Spinal algetic-tonic seizures manifesting as paroxysmal "positive" Brown-Séquard syndrome

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ABSTRACT – We report on a patient suffering from symptomatic spinal attacks in the form of a paroxysmal "positive" (algetic-tonic) Brown-Séquard syndrome. A cervical cord lesion, presumably inflammatory-demyelinating in origin, was identified as the morphological correlate of these attacks. Their pathogenesis is discussed in the light of similar case reports from the literature. For the first time, this rare type of seizure is published with a video documentation. It may deserve consideration in the differential diagnosis of otherwise unexplained paroxysmal events that present in an "epileptic" manner.
[Published with video sequences]

Key words: spinal seizures, tonic spasms, paroxysmal dystonia, Brown-Séquard-syndrome, tonic seizures, algetic seizures

Tonic paroxysmal attacks – synonymously termed "tonic seizures" or "tonic spasms" or "paroxysmal dystonia" (Tranchant *et al.* 1995) – of spinal origin can be a relevant alternative diagnosis in paroxysmal events presenting in an "epileptic" manner, *i.e.*, among other features, in a monomorphic fashion and responding to drugs such as carbamazepine. We present the case of a patient, with video-documented attacks of obviously spinal origin.

Case study

This 41-year-old man with arterial hypertension but otherwise unremarkable medical history presented with a one-year history of monomorphic attacks occurring about eight times per day. Apart from rare occasions of paroxysmal temporal tingling in his left hip (the first element of the full-blown events), he has been free of these attacks since the introduction of carbamazepine treatment. Getting up with moving of the legs *(see video sequence 1)*, as well as hyperventilation *(see video sequence 2)* seemed to induce the attacks.

During prolonged video-EEG monitoring under carbamazepine withdrawal, two of these events with the following semiology were documented *(see video sequences and figure 1A)*: initially, paresthesias in his left hip; three seconds later, additional burning pain in the right hip which descends downwards through the right leg; three seconds later, additional left-sided tonic contractions in the
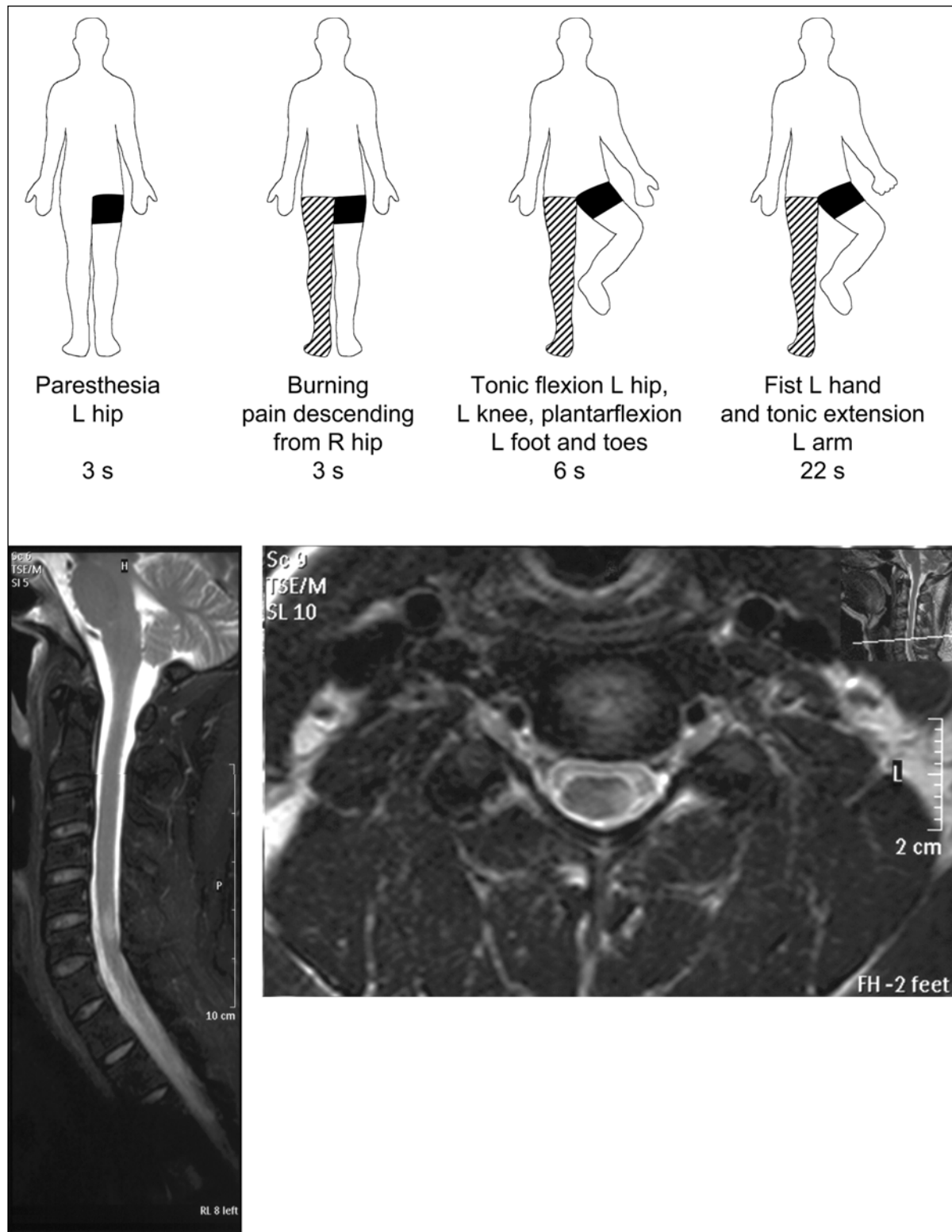
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Figure 1. A) Schematic representation of the course of the seizure symptoms in our case. **B)** Sagittal and **C)** transversal T2-weighted MRI images display a lesion within the left-sided and dorsal aspect of the spinal cord at the level of C6.

following sequence: plantar flexion of foot and toes - flexion in knee and hip; six seconds after onset of these contractions, making a fist plus tonic extension of the left arm; after another 22 seconds, the features resolve gradually over several seconds. There was neither cognitive impairment nor any epileptiform or rhythmic activity apart from muscle artifacts on surface EEG.

The patient's attacks were initially considered to be epileptic in origin. However, the lack of evidence of any pathological features of the brain substance (such as a cortical neurological deficit, an EEG abnormality or demonstration of a lesion on brain imaging) led to his transferal to this tertiary epilepsy center for confirmation of the epilepsy diagnosis.

Diagnostic work-up one year after onset of these attacks revealed that the patient's neurological status was normal. On spinal MRI, a left-sided, T2-signal intense, non-enhancing intramedullary lesion between C5 and C6 without mass effect was detected *(figure 1B, C)*. Thoracic and lumbar spine MRI were normal as were brain MRI, supra-aortal MRI angiography and ultrasound examinations (not shown). "Ictal" and "interictal" EEG recordings were always free of epileptiform features. However, during attacks a heart rate increase (from 75 to 120 bpm) was documented. The cortical latencies of somatosensory-evoked potentials (SEPs) indicated slowed dorsal column conduction in between the left tibial nerve and C6/C7. Motor-evoked potentials (MEPs) elicited by transcranial magnetic stimulation indicated a delayed central motor conduction time to the left arm and leg. Visual-evoked potentials and cerebrospinal fluid standard parameters were normal. A suggestive injection of 0.9% sodium chloride (Reuber *et al.* 2002) did not provoke a psychogenic response.

Follow-up has covered a period of two years since the onset of symptoms. The patient is free of attacks on 1 500 mg of oxcarbazepine. No new neurological deficits have evolved.

Discussion

This patient suffered from spinally originating "tonic seizures" [other synonymous terms: "tonic spasms", "paroxysmal dystonia" (Tranchant *et al.* 1995), "spinal sensorimotor seizures" (Osterman and Westerberg 1975), or "painful tonic seizures" (Shibasaki and Kuroiwa, 1974)] in the sense of a paroxysmal "positive" Brown-Séquard-syndrome [or a "Brown-Séquard-syndrome in reverse", as previously stated (Osterman and Westerberg 1975)]. To the best of our knowledge, this is the first case of this kind published with accompanying video documentation of habitual attacks. The semiology of such attacks is not always absolutely identical. The hip and knee flexion, which is documented in this case, is apparently less

frequently observed than an extension or posturing motor pattern (Osterman and Westerberg 1975, Shibasaki and Kuroiwa 1974). The causative lesion in our patient is obviously the one detected in the spinal cord. Its nature could not be definitely clarified. MRI and ultrasound studies do not support the notion of an ischemic event. The most likely diagnosis is transverse myelitis (since the diagnostic criteria of multiple sclerosis [MS] are not fulfilled). This diagnosis is supported by the fact that according to the literature, inflammatory demyelinating CNS disease (in the form of established MS or as a presenting symptom) is by far the most frequent cause of this type of paroxysmal events [for a survey of the published case reports with new patients, see (Tranchant *et al.* 1995)]. A relatively large number of cases have been observed in Japan with its well-known preponderance of neuromyelitis optica among the inflammatory demyelination disorders (Shibasaki and Kuroiwa 1974). Despite the seemingly "epileptic" features of these attacks –short duration; monomorphic semiology; induction by hyperventilation; the ictal tachycardia; and the response to anti-epileptic drugs–, epilepsy in the most general meaning of the word can clearly not be diagnosed. Alternative diagnostic hypotheses (such as psychogenic attacks) could also not be corroborated.

The pathogenesis of obviously spinally originating paroxysmal events as described here is still under debate. An epileptic seizure is usually defined as a transient occurrence of signs and/or symptoms due to abnormal, excessive or synchronous neuronal activity in the brain (Fisher *et al.* 2005) as opposed to lower levels of the nervous system. Nevertheless, an "epileptic" or "positive" or "excitatory" event within the spinal grey matter has been proposed by several authors as underlying a tonic spasm [for a thorough review, see (Osterman and Westerberg 1975)]. Reports on subcortically-originating epileptic attacks have addressed cases of epilepsy caused by hypothalamic (Berkovic *et al.* 2003) hamartoma or ponto-medullar (Arzimanoglou *et al.* 1999) and – although not universally accepted - by lesions of the brainstem and diencephalon (Wieser *et al.* 1998). Alternatively, an ephaptic activation of axons, which spreads transversely within demyelinated areas of the spinal cord, has been proposed as the underlying mechanism (Osterman and Westerberg 1975) – in the present case, this activation would have occurred strictly within the left half of a transverse section of the spinal cord. It may be noted that massive depolarisations, apart from synaptic events, can also be transmitted via membranes, from one dendrite to another. Gap junctions support this effect (Jefferys 1995a, Jefferys 1995b). Similar phenomena are found in terms of ephaptic events on peripheral nerves. At present, it is probably not possible to differentiate a tonic spasm from a "true" epileptic seizure in the spinal cord. □

Legend for video sequences

Two habitual attacks of the patient, apparently provoked by leg movements *(sequence 1)* and hyperventilation *(sequence 2)*. Apart from muscle artifacts, EEG activity is normal throughout the events. Note the increase of the heart rate in both attacks. For a full description of the events see manuscript.

EEG channels are shown at right.
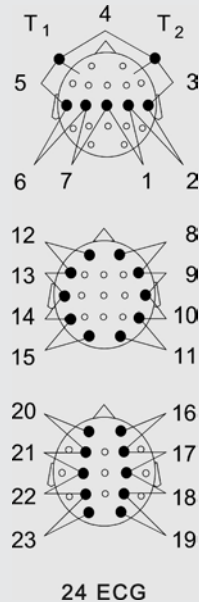
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