Episodes of loss of consciousness in a patient with a background of cerebral venous thrombosis

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ABSTRACT – Episodes of loss of consciousness are common, even in young, healthy people, and can sometimes represent a diagnostic challenge. The main diagnosis to consider are syncope and epileptic seizures, both of which may have similar symptomatology such as dizziness, loss of consciousness, falls, or “convulsive” phenomena. We present the case of a young male patient with a background of two venous thrombosis episodes (superior vena cava thrombosis and cerebral venous thrombosis), attributed to protein C and S deficiency and complicated by high intracranial pressure. A lumboperitoneal shunt was performed and anticoagulant therapy was initiated. He did not experience any medical problems until several years later, when he suddenly began to develop frequent, repetitive, transient episodes of dizziness, followed by loss of consciousness. Simultaneous video-EEG and ECG performed during these events showed a typical pattern normally observed during syncope. Due to the absence of changes in heart rate or blood pressure, and taking into account his medical history, intracranial hypertension was considered as a possible cause of cerebral hypoperfusion. Cerebral arteriography demonstrated chronic thrombosis of all the cerebral sinuses, and the lumbar puncture an intracranial pressure of 47 mm Hg. The lumboperitoneal shunt was replaced and the patient has since not presented with any episodes. The use of simultaneous video-EEG and ECG is a reliable and efficient approach to differentiate between syncope and seizure and in this case, was the key to finding the cause of these episodes.

Key words: syncope, intracranial hypertension, venous cerebral thrombosis, loss of consciousness, seizure, video-EEG

Syncope is an abrupt, transient loss of consciousness and postural tone characterised by complete and spontaneous recovery, which is caused by sudden and global cerebral hypoperfusion (Kapoor, 2000). The pressure of blood flowing to the brain (the cerebral perfusion...
pressure (CPP) depends on two parameters: the median arterial pressure (MAP) and the intracranial pressure (ICP), which is the pressure inside the skull and thus in the brain tissue and cerebrospinal fluid (CSF).

Many disorders can cause syncope. The most common are classified as neurally-mediated syncope (vasovagal syncope), cardiac arrhythmias, and orthostatic hypotension (Brignole et al., 2004; Miller and Kruse, 2005). Uncommon aetiologies, such as metabolic or neurological disorders, should be considered when other causes have been ruled out (Jhanjee et al., 2006), including episodes of transient increase of intracranial pressure.

We present a case of a patient with recurrent loss of consciousness of unknown aetiology in which video-electroencephalogram (VEEG) monitoring indicated a diagnosis of syncope with uncommon aetiology.

Case report

A 37-year-old, right-handed man was referred to the Epilepsy Unit for the evaluation of episodes of unconsciousness that had begun two months earlier. He was born after an uneventful pregnancy, at term, to non-consanguineous parents, and his development was normal. There was no family history of epilepsy. He had suffered two episodes of venous thrombosis at the ages of 20 and 22, respectively. In the first episode, he experienced discomfort, dyspnoea, and facial redness; he was diagnosed with superior vena cava thrombosis and was prescribed anticoagulants for six months. Two years later, he started to have headaches, diplopia, and paresis of the sixth cranial nerve. Cerebral MRI showed a superior longitudinal sinus cerebral thrombosis and therefore anticoagulant treatment was reintroduced. Testing apparently revealed protein C and S deficiency and anticoagulant therapy was therefore continued indefinitely. During the following year, he remained asymptomatic, however, during a routine consultation, a bilateral papilloedema was discovered and a lumbar puncture demonstrated high intracranial pressure. Due to these findings, a lumbo-peritoneal shunt was inserted and during the subsequent 14 years, the patient did not experience any medical problems.

At the age of 36, the patient presented with paroxysmic, transient episodes of loss of consciousness. On presentation, these events were sporadic but they increased in severity and frequency, such that by the time he came to our attention, the episodes were occurring almost daily. During these episodes, he typically experienced malaise and dizziness, accompanied by generalised weakness. After several seconds, he lost consciousness but his eyes would remain open and the family reported that he stayed motionless, except for a discrete tonic flexion of his superior limbs. During some of the episodes, he yawned, snored, perspired profusely, and had bilateral mydriasis and facial redness. These episodes would last several minutes and afterwards the patient would recover consciousness with short or non-existent postictal confusion. These events would occur at any time of the day, whether sitting or standing, and the patient did not identify any triggers. At first, the episodes could be aborted at onset by lying down, but later they became unresponsive to postural change (see video sequence).

The patient’s neurological and physical examinations were normal except for a chronic bilateral papilloedema which was not resolved, despite the shunt. The results of various blood tests, cerebral MRI, interictal EEG, electrocardiography (ECG), echocardiography, and Holter-electrocardiography did not reveal any anomaly. During the tilt-test, and with the administration of nitro-glycerine, the patient experienced a loss of consciousness but reported that “this time it was different because I did not experience the usual symptoms”.

Different possible aetiologies were considered. Cerebral MRI venography (MRV) revealed a chronic partial superior longitudinal sinus thrombosis, and several EEGs did not show epileptiform activity. Nevertheless, epilepsy was considered probable and thus antiepileptic treatment was introduced (levetiracetam, up to 2000 mg a day). The patient experienced a reduction in the number of episodes, which did not completely disappear. VEEG monitoring recorded several events, identified by the family and the patient as “his usual epileptic seizures”. Arterial blood pressure, heart rate, and glycaemia were normal during the episodes. The patient recovered without confusion from each event, although he did not remember the episode. Simultaneous EEG showed a generalised, theta activity, followed by hypersynchronous, diffused, delta activity of high voltage with frontal predominance. When the patient recovered, this activity disappeared following a reverse sequence (each episode lasted for 5-8 minutes) (figure 1). In some of the episodes, a slow-flat-slow pattern was observed. The interictal EEG showed general and discrete slowing-down without any epileptiform abnormalities.

These EEG alterations are similar to the pattern observed during syncope but neither blood pressure nor heart rate changed, therefore cardiac arrhythmias, vasovagal episodes and orthostatic hypotension were ruled out. This led us to consider other less frequent causes of cerebral hypoperfusion, such as an increase in ICP. We performed a lumbar puncture which demonstrated an ICP of 47 mm Hg. Lumbar cisternography revealed that the lumbo-peritoneal shunt was not working properly. Cerebral arteriography showed partial and chronic thrombosis of all
**Figure 1.** EEG recording in an average montage during an epileptic seizure. (A) EEG showing the generalised theta activity at the beginning of the episode. (B and C) After a few seconds, this activity was followed by hypersynchronous, diffused, delta activity of high voltage with frontal predominance. (D) When the patient recovered, the activity disappeared and the EEG normalised, following a reverse sequence. There were no alterations on the ECG.
the cerebral sinuses and of both of the internal jugular veins. Venous return occurred through the vertebral plexus and external facial veins (figure 2). The superior cava vein, the brachiocephalic trunk, and the subclavian vein were also thrombosed, as seen on chest computed tomography.

Because the thrombosis was chronic (as demonstrated by significant collateral circulation), fibrinolytic endovascular treatment was rejected. The lumbo-peritoneal shunt was replaced and since then, the patient has been free of episodes. A 24-hour period of VEEG monitoring performed one month later was completely normal. Antiepileptic treatment was slowly withdrawn. A new coagulation test did not show protein C and S deficiency, but instead high levels of anticardiolipin antibodies were
Venous cerebral thrombosis and syncope

Figure 2. Cerebral arteriography: chronic and partial thrombosis of all cerebral sinuses with significant collateral circulation through the vertebral plexus, external facial vein, and diploic veins.

repeatedly found. Upon further questioning, the patient revealed a history of recurrent episodes of arthritis, mouth and genital ulcers, and thrombophlebitis. Taking into account the results of the clinical tests, and in the context of the new clinical data, Bechter disease was suspected and immunosuppressive therapy was initiated in addition to anticoagulants.

Discussion

We present the case of a young male patient with a background of two episodes of thrombosis (a cerebral venous thrombosis and a vena cava thrombosis) who presented with frequent, repetitive, transient episodes of loss of consciousness.

Obtaining information about a paroxysmal event in which a patient is unconscious represents a challenge. Furthermore, the patient is often asymptomatic upon examination and test results can be normal. Epileptic seizures and syncope are the two aetiologies most often considered (Britton and Benarroch, 2006). In our case, VEEG monitoring with simultaneous ECG allowed us to record and analyse several of these events in which we identified an EEG pattern typically described during syncope. This pattern is characterised by the early appearance of slow, high-voltage activity in the theta range, followed by a further slowing-down in the delta range, and is considered to be similar for all types of syncope, regardless of aetiology (Brenner, 1997; Mecarelli et al., 2004). If the hypoperfusion continues, the electrocerebral activity disappears and a flat EEG recording appears (convulsive syncope may occur at this time). After the syncope, the EEG activity normalises, following a reverse sequence (Ammirati et al., 1998). In our case, the pattern observed during these episodes showed generalised, diffuse, theta activity, followed by increased amplitude and reduced-frequency, brain-wave activity (delta activity.) There was also a flattening of the electrocerebral activity (during 2-4 seconds) in the middle of this delta activity. This flattening may indicate a further cerebral hypoperfusion and is more commonly associated with cardioinhibitory syncope than vasovagal syncope.

What is surprising in our case is the longer duration of the episodes compared with what is reported in the literature (less than a minute). This might be because the majority of syncopes with EEG correlates described in the literature are cardioinhibitory syncopes or vasodepressor syncopes, and the aetiology in our case was different.

We hypothesized that our patient was suffering from episodes of significant and sharp increases in ICP due to further cerebral sinus thrombosis and/or lumboperitoneal shunt failure (plateau-waves). Plateau-waves peak during several minutes before decreasing spontaneously and in each episode, a decrease of cerebral blood flow leads to global cerebral ischaemia, hypoperfusion, and syncope (Hayashi et al., 1991; Chazal et al., 1984; Jedynak et al., 1984). The EEG pattern observed during the plateau phase is similar to that during syncope (Naquet and Bostem, 1964). The results of the cerebral arteriography and cisternography confirmed our hypothesis regarding the increase in ICP.

The initial normal MRI can be explained by the fact that the appearance of the thrombus on MRI within the dural sinus or cortical veins is variable and largely dependent on its age. The diagnosis of the later stages of cerebral sinus thrombosis (chronic or partially recanalized) may be confirmed by MRI in some cases, but often there is neither a clear thrombus signal nor clear absence of blood flow (Filippidis et al., 2009; Isensee et al., 1994).

Although the patient had been taking anticoagulants appropriately for many years, this treatment had not been sufficient to prevent further cerebral venous thrombosis. High levels of anticardiolipin antibodies and the patient’s history pointed towards Bechter syn-
drome. Although we cannot conclude that our patient had Bećhtë disease (the major criteria and one of the minor criteria were met), this was highly likely given the large number of commonly associated symptoms presented (including dural sinus thrombosis) (Erdem et al., 2006).

**Conclusion**

Two issues are of particular interest in this case. Firstly, the usefulness of performing VEEG monitoring with simultaneous ECG in all patients who experience episodes of loss of consciousness of unknown cause, and secondly, the need to study all possible causes of cerebral hypoperfusion in a patient with repeat syncope.

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**Legends for video sequences**

**Video sequence 1**

The video shows a typical seizure of the patient. The event starts when the patient feels faint, warns the family, and lies down. He then loses consciousness, remains still, apart from occasional movements of his left hand and leg, and yawns occasionally. The episode lasts for seven minutes after which he recovers with no postictal period.

The EEG shows a generalised theta activity, followed by hypersynchronous diffused delta activity of high voltage with frontal predominance. A slow-flat-slow pattern is also observed. This activity disappears following a reverse sequence when the patient recovers.

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

Syndrome: non epileptic paroxysmal disorder

Etiology: venous thrombosis

Phenomenology: dizziness;
nonepileptic paroxysmal event;
consciousness (loss)

Localization: not applicable

**References**


