

The role of cognitive rehabilitation in limbic encephalitis

A case report

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Abstract

Rationale: Limbic encephalitis is a parenchymal inflammation caused by viral, bacterial, or other microbial and postinfectious agents, which is usually expressed by multifocal neurological signs and cognitive impairment.

Patient concerns: A 50-year-old female was admitted in postacute phase, at our rehabilitative Center, to undertake neuro-motor treatment for a period of 4 months.

Diagnoses: The patient was affected by limbic encephalitis. Clinical presentation revealed attention, memory and executive dysfunctions, as well as behavioral changes, emotional dysregulation and reduction of self-awareness.

Interventions: The patients received an intensive cognitive and motor rehabilitation training.

Outcomes: Neuropsychological assessment and magnetic resonance imaging were performed before and after rehabilitative training to evaluate the cognitive and cerebral changes induced by treatment. The patient showed an improvement in cognitive performances and behavioral aspects.

Lessons: The reducing cognitive deficits, especially memory deficits, could improve quality of life by using available cognitive resources.

Abbreviations: BDI = Beck depression inventory, CT = computed tomography, EEG = electroencephalography, ENB-2 = brief neuropsychological examination, HAM-A = Hamilton Anxiety Rating Scale, MRI = magnetic resonance imaging, NMDAR antibodies = N-methyl-D-aspartate receptor, ROT = reality orientation therapy.

Keywords: cognitive rehabilitation, limbic encephalitis, neuropsychological improvement

1. Introduction

Limbic encephalitis is a rare disorder that involved medial temporal lobe, sometimes associated with hippocampus and cerebellum atrophy and polyneuropathy.^[1] Diagnosis is typically performed by using clinical, laboratory, neuroimaging, and

electrophysiological evidences.^[2] Electroencephalography (EEG) is often abnormal and showed diffuse or focal epileptic activity in one or both hemispheres.^[3]

Limbic encephalitis leads to cognitive and behavioral progressive decline. It is characterized by amnesic syndrome, complex-partial and secondary-generalized epileptic crisis.^[4] Generally, patients show a rapid decline of short-term memory and severe deficits in learning and retention of new information and in remembering past events.^[5] Other cognitive symptoms may include aphasia and apraxia.^[4] Encephalopathy is also characterized by frontal behavioral symptoms such as irritability, depression, sleep disorders, epileptic crisis, and hallucinations,^[6] personality changes and deficits in problem-solving abilities.^[7] Encephalitis symptoms may evolve over a few days, weeks, or months.^[8] In several studies, use of external devices or mnemonic strategies seem to improve memory performance in encephalitis patients^[9,10]; for example, a camera that takes pictures automatically every 30 seconds was more effective than a written diary in improving autobiographical recall.^[11]

In addition, the use of a memory notebook was associated with improved memory strategies.^[12] However, there is no standardized protocol to cognitive rehabilitation in subjects with limbic encephalitis; it has been shown that the use of external devices to improve mnemonic strategies in cognitive rehabilitation.^[13] To date, only few studies describe the use of cognitive rehabilitative training in patients with postencephalitis.

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Informed Consent: The patient gave written consent to the study.

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The aim of this case report is to describe a specific neuropsychological rehabilitative treatment for verbal memory dysfunction observed after limbic encephalitis. We proposed a personalized, intensive and short-term training program by using multidimensional approach. We used a clinical approach that includes diagnostic and rehabilitative procedures based on psychotherapy and neuropsychological method, by using strategies to improve residual abilities. To evaluate the recovery of cognitive functions, the patient underwent magnetic resonance imaging (MRI) before and after rehabilitative training.

2. Case report

A 50-year-old female patient affected by inflammatory myelitis came to our attention in August 2016, due to the onset of inflammation of the perichirurgical and hyperpirexia area, she was hospitalized. Despite the administration of antibiotic therapy, symptomatology persisted. The patient underwent rachicentesis. She was positive for NMDAR (*N*-methyl-D-aspartate receptor) antibodies, despite computed tomography (CT) of the abdomen and pelvis excluded ovarian cancer (teratoma). Serum autoantibodies directed against dependent potassium voltage channels (VGSK, Voltage Gated K⁺ Channel) were found. The treatment with baclofen was discontinued

because of variable state of consciousness. In spite of the severe cognitive impairment, neurological deficits and psychiatric symptoms, the patient was admitted on November, in postacute phase, at our rehabilitative center, to undertake neuro-motor treatment for a period of 4 months.

The instrumental examinations confirmed the diagnosis of limbic encephalitis. Indeed, electroencephalography (EEG) results showed a typical epileptic pattern associated to intermittent rhythmic delta activity reactive to painful stimuli.

High-field- 3T MRI showed high-intensity signal on T2-weighted and fluid attenuated inversion recovery images of the hippocampus bilaterally. No abnormalities result from diffusion weighted imaging (DWI) and pathological enhancement after gadolinium administration were found (Fig. 1).

At baseline evaluation, the patient was awake but disoriented at temporal and spatial level, and she was appositive and slightly collaborating. In addition, the patient presented spastic paraparesis, a moderate control of head but no neck control.

Evaluations of motor and cognitive functions were performed at the beginning of the rehabilitative training (T0) and after 2 months (T1) the next follow-up was performed 8 months after discharge from the rehabilitative unit (T2).

The neuropsychological assessment was performed by using brief neuropsychological examination (ENB-2)^[14] (see Table 1).

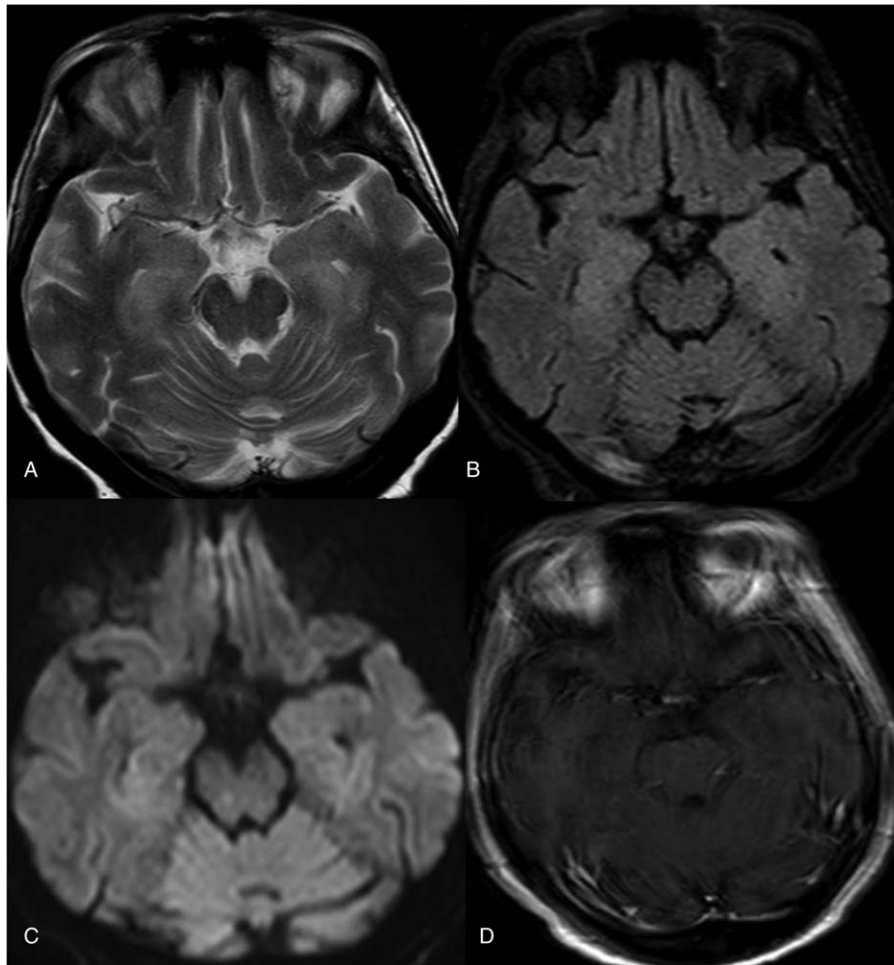


Figure 1. Magnetic resonance imaging (MRI) at 3 T showed high signal intensity on T2-weighted and FLAIR images of the hippocampus bilaterally. No signal abnormalities on DWI images and no pathological enhancement after gadolinium administration. DWI=diffusion weighted imaging, FLAIR=fluid attenuated inversion recovery, MRI=magnetic resonance imaging.

Table 1
Total score of neuropsychological assessment before, during and after rehabilitative training.

Test	T0	T1	T2	Cut-off
Digit span	2	4	5	4
Immediate recall prose memory	5	5	6	7
Delayed recall prose memory	7	9	9	11
Brown Peterson technique 10 sec	2	2	4	4
Brown Peterson technique 30 sec	3	3	4	3
Trail making test-A	129	99	90	86
Trail making test-B	NA	313	250	213
Token test	2	4	5	5
Word phonemic Fluency	5	6	8	8
Cognitive Estimation test	0	2	4	3
Intricate figures test	3	2	3	4
House figure copy	9	19	29	30
Daisy drawing test,	2	2	2	2
Spontaneous drawing	1	2	2	2
Clock drawing test	6	8	8	8
Ideomotor apraxia test	6	6	6	6
Total score	40	49	59	65

T0 (baseline); T1 (2 months after T0); T2 (8 months later after T0).

Table 2
Scores of clinical scales before, during, and after rehabilitative training.

Clinical cases	T0	T1	T2
Functional independence measure	52	60	75
Visual analog scale for Pain	8	5	2
Barthel index	40	40	40
Barthel index MOD	15	17	21
BRADEN	17	10	1
CONLEY	1	1	1
Beck depression Inventory	24	15	6
Hamilton anxiety rating scale	18	11	7

T0 (baseline); T1 (2 months after T0); T2 (8 months after T0).

We administered Beck depression inventory (BDI)^[15] and Hamilton Anxiety Rating Scale (HAM-A)^[16] to evaluate mood status. In addition, a pain scale (VAS) and a disability measurement scale (FIM) were used (see Table 2).

Neuropsychological evaluation (T0) showed a global cognitive impairment. In particular, the most critical aspects were disorientation on the spatial-temporal and autobiographic parameters. She showed alteration in attention, learning, and immediate and delayed memory. Language speech presented some abnormalities. Visuo-spatial abilities and executive functions were impaired (problem solving, planning and integration of information, decision making). Nevertheless, there were some residual cognitive abilities, such as automatic and imitation behavior, recognition and copying of simple visual stimuli and procedural memory. The mood was characterized by emotional lability. Neuropsychological rehabilitative training was carry out for period of 4 months, for a total of 24 sessions. Each session lasted 50 minutes. Cognitive rehabilitation was performed with exercises for temporal and spatial orientation, memory, and attention: reality orientation therapy (ROT), attention and vocal training, and pencil-paper exercises. In addition, external devices such as calendar, clock, family photos, city map, and diaries were used. Some exercises were focused on training of abstract reasoning (similes, metaphors, and proverbs). Overall, exercise difficulty gradually increased among the rehabilitative sessions. Each session was accompanied by emotional support and psycho-educational interventions to improve the problem-solving ability and her emotional self-regulation.

Motor rehabilitation was carried out twice a day for 6 days a week. This included standard physiotherapy exercises and verticalization by static bed for about 60 minutes. Two months after the rehabilitation treatment, at the second psychometric evaluation (T2), patient showed improvements on relationship's quality, language, and visuo-spatial immediate memory. A good recovery of cognitive and depression symptoms was performed at T2. Although the standing position is possible using a static bed, the patient improved her daily life autonomy and her quality of life.

MRI showed no physiological abnormalities on hippocampus bilaterally and both hippocampi were reduced in volume (Fig. 2).

3. Discussion

The limbic system includes thalamus, hypothalamus, amygdala and hippocampus, areas involved in the emotions, memory, social,

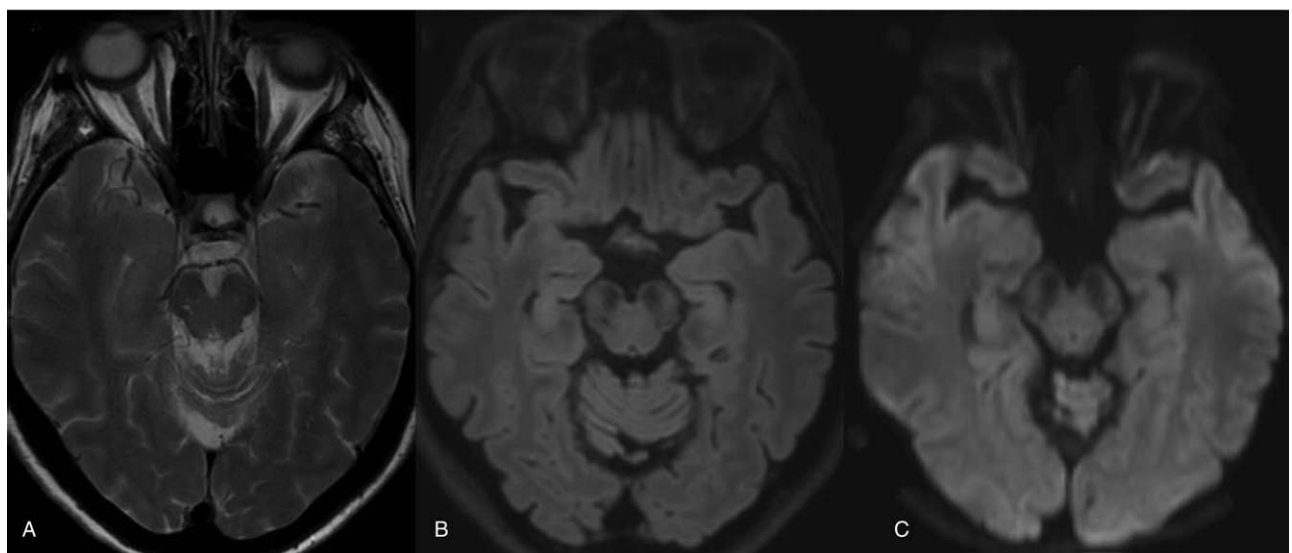


Figure 2. MRI showed no signal abnormalities of the hippocampus bilaterally and both hippocampi are reduced in volume.

and sexual behavior. Damages in these regions are associated with confusion, psychiatric symptoms, convulsions, memory deficit, apathy, anxiety, thought alterations, and myoclonus.^[17]

Memory deficits, after limbic encephalitis^[18] include autobiographic events.^[19] Cognitive impairment also affects the quality of life, social commitment, and daily activities.^[20] Compensatory approaches to memory rehabilitation minimize the impact of perceived disability.^[21]

In these patients, awareness deficit could obstruct the effective rehabilitative training worsening quality of life of the patient and their family members. For this reason, it is important to recognize these situations and develop appropriate tools to improve the patient's condition.^[22] Neuropsychological rehabilitation encourages metacognitive elaboration and improves motivational and cognitive processes.^[20–24]

Our patient rehabilitative program, improved cognitive functions and the enhancement of daily life activities through successful attention working memory and processing speed training.^[25,26]

Neuro-rehabilitative program was focused on strengthening of cognitive functions and on the development of alternative strategies. Objective measures showed an improvement of memory, attention, shifting, and metacognition domains. Moreover, relational aspects were improved, showing proactivity in seeking interactions with others. However, persisted some residual motor disturbances.

The multidisciplinary approach by using pharmacological therapy, motor, and cognitive rehabilitation were important for a global recovery of compromised abilities. Cognitive-behavioral psychotherapy contributed to enhancement emotional area of our patient. In particular, this treatment was based on reducing psychiatric symptoms, anxiety, and depression. Mood and behavioral changes contributed to improve autonomy and quality of life of the patient. Also caregiver's psychological support revealed useful for a returning to living his daily life. Despite rehabilitation objectives were achieved, a long-term healthcare assistance, with multidisciplinary interventions, by social services will be useful.

Author contributions

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Validation: Silvia Marino.

Visualization: Viviana Lo Buono, Caterina Formica, Daniela Florida, Concetta Pastura, Carmela Rifici, Giangaetano D'Aleo, Edoardo Sessa, Silvia Marino.

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