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Ventricular standstill disguised as epilepsy: A case report on Stokes-Adams attacks

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ABSTRACT

Background: Stokes-Adams attacks presenting as convulsions may be difficult to distinguish from epilepsy. Stokes-Adams Syndrome is a transient abrupt collapse into unconsciousness due to a sudden but pronounced decrease in cardiac output caused by change in heart rate and rhythm, resulting in syncope.

Case presentation: We report a patient who presented with multiple convulsive episodes managed as epilepsy, until she was found to have paroxysmal total atrioventricular block. Previously, she had been treated with anti-seizure medications without relief. Ventricular standstill was seen on cardiac monitoring and the convulsive episodes were determined as Stokes-Adams attacks. She underwent percutaneous coronary intervention and has been free of convulsive episodes since.

Conclusion: Awareness of distinction between seizures/epilepsy and convulsive syncope is important and may be life-saving. A good clinical history as well as simple non-invasive tests such as electroencephalogram and electrocardiogram are important in establishing correct diagnosis.

1. Introduction

Stokes-Adams Syndrome (SAS) is a condition in which there is sudden transient loss of consciousness (TLOC) from a sudden major decrease in cardiac output, caused by difference in the heart rate and rhythm [1]. It is usually caused by shifting from normal rhythm to high grade block, abnormal ventricular rhythm, and slowing of idioventricular rhythm during a complete heart block [1]. During these episodes, there is cerebral hypoxia that may lead to convulsive episodes and mimic epileptic seizures, contributing to the increasing rate of misdiagnosis of epilepsy. Ventricular asystole, also known as paroxysmal ventricular standstill, is a fatal arrhythmia that is one of the rarest causes of SAS [2].

Convulsive episodes may be non-epileptic events mimicking epileptic seizures caused by non-neurological conditions including metabolic and cardiac causes including transition from normal rhythm to high grade block, complete heart block, and other abnormal ventricular rhythm such as ventricular fibrillation [1]. It has been shown that around 20% of patients who are being treated with chronic use of anti-seizure medications and on long-term follow-up in epilepsy clinics do not have epilepsy [2]. These patients are shown to have cardiovascular problems that may cause syncope, complicated by abnormal movements from generalized cerebral hypoxia, which may appear like epileptic seizures, leading to a misdiagnosis of epilepsy [3].

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We present a case of a patient with Stokes-Adams attacks from ventricular standstill presenting with recurrent convulsive episodes and was misdiagnosed as epilepsy. This report aimed to highlight that syncope may present with convulsive episodes that are easily mistaken for epilepsy. Prompt recognition can be life-saving. It also aimed to discuss the importance of clinical evaluation, identification of danger signs, and work-up in such cases.

2. Case report

A 58-year-old otherwise healthy female presented at the emergency department for multiple convulsive episodes. She had a seven-month history of recurrent seizure-like episodes presenting as upward rolling of eyeballs, twitching of the mouth, and myoclonic jerking of all extremities, usually lasting for 10 seconds, preceded by dimming of vision, lightheadedness, and bradycardia. After each episode, the patient has immediate return of awareness. This prompted consultation to a local hospital where she was managed as a case of seizure disorder and was prescribed levetiracetam 250 mg three times a day. One year prior, the patient observed onset of occasional palpitations, lightheadedness, easy fatigability, and chest heaviness which prompted consult. An electrocardiogram (ECG) was done which revealed arrhythmia. She was diagnosed with acute coronary syndrome and was started on aspirin, clopidogrel, atorvastatin, ramipril, and nebivolol, which the patient took for four months only. She was also advised to undergo coronary angiogram and electroencephalogram (EEG), which the patient did not do.

In the interim, the patient had no recurrence of the convulsive episodes, however, she would have paroxysmal episodes of lightheadedness and easy fatigability, with recorded bradycardia as low as 32 beats per minute, using their own pulse oximeter. She sought consult at a private clinic where her medications were adjusted. During exertion, the patient's aforementioned symptoms became more severe, eventually leading to TLOC. During these episodes, the patient's eyes were open and directed upwards, associated oral automatisms, and mycolonic jerking. A few seconds after the event, the patient immediately gains back awareness and becomes verbally responsive. This prompted another consult and the patient was then advised to increase her levetiracetam dose to 3000 mg/day without relief.

On admission, the patient was noted to be an ectomorph. She had an adynamic precordium and an irregular heart rhythm. The rest of the systemic physical and neurological examination was unremarkable. The patient had upward rolling of eyeballs and mouth twitching with associated desaturations hence, the patient was hooked to oxygen support via nasal cannula. Recurrent attacks were observed every few minutes, prompting referral to the neurology service. She was hooked to a cardiac monitor, where it was observed that the heart rate would decrease and would eventually stop before every attack, lasting for about 10 seconds. After the attack, the heart rate returns to normal. It was also observed that the patient had spontaneous recovery within a few seconds without post-ictal confusion.

The complete blood count, blood chemistries, and other inflammatory markers were normal. Chest radiograph showed cardiomegaly and prominent interstitial markings which may be secondary to senescent changes. Transthoracic echocardiogram was also done which revealed normal left ventricular geometry with multisegmental wall motion abnormality and mildly depressed overall systolic dysfunction, with grade I diastolic function.

The ECG revealed complete left bundle branch block, normal axis, with prolonged corrected QT interval (0.51 ms) and prolonged QRS (160 ms) (see Fig. 1A), which eventually developed to ventricular standstill showing P waves without ventricular conduction (see Fig. 1B). There was no evidence of a myocardial infarction. Based on the patient's clinical presentation and ECG findings, the convulsive episodes were diagnosed as Stokes-Adams attacks from the paroxysmal total atrioventricular (AV) block. The patient was referred to the cardiology service. The patient immediately underwent insertion of a temporary pacemaker via a transcutaneous catheter. Eventually, she underwent coronary angiogram and permanent pacemaker insertion. Since the procedure, she has not been having bradycardia and has been free of convulsive episodes. Her repeat ECG findings now show sinus rhythm and normal axis, with left ventricular hypertrophy. On follow up one month after discharge, her levetiracetam was discontinued completely. A repeat EEG was not done. Currently, she has been off levetiracetam for eight months, with no recurrence of seizure-like episodes.

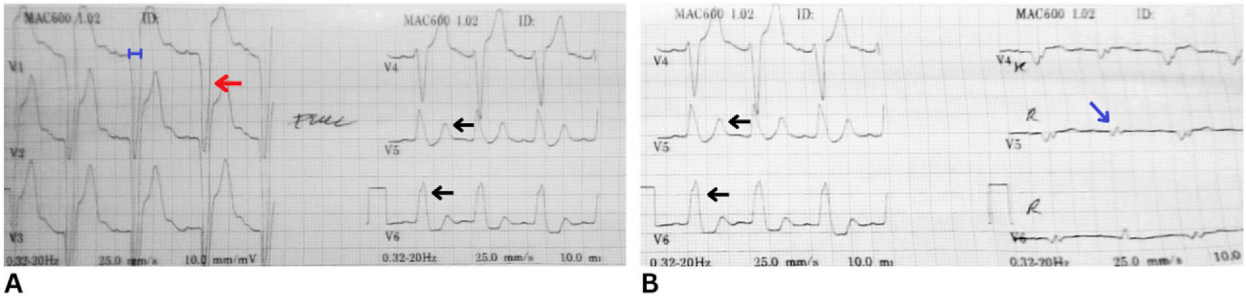

Fig. 1. Electrocardiogram (ECG) at the emergency department showing complete left bundle branch block **(A)** with QRS 0.16 (blue line), deep S waves in V1 (red arrow), and broad monophasic R waves in V5–V6 (black arrow), followed by ventricular standstill **(B)** during the seizure-like episodes. Note the P waves without ventricular conduction in ventricular standstill (blue arrow). (For interpretation of the references to colour in this figure legend, the reader is referred to the Web version of this article.)

3. Discussion

Our patient was initially managed as a case of epilepsy due to seizure-like episodes with no relief of symptoms. The preceding symptoms of lightheadedness and profound bradycardia, which led to asystole argue for syncope. Together with the acute cardiac events, the left bundle branch block and the absence of triggers, yielded a high suspicion for a cardiac cause. These episodes were proven to be Stokes-Adams attacks. The seizure-like episodes were observed during brief periods of the ventricular standstill.

Seizures may be manifestations of diseases with various etiologies including metabolic, cardiac, and neurologic [2]. These convulsive episodes may be non-epileptic events mimicking epileptic seizures caused by a non-neurological condition which are difficult to distinguish clinically [3]. This often results in misdiagnosis of epilepsy. As many as 30% of epileptics may have been misdiagnosed, with 25% having syncope-associated convulsive episodes [2]. Many cardiovascular problems may cause syncope, complicated by abnormal movements caused by generalized cerebral hypoxia, appearing similar to epileptic seizures, leading to a misdiagnosis of epilepsy [3]. Epileptic seizures are unprovoked and usually present with pre-ictal and post-ictal symptoms [4]. Presentations of epileptic seizures are varied and there are many imitators, including convulsive syncope, acute symptomatic seizures, and psychogenic events. Furthermore, the phenomenon most commonly mistaken for a convulsive seizure is syncope. A convulsive episode lasting for seconds only, with immediate return of consciousness is likely syncope, not epilepsy [4].

A Stokes-Adams attack is a transient abrupt collapse into unconsciousness due to a sudden but pronounced decrease in cardiac output caused by change in heart rate and rhythm, resulting in syncope with or without convulsions [1]. It is usually caused by a high-grade AV block (complete AV block and Mobitz type II AV block), pulseless ventricular tachycardia, and paroxysmal ventricular standstill, which is one of the rarest causes Stokes-Adams attack [1]. Ventricular standstill is a rare cardiac phenomenon that manifests as absence of ventricular activity despite normal atrial function, which appears as P waves, without accompanying QRS complexes on ECG [5,6]. It is often associated with conduction blocks, however, this may still occur without them [1]. Patients with ventricular standstill are at risk of profound bradycardia or asystole which may present with syncope or sudden cardiac arrest [6]. In addition, it may present as SAS, such as in our patient, which is a rare manifestation. Some reports have shown that ventricular standstill may present as seizure-like episodes [3,7]. In ventricular standstill, the cardiac circulation is practically arrested which may lead to generalized cerebral hypoxia, particularly reflex forms of syncope, which may appear like seizures [2,8]. Once recognized, immediate treatment with a temporary pacemaker and eventual insertion of a permanent pacemaker are necessary [1].

Table 1
Diagnostic tests and findings for patients presenting with transient loss of consciousness.

Diagnostic Test	Findings
Carotid sinus massage	Bradycardia, asystole, and/or hypotension that reproduce symptoms (in patients with clinical features of reflex syncope)
Orthostatic challenge	Symptomatic abnormal blood pressure fall
Active standing	Decrease in systolic blood pressure from baseline value $\geq$ 20 mmHg or diastolic blood pressure $\geq$10 mmHg, or a decrease in systolic blood pressure to $<$ 90 mmHg that reproduces spontaneous symptoms
Tilt testing	Reproduction of symptoms similar with clinical features of syncope
Autonomic function tests	
Valsalva maneuver	Absence of a blood pressure overshoot and absence of a heart rate increase during the Valsalva - > neurogenic orthostatic hypotension
Deep breathing	Blunted heart rate variability during deep breathing (<15 beats/minute)
24-hr ambulatory and home blood pressure monitoring	Nocturnal 'non-dipping' or 'reverse-dipping' blood pressure pattern
Electrocardiogram	
In-hospital monitoring (*for patients with high-risk clinical features*)	Arrhythmias associated with syncope
Holter monitoring (*for patients who have frequent syncope or presyncope - $\leq$ 1 episode/week*)	Arrhythmias associated with syncope
External loop recorders (*for patients who have inter-symptom interval for less than 4 weeks after the index event)*	Arrhythmias associated with syncope
Implantable loop recorders (*for patients with recurrent syncope of uncertain origin, patients with high-risk criteria in whom a comprehensive evaluation did not show any cause of syncope, patients with suspected epilepsy with treatment proven ineffective, patients with unexplained falls)*	Arrhythmias associated with syncope
Video recording	Clinical signs in relation to blood pressure and heart rate
Electrophysiological study	Correlation between syncope and: Asymptomatic sinus bradycardia (suspected sinus arrest causing syncope) Bifascicular bundle branch block (impending high-degree atrioventricular block) Suspected tachycardia
Cardiac biomarkers	Increased in cardiac syncope and structural heart disease
Echocardiography	Structural causes (i. e. aortic stenosis, obstructive cardiac tumors, pericadial tamponade, aortic dissection)
Exercise stress testing	Development of atrioventricular block after exercise (even without syncope); syncope immediately after exercise with severe hypotension
Coronary angiography	Presence of obstructive coronary artery disease (but this test alone is not diagnostic of the cause of syncope)

Stokes-Adams attacks are abrupt TLOC caused by cardiac pathology, hence, diligent history taking is important to determine its underlying mechanism. The TLOC can be mainly classified as syncope and its various forms (including cardiac syncope) and non-syncopal disorders such as epilepsy [9]. A 2008 review compiled various risk factors, patterns, and manifestations, that should be elicited in the history of patients presenting with TLOC. An accurate diagnosis can be obtained based on initial evaluation, including a thorough history from patients and eyewitnesses, physical examination, and ECG [9].

Convulsions caused by syncope and epilepsy are difficult to distinguish. The motor phenomena associated with these conditions have important diagnostic utility. In the seizure group, the myoclonic jerks were present significantly more often (100% vs 51%), lasted significantly longer (median in seconds 29.0 vs 3.6), and had a higher total number (median 48 vs 2) compared to the syncope group [10]. In convulsive syncope, myoclonic jerks are less (<10), in contrast with tonic clonic seizures (<20) [10]. Our patient's convulsive episodes lasted for only 10 seconds, with less number of myoclonic jerks.

In another study that described the semiology of tilt-induced reflex syncope in relation to the EEG changes, myoclonic jerking and tonic contraction were described as common semiology of syncope [11]. EEG findings during syncope showed either a "slow-flat-slow" or a "slow" pattern. General stiffening is a common manifestation of syncope which develops during the first slow phase, stays during flattening, and stops in the second slow phase, implying more profound circulatory changes – lower minimum blood pressure (BP), longer maximum RR-interval. Also, this group contained more cases with asystole and had a longer duration of loss of consciousness than the "slow" group [11].

As discussed in this report, TLOC from convulsive syncope is difficult to distinguish from non-syncopal forms like epilepsy and keen clinical eye is key to prompt diagnosis. Clinical features including danger signs, as well as mechanisms are important to note. Clinical manifestations of TLOC are usually derived from history taking. It has four specific characteristics: short duration, abnormal motor control, loss of responsiveness, and amnesia for the period of the LOC [12]. From the thorough history, if the TLOC is suspected to be syncopal in nature, initial syncopal evaluation is warranted. Physical examination, including supine and standing BP measurements. Based on these findings, additional examinations may be done when needed, including immediate ECG monitoring for suspicion of arrhythmia and echocardiogram when there is previously known heart disease [12], as performed in this case report. Other signs that can suggest a cardiac syncope on initial evaluation include symptoms during exertion, sudden onset palpitations immediately followed by syncope, family history of unexplained sudden death at young age, presence of structural heart disease or coronary artery disease, persistent bradycardia (<40 beats/minute) in awake state and in absence of physical training, unexplained systolic BP in the ED < 90 mmHg, and ECG findings of arrhythmia [12].

There are different diagnostic tests that are ideally performed for diagnosis of syncope as recommended in the 2018 ESC Guidelines (see Table 1) [12]. Some of these tests were not done to our patient, however, they are recommended to avoid unnecessary injuries and deaths. bundle.

The primary limitation of this report is the lack of simultaneous ECG and video-EEG monitoring during the attacks which is important in the diagnosis. Also, EEG documentation after the pacemaker insertion was also not done. Other diagnostic tests as previously discussed were also not performed. Despite these limitations, the ECG findings of complete left bundle branch block which eventually developed to ventricular standstill during the seizure-like episodes could point to a Stokes-Adams attack [1,3,6]. Moreover, the episodes have completely resolved after the pacemaker insertion which is required treatment for ventricular standstill although there is no clinical guideline available [1].

4. Conclusion

Awareness of distinction between seizures/epilepsy and convulsive syncope is important and may be life-saving. A good clinical history as well as simple non-invasive tests such as EEG and ECG are important in establishing correct diagnosis.

Author contribution statement

All authors listed have significantly contributed to the investigation, development and writing of this article.

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Data included in article/supp. material/referenced in article.

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Ethics approval

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Consent to participate

Not applicable.

Written consent for publication

Written consent was obtained from the patient before submission for publication of this case report. A copy of the written consent is available for review of the Editor-in-Chief of this journal.

Declaration of competing interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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