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CASE REPORT

Rasmussen's Encephalitis- Rare Epileptic Encephalopathy: A Case Report

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Abstract:

Rasmussens encephalitis is a chronic progressive neurological disorder of childhood. It is a rare inflammatory disease, characterized by intractable focal epilepsy, progressive hemiplegia, cognitive deterioration, unilateral inflammation of the cerebral cortex. Median age of onset is 6 year. Our case is a 7 year old boy presented in the department of Paediatric Neurology of BSMMU with the complaints of weakness of the right side of the body, focal onset seizure, cognitive declination and difficulty of speech. In addition to classical clinical presentation of Rasmussens encephalitis, MRI brain showed hemispheric atrophy of one cerebral hemisphere.

Key words: *Rasmussens encephalitis (RE), epilepsy, cortical atrophy*

Introduction:

Rasmussen's encephalitis (RE) is a chronic neurological condition characterized by progressive neurological deficits, refractory focal seizures and gradual loss of cognition. It was first described by Theodore Rasmussen in 1958. It is usually reported in children aged <10 years with median age 6 years, though it can be seen in adolescents and adults as well[1]. Aetiopathogenesis of RE is not clearly known, It is hypothesized that, It occurs due to chronic immunological inflammation to unilateral cerebral hemisphere leading to atrophy[2].Clinical staging are Prodromal stage- Characterized by low seizure frequency and rarely mild hemiparesis, Acute stage- Characterized by augmentation in the frequency of seizures often as epilepsiapartialis continua(EPC) and manifestation of neurological deterioration such as hemiparesis, hemianopia, cognitive deterioration, and aphasia. Residual stage- Characterized by permanent and stable neurological deficits. There is reduction in the frequency of seizure episodes neuroimaging has emerged as a useful marker of disease diagnosis. MRI of brain changes depends on duration of disease manifestation. MRI of brain showing initial swelling of brain volume with hyperintense signals then gradual loss of brain volume with hyperintense signals, finally progressive atrophy with normal signals. Immunotherapy is the one of the important treatment modality and later hemispherectomy may be required for pharmacoresistant seizure.

Case summary

A 7 year male child of consanguineous marriage parents presented with weakness of the right side of the body for 3 years. It is associated with repeated episode of tonic seizure involving right upper and lower limb. Each episode of seizure lasting for 1 to 2 minutes along with post ictal drowsiness. The seizure episode begins two and half months back. The seizure usually occur at night during sleep time. There is also history of speech difficulty, abnormalities of gait and recent memory loss. This child was delivered by normal vaginal delivery and cried immediately after birth. No history of neonatal seizure. According to parents statement there was history of trauma three years back after that he developed unconsciousness for ten days that was treated in local hospital with antibiotic and condition was improved, subsequently he

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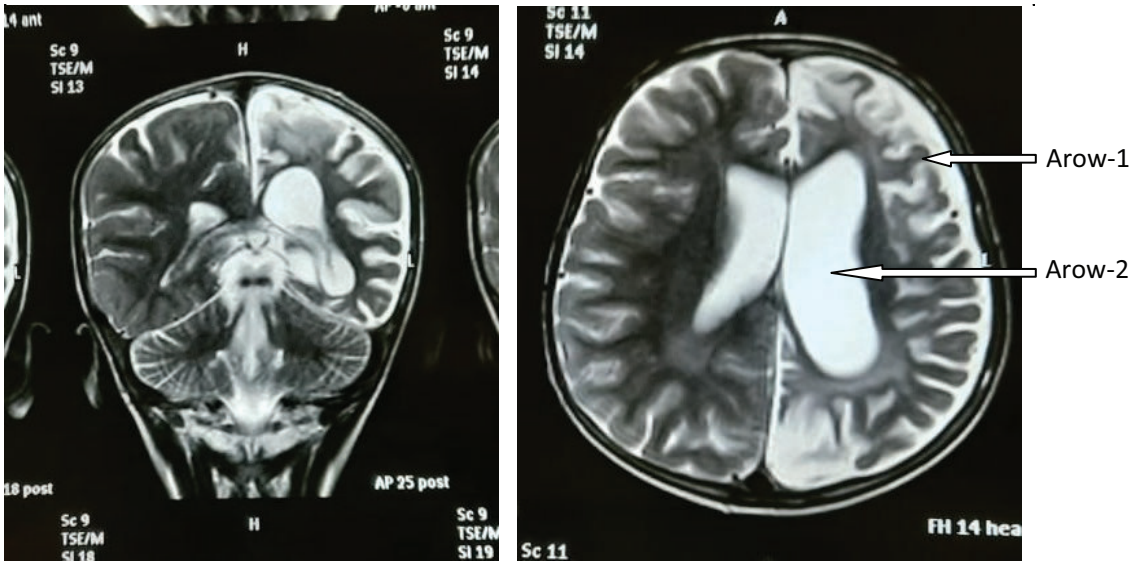

Fig.-1: *MRI findings shows T2WI hyperintense signal changes (Arow-1) noted at left cerebral hemisphere with volume loss and evacuee dilatation of left lateral ventricle (Arow-2). Left cerebral peduncle is smaller in size.*

developed weakness of right upper and lower limbs. On examination he was conscious, oriented. His vitals were stable. Nervous system examination shows right sided hemiplegia, exaggerated deep tendon reflexes (Biceps, knee, ankle) and extensor planter reflexes on right side. There was no sign of meningeal irritation. Cranial nerves were intact. Brain MRI T1WI shows hypo intense and T2WI & Flare shows hyper intense signal changes noted at left cerebral hemisphere with volume loss and exvacuo dilatation of left lateral ventricle. Left cerebral peduncle is smaller in size. Both lateral ventricles and 3rd ventricles are mildly dilated(Figure-1) .EEG report shows epileptiform discharges over B/L wide temporal and occipital regions (>Lt). There is localized dysfunction over left hemisphere. Background electrophysiological function is disturbed in left hemisphere. This patient was treated with multiple antiepileptic drugs and immunotherapy. He was improved gradually and discharged with advice. Now he was on regular follow up.

Discussion:

Rasmussen's encephalitis is a rare chronic inflammatory progressive neurological disorder. They usually present with focal motor seizures followed at varying time intervals by progressive loss of ipsilateral motor functions and cognitive impairment. The etiopathogenesis of RE is unknown. Etiologically, there are different theories for the pathogenesis.

This progression seems to be consistent with an immune mediated disease that is likely to be associated with both adaptive immune reactions, with T-lymphocyte responses, and innate immunity facilitated by both microglia and astroglia[3], autoimmune[4]against glutamate receptors or infective[5].Three stages have been proposed. The prodromal stage is manifested with mild signs (low seizure frequency and mild hemiparesis). The acute stage is characterized by frequent focal seizures, progressive hemiparesis, and cognitive deterioration. The residual stage is characterized by stabilization of neurological deficits and continuation of seizures, but less frequent than in the acute stage[6].Our patient had right sided hemiparesis, progressive cognitive impairment and focal motor seizure.

The diagnosis of RE is based on clinical, radiological, neurophysiologic and/or pathological features with emphasis laid on clinical and

radiological features[7].Diagnostic criteria were established by Bien et al. in 2005.(8).Brain MRI is an important tool for diagnostic assessment and follow-up in RE[7,8].The majority of patients show, at an earlier stage, unilateral enlargement of the ventricular system which is accentuated in the insular and periinsular regions. A T2/FLAIR hyperintense signal on MRI of brain is often present in the cortical or subcortical regions. Atrophy of the ipsilateral head of the caudate nucleus is a typical feature. In our case ipsilateral atrophy and T2 hyperintensity of left cerebral hemisphere including head of caudate nucleus are seen.MRI findings of our patient shows multiple T1WI hypointense and T2WI hyperintense signal changes noted at left cerebral hemisphere with volume loss and exvacuo dilatation of left lateral ventricle. Left cerebral peduncle is smaller in size. Both lateral ventricles and 3rd ventricles are mildly dilated.

Various abnormalities EEG are seen in patients with RE. Some unihemispheric findings such as impairment of background activity with persistent polymorphic delta waves and sleep spindles, focal slow activity, subclinical ictal discharges, and multifocal ictal discharges are strongly suggestive of RE[7,9]. The EEG of our patient showed multifocal epileptiform discharge Brain biopsy can also help the diagnosis, but it is not required in all RE cases.

RE can be treated by antiepileptic drugs, immunomodulator regimens, and surgery. The aim of these treatments is to reduce seizure severity and improve the motor and cognitive performance [7,8]. Pulses of high-dose methylprednisolone have been reported to be effective to stop disease progression[10].Intravenous immunoglobulin (IVIG) was used in some patients having RE with good results. The recommended dose is 2 g/kg monthly. The association of steroids and IVIG may be indicated when the two treatments alone are ineffective[11] .Plasmapheresis has good effects on seizures and neurological functions[11].

Medical treatments, such as using tacrolimus, rituximab, cyclophosphamide, azathioprine, and interferon,have been reported[10]. Our patient was treated by AED and methyle prednisolone and seizure was subsided gradually. Surgery seems to be the cure for the seizures and to improve cognitive outcome with consideration of inevitable sequelae (hemianopia, hemiparesis, and aphasia in the dominant hemisphere). Rehabilitation approach should be considered. It may improve the quality of life of RE patients.

Conclusion:

Rasmussen's encephalitis is a progressive disease involving one cerebral hemisphere, with management complicated by difficulties in making an early diagnosis. Neuroimaging has emerged as a useful marker of disease diagnosis. Further systematic randomized clinical trials are needed to clarify the efficacy of new nonsurgical treatment options that might be used to control seizures and preserve neurological function. Emphasis should be given on rehabilitation approach. It may improve the quality of life of RE patients.

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