Case Report

Non-convulsive status epilepticus with right arm apraxia: A case report

Barbara Ladisich a,b, Ferdinand Otto a, Lukas Machegger c, Waltraud Kleindienst a, Eugen Trinka a,d,e, Giorgi Kuchukhidze a,*

a Department of Neurology, Christian Doppler Klinik, University Hospital Paracelsus Medical University, Salzburg, Austria
b Department of Neuroradiology, Christian Doppler Klinik, University Hospital Paracelsus Medical University, Salzburg, Austria
c Department of Neurosurgery, Christian Doppler Klinik, University Hospital Paracelsus Medical University, Salzburg, Austria
d Centre for Cognitive Neuroscience, University of Salzburg, Austria
e Institute of Public Health, Medical Decision Making and Health Technology Assessment, University for Health Sciences, Medical Informatics and Technology (UMIT), Hall in Tyrol, Austria

Abstract

Non-convulsive SE (NCSE) is characterized by altered consciousness with or without slight motor manifestations or other phenomena such as aphasia, sensory, auditory, emotional, gustatory or other symptoms. A 69-year-old right-handed man developed the sudden onset of apraxia in his right arm. On admission, the patient was alert and well oriented. In his past medical history, an intracerebral hematoma (ICH) in the left temporo-parietal area was noted occurred five years before the current admission. An electroencephalography (EEG) showed rhythmic theta–delta activity with fluctuating frequency between 1.5 and 5 Hz in the left centro-parieto-temporal area, which promptly responded to the intravenous injection of 2 mg clonazepam and 1000 mg levetiracetam. Apraxia resolved completely and the EEG demonstrated intermittent non-rhythmic delta–theta slowing in the left temporo-parietal area. A cranial CT scan showed residual cystic encephalomalacia in the left temporo-parietal area due to the previous ICH. An MRI exhibited an old parenchymal defect in the left temporo-parietal area with a residual hemosiderin rim on the susceptibility weighted imaging (SWI) and no diffusion restriction on the diffusion weighted image (DWI). NCSE presented with right arm apraxia in our patient with a post-hemorrhagic residual parenchymal defect in the left temporo-parietal area.

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1. Introduction

Status epilepticus (SE) is defined as a condition, caused either by a failure of seizure termination or by initiation of mechanisms that provoke ongoing seizures [1]. It is a neurological emergency with possible serious consequences such as neuronal death, neuronal injury, and alteration of neuronal networks [1]. The diagnosis of SE is primarily based on clinical semiology and electroencephalography (EEG) changes [1]. Convulsive status epilepticus refers to excessive abnormal muscular contractions that classically occur bilaterally [2]. On the contrary, patients with non-convulsive status epilepticus (NCSE) demonstrate altered consciousness or behavior with or without slight motor phenomena [1,3–5]. According to the new definition and classification of SE by the International League Against Epilepsy (ILAE), NCSE may occur with or without coma. In comatose patients, symptoms of NCSE may be slight or non-existent (“subtle” SE) and therefore, an EEG plays an important role in diagnosing NCSE in such cases [1]. A NCSE without coma is subdivided into generalized, focal or unknown whether focal or generalized. Generalized forms of NCSE include typical, atypical or myoclonic absence SE seen mainly in patients with preexisting epilepsy [1]. Patients with focal forms of NCSE present clinically with sensory, visual, olfactory, auditory, emotional, gustatory, autonomic, psychic, experiential or other symptoms [3–5]. Focal forms of NCSE also include aphaemic SE [1].

In this report, we present an unusual case of NCSE manifested by right arm apraxia.

2. Case report

A 69-year-old right-handed man was brought to the emergency room of the Department of Neurology, Christian Doppler Klinik, Paracelsus Medical University of Salzburg, Austria by paramedics due to the sudden onset of apraxia in his right arm. The patient noticed early in the morning that he could not use his right arm properly; he was unable to carry out usual activities such as buttoning his shirt, tying his shoe- laces, and preparing his breakfast. He was disturbed by the fact that his right arm “did not obey him”. Prior to this episode, the patient had normal function of his right arm, was mobile and led a completely
independent life. On admission, the patient was alert and well oriented. In the patient’s history, an intracerebral hematoma (ICH) in the left temporo-parietal area was noted. ICH occurred five years prior and amyloid angiopathy was suspected as the etiology.

On neurological examination the patient demonstrated apraxia in his right arm — he failed to draw either a cross or circle with his index finger. The patient was unable to imitate how he brushes his teeth, combs his hair, drinks from a glass or uses a hammer. He understood the commands, was able to describe the movements, but could not carry them out and instead, he made some awkward movements. Right arm apraxia remained unchanged for about 2 h from the moment of the first neurological examination on admission to the time when the patient underwent an EEG after CT and MRI scans were performed where an acute stroke was ruled out.

The patient’s spontaneous speech was relatively fluent, but contained paraphasic and grammatical errors. The patient had difficulty finding some words and at times used neologisms. Comprehension, repetition and reading were intact. These elements of aphasia as well as homonymous inferior quadrantanopia to the right were unchanged since the ICH. There were no motor or sensory deficits and no Babinski sign was observed. Coordination was intact, tendon reflexes were normal and symmetrical. There was no apraxia in the patient’s left arm. A CT scan of the brain and the cervical spine showed no signs of fracture, new hemorrhage or early signs of ischemic stroke, but residual cystic alterations in the left temporo-parietal area due to the ICH. An MRI showed an old parenchymal defect in the left temporo-parietal area with a residual hemosiderin rim on a susceptibility weighted imaging (SWI) and no diffusion restriction in a diffusion weighted image (DWI) (Fig. 2). On a duplex sonography of the cerebral arteries, many small plaques with minimal effect on blood flow were detected. The ECG and routine laboratory results were unremarkable. On EEG, continuous rhythmic theta–delta activity with a fluctuating frequency between 1.5 and 5 Hz in the left centro-parieto-temporal area was demonstrated (Fig. 1A–B). There were no features of spatio-temporal evolution; no epileptiform discharges were recorded. The ictal rhythmic theta–delta activity on EEG resolved promptly after IV administration of 2 mg of clonazepam and 1000 mg of levetiracetam and transformed into low amplitude intermittent non-rhythmic delta–theta activity in the left temporo-parietal area and a sleep pattern (Fig. 1C). About an hour after treatment of NCSE, the patient woke up from medically induced sleep showing no signs of apraxia. His neurological status was the same as his baseline presentation that was documented in his medical report from five years prior. This included residual symptoms from the ICH: elements of sensory-motor aphasia and the homonymous inferior quadrantanopia to the right. The next morning, EEG showed intermittent non-rhythmic delta-theta activity in the left temporo-parietal area (Fig. 1D).

3. Discussion

We report an unusual case of NCSE manifest as right arm apraxia in a patient with rhythmic delta-theta activity in the left centro-parieto-temporal area on ictal EEG and post-hemorrhagic residual parenchymal defect on brain imaging.

Fig. 1. A and B. Rhythmic Delta-Theta activity on left centro-parieto-temporal region.
C. Non-rhythmic Delta-Theta activity in the left temporo-parietal region immediately after treatment with 2 mg of clonazepam and 1000 mg of levetiracetam. D. Intermittent non-rhythmic Delta-Theta slowing in the left temporo-parietal region the day after status epilepticus.

A. & B. Rhythmic Delta-Theta activity on left centro-parieto-temporal region.
C. Non-rhythmic Delta-Theta activity in the left temporo-parietal region immediately after treatment with 2 mg of clonazepam and 1000 mg of levetiracetam. D. Intermittent non-rhythmic Delta-theta slowing in the left temporo-parietal region the day after status epilepticus.
NCSE in non-comatose patients may have very heterogeneous and non-specific clinical manifestations such as acute confusion, aphasia, fear, acoustic and visual hallucinations. Therefore, an EEG is essential for diagnosing NCSE [1]. In our patient, the EEG fulfilled the Salzburg criteria for NCSE: continuous rhythmic theta–delta activity with fluctuating frequency and without definite evolution responded well to anti-seizure drugs [6]. As our patient had a de novo NCSE, there was no baseline EEG available. However, prompt resolution of the rhythmic EEG activity followed treatment with clonazepam and levetiracetam with return to his baseline neurological status that were highly suggestive of NCSE.

A cerebral CT scan and MRI showed a stable lesion compared with the scans from five years ago. No signs of a new ischemic stroke or hemorrhage were present.

Consciousness may be preserved in NCSE and the ictal semiology may present in the form of deficits in higher cognitive functions, such as apraxia in the case of our patient [1,3–5]. Cognitive deficits as ictal phenomena of SE may be under-represented in literature as diagnosis and treatment of SE (including NCSE) rarely occur in a setting where thorough neuropsychological testing is possible [4].

There are, however, reports on subtle behavioral or experiential presentations of NCSE, documented by neuropsychological examination [4].

Apraxia or dyspraxia is characterized by inability, in the absence of primary motor or sensory deficits, to correctly carry out learned purposeful movements such as brushing teeth or combing hair [7]. Apraxia develops due to loss or inaccessibility of a motor “program”, which “guides” the movement [7,8]. It may affect different parts of the body, face or limbs and is subdivided into ideomotor, ideational, constructional, oro-facial, oculomotor, limb-kinetic, speech apraxia, etc., depending on which part of the body and/or which “pathway” of executing motor action is affected [7,8]. Apraxia often develops due to lesions in parietal or frontal lobes as a result of stroke, brain trauma or neurodegenerative disease. Apraxia due to seizure or status epilepticus, as in our patient, is very seldom reported. In an observational study on ictal neuropsychological deficits, all six patients had signs of ictal ideomotor apraxia along with other cognitive deficits [4]. In our patient, ideomotor apraxia was not influenced by accompanied aphasia as the patient understood commands, was able to explain how to perform the action, but could not act out the movement. The elements of aphasia, which were residual and stable...
after the ICH from five years ago, were present the day after SE occurred, whereas the signs of apraxia completely disappeared shortly after the treatment with anti-seizure medication.

4. Conclusions

NCSE may manifest itself as right arm apraxia in a patient with post-hemorrhage residual parenchymal defect in a left temporo-parietal area.

Ethical publication statement

We confirm that we have read the Journal’s position on issues involved in ethical publication and affirm that this report is consistent with those guidelines.

Declaration of competing interest

None of the authors has any conflict of interest to disclose.

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