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Case Report

Rasmussen's Encephalitis: A Report of a Moroccan Pediatric Case

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Abstract: Rasmussen's encephalitis (RE) is a rare progressive inflammatory disease of the central nervous system. It is characterized by unilateral hemispheric atrophy, pharmacoresistant focal seizures, and progressive neurological deficit. The exact etiopathogenesis still remains unknown. Brain imaging plays an important role in diagnosis and follow-up. Fluctuation of lesions in brain imaging was reported in few cases. We report the case of a 12-year-old boy initially with intellectual development disorders presented to the pediatric emergency department with complaints of multiple episodes of seizures resistant to treatment. The MRI showing typical aspects of Rasmussen's encephalitis.

Keywords: Encephalitis, inflammatory disease, antiepileptic drugs, steroid.

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INTRODUCTION

Rasmussen's encephalitis (RE) was first described in the late 1950s [1]. It is a rare neurological disease of childhood characterized by unilateral hemispheric atrophy, pharmacoresistant focal seizures, and progressive neurological deficits. The exact etiopathogenesis still remains unknown. Brain imaging plays a pivotal role in diagnosis and control of disease progression [2]. We report on the clinical, imaging and electrophysiological, data of an additional pediatric case.

CASE DETAILS

A 12-year-old patient with impaired intellectual development (cognitive impairment) presented to the pediatric emergency department complaining of multiple epileptic seizures. There was no history of joint pain, rash, tuberculosis, diabetes mellitus, jaundice, head injury, or a family history of epilepsy. His mother reported that these episodes had begun when he was one year old. The child was put on triple therapy without further investigation, and did not improve on treatment.

According to his mother, the episode began with a focal tremor of the right upper limb, which spread to the lower limbs, progressing to a four-limb tremor and loss of consciousness. On examination, the left upper and lower limbs showed decreased tone and power (2/5). Brain MRI then showed cortico-subcortical atrophy of the right cerebral hemisphere responsible for dilatation of the homolateral lateral ventricle, which had a scalloped wall and several septa (Figure 1). A T2 and FLAIR hypersignal of the subcortical white matter is associated with gliosis. Hypotrophic appearance of right basal ganglia, corpus callosum and cerebral peduncle (Figure 2). Discrete subtentorial cortical atrophy more marked on the left (Figure 3). The electroencephalogram showed deterioration in background activity (Figure 4).

Given the clinical course and MRI finding, the diagnosis of RE was performed. Numerous regimens of antiepileptic drugs were prescribed (valproic acid 2 g/day, carbamazepine 1400 mg/day, levetiracetam 2500 mg/day, clonazepam 4 mg/day, and piracetam 1600 mg/day). A monthly steroid pulse at a dose of 1 g/day for 3 days was administered during 12 months. Azathioprine was prescribed at the dose of 100 mg/day. Partial control of seizures was obtained.

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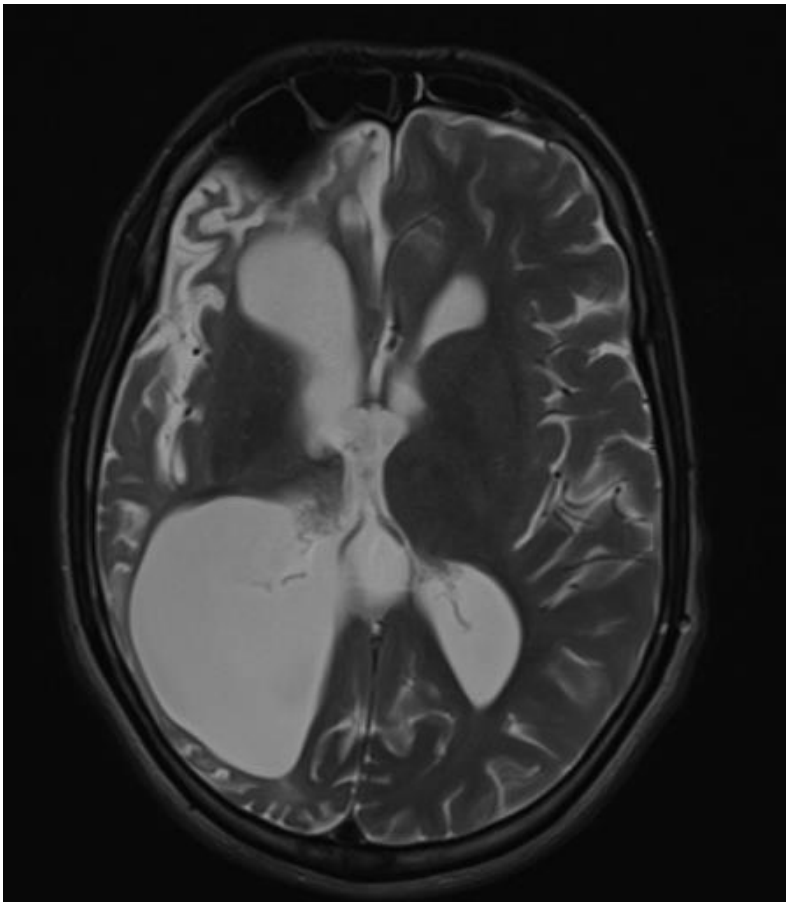

Figure 1: Cortico-subcortical atrophy of the right cerebral hemisphere responsible for dilatation of the homolateral lateral ventricle, which had a scalloped wall and several septa

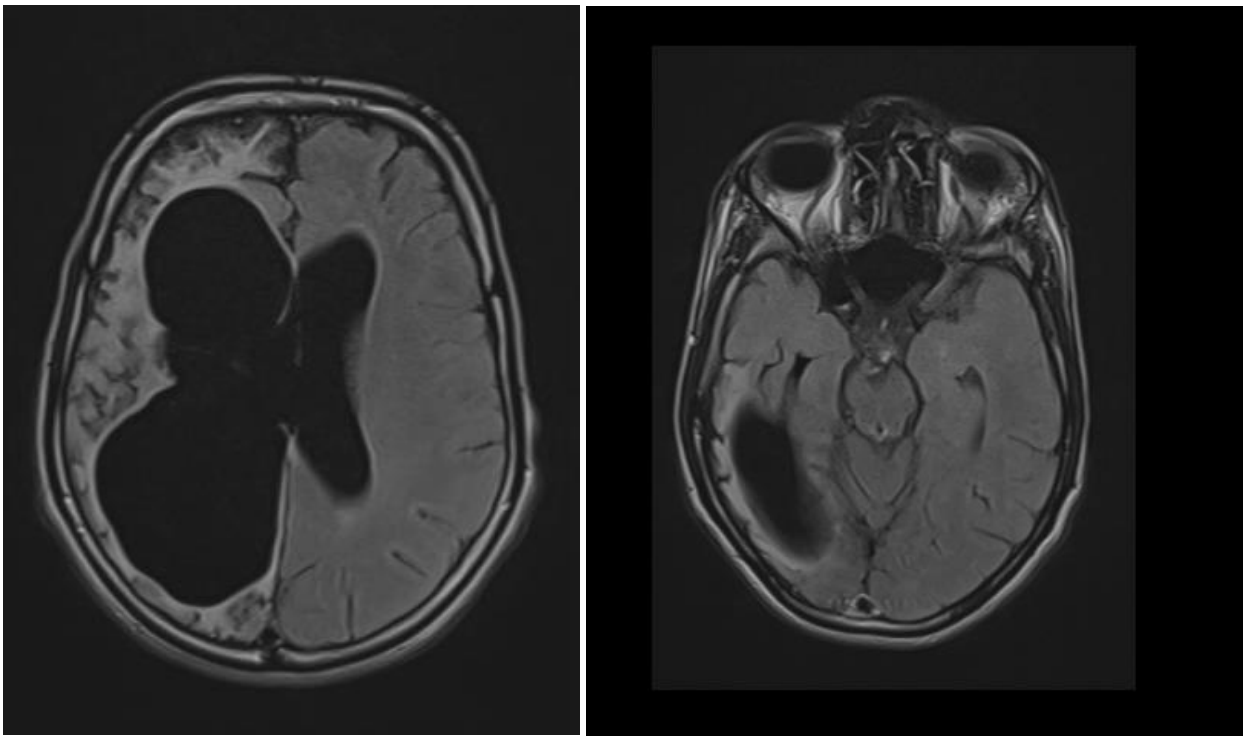

Figure 2: T2 and FLAIR hypersignal of the subcortical white matter is associated with gliosis. Hypotrophic appearance of right basal ganglia, corpus callosum and cerebral peduncle

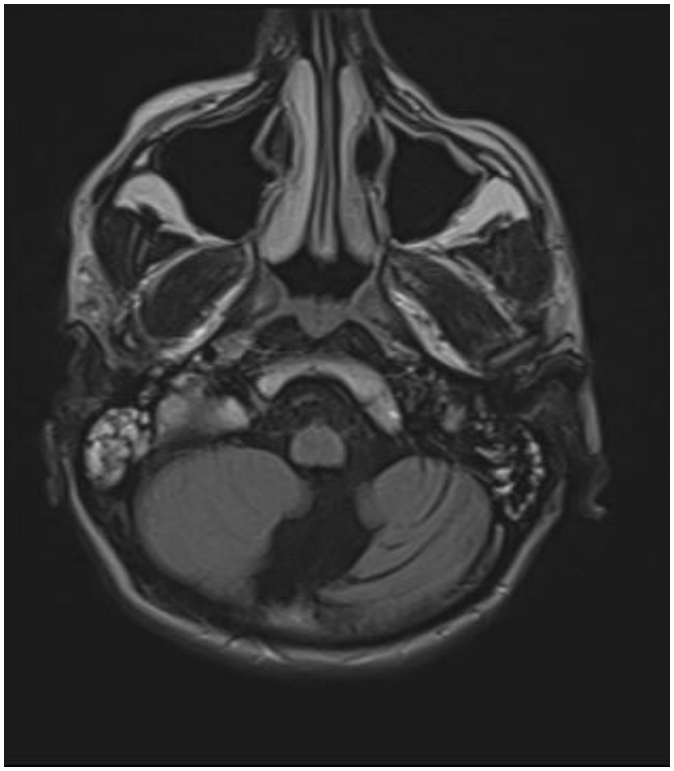

Figure 3: Discrete subtentorial cortical atrophy more marked on the left

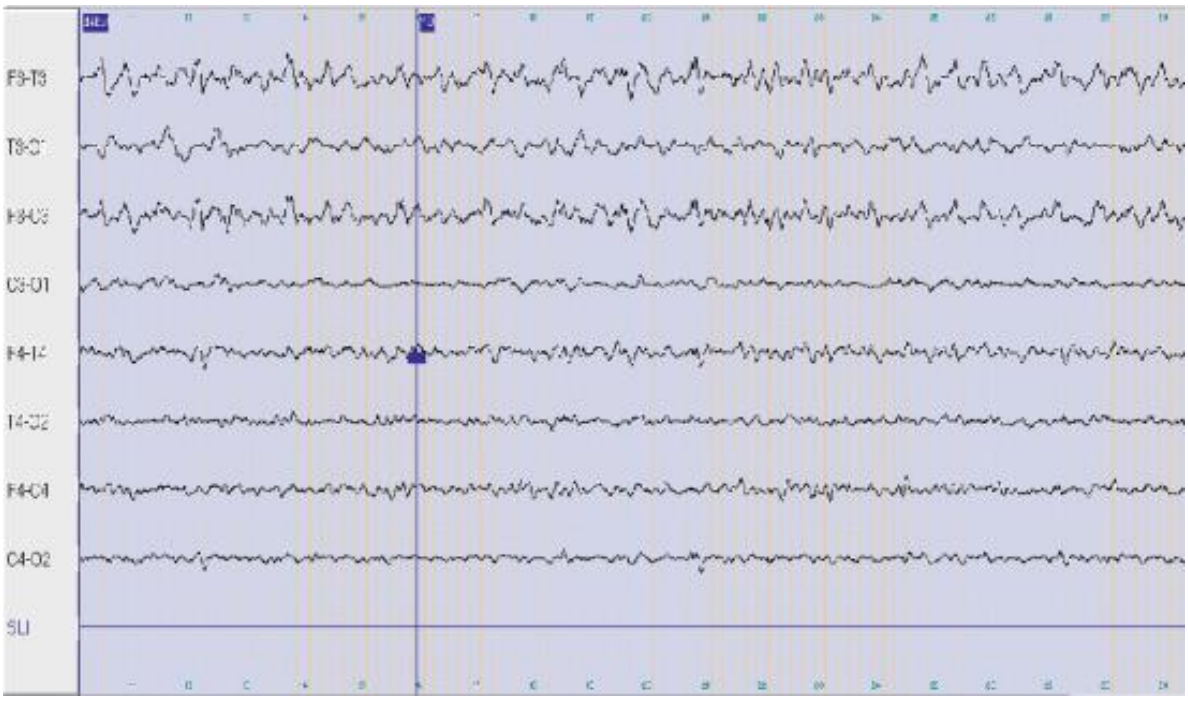

Figure 4: EEG showing asymmetric background activity

DISCUSSION

Rasmussen encephalitis is a chronic localized encephalitis of uncertain cause affecting one brain hemisphere (half) progressively. It usually begins in childhood between 6 and 8 years. It was first reported by the American neurologist Theodore Brown Rasmussen in 1958 when he suggested a viral etiology in his original paper [3]. Nevertheless, no significant signs of viral infections were neither excluded nor confirmed with inflammation, gliosis and loss of neurons being all found histopathologically [4]. Clinically, this disease is characterized by refractory epilepsy followed by progressive hemiparesis or cognitive decline which describes 3 phases (Figure 5).

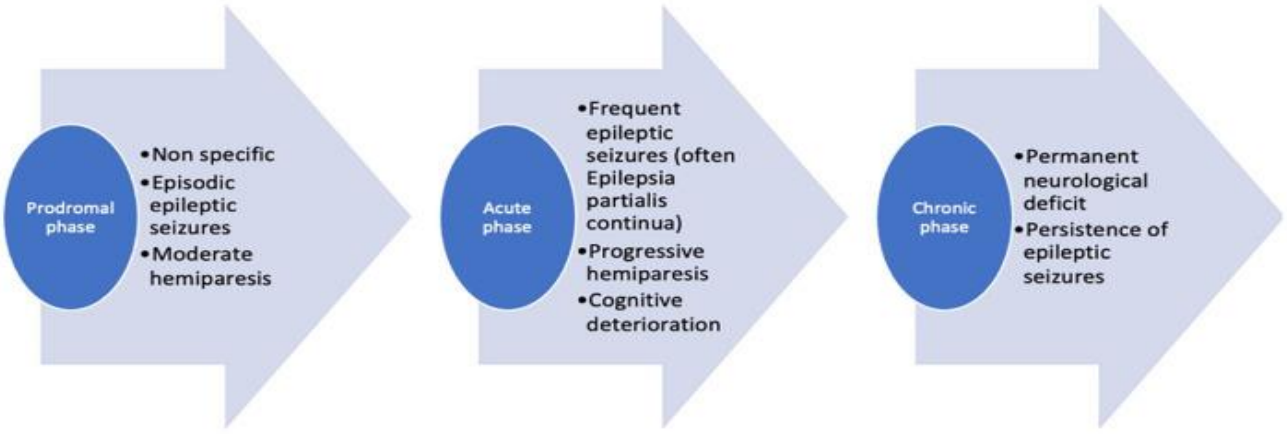

Figure 5:

MRI is the imaging of choice in evoking the diagnosis and findings vary with the disease stage [5]. Initially, unilateral focal cortical swelling may be seen. With time, T2/FLAIR hyperintensity develops in the cortex and subcortical white matter of the involved side. With the disease progressing into chronic stages, we may witness unilateral cerebral and basal ganglia atrophy (Figure 2). In the final phase, hyperintensities may disappear, leaving a marked hemi cerebral atrophy. Those stages were proposed by Bien *et al*., based on a retrospective study on 10 patients and their MRIs (initial and follow-up) and focus essentially on T2 sequences (Figure 6) [6].

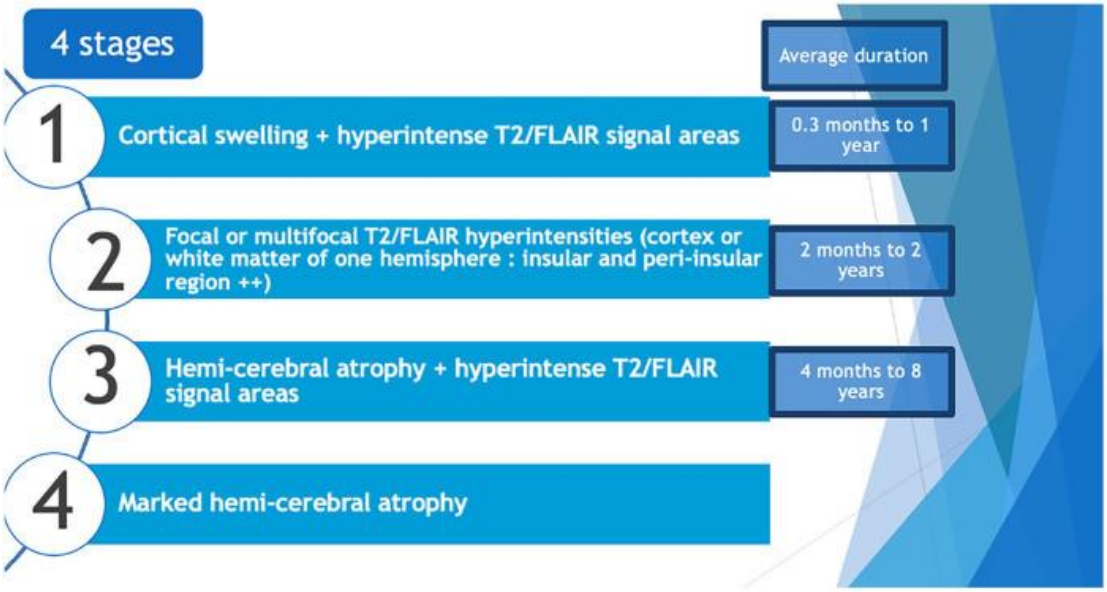

Figure 6:

We can conclude that our patient installed hemiparesis which means she was in the chronic phase with the MRI showing hemi cerebral atrophy and an area of T2/FLAIR hypersignal which matches stage 3.

In early disease stages, electroencephalograms (EEG) may contribute to the diagnosis of RE [7]. Various abnormalities 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 [2, 3]. The EEG of our patient showed deterioration in background activity.

Brain biopsy can also help the diagnosis, but it is not required in all RE cases. The characteristic histopathological features are microglial and lymphocytic nodules, neuronal loss, neuronophagia, and perivascular cuffing, confined to one cerebral hemisphere with frontoinsular predilection [4, 2].

Our patient was put on antipelpetics and A monthly steroid pulsen administered for 12 months. as well as immunosuppressant.

RE can be treated by antiepileptic drugs, immunosuppressive and immunomodulator regimens, and surgery. The aim of these treatments is to reduce seizure severity and improve the motor and cognitive

performance [4, 7]. Frequently, seizures are resistant to antiepileptic drugs [8]. Patients receiving immunotherapy had a beneficial effect on seizure frequency and delayed deterioration [9]. Steroids, prescribed in our case, are the most effective treatment and the most widely used. Pulses of high-dose methylprednisolone have been reported to be effective to stop disease progression [8].

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 [10].

Plasmapheresis has good effects on seizures and neurological functions. It contributes to assessing the mental and residual motor function before surgery. The frequency was three to six single volume exchanges on consecutive or alternate days every 2 to 8 weeks [10].

Other medical treatments, such as using tacrolimus, rituximab, cyclophosphamide, azathioprine, and interferon, have been reported [8]. Our patient was treated by azathioprine.

Surgery (anatomic hemispherectomy, functional hemispherectomy, perisylvian hemispherotomy, trans-sylvian hemispherotomy, and central/vertical hemispherotomy) seems to be the only cure for the seizures and to improve cognitive outcome. However, inevitable sequelae (hemianopia, hemiparesis, and aphasia in the dominant hemisphere) should be considered [4]. Rehabilitation approach should be considered. It may improve the quality of life of RE patients.

Cerebral hemiatrophy has other causes such as Dyke–Davidoff–Masson syndrome (DDSM), hemi-megalencephaly, and Sturge-Weber Syndrome, with them all generally associated with hemiplegia and epilepsy. Some clinical and MRI findings are of great help in differentiating these etiologies from RE.

CONCLUSION

Rasmussen's encephalitis remains a rare pathology which can be a diagnostic dilemma for some physicians. We should always evoke ER in front of an elevated frequency of complex partial seizures with a post-ictal deficit in patients between 1 and 15 years old), especially when associated with a "normal" initial imaging and a disease progression marked with drug-resistant epilepsy with progressive atrophy of a hemisphere and areas of T2 hypersignal. Thus, MRI findings can be highly suggestive of Rasmussen Encephalitis diagnosis and also in excluding other causes of cerebral hemiatrophy.

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