

Epilepsia partialis continua

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Abstract

Epilepsia partialis continua (EPC) is a rare syndrome of continuous focal jerking of a body part, usually localized to a distal limb, occurring over hours, days or even years. Etiology is multiple and diverse as well as the outcome is also variable. Here we report two cases of EPC admitted almost in a same period in our hospital in Khulna with different etiology and outcome. The first one, an adult diabetic patient, developed status epilepticus followed by right sided epilepsia partialis continua over a period of three days after a viral illness. He responded well with treatment. The second case, a three year old girl, progressively developed epilepsia partialis continua over a period of six months with no antecedent event. Her treatment response is not satisfactory.

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Introduction

Epilepsia partialis continua (EPC) is defined as a syndrome of "spontaneous regular or irregular clonic muscle twitching of cerebral origin, sometimes aggravated by action or sensory stimuli, confined to one part of the body, and continuing for a period of hours, days or weeks" and "recurring at intervals of no more than ten seconds". EPC is conceptualized as a special form of focal status epilepticus.¹ Etiology is multiple and diverse, like focal or multifocal brain lesions, systemic diseases affecting the brain and metabolic or other derangements. In children the main causes are Kozhevnikov Rasmussen syndrome and malformations of cortical development. Cerebrovascular disease and brain space occupying lesions are the main causes in adults. Nonketotic hyperglycemia are the most common of the reversible causes. An 'encephalitic process' is found in >50% cases.² Around 2/3rd patients have an abnormal brain MRI and EEG that become worse in progressive disorders such as Kozhevnikov Rasmussen syndrome. PET and SPECT scans often localize the abnormal region but they are not specific. Screening for metabolic and mitochondrial disorders may be needed. Epilepsia partialis continua is notorious with regard to lack of clinico - EEG correlations; epileptiform abnormalities may or may not be concomitant with the jerks.³

Prognosis of EPC with regard to neurological outcome and control of continuous jerking depend

largely on the underlying etiology. While EPC due to Rasmussen's encephalitis has been shown to be highly resistant to any kind of medical therapy, drug therapy may be successful in adult patients where EPC is often caused by local, non progressive lesions. Even in those patients, however, medical therapy of EPC remains a challenge and in most cases, control of twitching can only be achieved by polytherapy with various combinations of intravenously administered antiepileptic drugs, including diazepam, pentothal sodium and valproate.^{3,4}

Case 1

A 38 year old businessman, hailing from Bagerhat, diabetic for 1 year with irregular treatment, developed mild fever which persisted for 3-4 days (figure-1). After 7 days, in a fine morning, he noticed involuntary repeated contraction of right side of face and right upper limb. It was continuous with no impairment of consciousness. It continued for 3 days, progressively was increasing in intensity and affected right lower limb also. On the succeeding day the convulsion became generalized and he developed status epilepticus. At this stage he was admitted in our hospital on 16th April, 2013. We found him unconscious (GCS=6), eyes were open

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and fixed, neck stiff, respiration was irregular, BP=140/80 mmHg and temperature was 99.0F.



Fig-1. Photograph of case-1 showing eyes deviated to right

RBS was 13.1mmol/l, urinary ketone bodies were absent, serum electrolytes were normal. Treatment started with IV Phenobarbitone, Syp. Phenytoin, Tab. Oxcarbazepine, IV fluid, antibiotic, PPI, steroid (Methyl-Prednisolon), S/C insulin and oxygen inhalation. In the next day patient regained consciousness but continuous convulsion of right half of body (EPC) restarted. The patient was violent and aggressive. Tab.

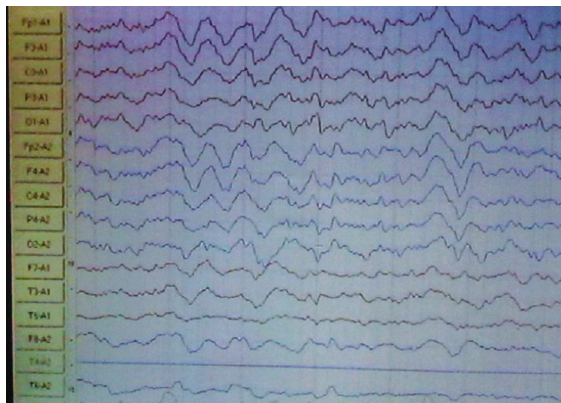


Fig-2. EEG of case-1 showing bilateral intermittent delta slowing predominantly over the frontal region and temporal region (left>right)

Clobazam, Inj. Haloperidol and Inj. Prochlorperazine were added. Biochemistry showed normalization of glycemic status and hypokalemia (for which oral supplementation started). After 1 day the intensity of EPC came down and for psychosis Tab. Lithium was added. CSF study revealed normal result except high glucose level (10.7mmol/L with corresponding blood sugar 13.3mmol/L). On the succeeding day the convulsion became very mild (was limited within the face only), MRI was done which showed T2

and FLAIR hyperintensity affecting both temporal region (Left>Right). EEG showed normal background activity with bilateral intermittent delta slowing predominantly over the frontal region and temporal region more marked over the left side (Figure-2). The convulsion was controlled, but right upper limb dystonia was seen. The patient became almost normal and was discharged after a total stay of 18 days with Oral hypoglycemic agents, Tab. Prochlorperazine, Tab Oxcarbazepine, Tab. Phenobarbitone, Tab Clobazam, Tab Lithium.

Case 2

A girl of 3 years, hailing from Bagerhat, had been suffering from continuous convulsion of right side of body for last 6 months (Figure-3). The convulsion started from right lower limb, gradually involved right upper limb and right side



Fig-3. Photograph of case-2 showing eyes, angle of mouth deviated to right, right upper limb jerking.

of face. Initially it was occurring for 10-20 minutes with an interval of half an hour without any loss of consciousness. She was admitted in our hospital on 18th April, 2013. On admission there



Fig-4. Ictal EEG of case-2 showing fronto-central delta slowing predominantly over the left side. (EKG artifacts are seen over the left temporal region)

were continuous right facial twitching, frothing and salivation, right eyelid repeated blinking, jerking of right upper and lower limb except during deep sleep. For previous two weeks speech became indistinct and she developed difficulty in walking due to weakness of right lower limb.

The patient was conscious, oriented and could follow command. The MRI showed no obvious pathology except mild atrophy of left temporo insular region. Ictal EEG showed normal background activity with fronto central delta slowing predominantly over the left side (Figure-4). Treatment started with IV Methyl Prednisolon, Oral Carbamazepine, Clobazam followed by Phenobarbitone with little improvement. The patient was diagnosed as a case of Rasmussen Encephalitis. She was discharged with carbamazepine, clobazam and phenobarbitone and was advised to come for regular follow up.

Discussion

The first case may be due to viral encephalitis on the background of fever, unconsciousness, generalized seizure with status epilepticus. MRI is showing hyper intensity changes in both temporal regions, although CSF study is essentially normal. In a case report on 'Herpes encephalitis presenting with an opercular syndrome and epilepsy partialis continua' the author has shown that the patient had normal initial EEG and normal CSF finding even though having encephalitis and EPC.⁵ Our case also has almost same scenario. Non ketotic hyperglycemia is another possibility. In a case report, the patient having focal seizures, it was found that his average plasma glucose level was 18.32 mmol/L (range: 15.24 mmol/L).⁹ In our patient glucose level is not very high. In another case report by Kamha A, a 52 year old lady presented in the emergency department with left sided continuous seizure. Her blood sugar was 96 meq/l, serum sodium was 123mmol/l, potassium 15.6 meq/l and serum osmolarity was 375 mOsm/l.¹⁰ The scenario of our patient does not match with these findings also.

The second case, considering the young age, continuous convulsion with preserved consciousness, MR1 feature of mild left sided atrophy with no response to any anticonvulsant suggest the possibility of Rasmussen Encephalitis. In a case report by NC Saha et al; two patients, both had intractable partial seizure, progressive hemiparesis and cognitive deterioration following an episode of encephalitis. Developmental milestones were age appropriate

till occurrence of encephalitis, thereafter started deteriorating with progressive deterioration of cognition, behavior, learning, memory and speech. Different anticonvulsants were tried in optimal doses, yet seizure remained uncontrolled. EEG showed unilateral slow waves and MRI revealed unilateral cortical atrophy with hyper intense signals in T2 and FLAIR. According to European consensus statement both cases were diagnosed as Rasmussen's Encephalitis (RE).⁸ Except the occurrence of encephalitis, rest of the features are almost same with our case. Rasmussen Encephalitis is a rare, chronic inflammatory neurological disease of unknown origin that usually affects only one hemisphere of the brain. It is common in children under the age of 10 with average age at disease onset around 6 years. The disease is characterized by intractable severe seizures, loss of motor skills and speech, paralysis on one side of the body (dysfunctions associated with the affected hemisphere).⁷ The most effective treatment of RE with regard to seizure freedom is hemispherectomy. This procedure, however, is usually performed only at later stages of the disease when a patient has developed a fixed hemiparesis with loss of fine finger movements.⁷

Epilepsia partialis continua is not very common problem. Prevalence is extremely small, probably less than one per million population.¹¹ We encountered these two cases presenting almost simultaneously from a common area. The first case improved nicely, but the second one is giving disappointing result. The only way to control the seizure is to do left cerebral hemespherotomy at the expense of dense right hemiplegia, loss of speech and incapacitated state.

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