CASE REPORT

Silent Storm: Ventricular Standstill Masquerading as Seizure in Adams-Stokes Syndrome

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ABSTRACT

Paroxysmal ventricular standstill is one of the rarest but life-threatening cause of Adams-Stokes (A-S) syndrome. We present the case of an elderly female who experienced recurrent episodes of convulsions, described as tonic movements of all limbs followed by brief loss of consciousness, due to paroxysmal ventricular standstill. The diagnosis was confirmed when an episode of convulsions coincided with an ECG strip showing ventricular standstill, and the convulsions stopped once the arrhythmia resolved, as observed on the cardiac monitor. The diagnosis was further supported by the absence of additional convulsive episodes after the insertion of a temporary transvenous pacemaker, which corrected the arrhythmia. Adams -Stokes syndrome refers to a lack of brain perfusion caused by inadequate or inefficient cardiac contractility due to a cardiac arrhythmia. This can result in abnormal brain activity, leading to convulsions or syncope.

INTRODUCTION

Paroxysmal ventricular standstill (PVS) occurs when there is no electrical or mechanical activity of the ventricles despite a normal functioning atria. This translates to presence of p waves followed absence of qrs complex in an electrocardiography strip. In turn, this leads to decreased cardiac output and significant decreased cerebral perfusion even when the episode lasts only for few seconds which manifest as Adams-Stokes (A-S) attack with seizures and loss of consciousness.

The common ethologies for ventricular standstill includes ischemic heart disease, electrolyte imbalances, drug toxicity and structural heart diseases. In rare occurrences ventricular standstill has also been seen in patient with acute gastroenteritis due to exaggerated vagal tone.[4] The prevalence of ventricular standstill worldwide and in Malaysia in unclear as this is a rare cardiac arrhythmia occurring sporadically around the world. We present a rare case of seizures caused by Adams-Stokes syndrome secondary to paroxysmal ventricular standstill. Other causes of Adams-Stokes syndrome are cases of both tachyarrhythmias, such as ventricular fibrillation, and bradyarrhythmias, such as heart blocks. Ventricular standstill is comparable to ventricular fibrillation in terms of severity of the arrhythmia as it is associated with high mortality therefore it is essential to recognize this disease. The prognosis is poorer with patients with longer duration of PVS. Hence, patients with recurrent and frequent episodes of PVS require immediate attention to improve morbidity and mortality associated with the arrhythmia.

CASE REPORT

An 88 years old lady with no known medical illness presented with multiple convulsive episodes described as tonic movement of bilateral upper limb and lower limb associated with loss of consciousness which lasted for ten to fifteen second each episode which self aborted with no drowsiness or confusion following the convulsive episode. Further history from the patient revealed that she did not have a prior history of ischemic heart disease or hospital admissions for any reason. She had no significant drug history and was not on any medications, including beta blockers. She also strongly denied experiencing chest pain

or any symptoms suggestive of acute coronary syndrome during this presentation. Upon arrival to the Emergency Department (ED) of Hospital Tawau her vital signs were stable. The patient had a similar episode of tonic movement of bilateral upper limb and lower limb which resolved spontaneously at the ED. Initial impression by the emergency team was seizure for investigation, and immediate CT brain done together with routine blood investigations and ECG. The CT brain was normal. Thyroid function test showed T4 level of 15.60 pmol/L and TSH level of 1.54 mIU/L.

Analysis of electrolyte showed potassium level of 4.4 mmol/L, sodium level of 141 mmol/L, serum calcium level of 2.23 mmol/L, magnesium level of 1.04 mmol/L and phosphate level of 1.22 mmol/L. Other blood investigations were unremarkable as well. ECG showed prolonged PR interval (Figure 1). The initial impression was breakthrough seizure to rule out epilepsy with a differential diagnosis of cardiac syncope. Patient given intravenous phenytoin and planned for admission to cardiac coronary unit with continuous cardiac monitoring. In the ward the patient developed further frequent similar episodes of convulsion and repeated ECG noted to have complete heart block (Figure 2). Transcutaneous pacemaker was inserted. Despite being on a transcutaneous pacemaker with maximum output of 200 milliamps, the patient continued to have convulsive episodes lasting few seconds, cardiac monitor showed there were no electrical capture from transcutaneous pacemaker and cardiac monitor strip revealed ventricular standstill (Figure 3). Temporary Transvenous Pacemaker (TPM) was inserted uneventfully after urgent consultation with cardiology team and manage to obtain electrical and mechanical capture (Figure 4). The patient had no more episodes of tonic movements following TPM insertion. Phenytoin which was started in emergency department was off. Patient was transferred to Hospital Queen Elizabeth, Sabah, Malaysia, tertiary center for permanent pacemaker insertion.

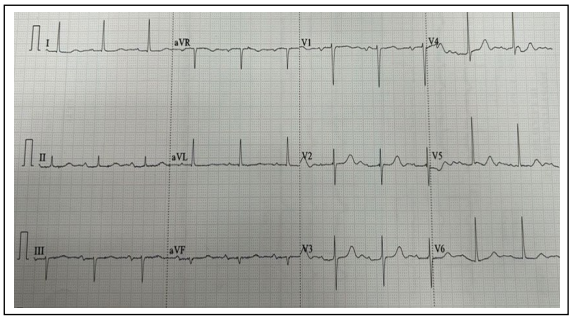

Figure 1 : ECG at Emergency Department showing prolonged PR interval, first degree AV block

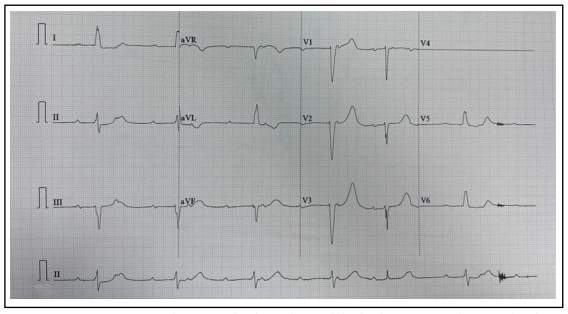

Figure 2 : ECG in ward showing third degree atrioventicular (AV) block

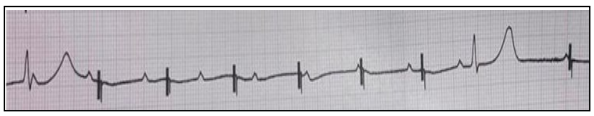

Figure 3 : Cardiac strip showing ventricular standstil and failure of transcutaneous pacemaker to have electrical capture

Figure 4 :ECG strip showing paced rhythm after transvenous pacing

DISCUSSION

Ventricular standstill is the absence of ventricular contraction due to the absence of impulse formation in ventricles or propagation of supraventricular impulse.[5] This translates to absence of ventricular rhythm with a normal atrial activity in an electrocardiography.[5] The exact pathogenesis of PVS is unclear. It is associated with conduction defects between the atria and ventricles. The etiology of ventricular standstill can be divided into primary causes which are due to structural, degenerative or idiopathic, while secondary causes include hypoxia, acidosis, electrolyte imbalance, hypothermia and drug induced causes.[1] Presentation of PVS can vary, ranging from being asymptomatic and incidentally detected during Holter monitoring, to presenting as seizures or syncope as part of Adams-Stokes syndrome, as in the case presented, and in rare instances, presenting as cardiac arrest requiring cardiopulmonary resuscitation. It is difficult to distinguish

seizures or syncope secondary to neurological conditions from those caused by Adams-Stokes syndrome, which has led to the over diagnosis of epilepsy. Approximately 20% of misdiagnosed epilepsy cases are due to cardiovascular causes.[9] In this case the patient's presentation of convulsive episodes presumed to be due to a neurological problem, and emphasis placed on a neurological workup, including a CT brain scan. A detailed history can help in making the diagnosis of Adams-Stoke syndrome which characterized by abrupt transient loss of consciousness (TLOC) from cardiac pathology. An immediate return of consciousness following a convulsive episode is due to cardiac syncope and not epilepsy.

Seizure secondary to epilepsy had significantly more myoclonic jerks (100% vs 51%) and had longer episodes (median in seconds 29.0 vs 3.6).[9] Other differentiating factor are myoclonic jerks were lesser (<10) in Adams Stokes syndrome compared to epileptic seizures (<20).[9] Our patient's convulsive episode lasted only for 10 to 15 seconds and only had tonic movement with no myoclonic jerks. "Slow-flat-slow" or "slow" pattens are seen in EEG findings during Adams-Stokes attack.[8] TLOC has four special characteristic pattern of history which are short duration, abnormal motor control, loss of responsiveness and amnesia for the period of LOC.[9] Investigation that should ideally be performed for diagnosis of syncope include carotid sinus massage, orthostatic challenges, electrocardiography, electrophysiological study, cardiac bio markers, echocardiography, exercise stress test and coronary angiography.[9]

Many of these tests were not done for this patient as we were in a resource limited centre and diagnosis made at the early stage with demonstration of symptoms correlating with observed ventricular standstill in cardiac monitor. Following identification of cause of the Adams Stokes attack, it is essential to treat the primary cause of this attack which in this case was paroxysmal ventricular standstill. Based on the 2018 ACC/AHA/HRS Guideline on the Evaluation and Management of Patients With Bradycardia and Cardiac Conduction Delay the management of a patient with PVS, which is a form of atrioventricular (AV) block, is to rule out structural heart disease, either through transthoracic echocardiography or other advanced imaging modalities, depending on the clinical suspicion of structural heart disease. In cases of complete heart block or advanced AV block such as in this case, the patient would require permanent pacemaker insertion.[10] Subsequently, decision on the use of a single or dual-chamber pacemaker and the placement of the pacing lead made based on factors such as heart function, frequency of pacing, and the presence of atrial fibrillation.[10] In this case, a temporary pacemaker inserted to urgently abort the PVS episodes and patient transferred to a tertiary cardiac centre for further management.

CONCLUSION

This case is a reminder to physicians that ventricular standstill may present as convulsive episode mimicking possible neurological disorder or epilepsy. Early recognition and prompt steps such as regular ECG and continuous cardiac monitoring would allow early diagnosis and initiation of appropriate treatment. Although ventricular standstill is a dangerous arrhythmia associated with high mortality, this case showed that prompt treatment with cardiac pacing reduces chances of mortality and resolves the symptoms.

CONSENT

Written and verbal consent were obtained from the patient for publication of this case report

CONFLICT OF INTEREST

The authors declare that they have no conflict of interest

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