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CASE REPORT

Syncope in a Patient with H/O Kearns Sayre Syndrome

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Abstract

Patients with history of mitochondrial disorders are at increased risk of having conduction disorders and cardiomyopathy and should have low threshold for pacemaker and implantable cardioverter defibrillator placement. Kearns Sayre syndrome is the result of deletions in mitochondrial DNA which causes bilateral pigmentary retinopathy and conduction abnormalities. Judicious use of implantable cardioverter defibrillator in this subset population with cardiomyopathy or prolonged QT interval is required in addition to pacing to prevent risk of sudden cardiac death. A subset of these patients might continue to experience life threatening arrhythmias including torsade de pointes and ventricular fibrillation despite a functional pacemaker.

Keywords

Kearns Sayre syndrome, Atrioventricular block, Implantable cardioverter defibrillator, Polymorphic ventricular tachycardia

Patients with history of mitochondrial disorders are at increased risk of having conduction disorders and cardiomyopathy and should have low threshold for pacemaker and implantable cardioverter defibrillator placement as compared to the general population.

The patient is a forty-five-year-old female who presented with multiple syncopal episodes in the last one month with each episode lasting for a few seconds without any associated palpitations, seizure-like activity, or chest pain. She had a history of Kearns Sayre syndrome [KSS], diagnosed at the age of twenty-two with bilateral visual and hearing loss, ptosis, ataxia, and progressively worsening muscle weakness and was wheelchair dependent. Family history was not significant for mitochondrial disease or any other disorder. Her pulse was 35 beats per minute and BP was 120/70. The cardiac exam was significant only for bradycardia. She denied any intake of beta-blockers, recent travel to the New England area, or insect bite. EKG showed bradycardia [heart rate of 48 bpm] with a 1st-degree heart block [PR interval of 352 ms] and incomplete right bundle branch block (Figure 1) while echocardiogram showed no abnormalities and preserved left ventricular function. Left-sided dual-chamber along implantable cardioverter-defibrillator was placed and she remained asymptomatic afterward at one year follow up.

Discussion

Kearns Sayre syndrome is a rare sporadic mitochondrial myopathy with the onset of disease before the age of twenty years [1] and is characterized by the triad of progressive external ophthalmoplegia, pigmentary retinopathy, and cardiac conduction system disturbances [2]. Prognosis depends mainly on cardiac involvement which occurs in about 50% of the patients with the most common presentation being atrioventricular block. The patients with syncopal episodes are presumed to have underlying heart block [3]. The other common arrhythmia is bradycardia related polymorphic ventricular tachycardia [4] and insertion of a pacemaker should be curative for both of these arrhythmias. In an asymptomatic patient with KSS, routine screening EKG or Holter monitor should be done at regular yearly intervals to detect life-threatening arrhythmias. Despite of 1st degree heart block which is not a classic indication for general population, patients with KSS like our patient should be

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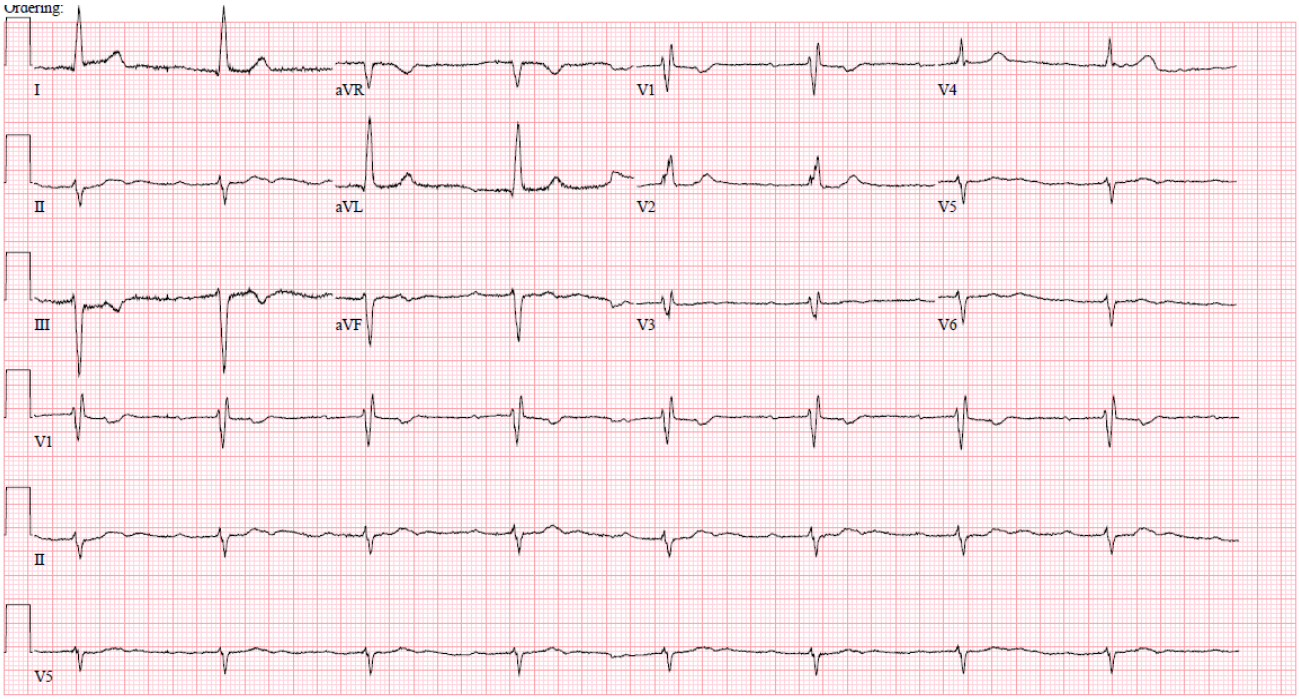

Figure 1: EKG at time of presentation showing 1st degree AV block.

paced due to rapid progression to AV block and then to complete heart block in 50% of the patients. It has been recently recognized that a subset of patients might continue to experience life-threatening arrhythmias including torsades de pointes and ventricular fibrillation despite a functional pacemaker [5-7]. Patients with structurally normal hearts on transthoracic echocardiogram may have subclinical disease only detectable by cardiac MRI [4], However, a causative relationship between myocardial fibrosis and life-threatening arrhythmias has not been established yet. Thus, judicious use of implantable cardioverter-defibrillator in this subset population, especially ones with cardiomyopathy or prolonged QT interval is required in addition to pacing to prevent the risk of sudden cardiac death [7] like done in our patient. The use of pacemaker alone may not have mortality benefit in this subset of population as was previously believed.

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None.

Conflict of Interest

None.

All the authors have access to the data and a role in writing of the manuscript.

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