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Syncope with atypical trunk convulsions in a patient with malignant arrhythmia

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ABSTRACT – Syncope is a condition often misdiagnosed as epilepsy. Syncope caused by cardiac disturbance is a life-threatening condition and accurate diagnosis is crucial for patient outcome. We present a case study of a 71-year-old woman who was referred to our epilepsy centre with a diagnosis of refractory epilepsy. We diagnosed convulsive syncope caused by malignant cardiac arrhythmia based on the presence of cardiac asystole lasting for 20-30 seconds, which was caused by sick sinus syndrome combined with third-degree atrioventricular block. The most prominent feature of this syncope was atypical trunk (abdominal or thoracoabdominal) convulsions, which were accompanied by other motor signs (head and eye deviation and brief jerks of the extremities). In the periods between attacks, all investigations, including standard 12-lead ECG and 24-hour ECG monitoring, were normal. This case study highlights the challenge in differential diagnosis of sudden loss of consciousness. [*Published with video sequences*]

Key words: syncope, epilepsy, arrhythmia, asystole, trunk convulsion, sick sinus syndrome, central pattern generators

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The misdiagnosis of epilepsy is a relatively common occurrence. Of patients diagnosed with epilepsy who were referred to epilepsy centres, 20-30% were found to be misdiagnosed (Benbadis, 2009). The most common conditions misdiagnosed as epilepsy were psychogenic non-epileptic attacks, followed by syncope (Benbadis, 2009). Five basic mechanisms to describe the genesis of syncope have been described: syncope may be neurally mediated, sometimes referred to as a reflex (vasovagal syncope, carotid sinus syncope, and situation syncope), caused by orthostatic hypotension, cardiac arrhythmias, structural cardiac or pulmonary disease, or mediated by the central nervous system (ictal bradycardic syncope) (Crompton and Berkovic, 2009). Cardiac arrhythmias, in particular, can cause life-threatening events.

We present a case study of a patient who was referred to our epilepsy

centre with a diagnosis of intractable epilepsy and later diagnosed with syncope and atypical trunk convulsions induced by malignant arrhythmia.

Case study

We present the case of a 71-year-old female who was treated for coronary artery disease, arterial hypertension, and type 2 *diabetes mellitus*. Her family and social history were insignificant. The first episode of unconsciousness occurred at the age of 70 without any triggering factors. Witnesses reported a sudden loss of consciousness, accompanied by brief jerks of the extremities and contractions of the abdominal wall. The patient was admitted to a local hospital, where physical examination and other investigations (basic laboratory examination, 12-lead ECG, 24-hour ECG monitoring, EEG, and brain CT) were performed. The laboratory examination and ECG monitoring were described to be normal. On EEG, there was a greater representation of theta waves in both temporal regions, but this was interpreted to be normal based on the age of the patient. Based on CT, there was only mild cortical atrophy with moderate ischaemic/degenerative changes of white matter. The frequency of seizures was approximately one seizure per month. After the second attack, antiepileptic medication with lamotrigine (LTG) was started, but was ineffective. Carbamazepine (CBZ) and levetiracetam (LEV) were then introduced without any significant influence on seizure frequency.

One year later, the frequency of seizures suddenly increased and started to appear daily. For this reason, the patient was hospitalised in a local hospital and then referred to our epilepsy centre with a diagnosis of intractable epilepsy. She was admitted to our video-EEG unit in the late afternoon for urgent care and during the first night, three habitual paroxysmal events were recorded. For video-EEG monitoring, standard scalp electrode positions (10-20 system), one-channel ECG, and simultaneous video recordings were performed.

The attacks always started with heart asystole, followed by diffuse slowing-down and attenuation of EEG, accompanied by muscle and movement artefacts (*figure 1*). Within approximately 10 seconds, the patient lost consciousness. Head and eye deviation appeared, followed by brief jerks of the upper and lower extremities, and subsequently distinct abdominal or thoracoabdominal convulsions developed. These convulsions lasted for 3-5 seconds. After spontaneous restitution of heart activity, the patient almost immediately regained consciousness without any signs of confusion, but nausea and occasional vomiting were present at the end (*video sequence 1*). We recorded one seizure in which only abdominal/thoracoabdominal contractions were present (*video sequence 2*). The duration of asystole ranged from 20 to 30 seconds. The patient was immediately admitted to our cardiology department. Based on the 12-channel ECG results, she was diagnosed with sick sinus syndrome, probably combined with third-degree atrioventricular block, and was urgently implanted with a Biotronic Talos DR pacemaker. After discharge from the hospital, the antiepileptic medication was withdrawn. The patient has now been free of syncopal attacks without any antiepileptic drugs for almost two years.

Discussion

This case study highlights the challenge in differential diagnosis of a loss of consciousness. An incorrect diagnosis may have severe consequences; the patient may not be treated appropriately, moreover, some antiepileptic drugs can be potentially

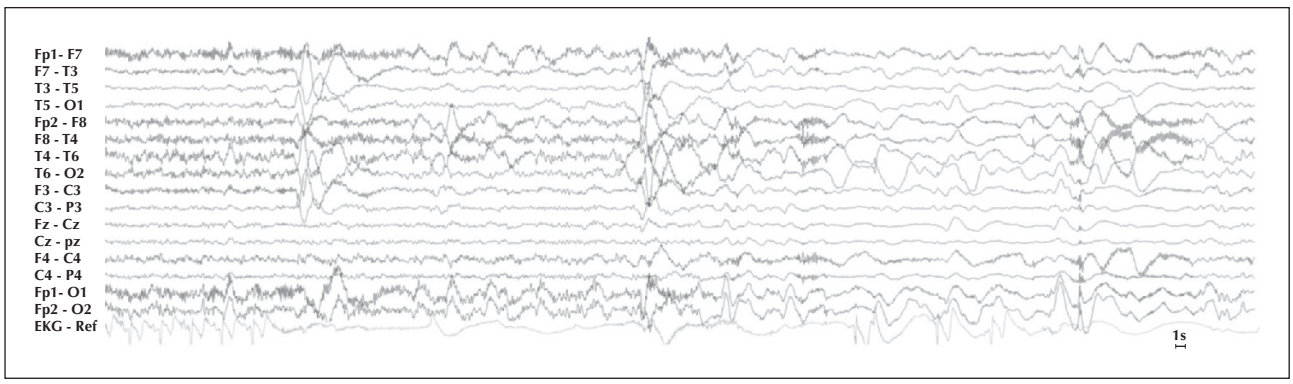

Figure 1. EEG during heart asystole.
The heart asystole is followed by the deceleration and subsequent attenuation of background activity. The EEG is contaminated with muscular artefacts.

harmful to patients with cardiac disease. The use of sodium channel blockers (for example, phenytoin and carbamazepine) in patients with cardiac-related syncope may worsen undetected heart disease through their negative chronotropic and dromotropic effects (Kenneback *et al*., 1991).

The percentage of syncopes with motor manifestations is reported to vary; some convulsions were reported in 12% of blood donors who experienced vasovagal syncope, compared with 45% of patients with malignant ventricular arrhythmias and implanted defibrillators (Lin *et al*., 1982; Aminoff *et al*., 1988). In a study by Lempert *et al*. (1994), almost 90% of healthy volunteers, in whom syncope was provoked artificially by hyperventilation, squatting, and Valsalva manoeuvre, presented with syncope with some type of convulsion. However, we presume that this is an over-representation, influenced by the study design.

The pattern of movement usually consists of multifocal arrhythmic jerks of the extremities, generalised myoclonus, and some additional movements such as head turns, oral automatisms, and righting movements (Crompton and Berkovic, 2009). In our patient, the most prominent motor characteristics were atypical trunk convulsions (we were unable to distinguish reliably between abdominal and thoracoabdominal contractions based on video recordings), a syncopal feature which has probably not been previously reported. Other similar reports include those of Ambrosetto *et al*. (2009) and Gasparini *et al*. (2011) who reported case studies of syncope with bipedal activity and gestural automatisms. The underlying pathological mechanism is believed to be the release, most likely disinhibition, of central pattern generators by anoxic cortical inactivation. Central pattern generators located at the subcortical level (mainly the brainstem and spinal cord) are responsible for rhythmic behaviour and are "normally" under neocortical control (Tassinari *et al*., 2009).

In the literature, the characteristics of patient history and description of attacks, which enable physicians to differentiate between syncope and epileptic seizures, as well as identify different aetiologies of syncope, are summarised (Sheldon *et al*., 2002; Crompton and Berkovic, 2009). We would like to point out that history or physical signs of cardiac disease should lead to a high suspicion of cardiac aetiology (Alboni *et al*., 2001). In our patient, a history of coronary artery disease was known.

It is important to stress that establishing a correct diagnosis can be complicated by the possible coexistence of syncope and seizure within one attack (Crompton and Berkovic, 2009). Attacks in which an initial syncopal event triggers an epileptic seizure are rare and occur mainly in children (Horrocks *et al*., 2005). Conversely, focal-onset seizures can cause bradycardia-induced syncope as a result of the disruption of cardiac autonomic neural discharges (Crompton and Berkovic, 2009).

This case study is an illustrative example of the importance of differential diagnosis in patients with loss of consciousness. The differential diagnosis is crucial mainly in cases with atypical course progression or clinical manifestation and in elderly patients with somatic comorbidities. In our patient, establishing a correct diagnosis and the subsequent implantation of a cardiac pacemaker were probably lifesaving. □

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Legends for videosequences

Video sequence 1

00:00 development of asystole
00:10 head and eye deviation to the left, followed by brief myoclonus of right-sided extremities
00:14 trunk convulsions
00:24 heart rate reappears spontaneously
00:29 patient regains consciousness and is oriented

Video sequence 2

00:00 development of asystole
00:10 trunk convulsions
00:20 heart rate reappears spontaneously
00:23 patient regains consciousness and is oriented

Key words for video research on *www.epilepticdisorders.com*

Syndrome: non epileptic paroxysmal disorder
Etiology: syncope (cardiac)
Phenomenology:
head deviation;
motor seizure (complex);
nonepileptic paroxysmal event
Localization: not applicable

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