

Rasmussen Encephalitis with Crossed Cerebellar Diaschisis in An 18 Years Old Female – A Case Report.

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Abstract:

Rasmussen encephalitis is a chronic neurological disorder, characterized by unilateral inflammatory involvement of the cerebral hemisphere, resulting in progressive neurological and cognitive deterioration. Imaging (CT and MRI) is extremely useful in disease detection, differentiation from other entities affecting unilateral cerebral hemisphere and staging of the disorder.

Keywords:

- Encephalitis (MeSH unique ID: D004660).
 - Rasmussen Encephalitis (MeSH unique ID: D004660).
 - Epilepsia Partialis Continua (MeSH unique ID: D017036).
 - Magnetic Resonance Imaging (MeSH unique ID: D008279).
 - Epilepsy (MeSH unique ID: D004827).
 - Seizures (MeSH unique ID: D012640).
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I. Introduction

Rasmussen encephalitis is a chronic inflammatory disorder of the central nervous system commonly affecting the paediatric age group. In 1958, it was initially reported by Theodore Rasmussen. Mean age of presentation of this entity is from 6 to 8 years and it affects both genders equally.^[1]

II. Case Report

An 18 years old female patient presented with a history of repeated episodes of tonic clonic seizures involving left upper and lower limbs. Each episode of seizure lasted for approx. 10 – 15 minutes. The first episode of seizure was recorded at the age of 4 years. There was no history of neonatal seizures, blood transfusion or family history of epilepsy. On examination, left sided hemiplegia with upper motor neuron type of facial palsy, exaggerated tendon reflexes on right side. No signs of meningeal irritation were noted. Magnetic resonance imaging of the brain was performed which revealed diffuse atrophy of right sided cerebral hemisphere with cystic encephalomalacia-gliotic changes. Gross ex-vacuo dilatation of the right lateral ventricle was noted (**Figure 1**). Reduced volume of right sided cerebral peduncle suggestive of Wallerian degeneration was noted (**Figure 2**). Atrophy of contra-lateral left cerebellar hemisphere was noted (**Figure 3**). Based on the imaging findings and clinical profile of the patient, a diagnosis of Rasmussen encephalitis with crossed cerebellar diaschisis was given.

Figure 1

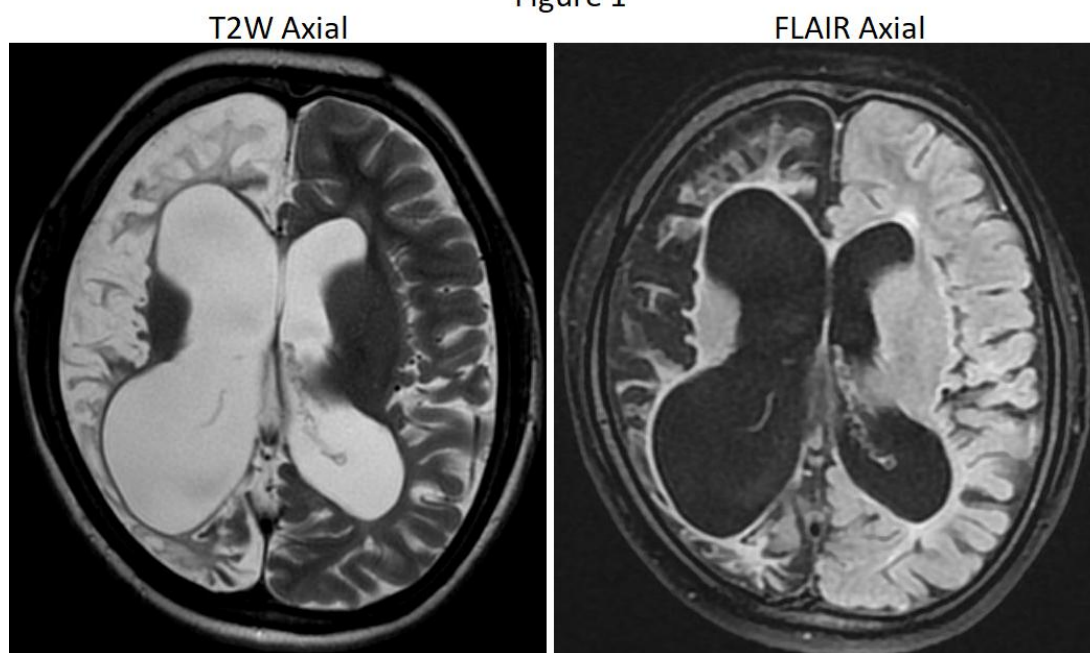


Figure 1: Axial T2WI and FLAIR showing diffuse atrophy of right sided cerebral hemisphere with cystic encephalomalacia-gliotic changes. Gross ex-vacuo dilatation of right lateral ventricle was noted.

Figure 2: T1W Axial

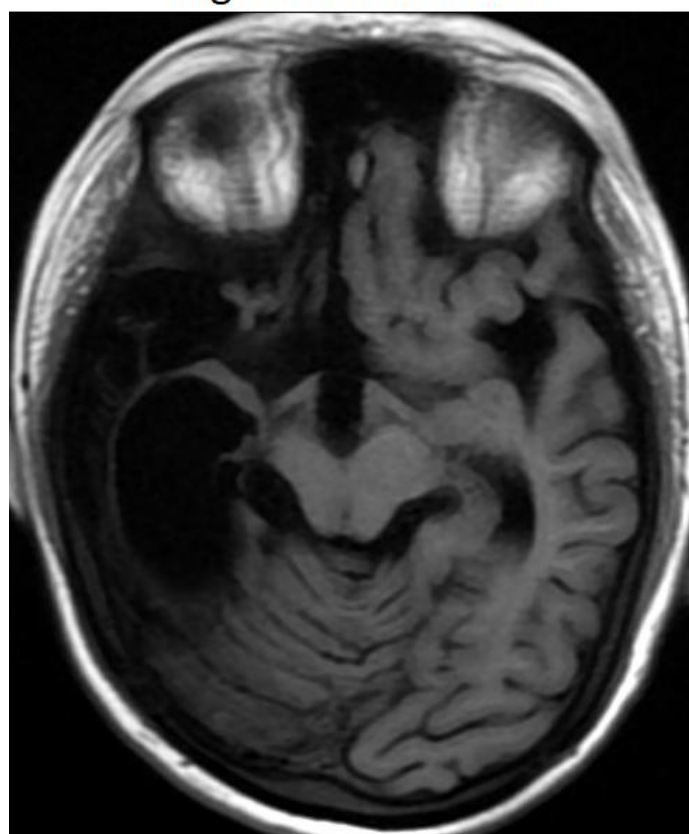


Figure 2: Axial T1WI showing reduced volume of right sided cerebral peduncle suggestive of Wallerian degeneration.

Figure 3

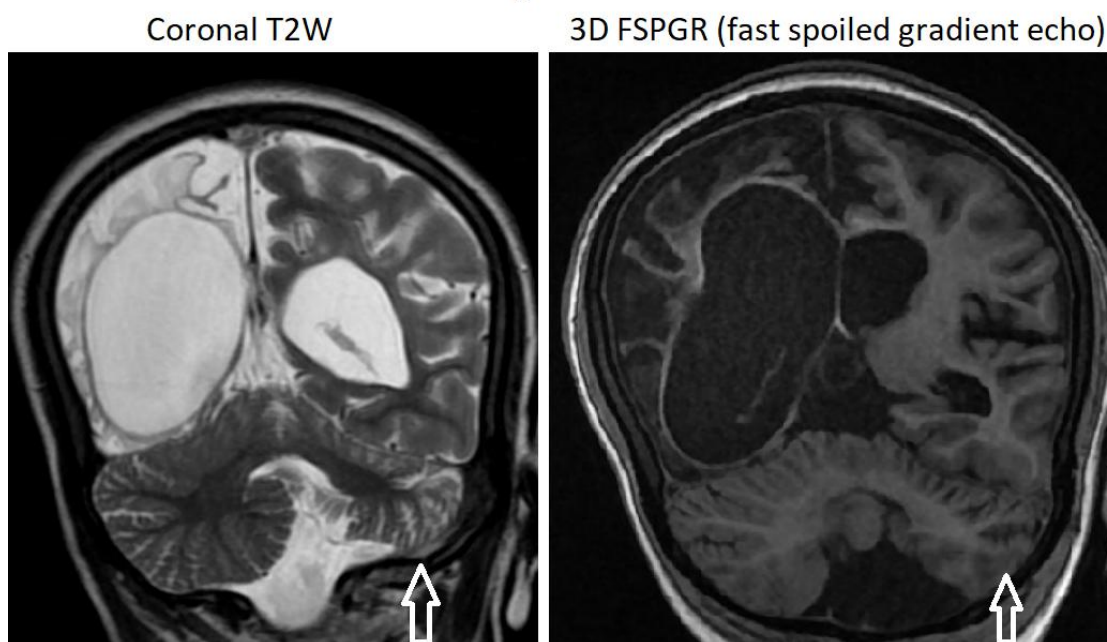


Figure 3:Coronal T2WI and 3D FSPGR (Fast spoiled gradient echo) showing atrophy of contra-lateral left cerebellar hemisphere (arrows).

III. Discussion

Rasmussen encephalitis is a chronic inflammatory disorder of the central nervous system commonly affecting the paediatric age group. In 1958, it was initially reported by Theodore Rasmussen. Mean age of presentation of this entity is from 6 to 8 years and it affects both genders equally.^[1]

The onset of this entity is in childhood, however adult onset cases have been reported.^[2]

In this disorder, focal onset seizures (characteristic of *epilepsia partialis continua*) following a progressive course are seen with neurological deterioration (progressive hemiparesis). As this condition is chronic progressive *epilepsia partialis continua* of childhood, it is included in the international classification of epilepsies and epileptic syndromes.^[3]

Manifestations of Rasmussen encephalitis include: polymorphous intractable seizures, progressive hemiparesis, cognitive deterioration, hemianopia and aphasia (due to involvement of dominant hemisphere).^[4] Eventually, there is residual stage consisting of persisting difficulty to treat relapsing epilepsy.^[5]

Disorders causing chronic unilateral epilepsy and hemiplegia are mainly development anomalies, lesions caused by anoxic-ischaemic damage and Rasmussen's syndrome. Lesions caused by anoxic-ischaemic encephalopathy in the perinatal or prenatal period are most common.^[6]

Characteristically, the disease is restricted to unilateral cerebral hemisphere.^[2]

Its etiology is unknown. There have been suggestions like auto-immune process related to autoantibodies to the GluR3 proteins or immune mediate disorder with viral infection. The diagnosis is formed in advanced cases of the disease based combined clinical-radiological imaging approach consisting of intractable seizures and unilateral cerebral hemispheric atrophy.^[2]

Based on a retrospective study of 39 patients, Bien et al proposed a MRI model of Rasmussen encephalitis consisting of five stages. Stage 0 showed normal brain parenchymal volume and normal T2W/FLAIR signal intensity. Stage 1 showed cortical swelling with T2W/FLAIR hyperintense signal intensity. Stage 2 showed normal brain parenchymal volume and multifocal areas of T2W/FLAIR hyperintense signal intensity affecting white matter or cortical grey matter of unilateral hemisphere, more prominent at peri-insular and insular region. Stage 3 showed unilateral hemisphere parenchymal atrophy evident as widened cortical sulci and dilated ipsilateral lateral ventricle. Associated T2W/FLAIR hyperintense signal intensity was also noted. Stage 4 showed prominent parenchymal atrophy of unilateral hemisphere with normalization of T2W/FLAIR signal abnormalities.^[1]

In 1992, mild to moderate atrophy of the putamen with periventricular hyperintense signal intensity were initially reported by Tien et al. Cendes et al reported MR spectroscopic findings consisting of reduced N-acetylaspartate in the periventricular region of frontal horn of lateral ventricle. One of the prominent changes that has been reported in Rasmussen encephalitis is temporo-insular atrophy.^[6]

Ipsilateral atrophy of the caudate nucleus head is typical, but not a constant feature and can be seen in the early phase of the disorder.^[5]

As described by recent radiological volumetric approach, highest rate of volume loss is seen in the acute clinical phase in the first 8 months of the disease.^[5]

Even when MRI detectable changes of atrophy are minimal, functional studies using 18F-FDG PET demonstrate diffuse unilateral cerebral hypometabolism.^[5]

Differential diagnosis of Rasmussen encephalitis are: Sturge Weber syndrome, Dyke Davidoff Masson syndrome, unihemispheric cerebral vasculitis and hemimegalencephaly. Sturge weber syndrome is a neurocutaneous syndrome characterized by leptomeningeal venous angiomatosis, capillary venous malformations in areas of trigeminal nerve distribution, seizures, hemiplegia and dementia. On imaging, it is evident as cerebral atrophy with gyriform calcifications, ipsilateral enlarged choroid plexus and enhancing angiomas. In hemimegalencephaly, neuronal migration defects (focal/diffuse) with pachygyria, polymicrogyria, heterotopia, and enlarged unilateral hemisphere with ventriculomegaly.^[1]

IV. Conclusion

In patients presenting with seizures in childhood period, imaging plays an essential role in narrowing down the differential possibilities from a broad spectrum to a limited number of conditions. Cases presenting as seizures with childhood onset coupled with imaging features like unilateral cerebral hemispheric atrophy without changes of calvarial thickening or gyriform calcifications, imaging plays a crucial role in establishing the diagnosis of Rasmussen encephalitis and follow up of these patients for progressive cognitive function deterioration.

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