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Case Report

Cognitive Rehabilitation of Language and Speech Deficits in a Case of Herpes Encephalitis Sequelae

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ABSTRACT

Acute encephalitis is a debilitating syndrome of global brain dysfunction that develops as a rapidly progressive encephalopathy (usually in less than 6 weeks) which can be caused by brain inflammation, both systemic, secondarily affecting the brain, and primary in the brain itself. Patients of all ages may get affected by encephalitis which represents a significant burden to self, family, and society as they usually follow a more chronic course. Initial or predominant symptoms consist of cognitive deficits as well as behavioral changes, emotional dysregulation, and reduction of self-awareness. A 32-year-old, right-handed married female, belonging to MSES was brought for consultation in the post-acute phase after around one and a half years of treatment for Herpes Encephalitis. After further stabilization of her complaints of pain in her right side, tremors in her hands and seizures; was referred to the neuropsychology unit for rehabilitation in December 2014. The clinical presentation revealed a loss of expressive language skills, significant deficits in memory, regression in terms of behavioral changes, and seizures controlled with medicine. Based on the findings of the neuropsychological assessment, an individualized tailored rehabilitation plan was formulated using a multidimensional rehabilitative approach with the aim to promote language reconstruction to integrate the training into daily life communication and enhance the recovery of the patient. The patient received intensive cognitive remediation training for initial 6 months followed by spaced sessions for the next year. Neuropsychological and functional assessments were performed again after six months and the next one-year rehabilitative training showed an improvement in different cognitive functions specifically speech and language, and overall behavioral aspects. Also, maintenance of these treatment effects at around 5-year follow-ups is extremely encouraging.

Keywords: *Acute encephalitis, cognitive rehabilitation, Aphasia, Multimodel Intervention*

INTRODUCTION

Acute encephalitis is a debilitating syndrome of global brain dysfunction that develops as a rapidly progressive encephalopathy (usually in less than 6 weeks) (Venkatesan et al, 2013) which can be caused by brain inflammation, both systemic, secondarily affecting the brain, and primary in the brain itself. Patients of all ages may get affected by encephalitis and represent a significant burden to patients, families, and society (Imor et al, 2008; Vora et al, 2010) as they usually follow a more chronic course, and the initial or predominant symptoms of cognitive as well as behavioral changes, emotional dysregulation, and reduction of self-awareness.

The literature review indicated a dearth of studies describing cognitive and language impairments following encephalitis in adult patients whereas language and speech impairments have been suggested mostly in children with severe encephalitis. Consequently, there has been limited going-over to see possible impacts of improvement in speech on quality of life or participant's satisfaction as such information is necessary to minimize unnecessary healthcare spending and improve patient outcomes. Progressively, findings are supporting the significance of residual non-linguistic cognitive abilities in general in rehabilitation after acquired brain injury (Lin et al, 2022) and thus can be key predictors in the outcome of language functions also (Brownsett, 2014, Lin et al, 2022, Simic,2022). Memory, attention and working memory (WM) are prerequisite cognitive processes for language and other cognitive functions, while other language difficulties may stem from impaired motor function(Pennington, et al., 2009). This directs attention to the notion that aphasia rehabilitation must focus not only on content (language representations) but also on the process (non-linguistic cognitive structures) as aphasia treatments eventually aim to improve communication, rather than reduce language impairment (Carragher, Conroy, Sage, & Wilkinson, 2012).

Till date cognitive rehabilitative training in patients with post-encephalitis sequelae has not received much attention thus, no standardized protocol was examined extensively. The aim of this case report is to describe a specific neuropsychological rehabilitative treatment for a patient, focusing on her impaired language function and behavioral changes after encephalitis. An individualized tailored rehabilitation plan was formulated using a multidimensional rehabilitative approach based on psychotherapy and neuropsychological method with the aim to promote language reconstruction to integrate the training into daily life communication and enhance the recovery of patient by reintegration with her family and society.

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The Case:

Ms. R.M. (name changed for maintaining the confidentiality), a 32-year-old, right-handed married female, belonging to MSES was brought for consultation in the post-acute phase after around one and a half years of treatment for Herpes Encephalitis. After further stabilization of her complaints of pain in her right side, tremors in her hands and seizures; was referred to the neuropsychology unit for rehabilitation in December 2014. The clinical presentation revealed loss of expressive language skills, significant deficits in memory, regression in terms of behavioral changes, and seizures controlled with medicine. MRI finding was suggestive of large gliotic encephalomalacia in temporoparietal region with ex-vacuo dilation of the underlying left lateral ventricle. R.M. was active, and mostly cooperative during the sessions but not oriented to place, person and time. Her visual fields, auditory abilities and motor functions were grossly intact. The initial interview revealed that her spontaneous language production was non-fluent and would mostly respond by echoing the question itself. She had difficulty finding words and using conventional grammar in constructing simple spoken sentences correctly, and difficulty identifying or writing letters, numbers and objects. After much prompting and cueing, she would repeat her personal and family information vaguely. In her interaction and behaviour, she appeared like a child. The patient could not perform any structured neuropsychological assessment. It took around one month to make her cooperative and prepare for assessment and intervention. Written informed consent was obtained from the patient and her primary caregiver (mother) for carrying out assessments and interventions.

In September 2012, R.M. developed sudden onset of fever. Two days later she was taken to a private hospital as she had a severe headache, vomiting and disturbance of consciousness. Based on a detailed clinical history, complete general and neurological examination, routine blood and CSF analysis, and brain MRI (ill-defined hyperintensities seen in patients with white matter nonspecific ischemic changes) she was diagnosed and treated for acute encephalitis. After 25 days, when she was discharged from the hospital she was completely dependent for her basic and instrumental ADLs due to significant loss of speech and language, memory functions and personality changes with regressive behaviour. Although none of her admission symptoms appeared again in last18-month, her mother reported no significant improvement in expressive speech, reading, writing arithmetic skills and needs prompting/assistance for her ADLs. Before contracting encephalitis patient was working as senior manager in a corporate sector with a professional master's degree. Her competence in all three languages i.e. English, Hindi, and Punjabi was reported by her mother to be very high and was managing her personal and professional responsibilities very well. Due to above problems discord erupted in marital relation and mother was the only care giver. That was the added stressor and burden to their lives.

Assessments:

A range of personalized standardized assessments (Table 1) was carried out to ascertain the type and severity of aphasia and pattern on selected cognitive functions. The subtest from the Western Aphasia Battery (used both Hindi adaptation and English version) (WAB; Kertesz, 1982) which was found to be useful to quantify the severity of deficit, assess prognosis, monitor progression, and planning rehabilitation (John et al., 2017) and the Boston Naming Test 15 item short form (Kaplan, Goodglass, & Weintraub, 2000) which is sensitive to deficits in semantic retrieval were used to assess speech and language deficits. The standard test criteria were used to apply and correct the items named spontaneously plus additional items named correctly after semantic cues. Subtests of the WAB include spontaneous speech, auditory verbal comprehension, repetition and naming. The assessment revealed severe aphasia (WAB AQ-33.4), comparatively intact auditory comprehension, and intact repetition yet nonfluent oral speech, reading and writing, more indicative of mixed trans-cortical aphasia. Her speech was characterized by abnormal intonation, dysarthria, effortful Short (1–2 words, telegraphic style) < 10-15 words, no grammar yet content and meaning.

Table 1: Pre and Post Remediation training Findings on Neuropsychological Assessment

Variable	Baseline assessment	1st Post Remediation training (After 6 Month)	2nd Post Remediation training (After 18 Months)
WAB AQ	33.4	50	73.7
Spontaneous Speech	3	11	16
Auditory Comprehension	7.2	10.2	12.9
Repetition	3.4	4.8	7.2
Naming	2.5	4.1	5.7
Reading &Writing	.6	4.1	5.9
Apraxia	5	7.5	12.5
Constructional & Calculation	3.4	10.8	11.3
Boston Naming Test	2	9	13
PGI Memory Scale	Below 20th Percentile	20-40 Percentile	40-60th Percentile
N-Back Visual	8.08	6.3	5.72
N-Back Verbal	10.92	8.07	5.97
Rey-Osterrieth Complex Figure Test	24	28	31
FIM™(Motor & Cognitive Items)	6,3	6,4	7,6

WAB AQ- Western Aphasia Battery Aphasia Quotiont, FIM™- Functional Independence Measure

Performance on subscales of PGI Memory Scale (Pershad & Wig, 1977), Rey-Osterrieth Complex Figure Test (Rey& Osterrieth, 1993) and N-Back test showed significant impairment in linguistic aspects while comparatively preserved non-linguistic functions in

terms of memory and attention. In N-Back test stream of information is monitored with the goal of deciding whether the current item matches an item that was "n" number of trials ago in the sequence for which a simple recognition response is required (Baddeley,2021). Thus a variety of stimuli, from letters, words to spatial locations can be presented making it easier to differentiate linguistic and non-linguistic deficits. The neuropsychological tests showed significant impairment in all cognitive domains with particularly enhanced deterioration of semantics. Successful attempts of repetition test (simple one), which requires an intact working memory, suggested that impairment of language functions cannot be solely explained by the deterioration of attention and short-term memory.

The Functional Independence Measure (FIM™,2009) evaluates the functional skills of individuals in six domains of function i.e. self-care, sphincter control, transfers, locomotion, communication, and social cognition on a 7-point ordinal scale, with scores of 1 to 5 indicating a need for caregiver assistance. The FIM™ was found to be more responsive to different client populations and improvement over the course of rehabilitation (Lee, 2022).Patient's scores on motor functions indicated modified dependence (Average score of 6) whereas communication and social cognition scores were in the complete dependence category (Average score of 3).The complete battery of tests for neuropsychological assessment was administered in three sessions.

Intervention:

The patient received an intensive cognitive remediation training for initial 6 months (twice weekly for 3 month then once in a week) followed by spaced sessions for next one year (once in two week).Based on the findings of the neuropsychological assessment, individualized tailored rehabilitation plan was formulated focusing on patients' impaired language function to promote language reconstruction and integrate the training into daily life communication. Eventually, to achieve the goal of best recovery of patients' language communication skills and enhance her reintegration with her family and society. Her caregiver was primarily the mother who was psycho-educated to help make sense of the patient's behavior and facilitate home assignments for the transfer of training in daily living subsequently. Speech and language functions, working memory and its extension in ADLs were targeted as interventions for this patient.

Considering the psycholinguistic properties known to influence word acquisition and retrieval (e.g., imageability, visual complexity and naming latency; Mätzig, Druks, Masterson, & Vigliocco, 2009) 40 items consisting of 20 nouns and 20 verbs (2 sets of 20 items, 10 nouns and 10 verbs)were selected initially to train the patient. For the selection of these items in consultation with her mother, different categories of nouns were utilised consisting of food, body parts, household objects, shapes, occupations, clothes, animals, etc. For practice verbal targets progressed along a hierarchy of stimulus complexity and task difficulty in the sequence of simple drawing of items, writing and reading the word with fading cues to assist verbal production. One hour practice session included casual interaction with the therapist and a short break for refreshment to sustain the participants' interest and motivation during the intensive treatment program.

The patient was encouraged to make a simple drawing of the item followed by the writing step wherein initially stimulus word was shown completely with gradual fading of cues overtime. At this stage patient was introduced to three tasks described by Luria (1970) as retraining strategies for reading in English: (a) counting the letters in individual spoken and written words, (b) counting syllables in an individual spoken and written word, and (c) synthesizing words from individually pronounced/written letters (i.e., recognizing word). Her performance on these tasks was better for short words (three to five letters) in comparison to increasing word length. Her strategy involved both letter-by-letter reading and counting phonemes referring to the sounds. It was interesting to observe that patient was able to count phonemes referring to the sounds rather than letters in written words which were presented in the English language. Even when these strategies failed to result in literal paralexias (thumb-th, m, d), with training gradually she would often correct herself and recognize the short word. The mother was encouraged to make her practice a similar task as the retraining strategy for auditory items in Hindi as the patient's efforts to communicate with the mother was in this language mostly. In the next step in reading and writing when she seemed to use a direct visual strategy in immediately recognizing the single words, a word combination of noun and verb was added gradually (I take my meal, I made a coffee) increasing the complexity.

Along with this WM training, consisted of practice on an n-back task (either with pictures or spoken words) in the same frequency during sessions and thrice a week at home. Some other tasks included in WM training consisted of digit span tasks (backward and forwards) of varying difficulty levels, as well as a paced auditory serial addition task (i.e., adding the last two numbers heard in a continuous list). For transfer of training in relevant settings client was encouraged to use at least one word each in the home setting (e.g., naming an item during meal preparation or making a request for an item e.g., during traveling, in a local shop) aiming shaping of socially driven communication task. Also patient informed that she has started reading simple picture

books, interaction-based games and listening to English news suggesting her motivation for therapy.

Outcomes:

Neuropsychological and functional assessments were performed again after six months and the next one-year of the rehabilitative training to evaluate the cognitive and behavioral changes that resulted from treatment. The changes in scores and reported functional improvement (Table-1) indicated patient showed an improvement in cognitive tasks as well as behavioral aspects. She could respond verbally and write to dictation at the word-by-word level for short sentences. Western Aphasia Battery indicated significant improvement in clients' everyday communication from severe to progressing towards mild aphasia (WAB AQ: 50 and 73.7 in 2nd and 3rd assessments). Multi-modal cues (drawing, writing and reading the word while not restricting the use of gestures) assisted verbal production. A considerable change was observed in her speech quality in terms of better production (<40-50 words/min) and normal intonation, effortful telegraphic style normal phrase length. Response during functional communication/discourse (Can you tell me about your family? How do you prepare tea?) and quality of life results (Cognitive score on FIM™) further add weight to the efficacy of remedial training. Also, maintenance of these treatment effects at around 5-year follow-ups was extremely encouraging although improvement gained further was slow-paced.

DISCUSSION

Acute encephalitis (AE) may present with a wide variety of symptoms including cognitive regression accompanied by loss of language skills. Despite high prevalence where around 50 percent of patients presented with language impairment in AE(Guevara-Silva et al, 2022)primarily linguistic functions have not been studied in detail. The dual stream model of language processing postulates that the bilateral posterior-medial temporal gyrus and posterior-inferior temporal gyrus regions is the seat for semantics regulation while auditory-motor transformation, repetition of words, verbal working memory and auditory attention are monitored through the left temporal plane and left posterior frontal lobe (Hickok & Poeppel, 2002).Index patient's patterns suggesting severe reading and writing difficulties were similar to mixed non-fluent and transcortical motor aphasia as fewer such difficulties were reported in amnesic, Broca and conduction aphasia (Gonzalez et al., 2020). Thus significant semantic and cognitive deficits in this client raised concern about pieces of evidence pointing to the negative impact of semantic and cognitive impairment on response to aphasia treatment (Lambon, Snell & Fillingham, 2010).The patient showed significant impairment in all cognitive domains but especially in semantics skills and relatively spared repetition and non-linguistic abilities which do not require semantics skills, suggesting somewhat preserved linguistic processes of encoding, storage, and production governed by phonological working memory. The outcome of the WAB, and Functional Independence Measure (FIMTM) was consistent with the report that with intensive comprehensive treatment more severe aphasia tended to show greater recovery in language ability and functional communication than those with mild aphasia (Persad et al., 2013). There are findings suggesting the use of external devices, memory notebooks, or mnemonic strategies to improve memory performance in encephalitis patients (Emslie, 2007; Langenbahn et al, 2013) which was initially not possible with this client due to limitations in writing skill and behavioral regression. This was tried to compensate using regular socially mediated practice contexts of learned tasks that may work positively to facilitate the transfer of training. Importantly, WM training which is considered to involve speed of processing focussed attention, task switching and updating comprehensively representing crucial parts of executive functions also helped in improvement in sentence comprehension and everyday memory activities as supported by a study where participants demonstrated improvements in functional communication (i.e.,understand ability and intelligibility of spoken messages on familiar everyday topics) (Zakariás, Salis & Wartenburger, 2018).It is evident that the WM task requires both maintenance and updating of information with each trial which activates a bilateral frontoparietal network. Neuroimaging studies indicate that activation of this region overlaps with language networks (Rottschy, 2012). Sreedharan et al., 2019, mentioned the role of plasticity of the isotopic brain area of the ipsilateral or contralateral brain replacing the function of the damaged core language area. Thus, further improvement can be explained on the basis of neuroplasticity and functional connectivity of the brain, replaced or supported by a cerebral language area different from the original one affected by encephalitis. A major limitation of this study was the lack of supportive findings in terms of any radiological or other investigations indicating structural change as the patient did not cooperate with the MRI later. Therefore, addressing cognitive deficits, especially language deficits, could improve quality of life by using available cognitive resources.

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