

The Ultimate Stage of Rasmussen's Encephalitis

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Abstract

Rasmussen encephalitis (RE) is a rare and progressive disease of childhood. We report the case of 14-year-old girl who presented with recurrent partial and generalized epileptic seizures with mental impairment and progressive left sided hemiplegia. EEG showed slowing of the background activity and paroxysmal discharges of slow waves in the right hemisphere. Serial MRI brain scans revealed an important atrophy of the right hemisphere with dilatation of the ipsilateral lateral ventricle. The residual phase, characterized by decrease in seizure frequency and stabilization in the hemiparesis, constitute the ultimate stage of RE.

Keywords: Rasmussen's encephalitis; Residual stage; Epilepsy; Hemiparesis; MRI

Introduction

Rasmussen's Encephalitis (RE) is a well-recognized cause of drug-resistant focal epilepsy, followed by progressive hemiparesis with cognitive impairment, and accompanied by unilateral cerebral atrophy due to chronic localized encephalitis of unknown etiology.

Case Report

We report the case of a 14-year-old, right handed girl, with no family history of epilepsy or neurological disease. She was born at 38 weeks following an uneventful pregnancy, labor, and delivery. She first became symptomatic with seizures at 4 years of age, in September 2002. The seizures started with a focal jerking of the left upper limb followed by spread to the left lower limb and cheiro-oral region. The patient was initially followed in a local general medicine center where a CT scan was performed and returned normal. Cryptogenic epilepsy diagnosis was therefore retained. Three to four years later, focal seizures progressed to a loss of consciousness and jerking of all the 4 limbs. Moreover, a progressive deterioration in her language function with difficulty in speaking and comprehending had been noticed by her family members. In July 2012, the patient was presented to our department because of an increase of seizure's frequency from the initial once a

month to almost daily, despite being on multiple antiepileptic drugs (Valproate, Carbamezipine and Clobazam) with good compliance. On examination, she was conscious, and she had a severely impaired comprehension with dysarthric output speech. There were no frontal cortical release signs. She also had a left sided proportional hemiparesis including a slight ipsilateral lower facial weakness with increased tone. Deep tendon reflexes were brisker on the left side with an extensor left plantar reflex. There were no cerebellar or extrapyramidal signs. Furthermore, systemic examination was unremarkable. The patient underwent a complete blood screening including blood count, erythrocyte sedimentation rate, C reactive protein, serum glucose, electrolytes, liver and kidney function test, calcium, lipid profile, serum angiotensin-converting enzyme levels, cerebro-spinal fluid analysis and antinuclear antibodies, all of which were normal. ECG and X-ray chest were normal. MRI revealed an important cortical atrophy of the right hemisphere (Figure 1) with dilatation of the ipsilateral lateral ventricle (Figure 2). There was no evidence of contrast enhancement or calcification. EEG showed slowing of the background activity and paroxysmal discharges of slow waves in the right hemisphere. After admission, Clobazam infusion was required for seizure control. The patient received also 3 IV bolus of methylprednisolone (500 mg each). On discharge, she was referred to a specialized epilepsy unit for neurosurgical intervention. Unfortunately, neurosurgical evaluation considered that surgery was useless because of the delayed stage of the disease.

Comment

RE was first described in 1958, as 'focal seizures due to chronic localized encephalitis' in three patients [1]. So far, a few hundred of cases have been reported in the literature. In fact, RE is a rare chronic inflammatory disease presenting as progressive focal cortical inflammation and tissue destruction usually involving a single brain hemisphere. It usually begins in the first decade of life, with rare

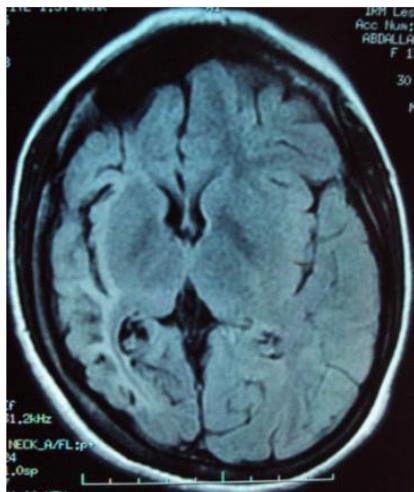


Figure 1: MRI-brain, Axial flair demonstrating severe cortical atrophy of the right hemisphere.

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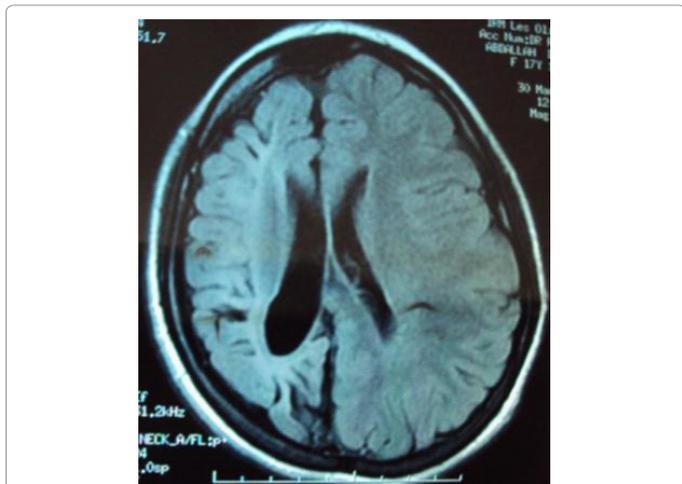


Figure 2: MRI-brain, Axial flair showing enlargement of the right ventricle.

reported onset in adolescence and early adulthood [2]. Neurologic symptoms are divided into three stages including intractable seizures, progressive hemiplegia, and mental deterioration before entering an ultimate phase of deficient neurologic state. The proposed clinical diagnostic criteria for RE include focal seizures, unilateral cortical deficits, unihemispheric EEG slowing with or without epileptiform activity and unihemispheric focal cortical atrophy with or without increased cortical or subcortical T2 and/or FLAIR signal in MRI [3]. Although our patient met all the criteria of RE, the MRI findings can be confusing with other neurological diseases such as anti-NMDA receptor encephalitis. However, the progressive evolution with

normal CSF composition seems to be against this diagnosis. It was demonstrated that ^{18}F -FDG PET is better than concomitant MRI for detecting the lesions and their extension early in RE [4]. In our case, this examination was not carried out because of the late stage of the disease, while being exceptionally used in our country. According to the different clinical manifestations and phases of the disease, the therapeutic strategy includes both medical and surgical options. Most patients with RE are refractory to conventional antiepileptic drugs, thus, corticosteroid and intravenous immunoglobulin treatment should be considered in the early stages of the disease [5]. The only recommended therapeutic approach is surgical hemispherectomy, which is performed successfully to treat medically intractable hemispheric epilepsy [6]. This option may not be useful in the ultimate stage of RE.

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