ELSEVIER

International Journal of Cardiology 101 (2005) 147–150

International Journal of Cardiology

www.elsevier.com/locate/ijcard

Letter to the Editor

Kearns–Sayre syndrome associated with trifascicular block and QT prolongation

Pavlos Karanikis, Panagiotis Korantzopoulos*, Evaggelos Kountouris, Vasiliki Dimitroula, Dimitrios Patsouras, Eugenia Pappa, Konstantinos Siogas

Department of Cardiology, "G. Hatzikosta" General Hospital of Ioannina, Leoforos Makrygianni, 45001, Ioannina, Greece

Received 1 December 2003; accepted 7 January 2004
Available online 18 May 2004

Abstract

Patients suffering from Kearns–Sayre syndrome (KSS) often develop conduction defects that may lead to syncope or sudden cardiac death. The association of conduction abnormalities with prolonged QT interval in these patients is very rare. We describe a patient with KSS and diabetes mellitus who suffered a torsades de pointes-induced syncopal attack, in the presence of trifascicular block and QT prolongation (QTc: 574 ms). The patient was successfully treated with permanent pacing. This case highlights that torsades de pointes represents a potential mechanism of syncope or sudden cardiac death in patients with KSS.

Keywords: Kearns–Sayre syndrome; Conduction abnormalities; Trifascicular block; Long QT; Torsades de pointes; Diabetes mellitus

1. Introduction

Kearns–Sayre syndrome (KSS) represents a mitochondrial encephalomyopathy that may involve several organ systems [1,2]. Notably, neuromuscular and cardiac conduction systems are the most frequently affected. The diagnosis is based upon the triad of progressive ophthalmoplegia, pigmentary retinopathy and cardiac conduction defects [1]. In this report, we briefly describe a patient with KSS who experienced syncopal attacks in the presence of trifascicular block and QT prolongation.

2. Case report

A 47-year-old woman was transferred to the hospital due to a syncopal attack that happened 1 h before, while she was lying down on her bed. Her husband noticed that the episode was sudden and lasted a few minutes. In addition, he stressed that during the attack she was pulseless and pale. The recovery from that episode was sudden and complete, and it was accompanied by vomiting. On arrival at the Emergency Department the patient was complaining only of a mild dizziness and nausea. There was no impairment in the level of consciousness. Her past medical history was significant for KSS that had resulted in ophthalmoplegia, blindness and mobility disorders, and for diabetes mellitus. She did not mention any syncopal attack since early childhood. Her medications included only insulin for diabetes control. On admission, her blood pressure was 140/80 mm Hg in both arms without orthostatic hypotension, with a heart rate of 50/min. Apart from her occular anomalies, the rest of the physical examination was normal. Complete blood count, blood chemistries (including glucose and electrolytes) and thyroid function tests were all within normal limits. Furthermore, arterial blood gases (while breathing room air) and acid–base balance were also normal. Interestingly, an electrocardiogram (ECG) disclosed sinus bradycardia, trifascicular block (right bundle branch block, left anterior hemiblock, first degree atrioventricular block) and prolongation of the QT interval (Fig. 1). Specifically, the corrected QT interval (QTc) was calculated to be 574 ms. The patient was then transferred immediately to the Cardiac Intensive Care Unit and a few minutes later she developed torsades de pointes (Fig. 2) with simultaneous loss of consciousness. Cardiopulmonary resuscitation was

* Corresponding author. Tel.: +30-26510-80350; fax: +30-26510-80353.
E-mail addresses: pkor@oneway.gr, pan-kor@mailbox.gr (P. Korantzopoulos).

0167-5273/$ - see front matter © 2004 Elsevier Ireland Ltd. All rights reserved.
doi:10.1016/j.ijcard.2004.01.027

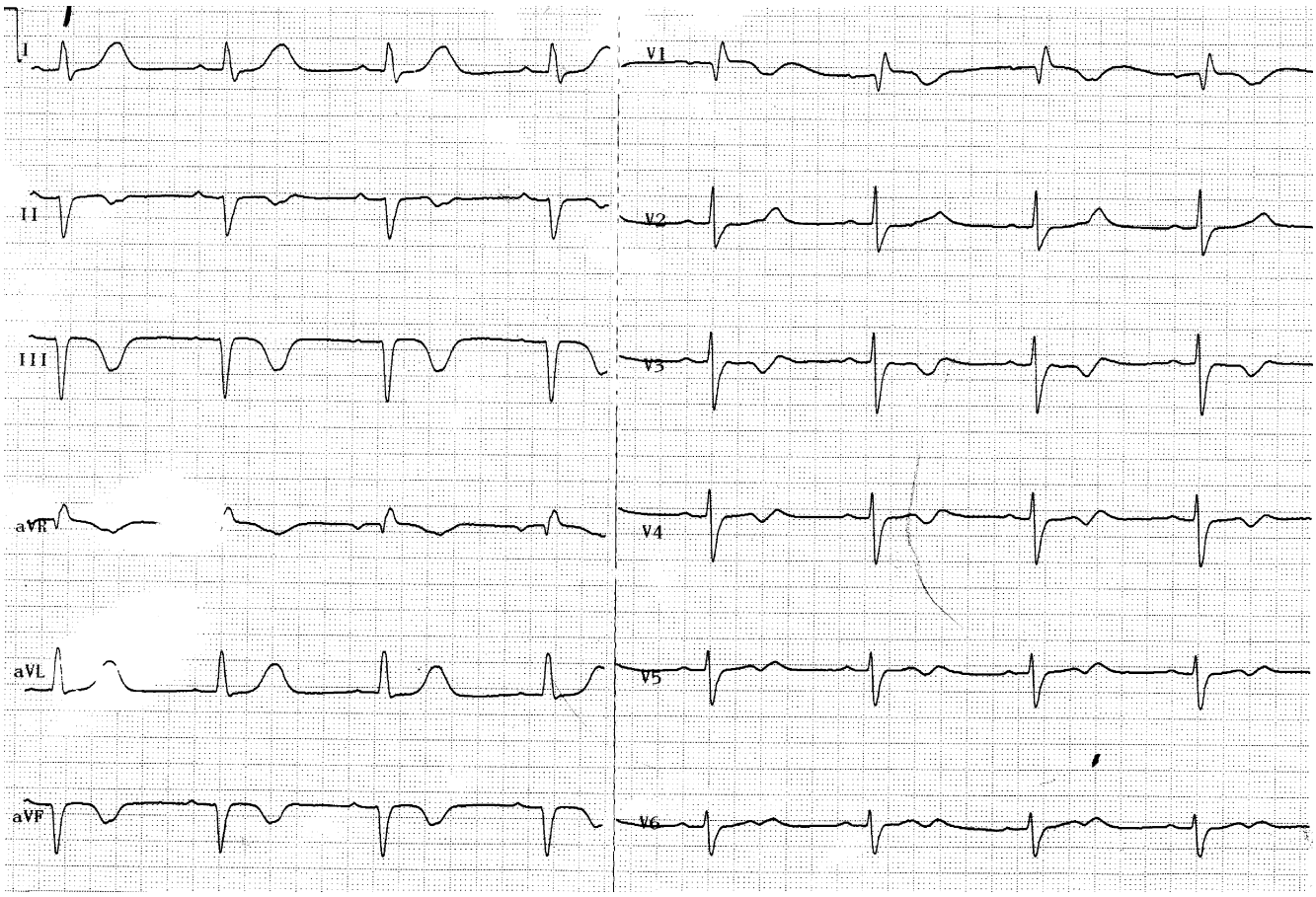

Fig. 1. Surface ECG on admission showing sinus rhythm, trifascicular block, and QT prolongation (QTc: 574 msec).

initiated and when the paddles of the defibrillator were placed on her chest sinus rhythm was automatically restored before electrical discharge. The patient became hemodynamically stable and subsequently a temporary pacemaker was successfully placed. An echocardiographic study at the bedside showed normal size of all cardiac chambers, an ejection fraction of 0.70, and a mild mitral and tricuspid regurgitation (Fig. 3). Two days later, a permanent dual chamber pacemaker (DDDR) was implanted. The patient was discharged after 4 days on good clinical condition. During follow-up, the patient remained asymptomatic but the pacing rate was adjusted to 80/min in order to normalize the QT interval.

3. Discussion

KSS typically affects the heart causing cardiac conduction defects, whereas in many cases the disease progresses to complete heart block manifesting as syncope or sudden death [1–4]. The principal pathological finding is the development of abnormal mitochondria that accumulate in myocardial cells before cardiac conduction abnormalities become overt [5]. It is believed that the presence of fascicular bundle branch block represents an important risk factor for the development of complete block [3]. Thus, prophylactic permanent pacemaker implantation in patients at high risk is imperative. The association of KSS with long

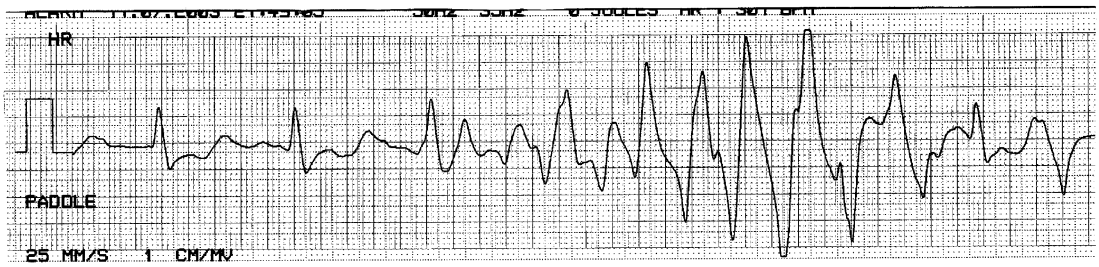

Fig. 2. ECG strip obtained from the defibrillator during the syncopal attack disclosing torsades de pointes.