

Case Report

# Sick sinus syndrome masquerading as idiopathic primary generalized epilepsy

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**Abstract.** Idiopathic primary generalized epilepsy (IPGE) is characterized by generalized convulsions with loss of consciousness (LOC) at times associated with lateral tongue bite and loss of bladder control. Interictal EEG record shows an organized background with a 9-12 Hz posterior dominant rhythm (PDR) and generalized spike wave and polyspike wave discharges. We present here a patient whose typical spells were characterized by abrupt LOC accompanied with tonic posturing and convulsive movements and occasional bladder incontinence. Interictal EEG was reported as intermittent generalized spike wave discharges leading to initiation of levetiracetam therapy. Marked sinus bradycardia was noted during examination during inpatient video-EEG admission. A typical event captured on video-EEG with simultaneous telemetry revealed sinus arrest leading to cerebral hypoperfusion and seizure as the underlying cause of his "epilepsy".

Key words: Sick sinus syndrome, sino-atrial arrest, seizure, idiopathic generalized epilepsy

## 1. Introduction

Sick sinus syndrome is characterized by a dysfunctional sinus node and can be caused by either intrinsic pathologies of the sinus node (e.g. amyloidosis, connective tissue disorders, Chagas disease, hemochromatosis, Diphtheria, leukemia, sarcoidosis) or by extrinsic causes. Its clinical presentation can at times mimic a generalized seizure disorder.

## 2. Case report

A-29-year-old right handed male was admitted for inpatient video-EEG for characterization of his epilepsy. His typical seizures were characterized by abrupt LOC lasting for 30-40 seconds accompanied with tonic posturing and low amplitude convulsive movements as per his girlfriend. During one such seizure he recalled suffering loss of bladder control. Post ictal state

was described as an exceedingly brief period of confusion and disorientation with rapid return to full awareness. At the time of admission patient was on levetiracetam 500 mg twice daily for presumed IPGE after interictal EEG carried out at an outside institution was reported as intermittent generalized spike wave discharges. MRI brain was normal. The first day of video-EEG recording showed an organized EEG background, PDR of 10 Hz in the awake state and paroxysmal frontally predominant delta discharge with a poorly defined spike wave component suggesting the diagnosis of IPGE (Fig. 1). Marked sinus bradycardia on auscultation prompted simultaneous telemetry monitoring. An event was recorded in the early morning (later confirmed to be typical of the patient's reported seizures by his girlfriend). On the telemetry abrupt sinus pause for 10 seconds was noted. The EEG showed high amplitude delta slowing followed by diffuse voltage attenuation (Fig. 2,3,4). On the video the patient was noted to have abrupt behavioral arrest followed by LOC, tonic posturing, and low amplitude convulsive movements with rapid return to consciousness on resumption of cardiac rhythm.

Patient was transferred to the coronary care unit where sick sinus syndrome (SSS) was diagnosed warranting permanent pacemaker (PPM) implantation.

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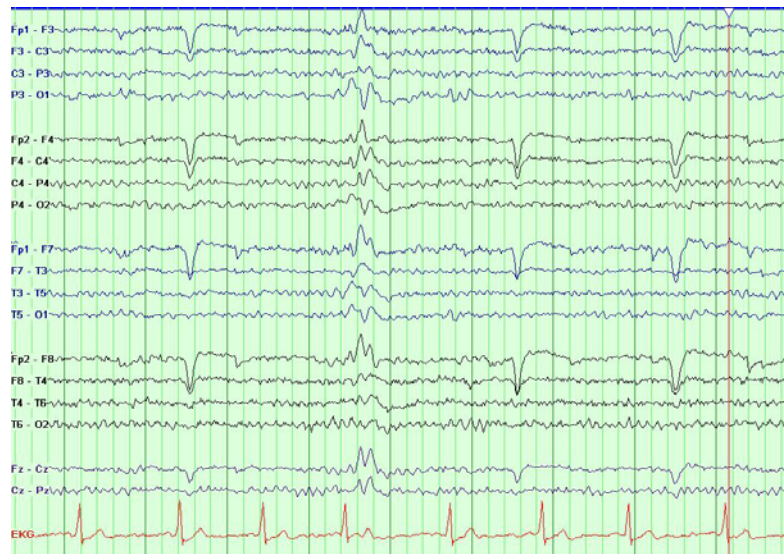


Fig. 1. EEG (longitudinal bipolar montage) showing an organized background with intermittent bursts of paroxysmal delta activity with ill-defined sharp wave component.

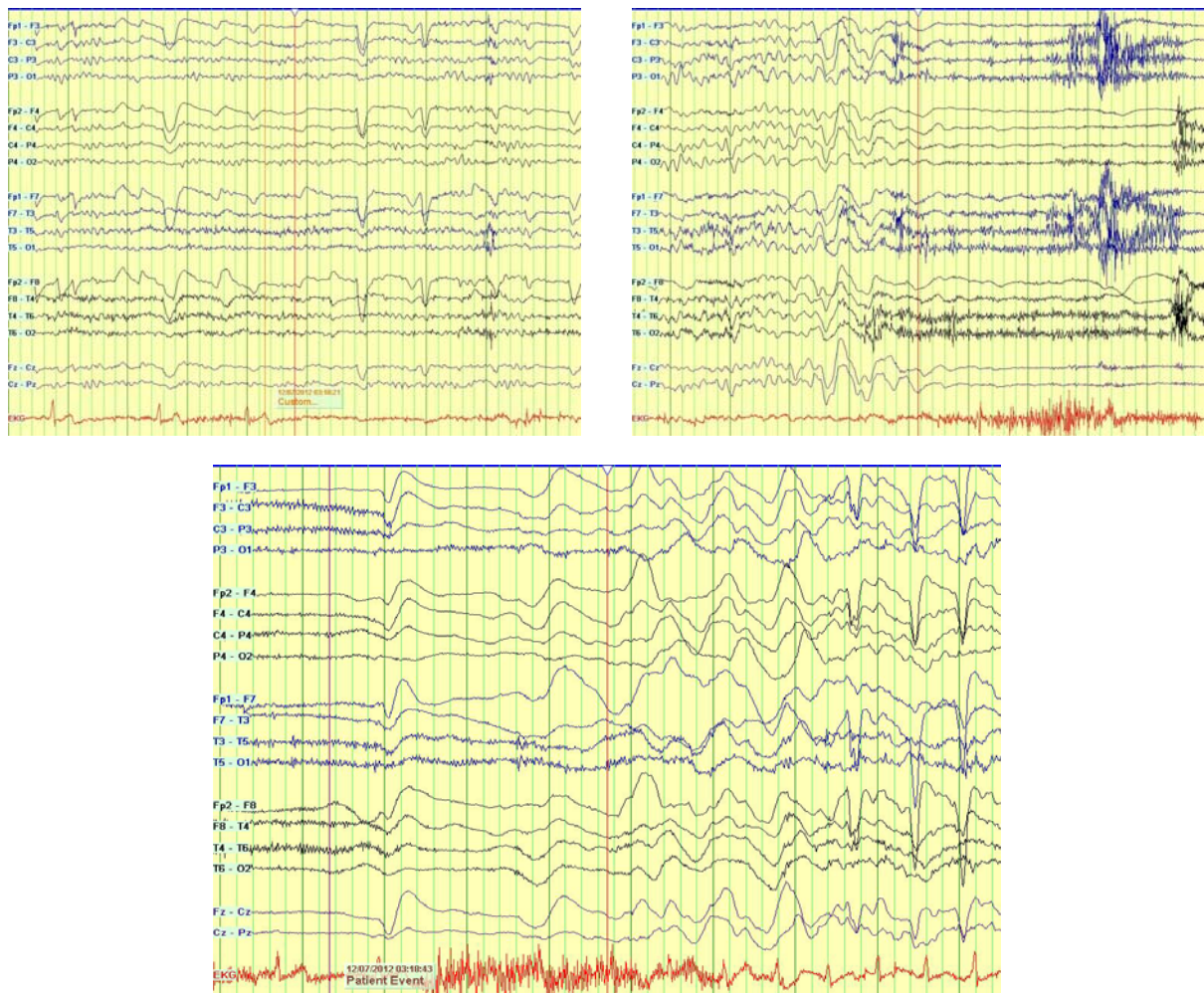


Fig. 2,3,4. Abrupt sinus pause for 10 seconds is noted on the EKG tracing. The EEG shows high amplitude delta slowing followed by diffuse voltage attenuation. On the video the patient was noted to have abrupt behavioral arrest followed by LOC, tonic posturing, and low amplitude convulsive movements with rapid return to consciousness on resumption of cardiac rhythm.

### 3. Discussion

Sick sinus syndrome is characterized by a dysfunctional sinus node and can be caused by either intrinsic pathologies of the sinus node (e.g. amyloidosis, connective tissue disorders, Chagas disease, hemochromatosis, Diphtheria, leukemia, sarcoidosis) or by extrinsic causes (e.g. drugs like beta blockers and digitalis, toxins, hyperkalemia) (1). It is important to remember that the term SSS encompasses a range of conditions characterized by sinus node dysfunction on electrocardiogram (EKG). The EKG patterns which have been described include sino-atrial arrest or block, severe sinus bradycardia, sino-atrial arrest with failure of subsidiary pacemaker resulting in cardiac asystole and bradycardia alternating with tachycardia among others (2). Stokes and Adams in 1846 described syncopal attacks related to paroxysmal atrioventricular block and sino-atrial blocker. In between the syncopal episodes patients may have a normal EKG and hence the diagnosis may be missed if the EKG is not recording during the spell itself. Spontaneous recovery usually ensues following an episode though sudden death due to fatal cardiac arrest has been reported. The state and duration of unconsciousness may range from momentary lapses of awareness to a complete loss of consciousness. Tonic or clonic seizures may occur if the period of cerebral hypoperfusion is prolonged (3). PPM implantation is indicated when episodes of symptomatic bradycardia occur (syncope, LOC, confusional states) or when the sinus arrest exceeds 3 seconds.

Idiopathic primary generalized epilepsy is characterized by generalized seizures (without any preceding aura) which cause abrupt LOC at times accompanied by a lateral tongue bite and loss of bladder control. Post ictal period is characterized by confusion and disorientation sometimes lasting up to an hour with gradual return to full awareness. Thus the duration of the post ictal period can at times help to differentiate a generalized tonic-clonic convulsion from a syncopal episode since tongue biting and even urinary incontinence can occur in severe episodes of syncope. Both neurogenic (vasovagal) and cardiogenic syncope may mimic IPGE with spells characterized by abrupt LOC. In vasovagal attacks there is usually a rapid return to full awareness on assumption of a supine posture. Our patient's spells were thought to represent IGE with interictal EEG showing organized background, PDR of 10 Hz and suggesting intermittent generalized spike wave discharges. As a result treatment with anticonvulsant was

initiated. During the video-EEG study at our institution we did not document any definite generalized spike wave or polyspike wave discharges. Only the paroxysmal discharges shown in Fig 1 were noted. We were unable to obtain the EEG carried out at the outside institution. So while it is possible that the EEG was simply misread at the outside institution, this in the end proved to be a red herring since the etiology revealed itself to be cardiac (sick sinus syndrome). The patient's stepbrother from his father's side had suffered similar episodes of LOC, had an abnormal EEG and was on anticonvulsant therapy. So it is possible that the paroxysmal discharges with an ill defined spike wave component visualized in our patient's EEG are indeed an expression of IPGE trait running in the family. Conversely it is possible that the stepbrother too has a cardiac pathology which has been misdiagnosed as epilepsy. Further EEG and cardiac studies may help in clarifying the diagnosis and at the time of writing this manuscript we have reached out to the patient's stepbrother.

There was no doubt that our patient warranted a PPM placement after investigations revealed no structural or infiltrative heart disease. Whether levetiracetam is still warranted stirred a healthy debate. His cardiologist and I favored discontinuing levetiracetam therapy after the captured event was confirmed as typical of the patient's reported seizures by his girlfriend. In retrospective it was the sinus bradycardia noted on auscultation which raised the first red flag that this was not simple IPGE.

Our case underscores the importance of an accurate diagnosis of paroxysmal episodes of loss of consciousness. A misdiagnosis of primary generalized epilepsy in our patient could have exposed him to anticonvulsants which theoretically carry an arrhythmogenic risk. For example lamotrigine which is fast gaining acceptance as the drug of choice for primary generalized epilepsy exerts its anticonvulsant effect by acting on presynaptic voltage-gated sodium channels to decrease glutamate release. While it does not prolong QT/QTc in healthy subjects, prolongation of QRS interval on electrocardiogram and complete heart block has been reported in patients following lamotrigine overdose (4). Other anticonvulsants too have been reported to have effects akin to the cardiac effects of other sodium channel blockers such as tricyclic antidepressants (TCAs).

Our case has implications on sudden unexpected death in epilepsy (SUDEP) research as little is currently known about patients with sick

sinus syndrome who manifest abnormal EEGs and present with loss of consciousness.

#### **4. Conclusion**

Sick sinus syndrome (cardiac syncope) may mimic IPGE in young people. Patients may harbor a PGE trait characterized by spike wave discharges on EEG and still not have seizures. It is important to remember the adage which has stood the test of time-treat the patient and not his EEG.

#### **References**

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