

CASE REPORT

Adenosine-induced sinus tachycardia in a patient with Myotonic Dystrophy type 1

VINCENZO RUSSO¹, GERARDO NIGRO¹, ANDREA ANTONIO PAPA¹, ANNA RAGO¹, NADIA DELLA CIOPPA², ANNA CRISTIANO¹ AND MARIA GIOVANNA RUSSO¹

¹ Department of Cardio-Thoracic and Respiratory Sciences, Chair of Cardiology, Second University of Naples, Monaldi Hospital, Italy; ² Department of Anesthesiology, Monaldi Hospital, Naples, Italy

We report the case of a 32-year-old man with Myotonic Dystrophy type 1 showing adenosine-induced sinus tachycardia during transesophageal electrophysiological evaluation.

Key words: sinus tachycardia, adenosine, proarrhythmic effect, myotonic dystrophy

Case report

A 32 year old man with Myotonic Dystrophy type 1 (DM1) was referred to our observation for recurrent palpitations at rest, lasting a few minutes. The medical history was negative for family sudden death, dizziness or syncope. Physical examination revealed a blood pressure of 120/70 mmHg, clear lungs and normal heart sounds. Haematological examination, except for CK values, urinary analysis and thyroid function were all normal. He had no other medical pathologies in the past, and denied intake of alcohol, tobacco or any medications.

Muscle involvement was characterized by slight weakness and atrophy of facial, axial, semi-distal, and distal compartments. No abnormalities of other organs and systems including eye, endocrine system, central nervous system, gastrointestinal system, respiratory apparatus and heart were present. Electrocardiographic (ECG) examination showed a sinus rhythm of 75 bpm, normal atrioventricular conduction (PR: 160 ms), mild slurred QRS upstroke in leads V3-V4, no ST segment or T wave changes (Fig. 1). Neither chest x-ray nor color-doppler echocardiography revealed any cardiac structural or functional abnormality. 24-hours ECG Holter monitoring and treadmill stress test did not show arrhythmias. He

underwent transesophageal electrophysiological evaluation: the atrioventricular node refractory period was 240 msec. During the test was not used anesthesia. The patient was conscious during the entire test and his O₂ saturation, measured by a pulse oximeter, was consistently 98-99%. Programmed atrial stimulation up to triple extrastimuli did not induce supraventricular arrhythmias. Intravenous adenosine (12 mg) was performed to slow AV conduction and unmask unapparent pathways. After a single-bolus, rapidly followed by saline flush, a sinus tachycardia at a frequency of 145 beats/min, was observed, self-terminating in approximately 50 seconds, without any change in QRS morphology (Fig. 2). After the second adenosine bolus, carried out about 5 minutes

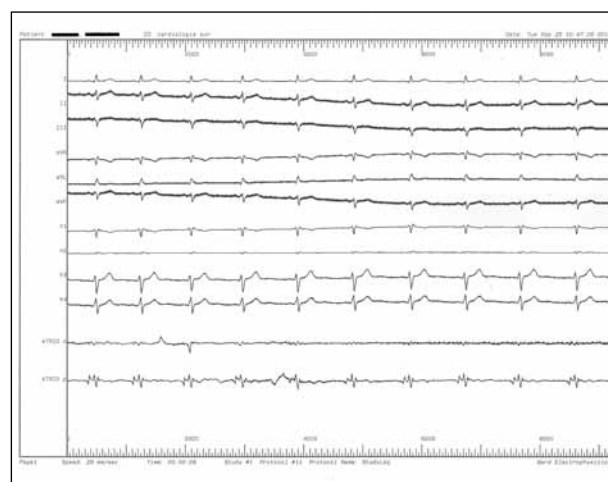


Figure 1. Basal electrocardiogram.

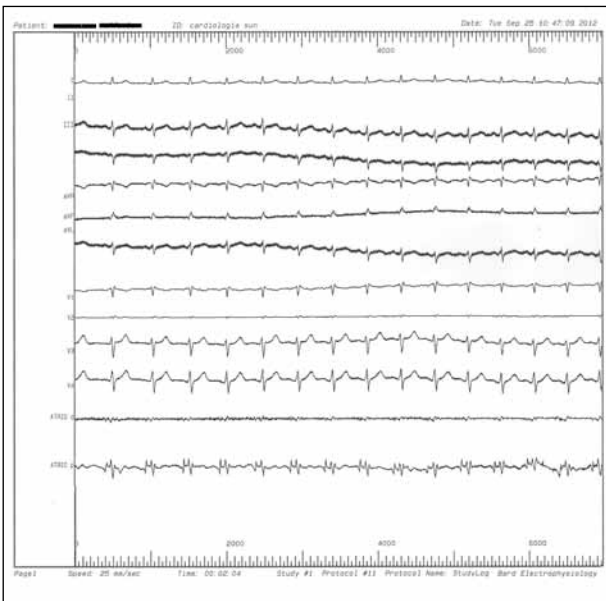


Figure 2. Adenosine induced sinus tachycardia.

later, a similar effect was observed. The patient remained conscious and asymptomatic during the entire period of tachycardia.

Discussion

Myotonic dystrophy type 1 (DM1) is a serious autosomal-dominant hereditary disease with an estimated incidence of 1 in 8000 births. The phenotype is characterized by myotonia and muscle weakness, but a multisystemic involvement with highly variable clinical manifestations is very frequent. Cardiac involvement, that often precedes the skeletal muscle one, occurs in 80% of DM1 patients and represents the second most common cause of death, after respiratory causes (1). Arrhythmic risk in DM1 patients may be related to the heterogeneity of ven-

tricular repolarization (2, 3), expressing by an increase in QTc and JTc dispersion, as reported in other congenital (4-8) or acquired (9-11) heart diseases and in neuromuscular diseases (12-16). Adenosine is an endogenous nucleoside whose actions were first investigated by Drury & Szent-Gyorgyi in 1929 (17). They described a slowing of sinus rate and a reduction of conduction through the atrioventricular node in the hearts of laboratory mammals. The adenosine test seems to have a good sensitivity for unmasking accessory pathways (18), because it extends the atrioventricular node refractory period, favoring the anterograde conduction. The electrophysiologic effects of adenosine on a specific AV bypass tract, depend on the type of cell that characterize the tract: nodal type cells (with decremental conduction) or atrial myocytes. In sinu-atrial nodal cells, the activation of a potassium outward current, results in a reduced rate of phase IV depolarization, thereby slowing sinu-atrial node automaticity. In the AV node, adenosine prolongs post-repolarization refractoriness and suppresses excitability of cells in the N region of the node, resulting in an AV nodal conduction block of variable degree.

Some authors reported serious adverse events related to the adenosine infusion, including supraventricular and life threatening ventricular arrhythmias (19, 20). To date, while the association between adenosine infusion and ventricular arrhythmias is well known (21), little is known about the adenosine induced supraventricular arrhythmias. The possible mechanisms underlying pro-arrhythmic effect of adenosine are summarized in Table 1.

The most common pro-arrhythmic effect of adenosine is the induction of atrial fibrillation (AF) (19). This is probably owing to the shortening of atrial refractoriness, that favours the induction of reentrant arrhythmias. Because the signal transduction pathways activated by adenosine and acetylcholine, converge on the same potassium channels and produce similar electrophysiologic effects in the atrial myocardium, adenosine-induced and vagus nerve-dependent AF are mechanistically similar.

Table 1. Mechanisms underlying pro-arrhythmic effect of adenosine.

Type of arrhythmia	Underlying mechanisms
Atrial fibrillation/atrial flutter	Shortening of atrial refractoriness Sympathetic activation
Rapidly conducted atrial flutter/atrial fibrillation	Sympathetic activation
Orthodromic atrioventricular reentry tachycardia	Critical prolongation of anterograde AV nodal and retrograde activation of the atrium via an accessory pathway
Pre-excited atrial fibrillation/atrial flutter	Critical prolongation of anterograde AV nodal and retrograde activation of the atrium via an accessory pathway
Sinus tachycardia/atrial tachycardia	Reflex sympathetic discharge Direct sympathetic activation by stimulation in the carotid body chemoreceptors

Both adenosine and vagus nerve activation cause a spatially and temporally heterogeneous shortening of atrial refractoriness (22).

A dangerous increase of the ventricular rate in patients with atrial flutter (23, 24), atrioventricular reentry tachycardia (25), narrow QRS complex tachycardia (26), and 'mild sinus tachycardia' following a brief period of bradycardia (27) have also been reported. The case here shown is the first report of sinus tachycardia induced by intravenous adenosine infusion in a DM1 patient without previous bradycardia, supporting the hypothesis of Biaggioni et al. (28) that adenosine may be responsible of a direct increase in circulating catecholamine levels and of sympathetic nerve traffic, by sympathetic stimulation in the carotid body chemoreceptors. However, in our patient, no bradycardia during the entire test was observed, as transesophageal electrophysiological evaluation was performed under continuous ECG monitoring. We are aware that the relation between adenosine infusion and sinus tachycardia is anecdotal, but we believe that all possible and unexpected pro-arrhythmic effects of antiarrhythmic drugs should be taken into account in clinical practice, particularly in patients affected by neuromuscular disorders.

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