Case reports

Successful treatment of electrical storm with oral quinidine in Brugada syndrome

Katia Bettiol, Lorella Gianfranchi, Salvatore Scarfò, Federico Pacchioni, Mario Pedaci, Paolo Alboni

Division of Cardiology and Arrhythmologic Center, Civic Hospital, Cento (FE), Italy

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A patient implanted with a cardioverter-defibrillator for symptomatic Brugada syndrome was referred to our hospital 17 months later because of recurrent shocks due to ventricular fibrillation (VF). Isoprenaline was intravenously infused and prevented VF episodes, but VF recurred after every attempt of drug discontinuation. A total of 34 shocks were recorded over 25 days. Subsequently, we treated the patient with oral quinidine and the drug suppressed the electrical storm and prevented VF episodes during a follow-up period of 3 years. This case report, together with few others reported in the literature, suggests a role of oral quinidine in the treatment of electrical storm in Brugada syndrome.

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Address:

Dr.ssa Katia Bettiol

Divisione di Cardiologia
Ospedale Civile
Via Vicini, 2
44042 Cento (FE)
E-mail:
p.alboni@ausl.fe.it

The Brugada syndrome is an inherited arrhythmogenic disease associated with a risk of sudden death due to ventricular fibrillation (VF)[1]. The implantable cardioverter-defibrillator (ICD) appears to be the only effective treatment for prevention of sudden death, since antiarrhythmic drugs are often harmful[2,3]. A case of electrical storm (recurrent VF) occurring in a patient with Brugada syndrome, implanted with an ICD is reported, in whom oral administration of quinidine was effective in abolishing the electrical storm and preventing VF episodes during a 3-year follow-up period.

Case report

A 57-year-old woman had been implanted with an ICD in our Division of Cardiology in June 1999 because of Brugada syndrome with syncopal episodes and a cardiac arrest due to VF just after hospital admission. She was again referred to our hospital 17 months later because of recurrent syncopal episodes followed by electrical shocks (5 episodes in the last 10 days). During the first 5 days of hospitalization the patient received other 10 shocks due to VF (Fig. 1). Then, isoprenaline was intravenously infused at the dosage of 0.02 mg/hour. Drug infusion reduced ST-segment elevation in the right precordial leads and VF episodes were prevented, but VF recurred after every attempt of isoprenaline discontinuation. Other 19 ICD shocks due to VF were recorded in the next 10 days after four attempts of drug discontinuation. So, a total of 34 electrical shocks were recorded over 25 days. Subsequently, we treated the patient with quinidine bisulphate at the dosage of 500 mg/day and during an in-hospital observation period of 6 days the patient remained free of VF episodes. Quinidine markedly reduced ST-segment elevation in the right precordial leads (Fig. 2). During a follow-up period of 3 years the patient has always assumed quinidine at the same dosage and she remained free of ICD shock. The patient has often been advised to discontinue the drug, at least temporarily, but she refused for fear of electrical shocks.

Discussion

Electrical storm seems to be a rare phenomenon in ICD implanted patients with Brugada syndrome; however, its prevalence is unknown. Few data suggest that isoprenaline infusion may be a valuable treatment in case of electrical storm in this syndrome[4-6]. In our patient it was effective but VF episodes recurred after every attempt of drug discontinuation. We started a

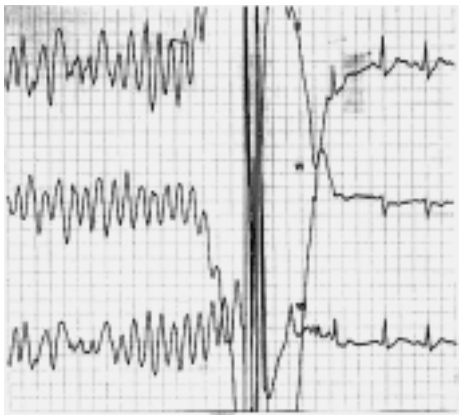

Figure 1. *Episode of ventricular fibrillation during in-hospital monitoring, interrupted by an implantable cardioverter-defibrillator shock.*

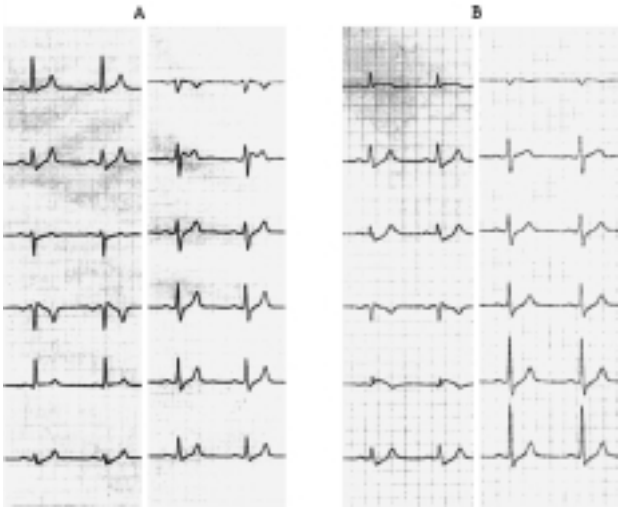

Figure 2. A: *electrocardiographic tracing of the patient; a typical ST-segment elevation is present in the right precordial leads.* B: *ST segment normalizes after quinidine administration.*

treatment with oral quinidine on the basis of a few pretherapeutic reports[7-9]. In fact, it had been reported that oral quinidine can reduce ST-segment elevation in the right precordial leads[7,8] and prevent reinduction of VF during electrophysiological study[9]. In our patient the drug reduced ST-segment elevation, as well. Very recently, 5 patients with Brugada syndrome and electrical storm, successfully treated with oral quinidine, have been reported; 4 by Hermida et al.[10] and 1 by Mok et al.[11]. The proposed mechanism of ventricular arrhythmias and ST-segment elevation in Brugada syndrome involves the imbalance between the inward (I_{Na} and I_{Ca}) and outward currents, mainly the I_{to} at the end of phase 1 action potential of the right ventricular epicardium[4,12]. Experimental studies have shown that quinidine, by blocking I_{to}, is effective in restoring the epicardial action potential dome and thus normalizing the ST segment and preventing phase 2 reentry and ventricular arrhythmias in experimental models of Brugada syndrome[12]. We cannot exclude with certainty that interruption of electrical storm in our patient was a chance and not secondary to quinidine treatment. However, our case report, together with the few others reported in the literature[10,11], suggests a role of oral quinidine in the treatment of electrical storm in Brugada syndrome, although a randomized trial should be carried out.

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