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Case Report

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An unusual reason for complete atrioventricular block; Kearn Sayre Syndrome

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Abstract

Kearn sayre syndrome (KSS) is a rare disease with a typical onset before 20 years and characterized by triad of chronic progressive external ophthalmoplegia, retinitis pigmentosa and progressive cardiac conduction abnormalities. The most important prognostic factor in KSS patients is the involvement of heart characterized by progressive degeneration of the conduction system. In patients with KSS, early diagnosis of cardiac involvement is very important because complete atrioventricular (AV) block may develop and it can be cause of sudden cardiac death. Herein we report a patient with KSS who presented with syncope due to complete AV block.

Keywords: Kearn sayre syndrome, atrioventricular block, disease

Introduction

Kearn Sayre syndrome (KSS) is a rare mitochondrial myopathy. The exact prevalence of Kearns-Sayre syndrome is unknown but it has been estimated 1 to 3 per 100,000 individuals [1]. The characteristic diagnostic triad consists of progressive external ophthalmoplegia, retinitis pigmentosa and cardiac conduction abnormalities [2]. We report a patient with 3rd degree atrioventricular block which due to KSS.

Case Presentation

A 20-year-old male patient presented to cardiology outpatient clinic with complaints of dizziness and weakness. The patient had these complaints for several years and but recently his complaints had progressed and also new onset fainting was added. The patient had 3 syncope episodes during the last month. He described these episodes as loss of consciousness following blacking outs. His medical history was unremarkable and he was not on any medication.

On inspection bilateral ptosis was remarkable and neurologic examination revealed bilateral limited lateral eye movements (figure 1). Other system examinations were normal. The patients ECG showed normal sinus rhythm, 65/min, without any conduction abnormality and the QT interval was normal. Transthoracic echocardiogram revealed normal biventricular dimension and functions without any stenotic pathology which might be explain the patient's complaints. Results of laboratory tests were all normal. 24-hour Holter ECG monitoring was performed. Also the patient was referred to neurologist and ophthalmologist for further investigations of bilateral ptosis and limited eye movements.

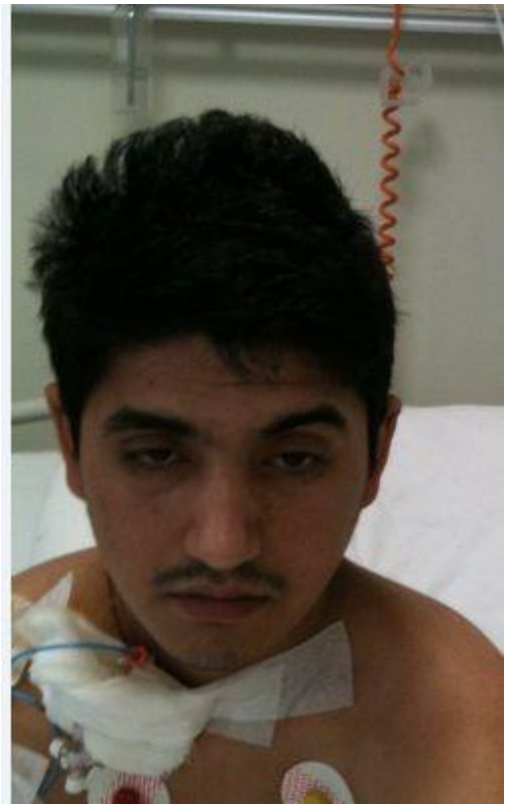

Figure 1. Bilateral ptosis and opthalmoplegia

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Three days later he dropped suddenly while he was waiting in cardiology outpatient clinic. His blood pressure was 70/40mmHg, and heart rate was 36/min. ECG showed complete AV block (Figure 2). The patient admitted to coronary care unit and temporary pacemaker insertion performed through the right internal jugulary vein. Following pacemaker insertion, the patient's haemodynamic parameters normalized and he was fully conscious. He reported that previous syncope episodes had been identical with this last episode. His Holter ECG monitoring showed multiple episodes of 2nd and 3rd degree AV block. Neurologist evaluated the patient and the final diagnosis was KSS. Cerebral MRI was performed and revelead normal. The patient was evaluated by an endocrinologist and they did not detect any problem. A Medtronic VDD pacemaker was implanted on the fourth day of hospital admission. Following pacemaker implantation, the patient was discharged without any complaints of dizziness or weakness.

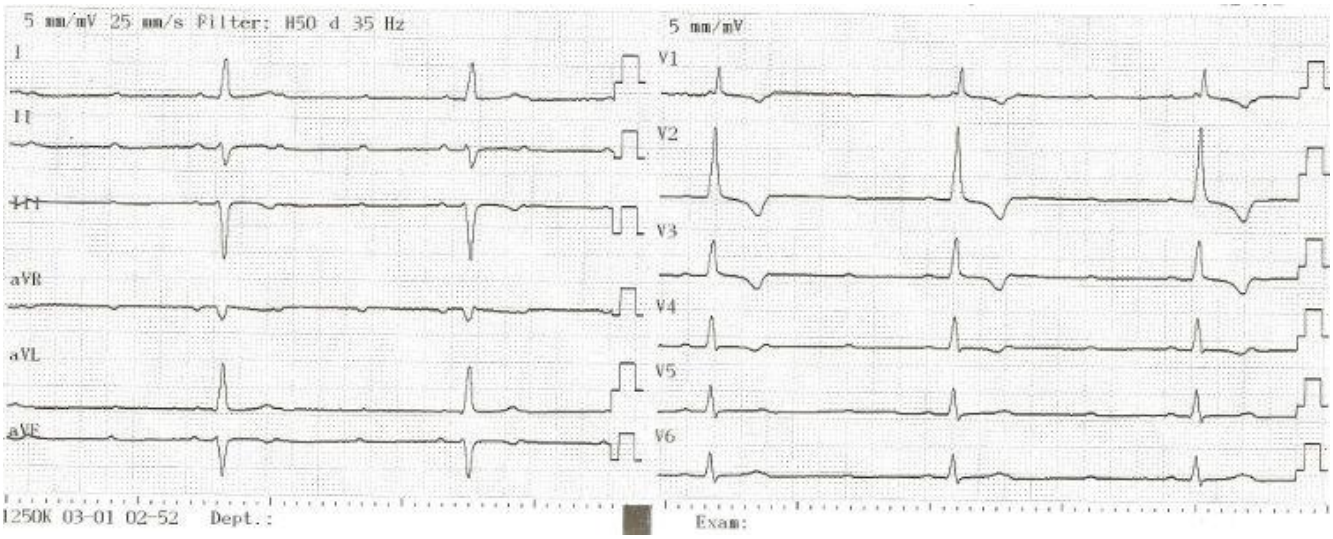

Figure 2: ECG shows complete AV block

Discussion

The most important prognostic factor in KSS patients is the involvement of heart characterized by progressive degeneration of the conduction system. Cardiac manifestations occur in 57% of patients with KSS which also include syncope attacks, heart failure and cardiac arrest. It may also be associated with mental retardation, ataxia, deafness, muscle weakness, and endocrine disorders [3-4] however our patient has not involved neuro-endocrine disorders.

Although KSS presumably reduces life expectancy, there is no numerical data in the literature. Morbidity depends on severity and the number of systems or organs involved, which widely varies from patient to patient. Cardiac involvement and especially disturbances of conduction system are significant and preventable causes of mortality [4-5].

The largest case series of KSS reported by Khambatta et al [3] of 35 patients with KSS, cardiovascular disorders of the patients included syncope (6 patients; 17%) and sudden cardiac death (4 patients; 11%). Other cardiac disorders included atrioventricular block (11 patients; 31%) and conduction abnormalities (23 patients; 66%)

Cardiac conduction abnormalities include fascicular blocks, complete or incomplete bundle branch blocks and atrioventricular blocks. Sudden cardiac death can occur in 23 % of KSS patients due to complete heart block [3]. In our case the patient had recurrent episodes of syncope and his ECG during one of this episodes showed complete AV block and 24-hour Holter ECG monitoring demonstrated multiple episodes of 2nd and 3rd degree AV block. Because of the progressive nature of the disease and high risk of sudden cardiac death, the threshold for permanent pacemaker implantation in patients with KSS is very low. It is recommended implantation of a permanent pacemaker even in asymptomatic cases KSS due to possible unpredictable progression in any of the AV blocks [6].

Conclusion

In patients with KSS, early diagnosis of cardiac involvement is very important because complete AV block may develop and it can be cause of sudden cardiac death. Pacing even in asymptomatic patients who have 2rd and 3rd degree AV block is necessary because it improves survival and symptoms of the patient. Regular and long term cardiovascular follow up is essential.

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