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Rapidly progressive cognitive impairment with neuropsychiatric symptoms as the initial manifestation of status epilepticus

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ABSTRACT

The purpose of this article is to describe the clinical and electroencephalographic features of patients diagnosed with non-convulsive status epilepticus (NCSE) with uncommon cognitive and behavioral involvement. We present two cases with sub-acute cognitive impairment and neuropsychiatric disorders (including anxiety and transient behavioral changes) as their first manifestation of NCSE. A neuropsychological profile demonstrated executive dysfunction. In addition, the neurological examination revealed automatisms and 24-hour video EEG showed epileptiform activity. Although neuroimaging studies showed frontotemporal abnormalities, both neurophysiological and cognitive improvement after specific antiseizure drug treatment confirmed the diagnosis of non-convulsive status. Theoretical considerations between mental status changes and focal epilepsy will be reviewed. Our cases raise awareness of the importance of considering NCSE, a treatable condition, in the differential diagnosis of rapidly-progressive cognitive impairment with neuropsychiatric symptoms.

1. Introduction

Status Epilepticus (SE) is a medical emergency with significant morbidity and mortality, affecting mainly people with epilepsy on low-dose antiseizure drugs [1]. According to the ILAE, SE is defined either as a continuous seizure lasting 5 min or more, or two or more sequential seizures without full recovery of consciousness between them [2]. The prevalence of SE in epilepsy patients ranges from 1 to 16%. In the United States, its incidence is 6.2–18.3 per 100,000 [3].

Cognitive and emotional disorders in epilepsy patients are common and have an impact on quality of life and adaptive social behaviors. Chronic epilepsy, including an accumulation of single attacks, may lead to neurophysiological consequences, as well as mental decline [4,5]. It is well known that cognitive dysfunction can be associated with drug-resistant epilepsy. Additionally, several studies indicate that SE may generate cognitive impairment through selective neuronal loss in vulnerable brain regions [6].

An increased prevalence of psychiatric disorders in epilepsy patients has been reported in epidemiologic studies, compared to the general population [7]. It is likely that focal areas involved in generating seizures interfere with brain structures which control emotional behavior. For instance, memory and anxiety disorders have been related to dysfunction of the hippocampus and amygdala in temporal lobe epilepsy, while executive and dyscontrol/impulsivity problems have been related to frontal lobe epilepsy [8]. The bidirectional relationship between behavioral disorders and epilepsy is probably a combined consequence of emotional side effects from antiseizure drugs, psychosocial factors, and a common neurobiological mechanism. Affective disturbances associated with non-convulsive status epilepticus (NCSE) have previously been reported by Geier and Profitlich [9,10]. Emotional symptoms in these patients includes dysphoria, irritation, and anger, as well as agitation, increasing anxiety and panic attacks. Depression and suicidal ideation have been apparent as well.

Whereas convulsive SE is a medical emergency, NCSE can be insidious, and remain undiagnosed for prolonged periods of time. Taking into account the infrequent primary presenting symptoms and the challenges associated with the diagnosis, there are few clinical reports which provide a sufficient description regarding cognitive and psychiatric manifestations as the initial manifestations of NCSE. The mental status changes may mislead the clinician to alternative diagnoses in the absence of physical signs suggesting seizures. We present two cases in which cognitive and behavioral involvement, as well as uncommon neuropsychological findings, were the first clinical manifestation of NCSE. Specific neuropsychological and mental tests were applied prior to, and following treatment, and suggest a causative role for sustained seizure activity.

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2. Case 1

2.1. Medical history and clinical features

A fifty-two year old male was admitted to the emergency department (ED) and was assessed by our neurology service (Hospital Mayor Mederi - Bogotá, Colombia) for a first episode of guttural sounds, eyeballs rolling back, hypersalivation, and generalized rigidity. The patient also had loss of consciousness during the attack with full spontaneous recovery, having over 5 min of amnesia following the event. The patient had a history of anxiety disorder four months prior, and obstructive sleep apnea (OSA) two months ago. A family member also reported jerking movements while asleep. These had worsened in the last week, and they persisted somewhat during wakefulness. His previous studies included normal brain CT and MRI scans, normal EEG and a polysomnogram study showing OSA (apnea-hypopnea index (AHI) 27.3; average desaturation 85%). He was on a SSRI antidepressant medication (Sertraline), 50 mg per day.

On neurological examination he was alert and oriented, having disorganized and incoherent thought content, hypoprosexia with defective fixing attention on a stimulus and bradylalia. His physical exam showed no cranial nerve abnormalities, no sensory or motor deficits, and no meningeal irritation signs. Initial neuropsychological assessments (Table 1) before antiseizure drugs (ASDs) showed low performance in both episodic declarative memory and verbal semantic fluency, according to the RAVLT and Isaac tests respectively. A dysexecutive syndrome was suggested by low performance in procedural and working memory, according to the WAIS III test.

2.2. Paraclinical tests

A brain MRI showed bilateral mild atrophy of the temporal lobes (Supplementary Fig. 1A). He had a non-reactive VDRL, and normal vitamin B12 and folic acid. There were no relevant findings in his CSF study (neither infectious nor inflammatory changes). Twelve seizures were recorded during the 24-h video-EEG monitoring. Electrographic seizures were manifest as rhythmic delta and theta rhythmic frequencies with amplitudes of 30-to 60-uV activity arising from rhythmic activity of 30–60 μV increased in frequency and amplitude arising from the right fronto-central region subsequently involving a bihemispheric field. This activity was correlated with hand automatisms of 20 to 40 s in duration (Fig. 1B). The paroxysmal events were followed by left gaze deviation. Background suppression of the EEG activity with a symmetric posterior dominant rhythm of 9 Hz was observed during the postictal state.

2.3. Treatment

Given the evidence of seizure activity, a phenytoin loading dose of 1200 mg was given intravenously (IV) and followed by 300 mg Oral Administration (P.O.) daily; 2000 mg of IV valproic acid was added and followed by 500 mg P.O. every 12 h.

2.4. Outcome and follow-up

Given the adequate clinical response, the antiepileptic medication were switched from the IV to oral route, and sertraline was discontinued. The patient showed significant clinical improvement with coherent thought content and euprosexia, and full resolution of both seizure activity and anxiety. Compared with the initial neuropsychological profile, there was improvement in procedural (below the 90th percentile), working memory and episodic declarative memory. On the other hand, impairment in verbal semantic fluency remained Table 1 after ASDs. Follow up examination two months later revealed that the patient was seizure-free, and return to his baseline work activity. A second video EEG showed no seizures, but right fronto-central cortical dysfunction and rhythmic slow theta activity at 6 Hz was found (Fig. 1C).

3. Case 2

3.1. Medical history and clinical features

A sixty-two year old female was admitted to the ED with a 10-day history of aggressive behavior, working memory impairment and transient dyscognitive episodes. Over the last week she had complained of moderately-strong tension-type headaches.

She had a history two years prior to admission, of a right middle cerebral artery aneurysm clipping complicated by a right temporal

Table 1
Neuropsychological (NP) findings before and after ASDs in patients with focal NCSE.

NP test	Case 1			Case 2		
	Pre ASD	Early post ASD[a]	Late post ASD[b]	Pre ASD	Early post ASD[a]	Late post ASD[b]
WAIS III - Digit Span	Digits forward: 4 Digits backwards:2 Raw score:7 Scaled score: 4	Digits forward: 4 Digits backwards:2 Raw score:7 Scaled score: 4	Digits forward: 4 Digits backwards:4 Raw score:7 Scaled score: 6	Digits forward: 3 Digits backwards:2 Raw score:6 Scaled score: 5	Digits forward: 5 Digits backwards:2 Raw score:7 Scaled score: 5	Digits forward: 4 Digits backwards:2 Raw score:7 Scaled score: 5
Isaac test set	13 (Es: 29)	11 (Es: 29)	11 (Es: 29)	32 (Es: 29)	27 (Es: 29)	29 (Es: 29)
ROCF	Nt	Nt	Nt	Score 36, Es: 31.19 (SD 3,68)	Score 36, Es: 31.19 (SD 3,68)	Score 36, Es: 31.19 (SD 3,68)
Stroop test	Nt	Nt	Nt	word (w): 86; color (c): 56; word-color (wc): 22; Es: w:119 (SD:20) c: 79 (SD:14), wc:50 (SD:11)	word (w): 89; color (c): 50; word-color (wc): 29; Es: w:119 (SD:20) c: 79 (SD:14), wc:50 (SD:11)	word (w): 95; color (c): 62; word-color (wc): 32; Es: w:119 (SD:20) c: 79 (SD:14), wc:50 (SD:11)
RAVLT	5 stimuli retrieval in 3 trials (Es:25,2)	11 stimuli retrieval in 3 trials and 23 stimuli retrievals in 5 trials (Es:25,2)	14 stimuli retrieval in 3 trials and 25 stimuli retrievals in 5 trials (Es:25,2)	43 stimuli retrievals (Es: 47,7, SD: 7,7)	49 stimuli retrievals (Es: 47,7, SD: 7,7)	51 stimuli retrieval (Es: 47,7, SD: 7,7)
TMT	Nt	Nt	Nt	TMT A: 212, Es: 35,1 (SD:10,6); B: 275, Es: 77,7(SD:23,8)	TMT A: 79, Es: 35,1 (SD:10,6); B: 178, Es: 77,7(SD:23,8)	TMT A: 85, Es: 35,1 (SD:10,6); B: 169, Es: 77,7(SD:23,8)

Abbreviations: expected score (Es); Not Tested by fluctuating clinical condition of the patient (Nt); Non Convulsive Status Epilepticus (NCSE); Rey Osterrieth Complex Figure (ROCF); Rey Auditory Verbal Learning Test (RAVLT); Trail Making Test (TMT); Antiseizure Drug (ASD).

[a] Early NPtest after ASD treatment (early post ASD).

[b] Late NPtest after hospital discharge with oral ASD treatment (late post ASD).