
CASE REPORT

ICTAL SYNCOPE

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Abstract

Isolated syncope is a rare manifestation of seizures in children. Many times, such episodes are mistaken as primary cardiac dysarrhythmias and we report one such case in an infant, who succumbed to the illness, despite treatment.

Key words: Ictal syncope, Electroencephalography (EEG), Electrocardiography (ECG), Sick Sinus Syndrome (SSS)

Introduction

Episodic loss of consciousness presents as a diagnostic challenge to the physicians as well as to the neurologists, many a time. Often, such episodes are named "epileptic" and even treated wrongly, depending upon the specific and non-specific inter-ictal EEG changes. When symptoms keep recurring, despite treatment, one would consider other organic and non-organic causes and invariably subject the patients to many diagnostic procedures with very little positive results. Changes in cardiac rhythms are known to occur during clinical seizures, but in situations where there is an epileptic discharge in EEG along with bradycardia and syncope, it is often referred as ictal bradycardia syndrome or ictal syncope. (1) Seizure related cardiac arrhythmias are mostly encountered in older children and adults only and very rare in infants. (2) The given case in an infant describes the initial difficulty in arriving at a diagnosis and the subsequent outcome.

Case Report

A six months old girl was referred from a primary physician for recurrent breath holding attacks since the age of 4 months. She was born as term, IUGR and at 4 months age, she started developing frequent apnea, following prolonged incessant cry with momentary stiffness and up rolling of eyes. These episodes were diagnosed as "breath holding spells" even though there were no definite precipitating events. Routine ECG and

EEG could not reveal initially any specific abnormality except bradycardia; she received oral iron syrup for some time with no improvement of symptoms and so parents stopped the drug. At six months of age, symptoms became more severe and of prolonged durations; at one instance she was hospitalized when she developed prolonged apnea, needed oxygen inhalation and bag and mask ventilation. At that time, ECG revealed, prolonged sinus bradycardia, asystole, junctional escapes preceded by sinus tachycardia. Sick sinus syndrome (SSS) was contemplated on this basis and oral beta-blocker (propranolol) treatment was started. Later permanent cardiac pacemaker was implanted (Intermedics S.A., Mode VV1 DASH rate adaptive pulse generator with rate of 80/mt) following left thoracotomy with an intramyocardial unipolar lead. With this, her symptoms improved and so she was labeled as SSS. After 2 weeks, she had a severe life threatening event; she collapsed with bradycardia and lost consciousness suddenly. She was intubated and ventilated for 48 hours. Pacemaker function was checked and found to be normal. Phenobarbitone was given as loading dose with the probable diagnosis of autonomic seizures as, interestingly, a prolonged sleep EEG recording with video telemetry, at this time, revealed a left fronto temporal paroxysmal theta burst which was not documented before; this confirmed a diagnosis of ictal syncope. Carbamazepine was planned; however the next day, she developed one more prolonged episode of syncope, bradycardia and died as she could not be resuscitated.

Discussion

Syncope as an isolated manifestation of seizures in children has a poor localizing value and the term ictal syncope is proposed for transient loss of consciousness and posture that occurs in a seizure before or even without convulsions. (1) Seizure related cardiac dysrhythms are well recognized and present mostly as tachyarrhythmia and bradycardia; asystole, on the

other hand are exceptional and cause life threatening hemodynamic disturbances. (3) They invariably occur along with clinical seizures or as late as 60 minutes after the seizures. (4) The present case of ours, presenting as "breath-holding spell" was diagnosed initially as prolonged QT interval syndrome leading to syncope, which is often misinterpreted as the possible etiology. Long QT syndromes are associated with genuinely life threatening syncope, which may be simple or convulsive. (5) Later, sick sinus syndrome was diagnosed on her with the characteristic ECG abnormality. There was no history suggestive of typical clinical seizures in our patient, which prompted us to diagnose this as primary cardiac rhythm disorder, as the primary event was apnea. The diagnosis of ictal bradycardia and syncope was contemplated, only, when a prolonged EEG trace was done, after one life threatening episode which showed left temporal focus, although routine EEG was normal before. EEG findings in syncopal events are non-specific with slowing of the background rhythms, followed by high amplitude delta waves and then absolute flattening, depending upon the severity of cerebral hypoperfusion, but tend to improve in the reverse sequence as clinical improvement sets in gradually. (6) Ictal bradycardia syndrome may occasionally mimic syncope and so unless simultaneous EEG and ECG are not performed, the diagnosis of this unique disorder could be missed. Review of the literature also emphasizes the difficulties in the diagnosis and treatment of this rare condition. (7,8) A possibility of dominant parasympathetic ictal stimulus following excitation of reticular formation, precipitating ictal bradycardia and sinus arrest cannot be ruled out. When autoregulation gets disrupted because of missing sympathetic innervations, especially in presence of associated cortical dysplasia, sudden cardiac ictal death might occur which could have been the reason in our patient. (9) Paroxysmal modification of autonomic parameters may be responsible for the cardiac rhythm and asystole. (10) Besides, right insular irritation could cause tachycardia and left insula, bradycardia. (11) Ictal sinus arrests with temporal lobe focus and seizures (TLE) have been reported in the past that became symptom free after epilepsy surgery. (12) This child, in spite of cardiac pacing and specific anticonvulsant treatment, succumbed to the illness probably because of the underlying cortical abnormality controlling autoregulation. MRI study of brain was planned to delineate any dysplastic abnormality of the brain but could not be done.

Conclusion

The ictal bradycardia / syncope syndrome should be considered in patients with unusual or refractory episodes of syncope or in patients with history of

both epilepsy and syncope. Correct diagnosis may be attained by simultaneous EEG / ECG at the time of syncope. At times, when the diagnosis could not be achieved, a video EEG recording of the ictal events becomes mandatory which could throw more light. Cardiac pacemaker implantation along with antiepileptic drug therapy may be necessary to minimize the attacks although permanent cure could be claimed with epileptic surgery. Pacemaker alone may correct the malignant form of bradyarrhythmia in the heart and thereby decrease seizure occurrence but may not prevent the death due to seizures totally.

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E-published: 1st March 2012. **Art#**15

DOI No. 10.7199/ped.oncall.2012.15
