Case Report

Prefrontal disturbances as the sole manifestation of simple partial nonconvulsive status epilepticus

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Abstract

We describe a case of frontal lobe epilepsy with rare nocturnal generalized tonic–clonic seizures and repeated prolonged episodes of altered behavior lasting 1 to 2 days. The changes consisted of poor organizational strategies, impaired set shifting, emotional indifference, reduced motivation, and impairment of emotional decision making. Memory and consciousness were undisturbed, as she was able to follow her profession as a teacher. During the episode, the EEG was marked by right frontal rhythmical spikes and waves with spread to the homologous left region. Behavioral abnormalities and EEG changes were successfully treated with intravenous diazepam. The clinical representation can be ascribed to alterations of prefrontal-subcortical circuits, especially the anterior cingulate and orbitofrontal circuits. This unique case is classified as simple partial nonconvulsive status epilepticus with prefrontal disturbances as the sole manifestation.

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1. Introduction

The term nonconvulsive status epilepticus (NCSE) includes several different epileptic conditions. Criteria for diagnosis are a diminished level of consciousness or other neurological deficits, electroencephalogram (EEG) changes, and, with important exceptions, response to antiepileptic drugs (AEDs). Setting aside comatose states in hypoxic, toxic, or metabolic encephalopathies, NCSE comprises two main categories: absence status and complex partial status [1]. Complex partial status epilepticus is mostly of temporal origin. Several cases have been published on complex partial status of frontal origin [2]. For both conditions, absence status and complex partial status, reduction or abolition of consciousness is mandatory by definition.

Further types of NCSE are characterized by ongoing sensible, sensory, vegetative, or psychic symptoms without disturbances of consciousness (aura continua or simple partial NCSE) [1]. We describe a case of NCSE of frontal origin without reduction of consciousness and with several features considered typical deficit symptoms of frontal-subcortical circuits.

2. Case report

A previously healthy 46-year-old primary school teacher and master of the local church choir had started having nocturnal generalized tonic–clonic seizures (GTCS) at the age of 15. Birth history was uneventful and psychomotor development normal. Neurological examination and formal neuropsychological tests were within normal limits. The EEG exhibited right frontal sharp waves without slow activities in that region. Repeated CT and MRI examinations yielded normal results. The diagnosis of right-sided
nonlesional frontal lobe epilepsy with rare nocturnal GTCS was made, and AED treatment was started. Despite obsti-
nate noncompliance with the AEDs, her seizure disorder remained mild with sporadic seizures for a year.

At the age of 36, she complained about episodes of changed behavior. She called these episodes “dawdling days.” Such days increased markedly in frequency to one per month after a gynecological operation in her 41st year. The most prominent feature of the episodes was carelessness with respect to daily demands. Originally rigidly conscious of her duties, on these days she felt she “had all the time of the world” and would begin other activities although she realized she was late for her lessons or a choir rehearsal. She felt comfortable and pleasant, started shopping tours, and bought expensive goods she could not afford. She was permissive in sexual activities with her partner, but she herself did not have an increased sexual appetite. She would continue an activity without adapting to changed circumstances; for example, she resumed cross-country skiing despite a stretch of snowless slope, which ruined her equipment. She never lost consciousness and remembered all the details. She was able to give lessons in school and to perform as a harpist in a concert. During the concert she realized some minor mistakes she had never made before but she did not care. Moreover, she was also able to correct her students’ tests. When she rechecked these corrections after normalization of her condition, she realized that she had correctly marked all spelling errors (“it goes automatically”), but her overall judgment was “too mild.” Known as an extraordinarily demanding person, her acquaintances found her unexpectedly friendly and calm. However, most of the behavioral changes were not noticed, even by her relatives. Only her partner realized them regularly as she found her “unnaturally friendly” and recognized that she reacted routinely, but sloppily in her habitual environment. The condition lasted hours or days and ended abruptly. She realized it immediately and was horrified when considering the consequences. She could not sleep the next night and slipped into a depressive mood.

During one of these episodes we were able to record an EEG (Fig. 1). She was under self-chosen therapy with pheno-
obarbital (serum level, 16 mg/ml) at that time. Her part-
ner had insisted on admission to the hospital because he was convinced that she was in an abnormal condition known to him from previous episodes. The patient herself also confirmed that this was a “dawdling day.” In a relaxed and friendly way she told us that she would never have come to the hospital without her partner’s advice. She iron-
ically complained about the interruption of a good time. Bedside neuropsychological testing before and during the EEG revealed no major abnormalities. No motor signs could be observed. She was able to follow sequential commands and to present a detailed report about her activities before and after the onset of the changed condition. She missed her school lessons and intended to go on a shopping tour together with her partner. On the basis of the ictal EEG (Fig. 1) the diagnosis of NCSE was made. We refrained from performing extensive neuropsychological testing during the NCSE because of the need for urgent treatment after diagnosis. The treatment, 20 mg diazepam

![Fig. 1. EEG recorded during NCSE. Continuous rhythmical 2.5–3/second spikes and waves over the right frontal region with spread to the left anterior region. Note the ongoing 8–9/second alpha rhythm.](image-url)
intravenously, resulted in an EEG change after 1 minute (Fig. 2), and termination of NCSE and normalization of the EEG (Fig. 3) after 10 minutes. Clinically, the patient was drowsy for 1 hour. Afterward she was in her habitual condition and relieved by the fact that a diagnosis of her “dawdling days” was possible. Although she had played down the behavioral abnormalities during NCSE, she now was able to seriously discuss the nature and therapeutic consequences of these events.

Standard psychometric tests were used in the interictal neuropsychological assessment. The patient participated with motivation and inconspicuous behavior. Language functions, verbal and figural episodic memory, psychomotor speed, and executive functions including category and phonematic fluency and the Trail B test were all in the average range. A computer-based measurement of attentional functions revealed normal reaction times on tasks of alertness and divided attention.

The patient was prescribed valproic acid, finally became compliant, and has been free of seizures and episodes of NCSE for more than 1 year.

3. Discussion

We have described a woman with simple partial NCSE with disturbances of higher cortical functions and preserved consciousness and memory. Several authors have described comparable features, namely, cognitive deficits [3], psychic seizures [4], frontal syndrome [5], ictal catatonia [6], and “type 1 NCSE” of frontal origin [7]. If consciousness was well preserved, Thomas et al. [7] classified these as simple partial NCSE.

The prominent symptoms of the case described resemble behavioral changes observed in patients with frontal lobe lesions. In addition to the control of motor and oculomotor movements, the frontal lobe serves higher cortical functions. Prefrontal regions are connected to the basal ganglia and the thalamus. A dorsolateral prefrontal circuit, a lateral orbitofrontal circuit, and an anterior cingulate circuit have been distinguished [8]. These prefrontal-subcortical circuits control executive functions, socially critical restraint, empathy, and motivation. Lesional abnormalities result in disturbances of cognitive and executive functions, personality changes, impaired motivation, and mood disorders, among numerous other neuropsychological deficits [8].

The symptoms of frontal lobe dysfunction in our patient were poor organizational strategies, impaired set shifting (continued cross-country skiing despite a snowless slope), emotional indifference (“unexpectedly friendly and calm”), carelessness as well as reduced motivation (“dawdling days”), and impairment of emotional decision making (expensive shopping). Other frontal functions like basic attentiveness, selection, sequence, maintenance of actions, as well as performance of learned routine skills, remained undisturbed as the patient was able to correct school tests,

![Fig. 2. EEG 1 minute after start of intravenous administration of 20 mg diazepam. Intermittent transition of spikes and waves into 10/second rhythmic activities over right frontal region superimposed by harmonic 20/second rhythms.](image-url)
teach school, and give a concert as a harpist. The peculiar ictal behavioral symptoms of our patient could therefore be ascribed to disturbances of prefrontal functions with accentuation of anterior cingulate (performance monitoring) [9] and orbitofrontal (changes in subjective emotional state and personality) circuits [10].

Besides the clinical symptomatology, the EEG was conclusive for a frontal origin (Fig. 1). We assume that EEG patterns similar to those recorded led to the prior episodes. The transition from rhythmical spikes and waves to high-frequency fast activity after intravenous administration of benzodiazepines (Fig. 2) is also observed in cases of absence status [11]. Rhythmic generalized alpha activity has been described during NCSE [12]. Niedermeyer et al. [13] reported one patient with decreased responsiveness during an episode characterized by “mu-rhythm status” in EEG. Repeated interictal EEGs of the latter case did not exhibit any focal slow activity. This unusual finding in focal epilepsies, the strict rhythmicity of spikes and waves during the NCSE and the transition to fast activity with intravenous benzodiazepines, together with normal neurological and structural findings, corresponds to the observations in type 1 NCSE of frontal origin [7] and is a reason to assume an idiopathic epilepsy.

Several attempts have been made to organize the bewildering semiology of seizure types of frontal lobe origin. All descriptions contain motor, autonomic, and affective symptoms, as well as impairment of consciousness in various combinations. The network organization and the rapid spread of epileptic activities within the frontal lobe make a close topographic correlation unreliable. Single seizures with pure prefrontal symptoms are not reported. In this regard, the case described here seems to be unique, although the patient exhibited prefrontal seizure signs only during the NCSE and not with single seizures. One might speculate that this seizure type escaped observation even by the patient herself and could be recognized only over a long duration as with NCSE, because the abnormalities became obvious only through the complex behavioral changes during daily life. Unrecognized seizures may act as an additional causative factor for interictal psychic changes. An influence of EEG discharges and short non-convulsive seizures on cognitive function or behavioral alterations has been postulated [14]. However, the behavior of our patient was inconspicuous outside the episodes of NCSE.

References


