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Case Report

Adult-Onset Rasmussen´s Syndrome-Case Report

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Abstract

Adult-onset adult Rasmussen´s syndrome is a rare, chronic and progressive disease of the central nervous system characterized by unilateral cerebral cortex atrophy due to immune-mediated inflammatory response. The disease is characterized by focal drug-resistant epilepsy, contralateral progressive hemiparesis, and cognitive impairment. Despite the higher prevalence and earlier precocity in children, cases of adult-onset are reported and may present atypical clinical course.

Case Report

A 54-year-old female patient presented with a 10-year history of slowly progressive left hemiparesis, cognitive impairment and epilepsy (complex partial seizures with secondary generalization). The EEG demonstrated slow base activity with delta waves in the right cerebral hemisphere. Brain MRI confirmed the presence of right cerebral hemisphere atrophy associated with suggestive ipsilateral temporal mesial sclerosis (Figure 1). Laboratory tests, including NMDA and VGKC receptors, as well as paraneoplastic investigation were normal. Treatment with antiepileptic drugs (VA, CBZ, CLB) were initiated and presented with partial control of seizures. The use of corticosteroids in oral therapy and in intravenous therapy (pulse therapy) was introduced with suboptimal response [1,2]. This case report alerts to the possibility of adult-onset Rasmussen´ssyndrome in the differential diagnosis of patients with progressive hemiparesis, cognitive deterioration and drug-resistant focal epilepsy.

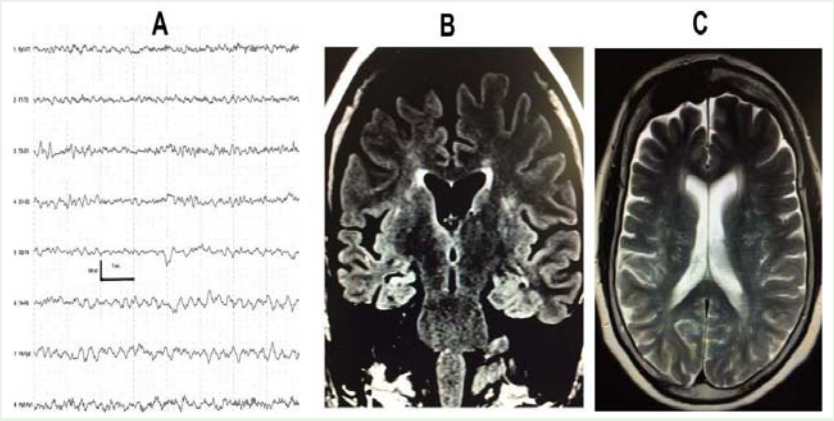

Figure 1: A-Electroencephalogram- Slow-wave activity- delta waves in the right cerebral hemisphere, B and C- RNM-FLAIR Coronal / T2 Axial- Cortical-subcortical atrophy of the right cerebral hemisphere associated with ipsilateral mesial temporal sclerosis.

References

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