

OPERCULAR MYOCLONIC-ANARTHRIC STATUS EPILEPTICUS: FIRST MANIFESTATION OF MITOCHONDRIAL DISORDER?

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A 38-year-old woman presented at age of 32 with epilepsia partialis continuum characterized by spontaneous, regular and focal clonic muscular twitches, that were continuous for days and involved the right hemibody (face and distal limb muscles). Dysarthria and synchronous myoclonic movements of palate and tongue accompanied these movements. Brain MRI showed a cortico-subcortical lesion in posterior part of frontal inferior left gyrus, hyperintense on T2- weighted images with cortical restricted diffusion. Electroencephalogram was normal. She became asymptomatic with antiepileptic drugs, later tapered on. Brain MRI and PET scan were performed three months later and were both normal. Infections and inflammatory disorders were excluded. The patient remained asymptomatic until the age of 37. At that age she was readmitted with a subacute, distal and symmetrical tetraparesis with brisk reflexes without sensory or sphincter involvement. A new brain MRI showed bilateral T2 hyperintensities in both caudate and lenticular nucleus. Medullar MRI and spectroscopy were normal. Laboratory investigations including blood count, serum biochemistry and cerebrospinal fluid analysis, electromyography, evoked potentials, body CT-scan and bronchofibroscopy were normal or negative. Muscle biopsy showed rare negative COX/ SDH positive fibers. Gradually, neurological symptoms disappeared. One year later, brain MRI was almost normal.

Opercular myoclonic-anarthric status epilepticus (OMASE) is an uncommon condition characterized by fluctuating cortical dysarthria associated with epileptic myoclonus involving glossopharyngeal musculature bilaterally. In the present case, OMASE was the first manifestation of the disease. Although the lack of definitive criteria for the diagnosis of mitochondrial disease, muscle findings suggest this etiology.