

XXXVIII COMU 2019 – Congresso Médico Universitário da FMUSP

Researches Classified – Panels Award – Case Report

Non epileptogenic Rasmussen's encephalitis: a case report

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Introduction: Rasmussen's encephalitis (RE) is a very rare chronic inflammatory unilateral encephalopathy with still unknown etiology. It predominantly affects children under the age of 10, but can also affect adolescents and adults. Current evidence on possible etiologies are divided in two main theories. The first one is based on brain inflammation caused by a reaction to a foreign antigen. The second one is based on an autoimmune disease that is limited to a single hemisphere of the brain. The common presentation is intractable and frequent focal motor seizures, often associated with progressive neurological decline, and progressive unilateral focal cortical atrophy. Progression of the inflammatory process in MRI is considered a good biomarker in RE.

Objectives: Report a rare case of a female infant presenting atypical manifestations of uncommon symptoms without seizures caused by Rasmussen's encephalitis.

Case Report: A ten-year-old right-handed female presented to the emergency department with sudden onset of abnormal involuntary movements of the right upper and lower limbs, face paresia of the same side and loss of attention without loss of consciousness. The involuntary contractions were brief, random, irregular, and worsened with activity, along with a positive milkmaid grip sign. She also displayed reduced movement speed and hyperactive biceps, triceps, patellar and ankle jerk reflexes. Associated with the motor abnormalities, the girl presented with a recent but progressive decrease in cognitive functions, which caused a decline in school performance and impaired reasoning, and affective blunting. She had no sign of previous neuropsychomotor development abnormality. She was taken to a quaternary hospital where a MRI and a CT of the brain were performed. MRI showed atrophy of the left cerebral hemisphere, predominantly in the operculum and head of the caudate nucleus, hypersignal in the insular cortex and adjacent white matter. CT presented with hypoattenuation of the left frontal lobe and parieto-occipital white matter associated with atrophy of the head of the caudate nucleus of the same side and enlargement of the adjacent fissures and sulcus. A subsequent PET-CT showed a left cerebral hemisphere hypometabolism, predominantly in the basal ganglia, temporal, frontal and insular lobes. Functional evaluation detected diffuse unilateral disorganization of brain electric activity, but no epileptic paroxysmal discharges were found on the electroencephalogram. After hospitalization, human immunoglobulin at the dose 2g/kg/day was used for five days. The girl remained stable, with no complications. After sixteen days, she was discharged with a rehabilitation program and follow-up care.

Keywords: Rasmussen's encephalitis; Autoimmune epilepsy; Magnetic resonance imaging.