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Unexpected etiology of syncope

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Abstract. Brugada syndrome is an autosomal dominant hereditary disease causing ventricular arrhythmias and sudden cardiac death, especially in young and otherwise healthy people. The incidence of this disease in central Europe is quite low, nevertheless we would like to introduce you a case report of a patient treated in our hospital for this disease. Fig. 3, Ref. 6, on-line full text (Free, PDF) www.cardiologyletters.sk

Key words: arrhythmia – syncope – sudden cardiac death

Brugada syndrome was first described by the Brugada brothers, who were researching syncopes and sudden cardiac death in young men (1). Brugada syndrome is an autosomal dominant hereditary disease with estimated incidence of about 5 per 10000 persons and the cause of 4-12% of sudden deaths (2). Typical electrocardiographic (ECG) findings are ST segment elevations with T wave inversion in right precordial leads, right bundle branch blockade and high incidence of sudden cardiac death in individuals with structurally normal heart (1, 3). The syndrome typically manifests during adulthood with a mean age of sudden death of 41±15 years (4). Mutation in the SCN5A gene encoding the α subunit of the sodium channel was described at molecular level and was identified in approximately 20% cases (2, 5). So far, the only established therapy remains implantable cardiac defibrillator (ICD) implantation (6).

Case report

A forty-six-year old, so far healthy, man was admitted to the coronary unit of our clinic because of syncope with a 2 minute loss of consciousness followed by dyspnea and vertigo. Sinus tachycardia with incomplete right bundle branch block, ST segment elevations in V1 to V3 precordial leads and negative T waves were recorded on ECG in the ambulance. Upon admission, the patient had physiological findings both on echocardiography and in laboratory results. After excluding the suspicion of pulmonary embolism or intracranial pathology, an ajmaline test was performed. Ajmaline infusion led to ST junction elevations in right precordial leads (type 1 Brugada syndrome characteristic pattern) and during higher ajmaline doses, frequent isolated ventricular extrasystoles occurred, some of them with very short coupling interval (R-on-T phenomenon) **(Figure 1)**.

Cardiac magnetic resonance examination (CMR) was performed to exclude possible myocardial scar **(Figure 2)**, extracellular edema **(Figure 3)** or lipomatous infiltration of the myocardium. After all these examinations, ICD implantation was recommended because of the high probability of Brugada syndrome diagnosis. No additional arrhythmia was recorded during the rest of the patient´s hospitalization and the patient was released with the suggestion for the cardiological examination of his relatives. His family, however, lives in another European country with less available public health care, so they were probably not examined.

Discussion

The most frequent cause of malignant syncopes in people over 50 years old is coronary artery disease. After the admission to the hospital, urgent coronary angiography was not performed, because of the normal findings on echocar-

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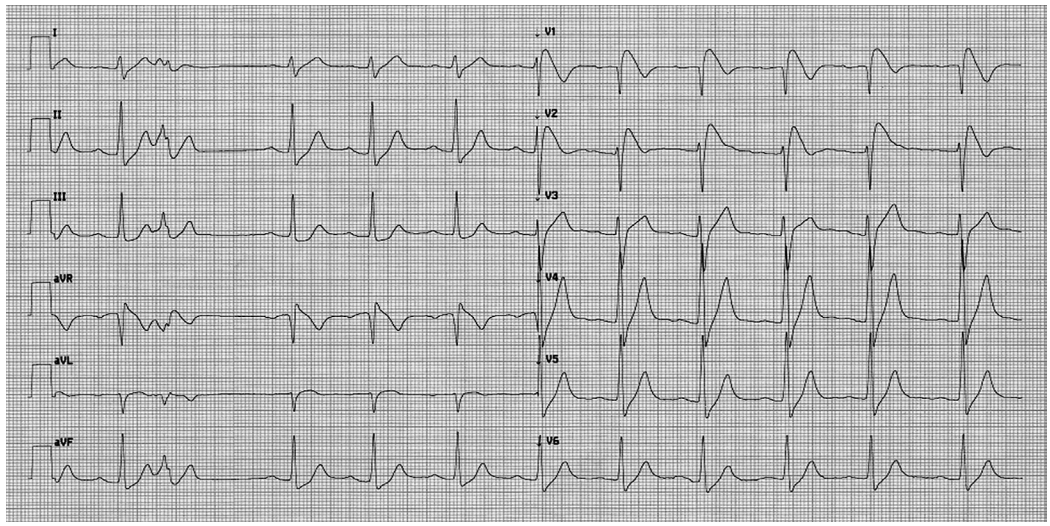

Figure 1 Electrocardiogram during ajmaline test: ST junction elevations in V1-V3 precordial leads (type 1 Brugada syndrome) and ventricular extrasystole with short coupling interval

diography and in laboratory results. Because of the dyspnea and right bundle branch block on ECG, pulmonary embolism was considered and soon excluded by computed tomography (CT) angiography. Cardiac magnetic resonance examination excluded older myocardial scarring or arrhythmogenic cardiomyopathy. ECG after loss of consciousness and ajmaline test showed a characteristic pattern, so the Brugada syndrome remains the most probable diagnosis.

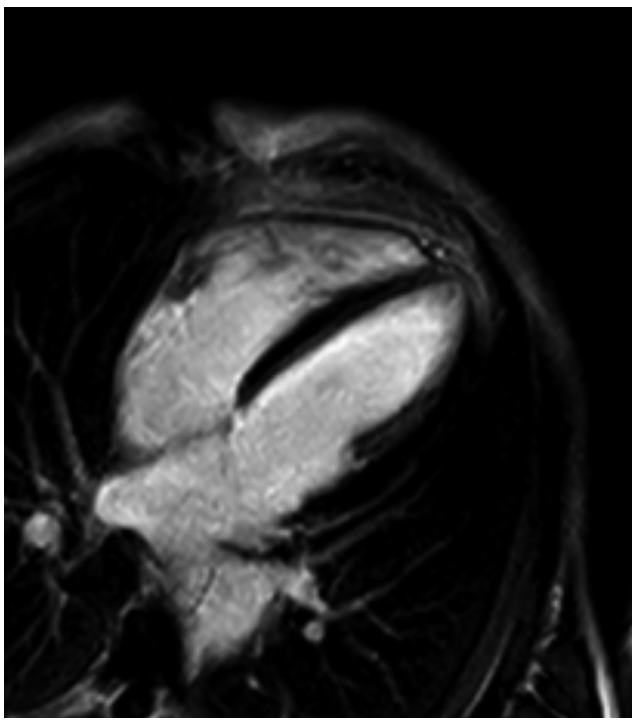

Figure 2 Cardiac magnetic resonance, 4-chamber view, delayed postcontrast inversion recovery images, no late gadolinium enhancement

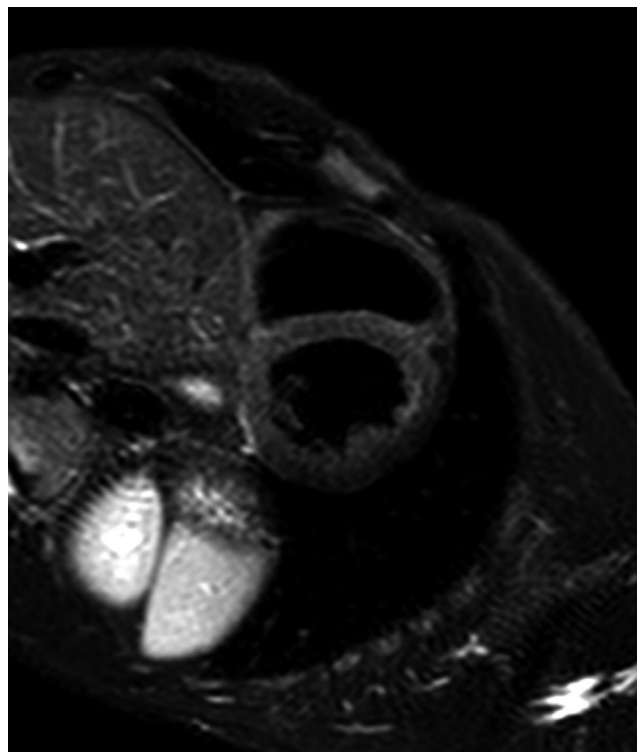

Figure 3 Cardiac magnetic resonance, T2-weighted sequences depicting absence of extracellular edema

Conclusion

Brugada syndrome diagnosis was made in a forty-six--year old patient after a 2 minute loss of consciousness with typical ECG pattern after ambulance arrival and during the ajmaline test. After excluding other possible causes of malignant arrhythmias (e.g. hypertrophic or arrhythmogenic cardiomyopathy, long- or short-QT syndrome) by a set of examinations (repeated ECG, echocardiography, CMR), an ICD implantation in secondary prevention of sudden cardiac death was performed. During 3 years of follow-up, no malignant arrhythmia was detected.

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