardial instability and an increased risk for severe ventricular arrhythmias and sudden death. Histologically there are two main types: fatty, associated with sudden death at rest, and fibrofatty, with sudden death during exercise. The purpose of this case report is to present a sudden death caused by an association of a fibrofatty type of arrhythmogenic right ventricular cardiomyopathy in a young athlete, associated with a hemodynamically significant myocardial bridging on the left anterior descending artery, the latter causing an atypical distribution pattern of the fibrofatty infiltration, that may pose diagnostic difficulties.

Key Words: Arrhythmogenic right ventricular cardiomyopathy, myocardial bridging, sudden death, sudden cardiac death in athletes.

Arrhythmogenic right ventricular cardiomyopathy (ARVC) is a genetically transmitted cardiac disorder, characterized by increased myocardial instability and an increased risk for severe ventricular arrhythmias and sudden death [1-6]. The main cause of the disease appears to be a desmosome defect causing ultrastructure changes including remodeling of the intercalated discs, increased intercellular gap, increased desmosome length, and paler desmosomes. These defective desmosomes can lead to myocyte uncoupling, especially during increased myocardial stress [7]. This in turn can cause unspecific changes that include inflammation, fibrosis, apoptosis, and adipocytosis [7] that can lead to an earlier onset of the clinical disease, and an increased risk of sudden death in patients with severe, sustained, myocardial stress like professional athletes [8].

The purpose of this article is to present the case of a professional athlete whose death was sudden, and caused by an undiagnosed ARVC.

CASE REPORT

A 25 year-old man collapsed on the football field and was pronounced dead by the ambulance about 30 minutes later. He has a professional football player until a year ago, when he interrupted this activity as he was diagnosed with hepatitis. Six months ago he had another blackout on the football field. As he was feeling often tired and had thoracic pains, he went to a cardiology hospital, seven and one month before death. Both examination revealed bradycardia, a QRS angle of 90 degrees, EVs, diffuse hypovoltage, and repolarization abnormalities in the anterior territory (V1-V4), and flat T waves in all derivations. The ECG suggested a high probability
of cardiomiopathy, either primary or ischemic, with a possible pericardial involvement, and the physicians recommended cardiac echography, myocardial scintigraphy, cardiac MRI, and coronarography. The patient refused these additional examinations. During the forensic autopsy was identified a left ventricle with an intense pale area, subendocardial, at the level of the interventricular septum. The thickness of the interventricular septum was 1.6-1.8 cm. The myocardial tissue contained numerous yellowish stripes, with areas of confluence and dissection (Figure 1). The right ventricle had a thickness of 0.1-0.3 cm, with an irregular pattern of the wall, with significant replacement of the myocardial tissue with adipose formations (Figure 1). The left anterior descendent coronary artery had a significant myocardial bridging, starting at about 2 cm from the origin and extending down about 7 cm, with a thickness varying from 0.2 to 0.6 cm (Figure 2). The circumflex artery finishes on the pulmonary face of the left ventricle. The right coronary artery, having a normal diameter, without atherosclerosis, ends on the diaphragmatic surface, in the superior third of the posterior interventricular groove. Cardiac histological examination showed interstitial vascular swellings, with hypoxic myocardial lesions, significant subepicardial lipomatosis, with extension in the adjacent myocardium (Figure 3). Trichrome Masson stain showed mild-moderate interstitial and perivascular lipomatosis (Figure 4).

**DISCUSSIONS**

Even if the sudden deaths of athletes is considered a rare phenomenon, with an estimated prevalence of between less than 1/100.000 cases [9] and about 3.5/100.000 cases [10], recent reports suggests that the cardiovascular risks associated with professional sports are significantly underestimated [11]. The presence of structural heart diseases, like hypertrophic...

**Figure 1.** Lipomatous infiltration of the myocardium.

**Figure 2.** Hemodynamically significant myocardial bridging on the Left Anterior Descending Artery.

**Figure 3.** Fatty infiltration of the right ventricle, with ischemic areas. HE, 50X.

**Figure 4.** Fibrofatty infiltration. Masson Trichrome, 50X.
cardiomyopathy or ARVC, even if only probable, should forbid the practice of performance sports [12]. As ARVC is one of the most frequent causes of sudden death in professional athletes [12], every athlete should be routinely checked for it, especially as most screening tests are readily available. The Recommendations of the European Society of Cardiology for screening professional athletes for ARVC include the following:

- **12-Lead ECG.** The presence of cardiac arrhythmias in a young athlete should always be further checked. In ARVC ECG abnormalities are present in more than 50% of patients, the most common being prolonged QRS (>110ms) with a pattern of right bundle branch block and inverted T-waves in right precordial leads, ε wave, premature ventricular complexes or ventricular tachycardia, usually with a left bundle branch block pattern and vertical axis [12].

- **Echocardiography:** enlarged right ventricle cavity, wall motion abnormalities, and segmental morphological abnormalities, including thinning, bulging, or aneurysms of the right ventricle wall [12].

  In our case the ECG suggested the probability of a severe cardiovascular disorder (most likely a cardiomyopathy), that would normally forbid that person to go on the football court. However, the patient has not followed the medical recommendation, nor in restricting strenuous physical activities, nor in further researching the cause of the electrical abnormalities.

**References**


