CASE REPORT

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Sudden sustained monomorphic ventricular tachycardia in a previously healthy adult with many causes for it, but which is the correct one?

Iznenadna dugotrajna monomorfna ventrikularna tahikardija kod prethodno zdrave odrasle osobe sa puno uzroka za to, ali koji je pravi?

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Abstract

Introduction. Sustained monomorphic ventricular tachycardia (VT) – SMVT is a rare, underdiagnosed pathology with a very poor prognosis. Along with ventricular fibrillation, SMVT is responsible for nearly all of the arrhythmic sudden cardiac deaths (SCD). The most common cause of VT is ischemic heart disease, but there are many other reasons, among which are arrhythmogenic right ventricular cardiomyopathy (ARVD) and myocardial bridging phenomenon. Treatment options include a hybrid approach consisting of antiarrhythmic drugs, catheter ablation, and implantable cardioverter defibrillators (ICD). **Case report.** We present a case of a 46-year-old man, a military officer, who experienced chest pain, palpitations, and nausea during regular physical activity at home. Due to the symptoms described, he was examined immediately and diagnosed with SMVT. Shortly after the diagnosis, he lost consciousness and was successfully resuscitated. A complete non-invasive and invasive cardiology examination was performed. It revealed that the patient had stable coronary disease and a muscle bridge on the left anterior descending artery. An ARVD was suspected after transthoracic echocardiography and heart magnetic resonance imaging. Genetic testing for ARVD was negative, but according to the Heart Rhythm Society expert consensus criteria, we had enough for a definitive diagnosis. The patient was hospitalized for ten days and treated with drugs without recurring VT or other disorders. We implanted an implantable loop recorder before the discharge and monitored the heart rhythm for one year. During a three-year follow-up, all of his electrocardiographic findings presented sinus rhythm without heart rhythm disorders. **Conclusion.** Sudden SMVT is the most common cause of SCD. It is of inestimable importance to carry out a detailed examination and determine the immediate cause of the arrhythmia and the right therapy, which, for these patients, is a life-saving form of treatment. Therapy includes medications, electrophysiology or ICD, or a combination of these treatment approaches.

Key words:
cardiomyopathies; coronary disease; defibrillators, implantable; diagnosis; magnetic resonance imaging; myocardial bridging; tachycardia, ventricular; ultrasonography.

Apstrakt

Uvod. Dugotrajna monomorfna ventrikulska tahikardija (VT) – DMVT je retka, nedovoljno dijagnostikovana patologija sa veoma lošom prognozom. Zajedno sa ventrikulskom fibrilacijom, DMVT je odgovorna za skoro sve iznenadne srčane smrti (ISS) nastale usled aritmije. Najčešći uzrok VT je ishemijska bolest srca, ali postoje i mnogi drugi razlozi, među kojima su aritmogena kardiomiopatija desne komore (AKDK) i fenomen miokardnog „mosta“ (*bridge*). Mogućnosti lečenja uključuju hibridni pristup, koji se sastoji od antiaritmijskih lekova, kateterske ablacije i implantabilnog kardioverter defibrilatora (IKD). **Prikaz bolesnika.** Prikazujemo slučaj 46-godišnjeg muškarca, oficira u vojsci, koji je tokom redovnih fizičkih aktivnosti kod kuće osetio bol u grudima, lupanje srca i mučninu. Zbog navedenih tegoba odmah je pregledan i dijagnostikovana mu je DMVT. Ubrzo nakon postavljanja dijagnoze bolesnik je izgubio svest i uspešno je reanimiran. Sprovedena je kompletna neinvazivna i

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invazivna kardiološka dijagnostika. Utvrđeno je da je bolesnik imao stabilnu koronarnu bolest i mišićni „most“ na prednjoj descedentnoj arteriji. Nakon transtorakalne ehokardiografije i magnetne rezonance srca, posumnjano je na AKDK. Genetsko testiranje na AKDK bilo je negativno, ali prema kriterijumima konsenzusa stručnjaka *Heart Rhythm Society*, imali smo dovoljno dokaza za definitivnu dijagnozu. Bolesnik je hospitalizovan tokom deset dana i lečen lekovima, bez pojave VT i drugih poremećaja. Pre otpusta iz bolnice, ugradili smo mu implantibilni *loop* rikorder i pratili srčani ritam tokom godinu dana. Tokom trogodišnjeg praćenja svi elektrokardiografski nalazi bili su normalni. **Zaključak.** Iznenadna DMVT je najčešći uzrok ISS. Detaljan pregled je od neprocenjive važnosti, kao i utvrđivanje neposrednog uzroka aritmije i primena odgovarajuće terapije, koja za te bolesnike predstavlja vid lečenja koji spasava život. Terapija uključuje lekove, elektrofiziologiju ili IKD ili kombinaciju ovih pristupa u lečenju.

Ključne reči:
miokard, bolesti; koronarna bolest; defibrilatori, implantabilni; dijagnoza; magnetska rezonanca, snimanje; miokardni mostovi; tahikardija, ventrikulska; ultrasonografija.

Introduction

Sustained monomorphic ventricular tachycardia (VT) – SMVT is a rare, underdiagnosed pathology with a very poor prognosis [1]. Along with ventricular fibrillation, SMVT is responsible for nearly all of the arrhythmic sudden cardiac deaths (SCD) [1, 2]. SMVT demonstrates a stable QRS morphology from beat to beat when the rhythm lasts longer than 30 s or hemodynamic instability occurs in less than 30 s [3]. SMVT may be idiopathic but occurs most frequently in patients with underlying heart disease of various types, including ischemic heart disease (IHD), dilated cardiomyopathy, hypertrophic cardiomyopathy, arrhythmogenic right ventricle (RV) cardiomyopathy (ARVC), myocarditis and complex congenital heart disease [4]. Other causes include myocardial bridge (MB) [5, 6], prolonged QT syndrome, infiltrative cardiomyopathy, Chagas heart disease, cardiac sarcoidosis, and left ventricular noncompaction [4, 7].

The most common cause of VT is IHD [1–3]. Another cause of VT is ARVC, a rare inherited disorder and underrecognized clinical entity manifested by ventricular arrhythmias and sudden death, especially in young athletes [8, 9]. MB or myocardial bridging is defined as the muscle overlapping the intramyocardial part of the epicardial coronary artery and is one more cause of VT [10]. The artery, which is enclosed by the myocardium, is called the “tunneled artery” [11]. In 70–98%, the tunneled artery is the left anterior descending artery (LAD) [10–11]. Autopsy studies show the frequency of MB ranging from 5% to 86%, with the largest one presenting a 26% rate [12]. Coronary angiography (CA) rates are much lower (0.5–12%) [11].

Diagnostic of all these diseases, which can lead to VT, includes medical and family history, clinical examination, laboratory analysis, 12-lead electrocardiography (ECG), and transthoracic echocardiography (ECHO) [13]. Computed tomography coronarography or invasive CA [14] and often endomyocardial biopsy, electrophysiologic testing, and cardiac magnetic resonance imaging (MRI) [15] are also needed.

Treatment of the known and variable cause of the arrhythmia can lead to its permanent cessation. If we have an unchangeable cause of the arrhythmia, like a genetic disorder or similar, treatment options include antiarrhythmic drugs, implantation of implantable cardioverter defibrillators (ICD), and catheter ablation. A hybrid approach consisting of antiarrhythmic drugs, catheter ablation, and ICD may provide an effective therapeutic approach in some situations [4, 16].

Case report

We present a case of a 46-year-old man, a military officer, who experienced chest pain, palpitations, and nausea during physical activity at home, mowing the grass. He immediately reported to the doctor at the nearest military facility. His heart rate was 245 beats/min, and his blood pressure was under 80/50 mmHg. ECG (Figure 1) demonstrated wide-QRS-complex VT consistent with sustained VT of the left bundle branch block with the superior axis. The patient was immediately transferred to the

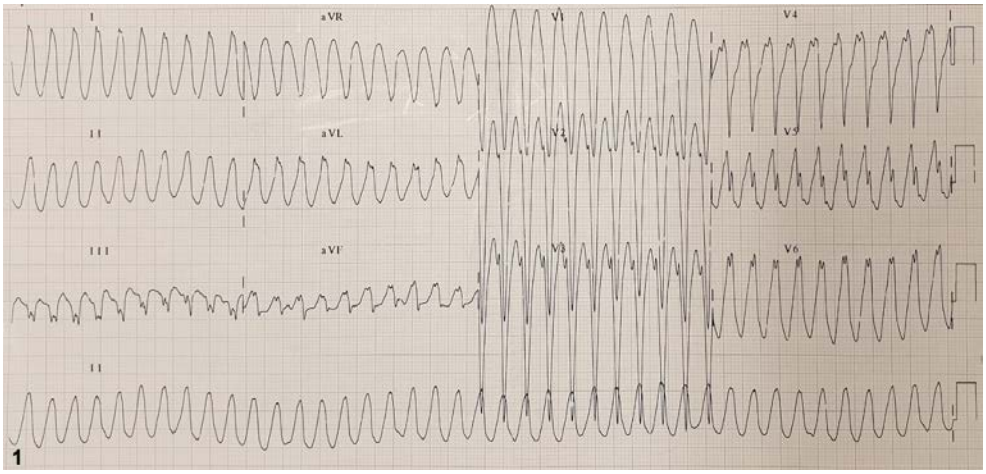

Fig. 1 – Electrocardiography during sustained ventricular tachycardia on admission.

Military Medical Academy (MMA), Belgrade, Serbia, and he was resuscitated during the transfer. Upon admission to the Emergency Unit of the MMA, he experienced a second VT occurring in a one-hour time frame, which required prompt reanimation and direct current cardioversion, after which his heart rate was 87 beats/min and blood pressure was 110/70 mmHg. The physical examination did not reveal anything extraordinary. Personal anamnesis unveiled hypertension, hyperlipidemia, glucose intolerance, active smoking for 25 years, moderate alcohol consumption, and playing recreational sports. Family anamnesis was negative for cardiovascular diseases.

Laboratory analysis revealed following increased findings on admission: leukocyte count 13.86 x 10^9/L [reference range (RR) 4–11 x 10^9/L)], glucose level 13.4 mmol/L (RR 4.1–5.9 mmol/L), creatinine 147 umol/L (RR 62–115 umol/L), creatine kinase 441 U/L (RR 32–300 U/L), and aspartate aminotransferase 51 U/L (RR 0–37 mmol/L). Electrolytes were within normal limits, and there was no suspicion of drug use. CA demonstrated non-significant stenosis of LAD in the bifurcation area with second diagonal branch and septal truncus (up to 50%) (Figure 2A), circumflex artery (ACX) in the ostial area (30–50%), and normal CA of right coronary artery (RCA) (Figure 2B). Intravascular ultrasound (US) displayed large calcified stenosis from proximal to medial part of LAD, 50–70%, with minimal luminal area (MLA) of 3.84 mm^2 (MLA < 4 mm^2 in LAD, ACX, and RCA vessels > 3 mm in diameter correlates with physiological significance) (Figure 3A), while stenosis in the ACX was 50–60% with MLA of 6.18 mm^2 (Figure 3B). Significant

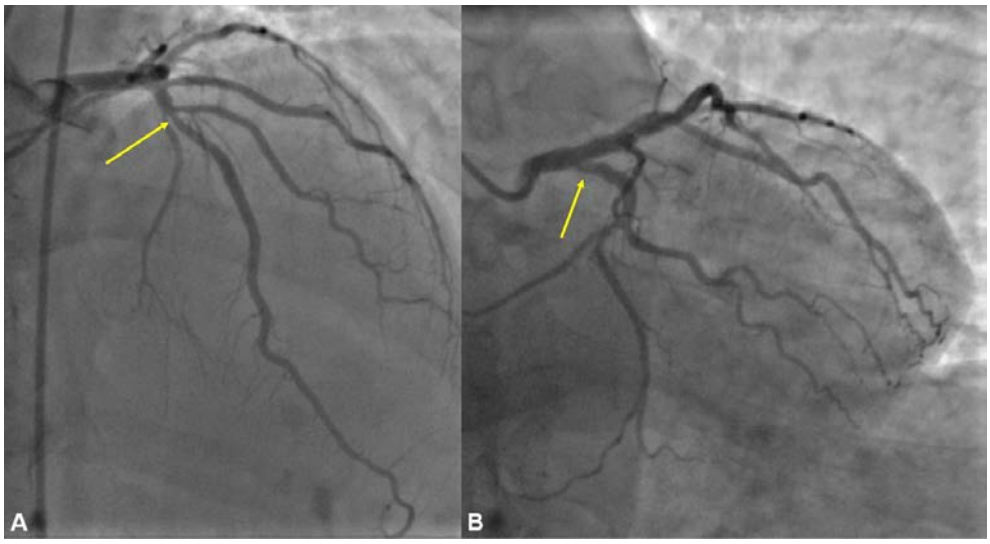

Fig. 2 – Coronary angiography of the left coronary artery reveals A) stenosis of 50–70% in the proximal part of the left anterior descending artery (yellow arrow) and B) stenosis of 30–50% in the ostial part of the circumflex artery (yellow arrow).

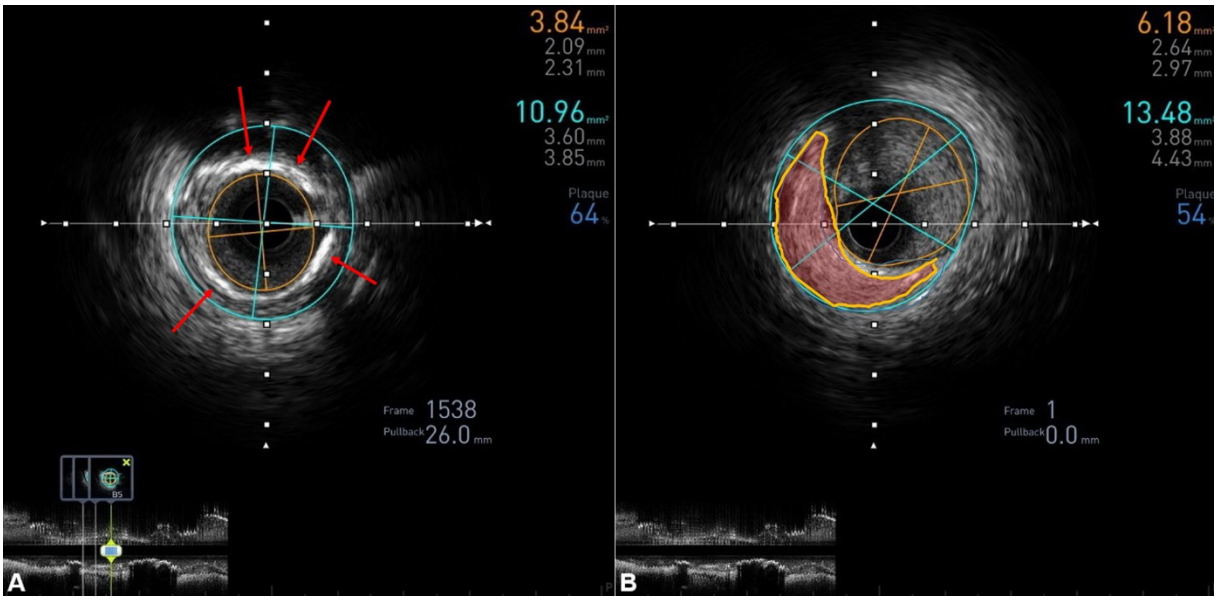

Fig. 3 – Intravascular ultrasound of left anterior descending (LAD) artery shows A) circular stenosis of 64% of mid LAD (minimal luminal area – MLA = 3.84 mm^2) with calcification (red arrows) and B) eccentric plaque of ostial part of circumflex artery with stenosis of 54% (MLA = 6.18 mm^2) (red field).

coronary compression in the mid to distal segment of the LAD was noted all along the systolic phase, attributing MB phenomenon (Figure 4A and 4B). Initially, ECHO showed unremarkable findings. Follow-up US showed good coronary flow reserve, resulting in 3.1 (reference values ≥ 2) in the LAD. Fractional flow reserve was 0.89 (reference values > 0.85) in LAD and 0.92 in ACX. The patient was hospitalized for ten days and treated with medicament therapy without recurring VT or other disorders. His therapy included the following medicaments: amiodarone 200 mg/day, bisoprolol 2.5 mg/day, valsartan 80 mg/day, aspirin 100 mg/day, rosuvastatin 40 mg/day, and trimetazidine 35 mg twice a day. Before the patient was discharged from the hospital, we implanted an implantable loop recorder and monitored the heart rhythm for one year. We performed a treadmill exercise test three months after discharge, which was negative for IHD. We did not discover any heart disturbances during the whole time of monitoring. Three years later, on ECHO, we found an enlarged RV with the greatest diameter of 4.56 cm (Figure 5A) and reduced strain of the free wall of the RV

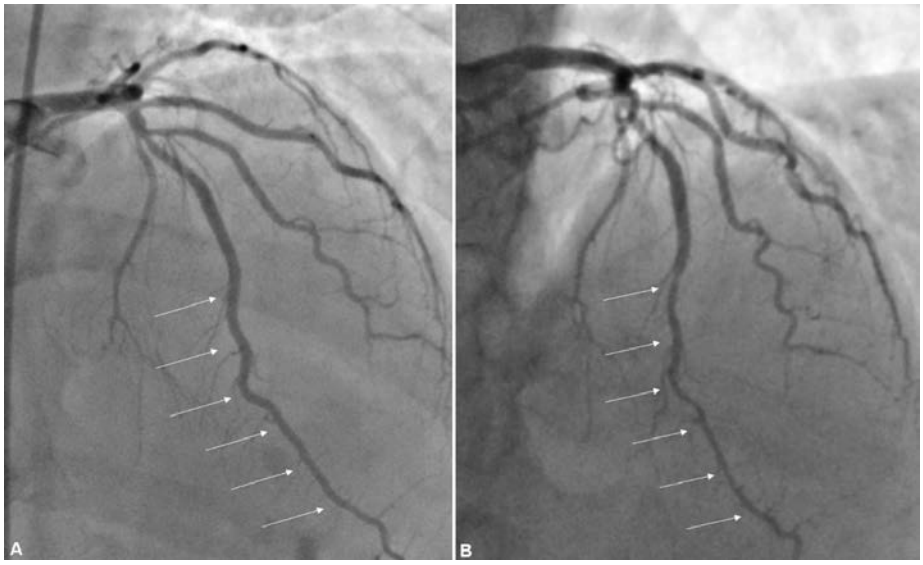

Fig. 4 – Myocardial bridge: A) left anterior descending (LAD) artery during diastole of the heart with marked part of the artery where the bridge will occur (white arrows); B) part of the LAD artery where the bridge occurred (white arrows).

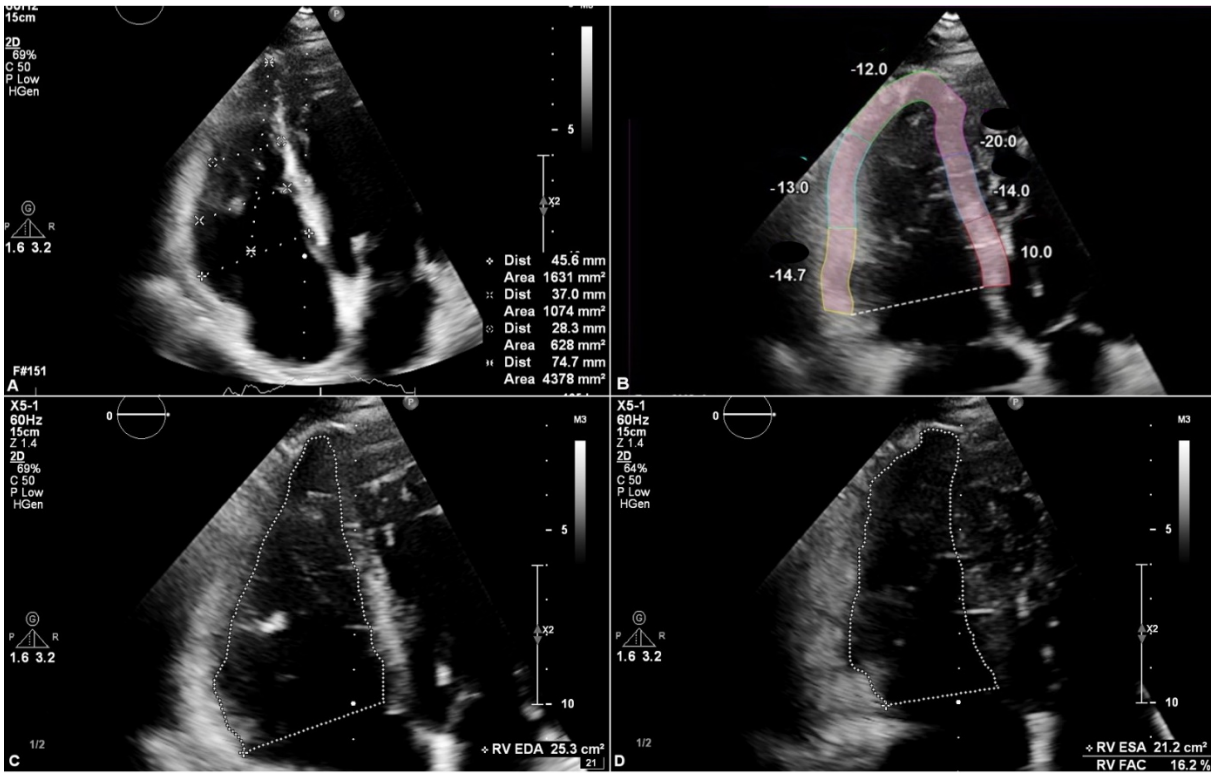

Fig. 5 – Transthoracic echocardiography: A) dimensions of the right ventricle (RV); B) the strain of the free wall of the RV (-13.3); C) right ventricular end-diastolic area; D) right ventricular end-systolic area and fractional area change (16.2%).

(-13.3) (Figure 5B). In addition, the parameter parasternal short axis – RV outflow tract was greater than 36 mm (result was 43 mm), and fractional area change was less than 33% (result was 16.2%) (Figure 5C and 5D). In the end, we decided to do an MRI of the heart. Results reveal suspicion of ARVC. Two smaller aneurysmal enlargements of RV were registered. The first was at the transition from the basal to the medial segment of the free wall forward 7.9 mm (Figure 6A), and the second was at the very apex of RV up to 6 mm in size (Figure 6B), which suggested electrophysiological testing as well as genetic screening. In order to continue with further diagnostic, according to current recommendations, we also performed genetic testing at the Institute for Molecular Genetics and Genetic Engineering, Belgrade, and the testing was negative. The patient has been followed up for three years, and all his ECG findings presented sinus rhythm without heart rhythm disorders (Figure 7). The patient's consent was obtained for this case report.

Discussion

Our case is complex, with the possibility of multiple causes for SMVT, which makes it difficult to determine the final cause of this malignant rhythm disorder.

During the examination, we diagnosed arterial hypertension, IHD, and an MB on the LAD, which were not of such a degree of severity that we could safely associate the occurrence of malignant rhythm disorders that the patient had. At this time, due to the absence of unchangeable causes for the development of malignant rhythm disorders, we made a conciliar decision to temporarily postpone the need for ICD implantation. It was also decided that severe MB is to be treated with drugs only and not surgery. During the long follow-up time, ECG as well as the exercise test were normal, but repeated ECHO revealed important changes. MRI of the heart showed suspicion for ARVD, and genetic testing on gnomAD Genomes, gnomAD exomes, and 1000 Genomes was negative, which does not rule out the existence of the disease [17, 18].

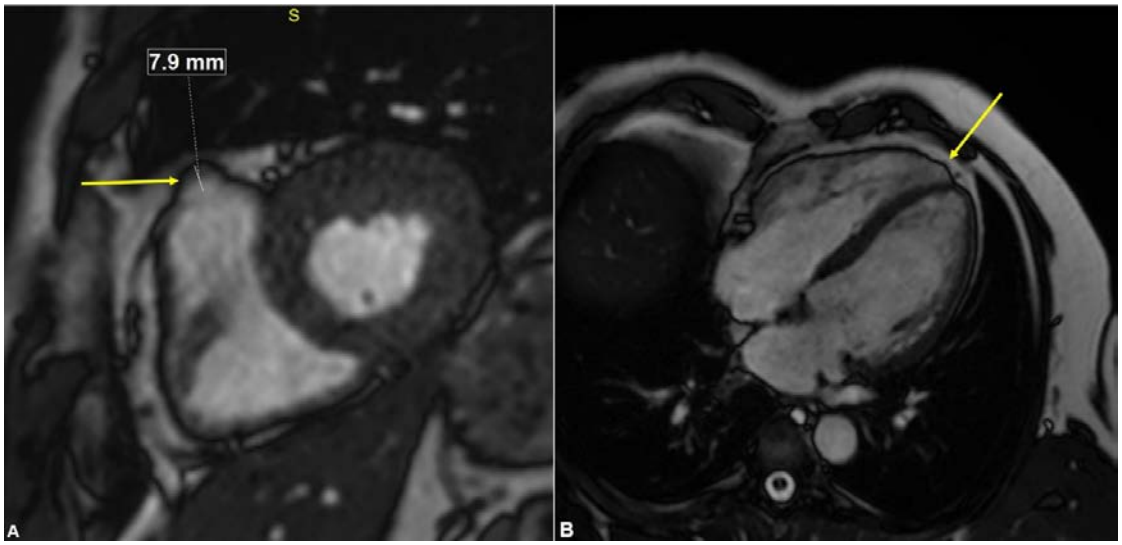

Fig. 6 – Heart magnetic resonance imaging shows A) aneurismatic dilatation of the right ventricle in the basal part with 7.9 mm (yellow arrow) and B) apex of the right ventricle (yellow arrow).

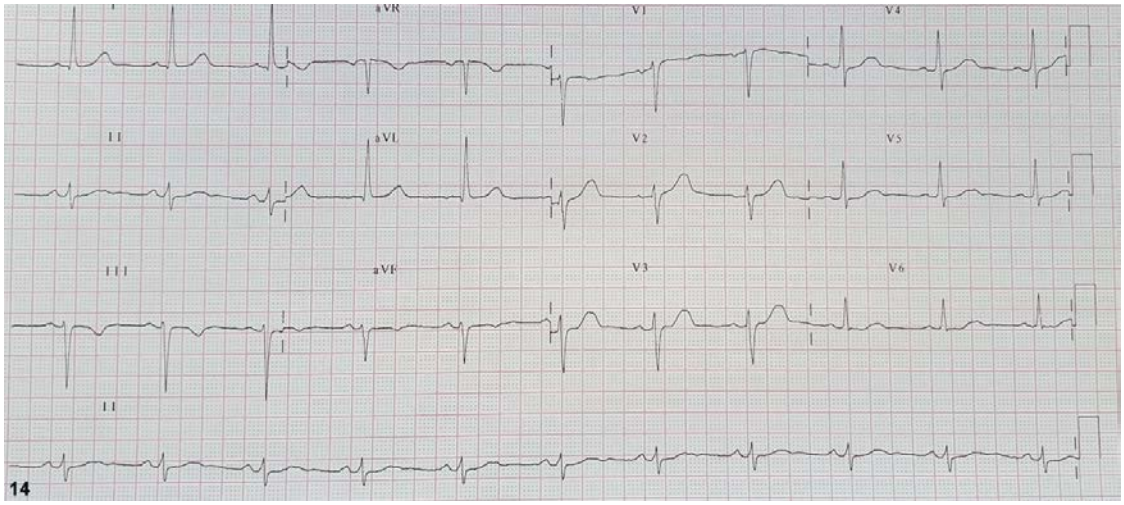

Fig. 7 – Electrocardiography three years after ventricular tachycardia shows sinus rhythm without heart rhythm disorders.

Including all these results, we had two major criteria (ECG and ECHO) for diagnosis of ARVD according to the recommendations of the Heart Rhythm Society expert consensus statement from 2019 [19]. Taking into account all the facts and based on current recommendations for the treatment of arrhythmias associated with SCD [1, 19], we decided to plan the patient's ICD implantation without previous electrophysiology testing.

Conclusion

Sudden SMVT is the most common cause of SCD. It is very difficult to diagnose, and it is often the first and last manifestation of the disease. If the disease is detected and the unwanted event is prevented, further determination of the true cause can be a problem, especially if there are several different reasons for the occurrence of the said arrhythmia. Due to the modality of treatment, which depends on the etiology of the arrhythmia, it is of inestimable importance to carry out a detailed examination and determine the immediate cause of the arrhythmia and thus determine the right therapy, which for these patients is a life-saving form of treatment. Further research in these areas will provide invaluable data for early detection and prevention of SCD.

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