Rasmussen's Encephalitis with Bilateral involvement presenting as pseudobulbar state: A Case Report

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ABSTRACT

Bilateral involvement in Rasmussen's Encephalitis is extremely rare. Only few cases have been reported so far. Here we report a case of rasmussen's encephalitis with bilateral involvement. Our patient was a 9 years old boy who was product of non-consangious marriage and presented with refractory partial motor seizures followed by left hemiparesis. Our patient progressed rapidly over 2 years and developed a pseudobulbar state with quadriparesis and pseudobulbar palsy.

Key Words: bilateral disease, Rasmussen encephalitis, pseudobulbar state

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Conflict of interest: None

INTRODUCTION

Rasmussen's encephalitis (RE) was first described by Theodore Rasmussen in 1958. It is a progressive encephalitis resulting in intractable seizures, cognitive decline and hemiparesis[1]. The clinical and pathologic findings are usually unilateral but bilateral cases have been reported[2-5]. The disease usually starts in childhood, with 85% of cases being less than 10 yrs of age. The death of RE is rare but can result from either brainstem involvement or complication of status epilepticus[6].

The typical end point of disease is dense hemiparesis, visual field defects and moderate to severe mental retardation[6]. Exact etiology of RE is not clear but inflammatory response seen on biopsy suggests a viral etiology, but so far attempts to isolate any virus have not been successful[7,8]. Roger et al suggested possibility of antibodies to GluR3 receptor as an etiology for RE[9].

Some researchers have suggested role of antibodies to munc-18, an intracellular protein in presynaptic terminals, in etiology of RE[10]. Some

researchers believe that RE may be precipitated by viral infection causing disruption of blood-brain barrier and stimulation of autoantibodies[11].

CASE REPORT

A 9 year old male was first seen in January of 2012 with partial motor seizures. The attack involved the entire left side of body followed by secondary generalization. He had four episodes over two days and he was started on carbamazepine.

Laboratory investigations:

His investigations revealed hemoglobin 12.7 g/dl, creatinine 0.6 mg/dl, bilirubin 0.5 mg/dl, AST 15 IU, ALT 25 IU and calcium 9.9 mg/dl. CT scan head was normal and EEG showed moderately organized symmetrical synchronous posteriorly dominant activity at rate of 6-7 Hz at 50-60 microvolt, better expressed over left posterior head region, there was polymorphic delta activity at 2-3 Hz over right hemisphere maximum over fronto-temporal regions and there were some focal epileptiform discharges over right frontal region (Figure 1).

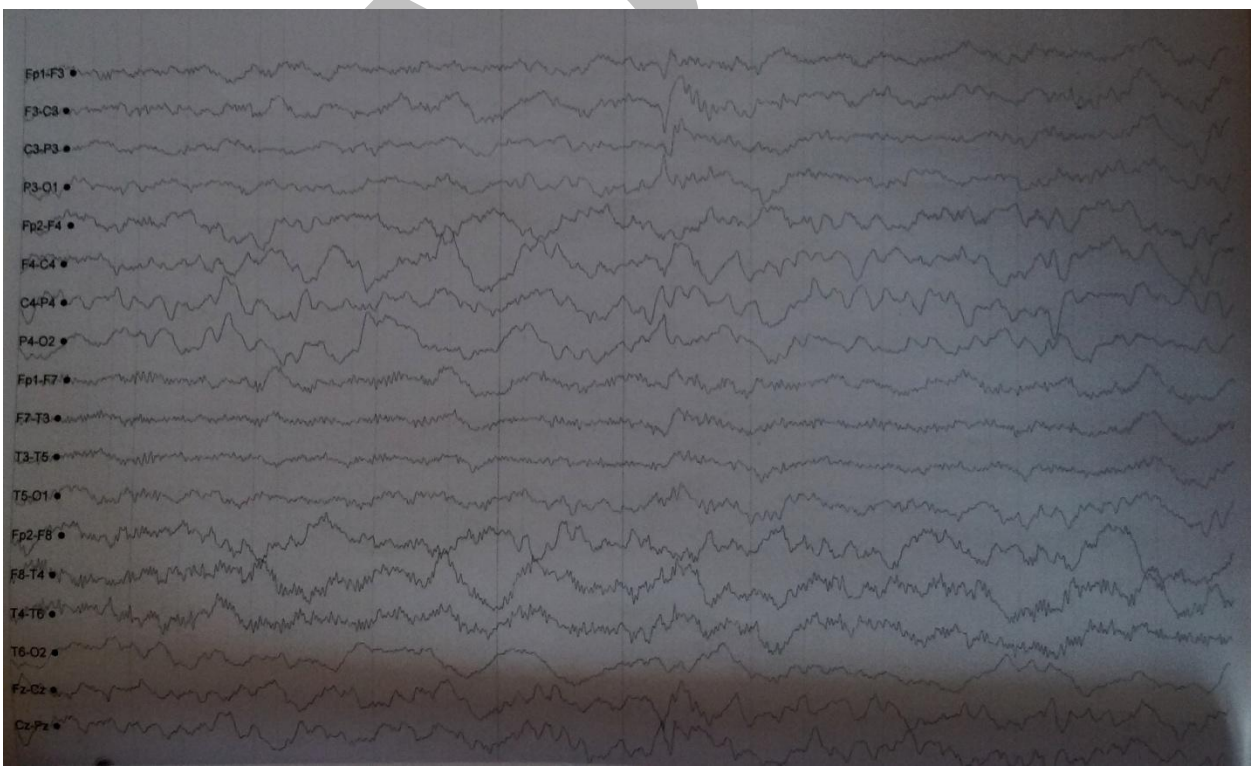

Figure 1: Electroencephalography revealing polymorphic delta activity at 2-3 Hz over right fronto-temporal region with some focal epileptiform disc.

Patient was discharged with advise to follow in OPD. After two months patient had another episode of seizure (left sided with secondary generalization) and he also noticed weakness of left half of body. His neurological examination revealed increased tone on left side with power of MRS grade 4+ and his left side deep tendon reflexes were brisk and left plantar was extensor. MRI brain revealed a hyperintense lesion in right internal capsule with no diffusion restriction or contrast enhancement (Figure 2).

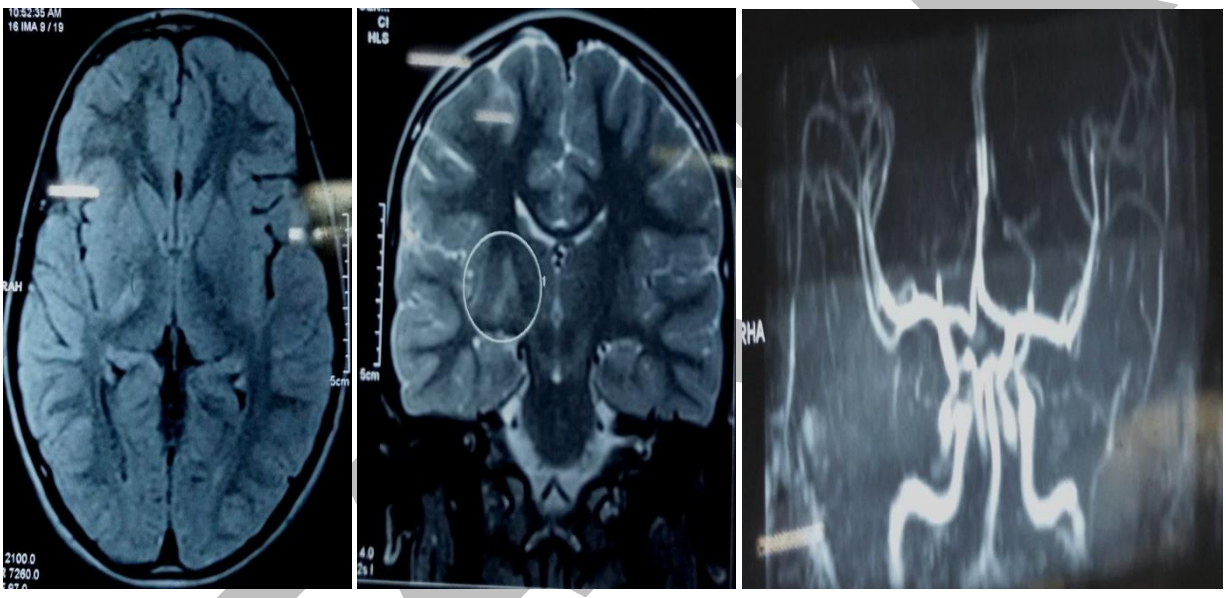

Figure 2 : MRI brain revealing hyperintense lesion in internal capsule and normal angiogram. There was no diffusion restriction or contrast enhancement.

His vasculitic profile (ANA, cANCA , pANCA) and prothrombotic work-up (factor v leiden, protein C , protein S and anti-phospholipid antibodies) were negative. Echocardiograghy was normal. MR angiography of neck vessels and circle of willis was normal (Fig. 2). Cerebrospinal fluid analysis revealed 2 lymphocytes with protein of 23 mg/dl and sugar 84 mg/dl. His lipid profile was normal. He was discharged on carbamazepine and aspirin.

Patient had poor control of seizures and was admitted again after 9 months with status epilepticus. He was put on two antiepileptic drugs (carbamazepine and levetaricetam). Neurological examination revealed left sided hemiparesis with worsening of

power to MRS grade 4-. His MRI brain was repeated which revealed atrophy of right cerebral hemisphere with subcortical hyperintensities (Figure 3).

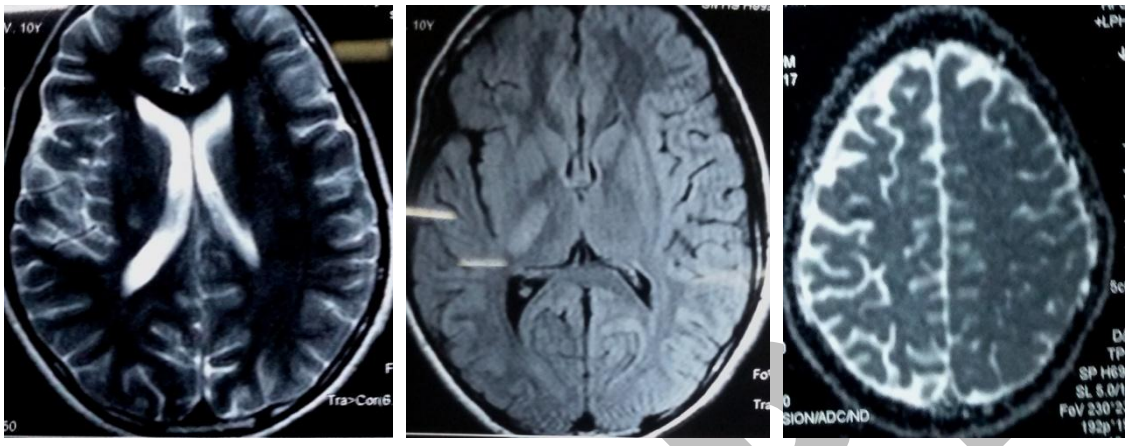

Figure 3: MRI brain showing atrophy of right hemisphere with ex-vacuo dilatation of ipsilateral lateral ventricle.

Keeping in view left hemiparesis with refractory seizures and atrophy of one cerebral hemisphere diagnosis of Rasmussen encephalitis was made and he was started on oral steroids (Prednisolone 2 mg/kg) and antiepileptics were continued. His guardians were given option of brain biopsy and hemispherectomy which they refused. After two months of follow-up patient showed improvement in hemiparesis and had better control of seizures. Then he was lost to follow-up and was admitted again after eighteen months with complaints of difficulty swallowing and weakness of right side of body. There was history of episodic inappropriate crying and laughing. Neurological examination revealed bilateral increased tone, brisk reflexes and extensor plantar response. His jaw jerk was also brisk. He continued to have generalized seizures intermittently. His MRI brain with angiogram was repeated which revealed atrophy of bilateral cerebral hemispheres (right more than left) and angiogram was normal (Figure 4).

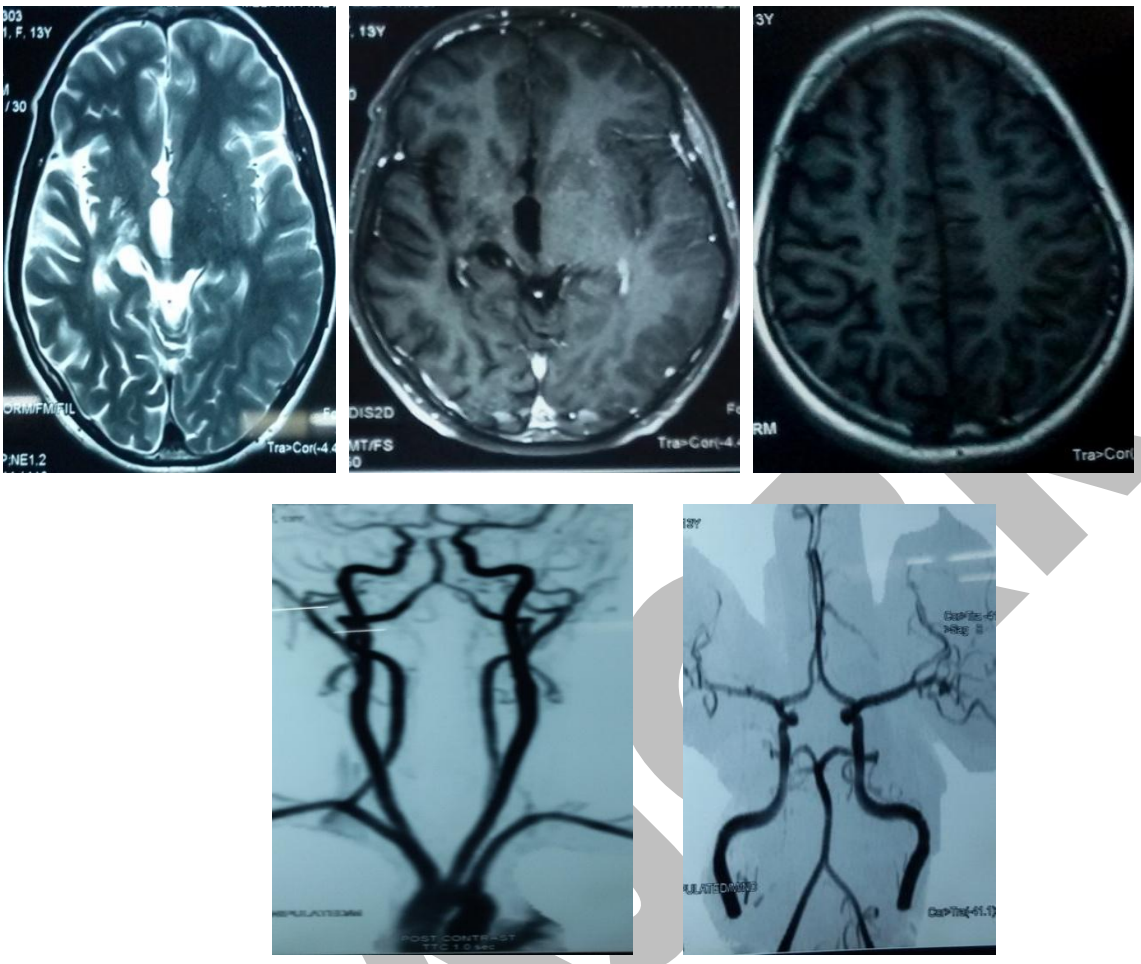

Figure 4: MRI brain revealing bilateral cerebral hemispheric atrophy (more on right side) and normal angiogram of circle of willis and neck vessels

EEG showed bilateral slowing with epileptic discharges (Figure 5). Cerebrospinal fluid analysis revealed 10 cells (lymphocytes) with protein of 73 mg/dl and sugar of 92 mg/dl. CSF was gram stain, AFB stain and fungal stain was negative. His vasculitic and prothrombotic work-up was negative. He was offered immunoglobulin (IVIG) therapy and received one cycle of IVIG (2 gm/kg) over five days and was advised to take IVIG doses monthly.

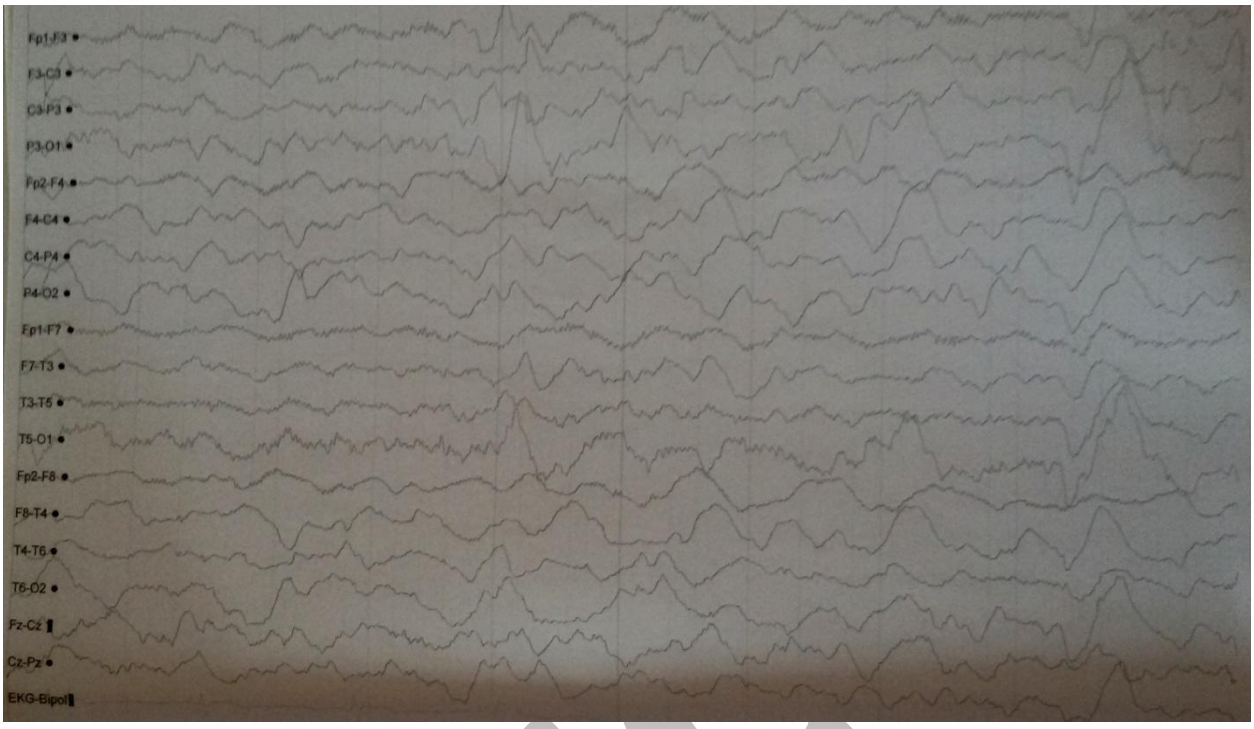

Figure 5 : EEG showing bilateral polymorphic delta activity at 2-3 Hz with intermittent focal epileptiform discharges suggestive of nonspecific neurophysiologic dysfunction.

DISCUSSION

Bilateral RE is rare, among 200 RE cases reported in literature, bihemispheric involvement has been suggested in nine[12,13]. Silver et al[5] had reported two siblings with bilateral RE. Both these patients had onset before age 2 years and both had severe disease. Consanguinity is believed to predispose to more severe bilateral variants of RE[5]. Adults are more frequently reported to have bilateral disease[4,12].

Bilateral RE in our patient resulted in pseudobulbar state with bilateral long tract signs and pseudobulbar palsy. Initial diagnosis of stroke and vasculitis were considered but were ruled out after investigation. During second admission when we suspected RE our patient fulfilled all criteria proposed by Hart et al 1994b[14]. We suspected bilateral involvement in our patient once he presented with pseudobulbar state. Option of brain biopsy and therapeutic hemispherectomy was given to parents but they refused.

Steroids started at high doses and slowly tapered down have been reported to have beneficial effects on seizures and neurological functions in

several series particularly once started early in course[14,2]. Our patient received steroids with improvement in seizure control and neurodeficit but patient did not follow us for almost a year then.

Finally we started our patient on IVIG therapy and is planned to receive monthly doses of IVIG. Beneficial effects of IVIG on seizure and neurological function were reported in some case studies and in Hart's large series where IVIG is recommended as first line immunotherapy[14].

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