Case Report

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A Rare Cause of Syncope Mimicking Refractory Seizure in a Child: Case Report

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Abstract

Background: Transient loss of consciousness may be due to cardiac cause. Arrhythmias may lead to syncope or to sudden death. Long QT syndrome (LQTS) is a genetically transmitted cardiac arrhythmia caused by ion channel protein abnormalities. It is characterized by electrocardiographic abnormalities and a high incidence of sudden cardiac death. *Case Report:* We report a case of long QT syndrome in a 10-year girl child who was treated for refractory seizures with multiple antiepileptic drugs. Simple electrocardiography confirmed the diagnosis. *Conclusion*: Child was treated with non-selective beta blocker and showed dramatic response.

Keywords: Arrhythmia; Beta blocker; LQTS; Refractory seizure; Syncope.

Introduction

Syncope is a transient loss of consciousness due to transient global cerebral hypoperfusion characterized by rapid onset, short duration and spontaneous recovery. Transient loss of consciousness can be due to syncope, neurologic or cerebrovascular disease, metabolic or psychogenic cause. Greater than 95% of all syncopal episodes involving healthy adolescents and young adults are innocuous. It is estimated that as many as 15% of children will have an attack between 8 and 18 years.[1] Before the age of 6, syncope is unusual except in the setting of breath holding spells, seizure disorders and cardiac arrhythmias.

Differential diagnosis of syncope include vascular (most common) and cardiac causes. Cardiac causes of syncope include obstructive lesions, myocardial dysfunction and arrhythmias. Arrhythmias (bradyarrhythmia or tachyarrhythmia) are the most common cardiac cause of syncope. Arrhythmias may lead to acutely decreased cardiac output and may degenerate into ventricular fibrillation and lead to sudden death. Non-traumatic sudden death in children are often due to specific cardiac cause. The incidence of sudden death varies from 0.8 to 6.2 per 100,000 per year[2] in children and adolescents as opposed to 1 per 1,000 in adults. Approximately 65% of sudden deaths are a result of heart-related problems.[2] Cardiac causes of sudden death could be due to structural lesions, acquired lesions and sometimes due to conduction system abnormality. Long QT syndrome (LQTS) is a cardiac channelopathy which may present in infancy or early childhood. The cardinal events of the cardiac channelopathies include syncope, seizures and sudden cardiac death. Sudden cardiac death may be the sentinel event.

Case report

An 11-year-old girl child, second born of second degree consanguinous parents, deaf mute had presented with complaints of multiple episodes of sudden unresponsiveness followed by spontaneous resolution within few minutes since one and a half years of age. There was no limb movements during such episodes and the recovery was dramatic. She was diagnosed to have seizure elsewhere and was started on antiepileptic drugs (AED). She had breakthrough seizures even with valproate and levetiracetam at maximum tolerated dose. Due to increase in frequency of episodes, clobazam and topiramate had been added. Neuroimaging and electroencephalography done elsewhere was normal. Despite multiple AEDs, frequency of episodes had increased (>10 episodes/month) and hence she came to our department for opinion. Detailed history revealed that the events comprised of vacant stare and unresponsiveness with loss of tone and posture without any involuntary limb movements. The events were usually precipitated by emotional stress, sometimes accompanied by palpitations and did not occur during sleep.

There was no similar illness, sudden cardiac death or pacemaker implantation in any of the family members. Drug compliance was good. She was delivered full term by labor naturalis with birth weight of 3 kg and there was no history of birth asphyxia. Infantile period was uneventful. General examination was normal with following anthropometric indices (weight: 26 kg, height: 133 cm, head circumference: 51 cm, BMI: 14.69). Her vital signs were within normal limits (Pulse rate: 95/min and regular, Respiratory rate: 16/min, BP: 110/80 mm Hg). CVS examination was normal. Other system examination were normal with no focal neurological deficit. Developmentally her motor milestones were normal and she was going to a special school for the hearing impaired with average scholastic performance.

The episodes were more in favor of cardiogenic syncope. Hence work up to identify causes of cardiogenic syncope was planned with the preliminary investigation being an electrocardiography. ECG showed heart rate of 90 beats per minute (bpm), normal QRS axis, PR interval of 140 milliseconds and QTc interval of 720 m/sec. Serial ECG showed consistent prolongation of QT interval. Hence a clinical diagnosis of long QT syndrome was made. Acquired causes of QTc prolongation were ruled out by history and investigations. This includes intake of precipitating drugs, dyselectrolytemias, and hypothyroidism. Echocardiogram was done to rule out any structural heart disease predisposing to syncope and was normal. Child was subjected to 24-hour Holter monitoring which confirmed the same finding of prolonged QT interval, QTc average being 564 msec (QTc min:464 m/sec, QTc max: 679 m/sec) and average heart rate was 92 bpm (max HR: 92 bpm, min HR: 61 bpm). Total ventricular ectopics during the study period was 803 (21.7%). ECG screening in both parents and elder brother was normal. Audiogram confirmed moderately severe bilateral sensorineural hearing loss. Prolonged QT interval on ECG and congenital deafness points to the diagnosis of Jervell and Lange-Nielsen syndrome type of LQTS. The child scored 5.5 on Schwartz diagnostic criteria for Long QT syndrome. Child was started on non-selective beta blocker propranolol 20 mg 12th hourly. Genetic testing was not done due to financial constraint.

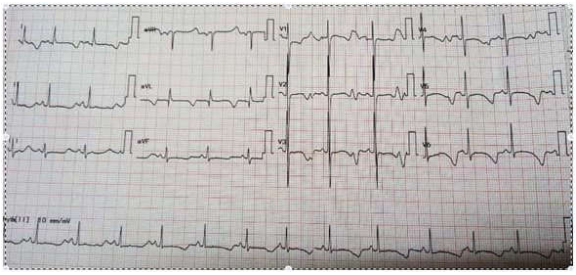

Fig. 1: Initial ECG before β-Blocker therapy showing prolonged QT interval.

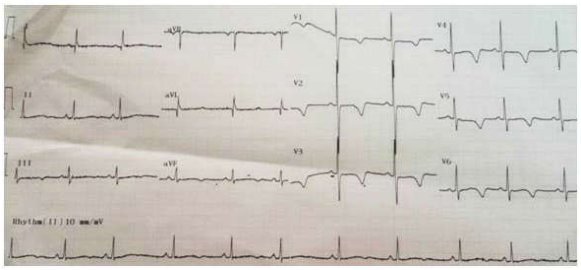

Fig. 2: Follow-up ECG after β-Blocker therapy showing shortening of QT interval.

Discussion

Long QT syndrome (LQTS) is a disorder of myocardial repolarization characterized by a prolonged QT interval on the ECG, ventricular arrhythmias and usually torsade de pointes that may result in sudden death. Ion channels that govern the electrical activity of the heart are defective in congenital LQTS. Hundreds of mutation in 13 distinct LQTS- susceptibility genes have been identified. Majority of LQTS

are due to LQTS1, LQTS2 and LQTS3. Prior to genetic mapping, LQTS was divided into two syndromes namely Romano–Ward syndrome and Jervell Lange-Nielsen syndrome. Jervell Lange-Nielsen syndrome (JLNS) is a rare inherited disorder (Autosomal Recessive) characterized by congenital deafness and conduction abnormalities of the heart.[3] Prevalence varies depending on the population studied (1/2,00,000–1/10,00,000). Romano–Ward syndrome has all the features of Jervell and Lange-Nielsen syndrome but without deafness. This syndrome transmits in an autosomal dominant manner and is much more common than Jervell and Lange-Nielsen syndrome. The severity of symptoms varies from being asymptomatic to syncope (26%), cardiac arrest (9%), and potentially sudden death. Physical activity, excitement, fright, or stress may trigger the symptoms. Hearing loss associated with JLNS is sensorineural, bilateral and profound. The family history is positive in about 60% of patients. A 12-lead ECG is the current screening tool for identification of LQTS. As per American Heart Association guidelines a QTc ≥ 450 msec in adult males and ≥ 460 m/sec in adult females must be considered prolonged QTc. In LQTS, ECG may also show abnormal T-wave morphology, bradycardia, AV block, monomorphic or polymorphic ventricular tachycardia.

All these ECG findings are considered risk factors for sudden death. Echocardiographic studies usually show a structurally and functionally normal heart. Genetic testing for known DNA mutations (KCNQ1 in LQTS1, HERG in LQTS2, SCN5A in LQTS3, KCNE1 in LQTS5) confirms the diagnosis with high specificity but low sensitivity. Hence, genotyping is not useful in ruling out the diagnosis in suspected cases. Noncompliance with medication is an important risk factor for sudden death. QT prolonging medications should be avoided and competitive sports should be restricted and if possible avoided. Swimming is particularly dangerous in LQTS1 patients. The initial treatment is β-blockers and lifestyle modification to avoid possible triggers. All symptomatic and asymptomatic children should be treated with β-blockers. β-blockers are effective in preventing cardiac events in approximately 70% of patients, and cardiac events sometimes continue to occur despite therapy. Even with β-blockers, sudden death can occur. Propranolol or nadolol is preferred and the response is LQTS type specific. β-blockers are extremely effective in LQTS1. High thoracic left sympathectomy removes the lower part of the stellate ganglion along with the first four thoracic ganglia, with almost no risk for Horner's syndrome. After this procedure, there is a dramatic reduction in the incidence of cardiac events, although sudden death may still happen (8%); the procedure has a 5-year survival rate of 94%. β-blockers are usually continued after the surgical procedure. Implantation of a cardioverter defibrillator (ICD) appears to be the most effective therapy. Appropriate shock from a cardioverter-defibrillator prevents dangerous ventricular arrhythmias resulting in sudden cardiac death. There is limited clinical experience of ICD with children. On follow up, this child is currently on regular oral propranolol 20 mg 8th hourly (Figs. 1 and 2). Her latest ECG showed heart rate of 64 bpm, normal QRS axis, PR interval of 144 m/sec and QTc interval of 509 m/sec. Child is symptom free for the past 10 months. Hence tapering of antiepileptic drugs is being done. Parents have been counselled for cervical sympathectomy and cardioverter-defibrillator implantation.

Conclusion

Long QT syndrome may masquerade as drug resistant epilepsy. This case highlights the importance of suspecting arrhythmia in a patient presenting with refractory seizures, as correct diagnosis is not only life saving but appropriate therapy improves quality of life. Electrocardiography is an effective screening tool for children presenting with transient loss of consciousness. There is potential high risk of sudden death in undiagnosed children.

Conflicts of interest: There are no conflicts of interest.

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