



A RARE CASE PRESENTATION OF RASMUSSEN ENCEPHALITIS

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ABSTRACT Rasmussen's encephalitis is rare neurological disorder, usually seen in children involving single cerebral hemisphere, characterized intractable seizure, cerebral atrophy and progressive neurological deficit. In classical clinical presentation of disease MRI shows hemispheric atrophy of one cerebral hemisphere. Treatment strategy varies among different patients at different stages. Treatment option is selected according to symptoms of the patients.

KEYWORDS : Rasmussen's encephalitis, Intractable seizures, Cerebral atrophy

INTRODUCTION:

Rasmussen encephalitis is a chronic inflammatory disease of unknown origin, where brain inflammation results in unilateral brain atrophy, as described by Rasmussen et al. in 1958¹. The disorder is rare and affects mostly children or young adult. The median age of onset is 6 years with range from infancy to adult hood. It is characterized by drug resistant focal seizure, progressive weakness, worsening of motor and cognitive function. Untreated children will develop hemiparesis, hemianopia, transient ischemic attack (TIA) and cognitive decline with in few year of epilepsy. Some cases of Rasmussen encephalitis have less common presentation, clinical course is slower and final deficit are less severe^{2,3}. Diagnosis is based on clinical history, imaging, histopathological findings and execution of other causes.

Case Report:

A young boy 5 years of age was referred to psychiatry OPD of tertiary care hospital affiliated to medical collage from pediatric OPD with complain of odd body movements and hallucinatory behavior. As per history given by patients grandfather and parents, since last 3 months patient had episode of odd body movements, in form of pointing his left hand in front and muttering "oh oh" with fearful facial expression lasted for approximately 5 to 10 seconds after which he would went to sleep. Initially these episodes were 1 to 2 in every 15 days and gradually increased 2 to 3 episodes per day for last 15 to 20 days and he had similar episodes also during sleep.

Patients had a past history of falling down from 2 to 3 feet height at the age of 8 month with history of unconsciousness for 2 to 3 days. At present, previous reports were not available. Patients had left sided weakness after fall and had difficulty in walking since then. Due to lack of health care facility in his remote village, he started physiotherapy at 3 years of age and it improved his weakness but still residual weakness remained in left hand and left leg. He had to walk with drag.

He had a history of dog bite 3 times in last one year. Vaccination for the same was taken for first 2 episodes. No vaccination taken for the last episode which occurred 6 month back He had history of inability to sit at one place for a long time and occasionally biting to his parents without reason.

On neurological examination, there was some residual left sided motor weakness. On mental status examination he had a cheerful affect, cooperated during the interview, responding to question asked. He did not report any anxiety feature and stranger anxiety was not evident.

All his routine laboratory investigation was normal. EEG finding was normal. His MRI brain findings are suggestive of possibility of "Rasmussen encephalitis" or "Dyke Davidoff Masson syndrome".

Pediatrician opinion was taken and he was put him on syrup Divalproex Sodium 1.25ml (250mg) twice a day. He did not improved in 5 days of treatment. So pediatric neurophysician opinion was taken, and Carbamazepine 100mg twice a day with gradual tapering of syrup Divalproex Sodium was considered, with concept of focal epilepsy. Patient significantly improved in 7 day and after that no episode occurred.

Psychiatric assessment was done IQ testing revealed mild intellectual disability. Children Apperception Test (Projective test) was not possible as patient would not cooperate or follow the instructions of test.

CONCLUSION:

The patient presented with odd body movements, fearful facial expression, mimicking transient hallucinatory behavior, resolving hemiparesis and normal EEG, was found to have Rasmussen encephalitis on radio imaging, which improved with carbamazepine.

DISCUSSION:

Diagnosis of Rasmussen Encephalitis is generally based on the characteristic clinical, radiological and pathological features. Typically remission is characterized by intractable seizure but in our case those seizure like phenomena improved with antiepileptic medicine. Some case reports also mention EEG observations as unihemispheric slowing with or without epileptiform activity and unilateral seizure onset. Instead of progressive hemiparesis there was resolving hemiparesis⁴. The child had certain attention deficit hyperactive syndrome not conforming to label it as a diagnosis of ADHD and psychometric testing revealed mild intellectual deficit.

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