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Case report

Electrical storm in a patient with implantible cardioverter defibrillator

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Abstract Electrical storm is the occurrence of at least three episodes of malignant ventricular arrhythmia (sustained ventricular tachycardia/ventricular fibrillation) during 24 hours, most commonly in patients with an implanted cardioverter defibrillator. We present a 48-year-old patient with an implanted cardioverter defibillator in the secondary prevention of sudden cardiac death and the occurrence of an electrical storm in the form of polymorphic ventricular tachycardia with degeneration into ventricular fibrilation treated by the delivery of 10 intracardiac DC shocks in a several hours period. A multimodality approach to the patient was applied, which included the initial search for precipitating factors, electronic interrogation and changes in device programming, antiarrhythmic drug therapy and patient sedation. The applied measures are discussed in the context of the available literature. A multimodality approach to patients with electrical storm in specialized centers improves outcomes of such complex patients.

Kew words electrical storm, inplantable cardiac defibrilator, multimodality imaging

Introduction

An electrical storm represents consecutive attacks of malignant arrythmia (sustained ventricular tachycardia - VT or ventricular fibrillation - VF) within a short period of time. The most widely accepted is three episodes of ventricular arrhythmia within 24 hours[1], although recent analyzes indicate an increase in mortality with as much as two episodes within a three-month interval[2]. An electrical storm is a medical emergency and is presented by repeated ICD shocks or activation of antitachycardia pacing in patients with an implanted cardioverter defibrillator (ICD) or repeated syncope, cardiac arrest or symptoms and signs of low cardiac output in patients without an ICD. Acute treatment consists of patient stabilization, removal or treating of precipitating factors, ICD programming, pharmacological antiarrhythmic therapy, catheter ablation of the arrhythmogenic substrate, and modulation of the autonomic nervous system[1]. We present a patient with an electrical storm three and a half years after an ICD was implanted in the secondary prevention of sudden cardiac death. Acute care modalities and a review of the relevant literature are presented.

Case presentation

A 48-year-old, extremely obese male patient was admitted to the Department of internal medicine, Health Center Zaječar due to syncope in the sitting position accompanied by sweating. There was a rapid recovery of consciousness in a lying position. He was observed during the first 24 hours in the Coronary care Unit, without any heart rhythm disturbances noted. Then, in the Department of Cardiology, as an inpatient, 24-hour ECG monitoring was carried out, during which a cardiac arrest with VF occurred, which was successfully treated by an asynchronous DC shock of 270J. Several episodes of nonsustained ventricular tachycardia and two episodes of sustained polymorphic ventricular tachycardia were seen on the Holter ECG. VF was triggered by the R on T phenomenon. A coronary angiography was performed, considering that the patient had an acute myocardial infarction in 2002, and it showed intermediate lesions of the OM1 branch, two tight lesions followed by occlusion of the medial segment of the right coronary artery, with distal segment visualised through well-developed heterocollaterals from the left coronary system. Myocardial necrosis markers were negative, so the arrhythmia was not figured to be induced by acute coronary ischaemia and an ICD was implanted according to the secondary prevention of sudden cardiac death protocol. The patient was regularly followed as outpatient at the Pacemaker Center and there were no ICD activations (shocks and antitachycardia pacing) over the next three and a half years. With the performed echocardiography, the left ventricular ejection fraction was estimated to be 45%.

15.5.2021. after finishing work (afternoon shift as a medical technician) and a large meal, the patient felt

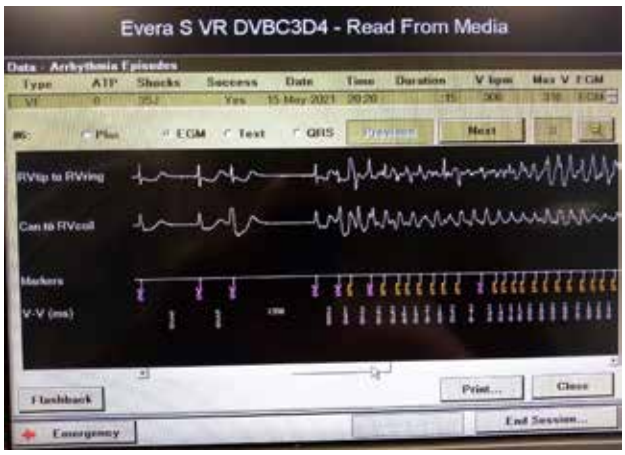

Figure 1

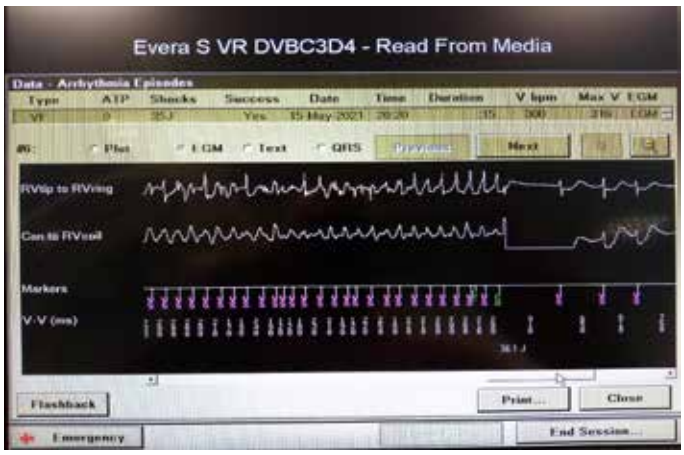

Figure 2

dizzy and experienced chest pain, which he thought was an intracardiac DC shock. The Emergency Medical Service was called, after which he was brought to the Internal Medicine Department. Interrogation of the ICD revealed 6 episodes of VF, which were caused by the degeneration of polymorphic VT initiated by the R to T phenomenon (pictures 1 and 2). In a short period, he had 4 more intracardiac DC shocks in the Coronary Care Unit. There was no hypokalemia that would promote arrhythmia - 4.7 mmol/L. Amiodarone i.v. (until then he was on a regular oral therapy with the drug) , lidocaine, metoprolol i.v. in repeated slow boluses of 5 mg and then bisoprolol 20 mg daily. The base rate of the ICD was raised to 80/min to promote ventricular pacing with sedation with midazolam i.v. Due to applied measures, the electrical storm stopped and the patient stabilized with signs of overt heart failure. Optimal medical therapy for heart faillure was instituted. Troponin was serially negative despite repeated DC shocks.

Recoronarography was performed, which showed stationary angiographic findings. Left ventricular ejection fraction was estimated at 42%. Subsequently, the patient returned to work with internal medicine outpatient visits and electronic ICD controls at the Pacemaker Center. In a two year follow up, there were no ventricular arrhythmia episodes detected. Obstructive sleep apnea was suspected as a significant comorbid condition contributing to arrhythmogenesis and referred to polysomnography, which has not yet been performed.

Discussion

It is not easy to estimate the incidence of electrical storm because it depends on the follow-up period in different studies, but in general in ICD patients ranges from 4-60%(3). Patients in whom an ICD is implanted for primary prevention usually have a lower incidence of electrical storms[4] compared to patients in whom secondary prevention of sudden cardiac death is carried out[5]. The frequency of occurrence is similar in patients with ischemic and non-ischemic cardiomyopathy (6). In the AVID study, 457 patients with an implanted ICD were observed, who were divided into three groups: the group with electrical storm; the group with non-clustered malignant ventricular arrhythmias that do not meet the criteria for electrical storm; group without any ventricular arrhythmias after implantation. Electrical storm is an independent risk factor for death, independent of left ventricular systolic function and other variables (OR 2.4), while non-clustered VT/VF is not (OR 1.0). The risk of death is highest in the first 3 months after an electrical storm and decreases over time[7]. In the study by Brigadeau *et al*[8] 307 patients with an implanted ICD were monitored. Independent predictive factors for the occurrence of electrical storm were chronic renal failure, ventricular tachycardia as an index arrhythmia and not taking hypolipemic drugs, while diabetics had a lower incidence of electrical storm (HR 0.49). The importance of electrical storm is best illustrated by a meta-analysis which showed that it increases mortality 2.5 times compared to patients with non-clustered ventricular arrhythmias and 3.3 times compared to patients without sustained ventricular arrhythmias[9].

One of the first line measures in the treatment of an electrical storm is the exclusion of inappropriate shocks. They may be the result of oversensing electrical potentials, atrial tachyarrhythmia or defibrillator lead fracture. If they are detected, ICD programming is carried out. Aggressive ICD programming with a lower heart rate threshold and shorter VF detection time and turning off antitachycardia pacing during ICD capacitor charging have been shown to be associated with the development of electrical storm[10]. Repeated intracardiac shocks alone increase morbidity and mortality[11]. In this context ICD shock burden should be reduced and antitachycardia pacing should be favored in order to terminate ventricular arrhythmia. This is achieved by increasing the heart rate, which represents the detection threshold, and by extending the arrhythmia detection time. Both interventions have been shown to reduce the number of shocks delivered, without increasing syncope, and also reduce mortality[12,13]. In some cases, the therapy delivered by the device is excluded, especially in a case of incessant VT that is well tolerated by the patient. If it is necessary and a subspecialist programming the defibrillator is not readily available, it can be turned off by placing a magnet over the device[14]. In the case of our patient, all delivered ICD shocks were appropriate, so an attempt was made to search for a precipitating factor. These are most commonly electrolyte disorders (hypokalemia, hypomagne-

semia), myocardial ischemia, worsening of heart failure, sepsis, discontinuation of antiarrhythmic drugs[1]. However, in most cases there is no identified reversible cause of an electrical storm. In the SHIELD study, a trigger was found in only 13% of cases[15].

In our patient, the potassium level on admission was normal, high-sensitivity troponin was negative even after multiple intracardiac shocks, which practically ruled out acute myocardial ischemia. Coronary angiography performed after the patient's stabilization during the same hospital episode indicated stationary angiographic findings on the epicardial coronary arteries. We were unable to routinely assess magnesium levels. The possible cause of the electrical storm in our patient could have been the heart failure worsening, as lung congestion signs were registered at the time of admission. Exacerbation of heart failure is one of the possible causes of electrical storm reported in the literature. In a prospective study by Guerra *et al*[16], 146 patients were followed, of whom 34 were initially hospitalized due to electrical storm, 82 due to heart failure worsening and 30 due to non-clustered VT/VF episodes. Patients from the first two groups had a similar and at the same time significantly higher mortality rate as well as a shorter time to rehospitalization compared to the third group. Among those initially hospitalized due to electrical storm, 25% were re-hospitalized due to electrical storm and 42% due to heart failure worsening. It was concluded that electrical storm in patients with heart failure and ICD can be considered a warning sign of impending pump failure and even overt heart failure, rather than as an independent event.

The ECG recording of arrhythmia during an electrical storm is significant because it indicates the possible etiology and thus the therapy of choice. Sustained monomorphic VT is the most common arrhythmia seen in an electrical storm and is caused by a re-entry mechanism on heterogeneous ventricular scar tissue as a consequence of ischemic and non-ischemic cardiomyopathy. On the other hand, polymorphic VT/VF storm is most often caused by myocardial ischemia, channelopathies or idiopathic VF in a structurally normal heart. A 12-channel ECG recording showing the morphology of ventricular premature beats that initiate an episode of polymorphic VT/VF is important for planning ablation of the arrhythmogenic substrate[17].

After confirming an electrical storm, a risk assessment is done. Patients with at least one of the following are treated as high-risk: hemodynamic instability, left ventricular ejection fraction below 30%, moderate to severe renal failure and the presence of chronic obstructive pulmonary disease. Such patients should be treated in intensive care units where sedation and mechanical ventilation are available[8]. Our patient did not meet the mentioned criteria, but due to the large number of delivered intracardial shocks in a short time frame and the development of congestive heart failure, he was treated in the Coronary Care Unit with constant electrocardiographic and hemodynamic monitoring.

Increased sympathetic tone plays an important role in the occurrence of an electrical storm[18]. Deep sedation with endotracheal intubation is recommended in high-risk patients in whom sustained arrhythmia with repeated intracardiac shocks is expected. These measures have positive psychological effects, reducing the patients discomfort. For sedation, benzodiazepines and short-acting opioid analgesics are recommended because they induce sedation and analgesia without negative inotropic effects. Anecdotally, the use of propofol as an effective means to terminate an electrical storm has been reported. It should be carefully used because of its cardiodepressant nature, and patients with an electrical storm usually have impaired left ventricular systolic function[17,19-21]. In our patient, we decided on the concept of so-called "conscious sedation" by giving repeated doses of modazolam of 2 mg i.v. so that relaxation and reduction of sympathetic tone in the patient was achieved without respiratory depression and the need for mechanical ventilation. The absence of a therapeutic effect would lead to us to escalate to deep sedation and mechanical ventilation, but this was not necessary in this particular case due to successful termination of electrical storm.

Antiarrhythmic drug therapy has historically been considered the mainstay of electrical storm treatment[17]. Although logical, this concept has its limitations. In a meta-analysis by Santagnelli *et al*[22], it was found that treatment with antiarrhythmic drugs reduces the probability of recurrent storm by 1.5 fold, but without decreasing mortality. When assessing pharmacological agents individually, only amiodarone was found to reduce the number of episodes of malignant ventricular arrhythmias and ICD shocks, while sotalolol, azimilide, and celivarone did not. On the other hand, unlike other antiarrhythmic drugs, amiodarone is associated with 3.36 fold higher mortality compared to a group of patients who received standard therapy, without specific antiarrhythmic drugs. The choice of antiarrhythmic drug depends on the cause of the electrical storm, the severity of the associated heart failure, and potential toxicity. In an electrical storm with VT, a vicious circle of events can occur when repeated intracardiac shocks induce new VT episodes and further shocks by increasing the sympathetic tone. Therefore, the suppression of increased sympathetic tone with the use of B blockers is an initial measure in the treatment of electrical storm[17]. In the MADIT II study, the use of B blockers in the treatment of ventricular arrhythmias reduced recurrent ventricular arrhythmias by 52%[23]. A recent study with a smaller sample size randomized patients with electrical storm to receive i.v. propranolol or metoprolol for the first 24 hours with mandatory coadministration of amiodarone i.v. in both groups. Patients who received propranolol had fewer recurrent ventricular arrhythmias in the first 24 hours (53% vs. 90%), fewer ICD shocks delivered, shorter time to termination of the arrhythmia, and shorter hospitalization time. The explanation for the effectiveness of non-selective B blockers in this context is the downegulation of β1 receptor seen in patients with heart failure. Instead of the usual β1:β2 receptor ratio of 70:30 to 80:20 in healthy cardiomyocytes, in heart failure this ratio is 60:40. In this study, propranolol

proved to be an effective and safe agent, especially considering that the subgroup receiving it had an average left ventricular EF of 25%[24]. This should be used in the context of its use in the acute phase of electrical storm, because propranolol itself is not indicated in the chronic therapy of heart failure[25]. Our patient was already on chronic oral therapy with amiodarone and 5 mg bisoprolol per day. In the acute phase, we re-administered amiodarone i.v. in the loading dose with simultaneous administration of lidocaine 120 mg i.v. bolus + 1200 mg/12h continuous infusion. In addition, we used 3 i.v. boluses of metoprolol 5mg every 5 minutes and then continued with 20mg bisoprolol per day. Although aggressive, this approach, with the use of midazolam and the programmed basic frequency of the ICD of 80/min, led to the cessation of further arrhythmias and stabilization of the patient. Overdrive pacing with a frequency faster than the patient's spontaneous heart rate acted by means of suppression of ventricular premature beats, because the record from the ICD programmer clearly indicated that these were episodes of polymorphic VT initiated by the R to T phenomenon, with further degeneration to VF and lead to the delivery of an intracardiac DC shock. The successful application of a similar therapeutic approach in individual cases was used by several groups of authors[26-28]. This strategy is also mentioned in the latest ESC guidelines for ventricular arrhythmias and the prevention of sudden cardiac death, in the algorithm after exhausting other therapeutic options in patients in whom bradycardia or post-extrasystolic pauses induce the occurrence of polymorphic VT/VF, without labeled class of recommendations and level of evidence[29].

One of the therapeutic measures in patients with electric storm is radiofrequency (RF) ablation of the arrhythmogenic substrate in specialized centers high volume centers. According to the latest ESC guidelines on this topic, it has a class IIa recommendation, level of evidence C for use in patients with repeated polymorphic VT/VF episodes resistant to antiarrhythmic therapy and revascularization[29]. After clinical stabilization we did not refer the patient to ablation , yet decided on clinical follow up. It has proven to be correct so far, because in the follow up period of two years, there were not any clinical episodes of VT/VF and also no episodes detected by electronic device interrogation. Considering the patient's habitus (body mass index of 38kg/m^2) and targeted anamnestic data on poor sleep quality and daytime sleepiness, it was suspected that obstructive sleep apnea in this particular patient may increase the arrhythmogenic potential. Such an association has been demonstrated in a number of smaller studies[30-31]. In a larger randomized study, it was shown that in patients with implanted CRT-D, sleep breathing disorders (obstructive and, even to a greater extent, central sleep apnea) are independently associated with shorter event-free survival, time to the first monitored ventricular arrhythmia and appropriate ICD therapies[32]. Our patient, for now, is not motivated for diagnosis and possibly treatment of sleep breathing disorders.

Conclusion

Electrical storm is the most dramatic clinical event in patients with an implanted cardioverter defibrillator that requires adequate diagnostics and multimodality approach in specialized centers with appropriate equipment and personnel. This approach improves the outcome of such complex patients.

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Sažetak

Električna oluja kod pacijenta sa inplantabilnim kardioverter defibilatorom

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Električna oluja predstavlja pojavu najmanje tri epizode maligne komorske aritmije (održiva ventrikularna tahikardija/ventrikularna fibrilacija) tokom 24h, najčešće kod bolesnika sa implantiranim kardioverter defibrilatorom. U radu se prikazuje bolesnik starosti 48 godina sa implantiranim ICD u sekundarnoj prevenciji naprasne srčane smrti i pojavom električne oluje po tipu polimorfne VT sa degeneracijom u VF prećenu isporučivanjem 10 intrakardijalnih DC šokova u periodu od nekoliko sati. Prikazan je multimodalitetni pristup bolesniku koji je uključivao inicijalnu potragu za precipitirajućim činiocima, elektronsku interogaciju i promene u programiranju uređaja, antiaritmijsku terapiju i sedaciju bolesnika. Primenjene mere diskutovane su u kontekstu dostupne literature. Multimodalitetni pristup bolesnicima sa električnom olujom u specijalizovanim centrima poboljšava prognozu ovako kompleksnih bolesnika.

Ključne reči: *električna pluja, inplantabilni srčani defibrilator, multimodalitetni imidžing*