CASE REPORT

Rasmussen's encephalitis

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Abstract

Background: Rasmussen's encephalitis is an uncommon chronic inflammatory disease disorder of unknown etiology.
Objectives: The aim of this report was to create an awareness of Rasmussen's encephalitis in our clinical practice in Africa.
Materials and Methods: We searched online for literature on Rasmussen's encephalitis with an emphasis on documented cases in Africa and in blacks all over the world.
Results: Master F.M., a 14-year-old male, presented with history of recurrent seizures on three occasions. The first was after an upper respiratory tract infection, the second was a post-complicated meningo-encephalitis and the third episode was associated with receptive aphasia, hemiparesis and intellectual impairment. Neuroimaging studies revealed cerebellar atrophy and infarction of territory of the middle cerebral artery. His electroencephalogram showed bi-frontal theta activity left>right and poly spikes left>>right, diagnostic of complex partial seizures. In a period of 12 months, his gait and speech had improved while his intellectual impairment permitted re-admission into a lower academic class. He is presently on phenytion and prednisolone tablets only.
Conclusion: Master F.M. had all the clinical features buttressed with neuroimaging results of a clinically probable Rasmussen's encephalitis. However, an advanced neuroimaging study is needed to detect and quantify hemispherical volume loss.

Key words: Rasmussen, encephalitis, disease

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Introduction

Rasmussen's encephalitis is a chronic inflammatory disease of unknown origin, usually affecting one brain hemisphere, described by Rasmussen and co-workers in 1958.[1] Locally, this is the first reported case in our 6 years of neurology practice at Lagos State University Teaching Hospital. It has been reported in adolescents, adults and more in children less than 10 years of age.[1,2] It is an uncommon unilateral hemispheric disease. Etiologically, there are different theories.

A school of thought support an autoimmune pathology.[3] Supporting this theory was presence of antibodies said to bind to type-3 glutamate receptor (GLUR3).[3] Another study documented cytotoxic T cells reacting against nerves taken from the affected hemisphere's brain tissue.[4] Decades back, it was postulated to be an infective process.[1] Recently, cytomegalovirus and herpes simplex virus 1 viral sequences has been detected at autopsy.[5] Trigger factors include viral infections, bacterial infections and past history of head injury. Clinical presentation is dependent on anatomical site and age at presentation. This includes the following: Epilepsy, focal or continuous, neurological deficits; aphasia (receptive or expressive); and cognitive deficits (memory intellectual impairment, neuropsychological deficits problems).

There is a clinical staging called the Montreal Neurological Institute (MNI).[6]

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Stage 1 is the period from the first epileptic seizure until the beginning of stage 2. Stage 2 is defined as the acute phase of the disease, with a rapid increase in seizure frequency (to >10/day) accompanied by the development or deterioration of a hemiparesis until the completion of neurological deterioration. Stage 3 is defined as a relatively stable state, with a permanent hemiparesis and a seizure frequency lower than that during stage 2. Neuroimaging studies have proven that brain tissue volume loss was significantly higher in the affected than in the unaffected hemispheres at a rate of 29.9 cm^3/year vs. 6.8 cm^3/year.[7] There is decreased cerebral blood flow and, generally, a hypometabolic state of the affected side.[8]

Case Report

Master F.M., a 14-year-old male, presented with a 3-week history of recurrent seizures and 1 week history of fever. The patient's medical history has been uneventful until March 2006, when he presented to the emergency unit with clinical features suggestive of sepsis with meningeal extension. He was first reviewed as a consult to the neurology unit, who made a diagnosis of meningo-encephalitis.

During the first review, cognitive functions were intact and he went back to school and was coping academically. He was re-admitted again in January 2008 after a 3-week history of left upper limb motor seizures, poor response to call and vomiting as a case of complicated meningo-encephalitis±cerebral abscess. These features were recurrent on admission and the neurology unit was invited again. Additional features included a low-grade fever associated with dry cough and weakness of the left half of the body. His genotype was AA. He had an undiagnosed allergic rhinitis that comes on twice in a year, which is relieved with one or two cough tablets. He had an uneventful minor fall before the age of 5 years.

Salient negatives

Blood transfusions, neurosurgery, peri- and post-natal events, developmental milestones, traumatic head injury, recreational drug use, proven allergies and family history of epilepsy. He has a younger brother in a monogamous family. His academic performance was satisfactory (form IV) until after the second ictus of which patient had to stop school. He did not exhibit neuropsychiatric features.

Clinical examination revealed a mildly pale, febrile, mildly dehydrated lanky adolescent. He was not icteric and his peripheral lymph nodes were not enlarged. He was conscious and alert with receptive aphasia. There was a decline in intellectual functions, evident by poor school reports. He could cooperate with the clinician for the Foinstein Minimental Status. There were no signs of meningeal irritation. All cranial nerves were intact. He walked with a mild hemiplegic gait. He had grade IV left spastic hemiparesis. Other systems were essentially normal.

Diagnosis

Focal encephalitis (Rasmussen's encephalitis)

The following differentials were entertained: Mollaret's meningitis, space occupying lesion (brain abscess, brain tumor) and epilepsy.

Results of his cerebrospinal fluid analysis: Xanthochromic, gram positive cocci and cell count $<5 \times 10/mm.^3$ Glucose was 59 mg/dl, protein 100 mg/dl and culture yielded no growth in 24 h. Antinuclear antibodies were 20.5 IU/ml. The cerebrospinal fluid sample was not sent for tumor markers and malignant cell examination.

Negative results were obtained for Venereal Diseases Research Laboratory, rheumatoid factor and retroviral I and II. Other results of patient's laboratory investigations, obtained at different times whilst on admission [between 2/082006 – 20/01/2008] revealed the following: his packed cell volume ranged from 34% - 38%; white blood cell count 7,800 – 11000mm^3 with a neutrophilic predominance; erythrocyte sedimentation rate 15 – 118 mm/hour; platelets count 175,000 – 504,000 109/L. His blood film picture showed anisocytosis and poikilocytosis whilst parasitology revealed few malaria parasites. However electrolytes, urea, creatinine and urinalysis were within normal limits

His electroencephalogram showed low amplitude 13 hertz/cycle alpha alfa rhythm, moderate β activity, bifrontal theta activity left>right and poly spikes left>>right [Figures 1]. The computerized tomogram revealed a wedge-shaped hypodense non-enhancing lesion in the right parietal lobe, which was indicative of infarction. Prominence of ventricles with widening of the cerebellar sulci was suggestive of cerebellar atrophy. (Patient till date has no cerebellar signs.) The radiologist concluded with infarction in the middle cerebral artery territory [Figures 2a and 2b].

He was placed on the following drugs: Phenytion, prednisolone, omeprazole and intravenous antibiotics. He is coping well in primary 2.

His total prednisolone dose is now 30 mg and phenytion total dose is 400 mg daily, all in divided doses.

The patient and his parents visit the neurology outpatient clinic twice a year. His last review was in September 2009.

Discussion

There was an 18-month interval between the 1st and the 2nd admission. His gait and speech improved after the

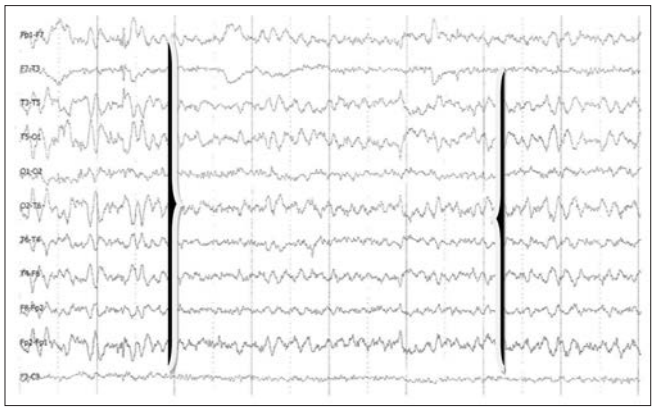

Figure 1: Interictal electroencephalogram of the patient

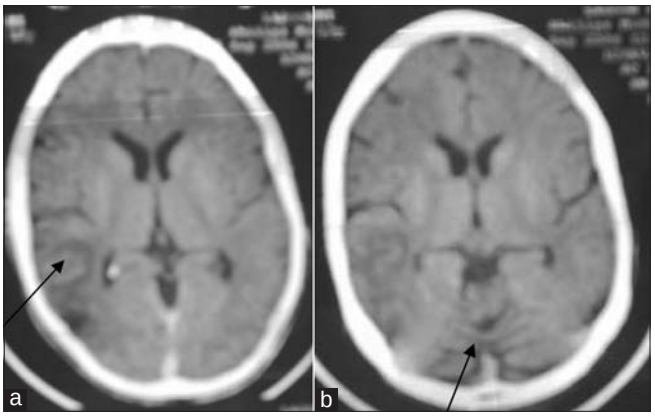

Figure 2: Computed tomograms of the patient. (a) Wedge shaped hypo dense non-enhancing lesion in the right parietal lobe. (b) Prominence of ventricles with widening of the cerebellar sulci

1st admission. Seizures abated soon, but progress in intellectual impairment was sluggish. He could not perform at the secondary school level. He was re-admitted into a primary school and demoted into primary class II. Our patient's cognitive decline cannot be explained with the differentials entertained (Mollaret's meningitis, brain abscess, brain tumor and epilepsy). ALthough epileptic seizures can start at this, they are not accompanied by recurrent infections and, when under early control with antiepileptic drugs, cannot explain patient's cognitive decline. Our patient's seizure history is more in keeping with the natural history of a Rasmussen's encephalitis.

Management of Rasmussen's encephalitis is tailored to control epilepsy and modify disease progression with the aid of the following: Antiepileptic drugs, immune suppressants, immune modulators, plasmapheresis and intravenous immunoglobulin.

Our patient manifested with different seizure types at different stages. He was actually managed with sodium valproate, carbamazepine and phenytion. These antiepileptic drugs were used at different times while on admission and were not used all together at once. Phenytoin was preferred due to its different routes of administration and it being a second choice for all manifested seizure types. Hemispherectomy might be of benefit to some young patients (partial/total).[9] This is usually performed only at later stages of the disease, when a patient has developed a fixed hemiparesis with loss of fine finger movements. Our patient does not qualify for surgical management.

Conclusion

Retrospectively, our patient presented with the natural history of a clinically probable Rasmussen's encephalitis. Although we could not perform advanced neuroimaging studies, which will confirm and quantify hemispherical volume loss, he is presently in MNI stage 3.

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