

Beating the Odds: Intact Neuropsychological Functioning despite TLE

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ABSTRACT

Patients with Temporal lobe epilepsy (TLE) frequently display cognitive comorbidity and can have widespread network abnormalities, which might affect a variety of cognitive and intellectual functions. As a result, refractory TLE seems to be associated with slow but ongoing cognitive deterioration. The case is of a 32 year old, right handed male, engineering graduate, diagnosed with TLE- right mesial, 12 years ago. A number of head injuries were caused due to the seizure present, which includes a fall from height of 12 feet in childhood. The neuropsychological tests administered were Gesell's Drawing Test, Mini Mental State Examination, PGI Memory Scale, Battery of Performance Tests of Intelligence, Verbal Adult Intelligence Scale, Hamilton Rating Scale for Depression, Bender Visual Motor Gestalt Test and Dysfunction Analysis Questionnaire. No impairment found on orientation; average cognitive functioning; above average attention and concentration, verbal working memory, visual and verbal memory; average practical ability, abstract ability, average verbal intellectual ability; superior ability on comprehension and average performance ability. Mild to moderate impairment on perceptuo-motor functioning and an evidence of depression were present. Patient showed high dysfunction in personal, social, vocational and cognitive areas. The study highlights that even despite chronic epilepsy, with a series of head injuries due to the seizures; an individual can still have average neuropsychological abilities. Holistic neuropsychological rehabilitation along with Vocational Retraining would go a long way in the functional independence of the patient. Neuropsychologists have a significant role in the assessment, treatment, and rehabilitation of people with epilepsy.

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Introduction

Temporal lobe epilepsy (TLE) is a form of focal epilepsy, a chronic neurological condition characterized by recurrent seizures.¹ It is the most common form causing refractory epilepsy.² There has been a suggestive link between febrile seizures, and subsequent TLE, but the exact role is unclear.³⁻⁷ Some studies have shown hippocampus abnormalities on Magnetic Resonance Imaging (MRI) in status epilepticus, which shows that prolonged seizures, damage the brain.⁸⁻¹⁰

Cognition refers to different mental processes which include attention, processing speed, memory, language, perception, reasoning ability, judgement, etc. that are used everyday. Persons with epilepsy frequently complain about deteriorating cognitive functioning, where memory tends to be the most affected. Other complaints also include reduced ability to pay attention and focus, slowed mental processing, and difficulty in expression. While all these problems can be directly related to the seizure activity, there are additional factors as well which are indirectly related to the diagnosis of epilepsy.¹¹

Neuropsychological evaluation is carried out to assess the extent of impairment of any particular ability or skill and to determine the damaged area of the brain. The neuropsychological evaluation includes detailed history, interviewing and assessment of a person's intellectual functioning, and cognition which can include perception, motor functions, attention, memory, processing speed, auditory and visual processing, language functions, executive functioning, planning, problem solving, organization, etc. These assessments

evaluate many areas of cognition and executive functioning to determine whether a patient's impairments/ difficulties in a particular function and behaviour have a neuropsychological basis.¹²

The assessment includes documenting specific effects of anti-epileptic drugs, as a part of clinical management.¹³ This evaluation also plays an important role in evaluating patients for temporal lobe surgery, since the temporal lobes have been implicated in memory functioning. Bilateral hippocampus excision is also associated with profound anterograde amnesia. Unilateral resections are traditionally associated with material specific memory dysfunction.¹⁴

Persons suffering from epilepsy, show that cognitive impairment, particularly memory, is a major complicating factor of epilepsy.¹⁵ Also, chronic TLE is associated with progressive memory impairment.¹¹ Intellectual functioning of these patients with a longer duration of refractory TLE are severely impaired. Consequently, refractory, TLE seems to be associated with slow but ongoing cognitive deterioration.¹⁶ Cognitive prognosis is poor for patients, where the severity of epilepsy, age, intellectual functioning and baseline abnormalities in quantitative magnetic resonance volumetrics play an important role.¹⁷

Methods

Case History

The patient S, is a 32 year old, right handed male, engineering graduate, diagnosed with Temporal Lobe Epilepsy (TLE)- right

mesial, in 2001 (12 years ago). The patient born of a non consanguineous marriage, as twin delivery born at 7.5 months, with normal birth cry but with delayed motor and speech milestones. The semiology includes neck turning to right side with screaming and tonic clonic movement of all four limbs. There is history of tongue bite. No other type of semiology has been reported. There is a history of a number of head injuries caused during/due to the seizure, which includes a fall from height of 12 feet in childhood. The patient had recent head injury due to a fall during an epileptic seizure (Figure 1). The patient has had a number of such injuries in the past There was no family history of seizures. Also, no history of focal weakness was recorded. The first episode of seizure was reported on 27 February 2001, with associated symptoms of fever before the onset. There is a history of slurring of speech, pain in the right side of the body after the seizure with pain in legs. A history of gait problems present as well. A substance use history of tobacco use present since the past 2–3 years. A detailed history regarding the patient's school performance and past academic achievement could not be elicited. Informed consent form was obtained from patient.



Fig. 1: The patient with a recent head injury due to a fall during an epileptic seizure. The patient has had a number of such injuries in the past.

Materials

The patient, diagnosed by the Department of Neurology, was referred to Clinical Neuropsychology, Neurosciences Centre, AIIMS, New Delhi, India, for a detailed neuropsychological assessment. The neuropsychological tests administered on the patient were Gesell's Drawing Test (GDT),¹⁸ Mini Mental State Examination (MMSE),¹⁹ PGI-Memory Scale (PGIMS),²⁰ Battery of Performance Tests of Intelligence,²¹ Verbal Adult Intelligence Scale (VAIS),²¹ Hamilton Rating Scale for Depression (HRSD),²² Bender Visual Motor Gestalt Test (BVMGT),²¹ and Dysfunction Analysis Questionnaire (DAQ).²³

Results

The patient was initially screened for intellectual functioning using Gesell's Drawing Test (GDT).¹⁸ Combine result for different scales are summarised in Table 1. The patient had a score of 12 which showed average mental age. The Mini Mental State Examination (MMSE)¹⁹ conducted on the patient revealed intact mental status with a score of 28, for which the maximum can be 30. The cognitive functioning of the patient Tab. 2 was assessed using Post Graduate Institute Memory Scale (PGIMS),²⁰⁻²¹ which is a part of the PGI Brain Dysfunction Battery. The overall score of 52nd PR indicated average cognitive functioning. The average for which can fall anywhere between 40th and 60th PR. Performance Quotient was calculated using a Battery of Performance Tests of Intelligence – Bhatia Short Scale (BSS),²¹ in which Performance Quotient (PQ) came out to be 107, Practical Ability was 101, and Abstract Ability – 103 which all come under the category of average performance ability. These scores were compared to an average of 90–110 IQ points (Wechsler, 1944). The Verbal Intellectual Quotient (VIQ) was assessed using Verbal Adult Intelligence Scale (Pershad and Verma, 1990). The score of 99.5 also indicated average verbal intellectual functioning Table 3.

Perceptuo-motor functioning was assessed using Bender Visual Motor Gestalt Test (BVMGT)²¹ which gave a score of 2, indicating mild to moderate impairment on perceptuo-motor functioning. The present Dysfunction assessment of the patient was evaluated using Dysfunction Analysis Questionnaire,²³ which

Table 1: The results of the entire neuropsychological assessment in detail

Test	Function	Result	Interpretation
Gesell's Drawing Test (GDT)	Intellectual Screening	M.A. = 12 years	Average
Mini Mental Status Examination (MMSE)	Orientation	28/30	No impairment
PGI- Memory Scale (PGIMS)	Cognition	52 nd PR	No impairment
Bhatia Short Scale (BSS)	Intellectual Functioning	PQ = 107 Practical Ability = 101 Abstract Ability = 103 Mean PQ = 103	Average
Verbal Adult Intelligence Scale (VAIS)	Verbal Intelligence Quotient (VIQ)	VIQ = 99.5	Average Verbal Intellectual Ability
Hamilton Rating Scale for Depression (HRSD)	Depression Screening	30	Very Severe Depression
Bender Visual Motor Gestalt Test (BVMGT)	Perceptuo-motor Functioning	Dysfunction Rating of 2	Mild to Moderate impairment on perceptuo-motor functioning
Dysfunction Analysis Questionnaire	Dysfunction Assessment	77%	High Dysfunction

Table 2: Detailed assessment for Cognitive Functioning using PGI- Memory Scale

S. No.	Sub-Test	Cognitive Area Assessed	Score	Interpretation
1.	Remote Memory	Ability To Remember personal/historical events of the past.	5/6	Mild Impairment
2.	Recent Memory	Ability To Remember relatively new information.	5/5	Average
3.	Mental Balance	Temporal Sequencing	8/9	Average
4.	Attention and Concentration	Attention & Concentration, mental control, working memory	13/16	Above average
5.	Delayed Recall	Short- term memory	9/10	Average
6.	Immediate Recall	Verbal working memory	11/12	Above Average
7.	Retention for Similar Pairs	Simple Learning Ability	4/5	Mild Impairment
8.	Retention for Dissimilar Pairs	New Learning Ability	11/15	Mild Impairment
9.	Visual Retention	Visuo-spatial memory	9/13	Mild Impairment
10.	Recognition	Visual & verbal memory	10/10	Above Average

Table 3: Detailed Intellectual Functioning (Verbal) Assessment

Test	Sub-Test	Function/Ability	Result	Interpretation
Verbal Adult Intelligence Scale (Pershad et al)	Information	General Knowledge	82	Low average
	Comprehension	Comprehension	125	Superior Ability
	Arithmetic	Working Memory	73	Borderline
	Digit Span	Attention & Concentration	118	High average
VIQ (Mean)			99.5	Average Intellectual Ability

Table 4: Detailed Dysfunction Assessment Using Dysfunction Analysis Questionnaire

S. No.	Area of Dysfunction	% of Dysfunctioning	Interpretation
1.	Social Area	72%	High
2.	Vocational Area	82%	High
3.	Personal Area	92%	High
4.	Family Area	58%	Average
5.	Cognitive Area	82%	High
Overall Dysfunction		77%	High

indicated an overall 77% dysfunction in everyday activities showing a high dysfunction Table 4.

Discussion

Intractable epilepsy is related to various transient and chronic electrical and neuro-chemical brain imbalances. Also, there is increasing evidence that chronic epilepsy brings about secondary neuronal metabolic and structural decline.¹⁶

TLE is usually associated with long term memory impairment. The frontal lobes are responsible for higher cognitive functioning like executive functioning and working memory which is vital for everyday living. Impairment in these functions has been reported on an increasing trend. Evidence from both the

neuropsychological and neuroimaging literature suggests that both executive functioning and working memory are impaired in the presence of TLE. Other executive functions like decision making and theory of mind have not received enough attention from previous studies. With regard to working memory, the medial temporal lobe structures appear to play a critical role, but with evidence of hippocampal dependent and independent processes.²⁴

The present case of patient S, shows that despite chronic epilepsy since more than 12 years, and repeated head injuries due to the falls during the seizures, a person can still have intact neuropsychological functioning. With extensive review of existing literature, it can be stated quite confidently that

neuropsychological impairment can be associated with chronic epilepsy. The patient had no complaint of memory loss or intellectual/cognitive deficits, whose detailed neuropsychological assessment revealed that aside from the dysfunction due to the seizures, the patient had no impairment on orientation, which was evaluated through Mini Mental Status Examination (MMSE).¹⁹ The patient was evaluated and assessed in detail on 10 different areas of cognitive functioning using Post Graduate Institute Memory Scale.²⁰ The cognitive assessment revealed mild impairment on Remote Memory (ability to remember personal/historical events of the past), Retention for similar pairs (ability for simple learning), retention for dissimilar pairs (Ability for new learning), Visual Retention (Visuo-spatial memory). He had average functioning in Recent Memory (Ability to remember relatively new information), Mental Balance (Temporal sequencing), and Delayed Recall (Short-term memory). However, the patient showed above average performance on Attention and Concentration, Immediate Recall (Verbal working memory), and Recognition (Visual and verbal memory). The cognitive functioning performance of the patient was compared to age and education matched norms. Overall, the cognitive functioning of the patient was at 52nd PR (Average = 40th to 60th PR) which again shows average performance.

Jokeit and Ebner studied if the duration of refractory TLE is related to cognitive and intellectual deterioration and impairment. The results revealed that duration of the TLE does affect cognitive ability. Patients with more than 30 years of TLE had more impairment as compared to patients with 15 or 30 years of TLE. Also, psychometric intelligence of patients with a longer duration of refractory TLE were more severely impaired, concluding that refractory TLE seems to be associated with slow but an ongoing cognitive deterioration.¹⁶

Detailed intellectual testing can be divided into verbal and performance subparts, which was assessed using Verbal Adult Intelligence Scale (VAIS).²¹ The patient's verbal functioning on Information (General Knowledge) was low average, on arithmetic (working memory) was borderline, above average on digit span test (attention and concentration) and a superior ability was found on comprehension. The overall VIQ (Verbal Intelligence Quotient) was found to be 99.5 which were again average. The Performance Quotient (PQ) was assessed using Bhatia Short Scale (BSS)²¹ which are a Battery of Performance Tests of Intelligence. The patient had an average Practical Ability which was 101, Abstract Ability of 103 PQ which was 107. The Mean PQ was again average (103). These categorizations of IQ/PQ were based on Wechsler's Classification of adult IQ scores.^{25,26}

In two independent cross sectional studies showcasing the global cognitive performance of patients, the first study observed the influence of the duration of epilepsy on the difference between a measure of pre-morbid intelligence and the current performance on an intelligence test, on 78 patients with refractory TLE. The second study observed the duration of epilepsy-dependent effects on the intelligence outcome of 209 patients with refractory TLE. Both the studies revealed that the duration of epilepsy plays a significant role in the intellectual measures of the patients. Those with a longer duration of TLE show greater variability in IQ measures, where the higher the duration, more the impairment. Age of epilepsy onset also plays an important role in inter-individual variability in IQ

measures of adult TLE patients. Consequently, refractory TLE seems to induce a very slow but ongoing cognitive deterioration and also chronic epilepsy is associated with worse cognitive abilities if the epilepsy exceeds more than 20 years. It can be confidently said that epilepsy-related noxious events exhaust the compensatory capacity of the brain functions.²⁷

Perceptuo-motor functioning is an important aspect which needs to be looked into for a patient with epilepsy/ head injury. In a study, 60 children with epilepsy of age 6–12 years with matched controls were compared for cognition, visual and spatial perception, visuo-motor organization and gross motor function. They found significant differences between these abilities in the experimental and control group. They also found a positive correlation between perception and gross motor dysfunctions.²⁸ In the present case, perceptuo-motor functioning was assessed using BVMGT. The patient had a dysfunction rating of 2 which indicated mild to moderate impairment on this area. The range of ratings can be from 0 to 3.

Up to 50 or 60% of patients with chronic epilepsy have various mood disorders including depression and anxiety. Anxiety is shown to have a deep influence in the quality of life of patients with epilepsy.²⁹ Depression is the most frequent co morbid psychiatric disorder in epilepsy.³⁰ The patient was assessed on Hamilton Rating Scale for Depression to screen for depressive symptoms, which showed that he had severe depression. The patient was referred to the neuro-psychiatrist for the treatment of the same.

Epilepsy impairs an overall cognitive, intellectual, social, personal, and vocational functioning of the affected individual. The dysfunction evaluation was done using Dysfunction Analysis Questionnaire which showed that he had high dysfunctioning in areas of social, vocational, cognitive and personal functioning. The overall dysfunction rating was 77%, which is again high. Epilepsy related stigma has three different levels; internalized, interpersonal, and institutional.³¹ This case highlights the high dysfunction in social, vocational and personal areas of the patient despite average neuropsychological functioning. This can be attributed to the stigma which is attached to this neurological disorder.

Significant negative attitudes prevail in adults and adolescents worldwide leading to loneliness and social avoidance. Patients with TLE are also wrongly viewed as being violent towards others. Twenty-five percent of adults having epilepsy describe social stigma as a result of their disorder. These patients fear rejection and often feel lonely consequent to diagnosis.³² This was also evident in the patient, as he mentioned about "not wanting to meet anyone and staying at home rather than going out". This can also be attributed to the severe depressive symptoms the patient is having.

This case is an inspiration and motivation to all the people suffering from chronic epilepsy that despite chronic TLE a person can have an intact neuropsychological functioning which would help them in carrying out the important day to day functioning and also the fact that chronic epilepsy is controllable.

Since it is a case study, therefore, no generalizations can be drawn. One of the limitations of the case report was that the exact intellectual and cognitive functioning of the patient prior to the onset of epilepsy, and a detailed history regarding the patient's school performance and achievement could not be elicited, so, the exact deterioration of the cognitive and intellectual decline/impairment could not be ascertained.

Conclusion

This case highlights that despite chronic epilepsy for 12 years, with a series of head injuries due to the seizures; an individual can still have average neuropsychological abilities. Holistic neuropsychological rehabilitation along with Vocational retraining would go a long way in the further rehabilitation and functional independence of the patient.

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