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Opsoclonus Myoclonus Ataxia: Two Idiopathic Cases [Version 1, Awaiting Peer Review]

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Abstract

Opsoclonus-mioclonus-ataxia (OMA) is a rare neurological disorder characterized by the presence of abnormal saccadic ocular movements, ataxia and myoclonus. It is an autoimmune disease probably caused by a dysfunction in Purkinje cells secondary to an altered humoral and cellular response. The most frequent etiologies in adults are paraneoplastic (breast, lung and gynecologic cancer), idiopathic and parainfectious.

We describe two patients that started suddenly with opsoclonus, ataxia and generalized myoclonic jerks. We confirmed the idiopathic etiology and they received immunotherapy. They had a good outcome, with complete recovery.

Keywords: Opsoclonus; Myoclonus; Ataxia; Idiopathic.

Introduction

Opsoclonus Myoclonus Ataxia Syndrome (OMAS) is characterized by opsoclonus, which is a universal component of this syndrome, with focal or diffuse myoclonus, and ataxia.

Opsoclonus is a disorder of ocular motility characterized by spontaneous, arrhythmic and conjugate saccades, occurring in all directions of gaze without a saccadic interval.

Myoclonus is a clinical sign characterized by brief involuntary movements that can involve limbs and trunk.

The physiopathology of this syndrome is autoimmune and includes a humoral and cellular dysfunction. There are two theories about the target of this autoimmune response. One of them states that cerebellar dysfunction leads to disinhibition of the fastigial nucleus by the Purkinje cells resulting in Opsoclonus Myoclonus Syndrome. Another theory proposes that the responsible of OMAS is the caudal pontina reticular formation through a dysfunction of omnipause neurons which inhibit gaze center burst neurons [1,2].

The most frequent etiologies of OMAS in adults are paraneoplastic, idiopathic and parainfectious diseases [3].

We describe the clinical presentation and diagnostic tests of two patients with idiopathic OMAS during a period of one year.

Case Reports

Case One

32 year/old woman who, a week after gastroenteritis, started suddenly with dizziness, abnormal eye movements, head nodding, abnormal brief movements in her arms and legs and unsteady gait .

On examination, the patient had abnormal eye movements with multidirectional saccades, with variable amplitude, present at rest and increased during pursuit. The saccadic movements were visible with closed eyelids.

The generalized myoclonus was spontaneous and associated with movements. It stopped only during sleep.

She had a severe ataxia and she couldn´t walk.

The remainder of her examination was normal.

We performed blood test, with rheumatology test, which were normal.

Polymerase Chain Reaction (PCR) for Epstein Barr, Herpes simplex and Varicella Zoster virus and serologies for HIV, hepatitis, syphilis, cytomegalovirus and Epstein Barr were negative.

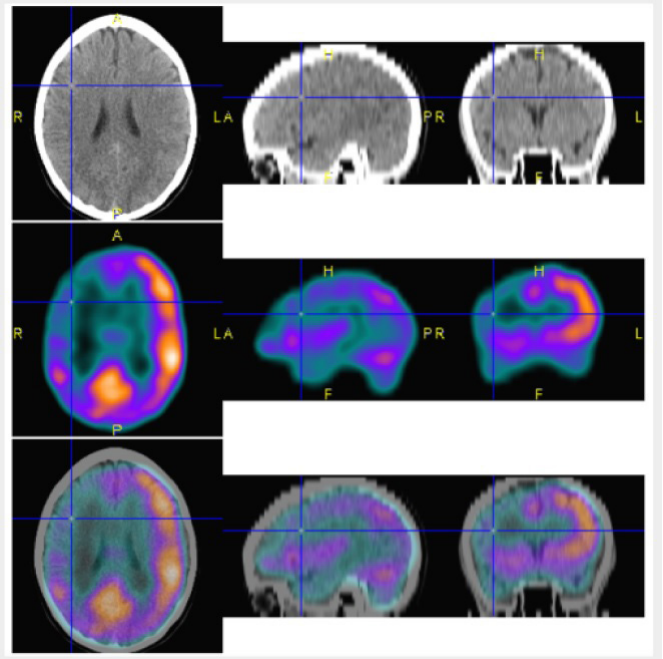
Onconeural antibodies (anti-Yo, anti-Hu, and anti-Ri) were negative too.

Cerebrospinal fluid analysis was normal.

Brain Magnetic Resonance Image and Magnetic Resonance Angiography were normal.

We excluded hidden tumor as a possible etiology with a whole body Possitron Emission Tomography. However, in this study, we observed changes in the brain metabolism, such as hypometabolism in right frontal and temporal lobes.

The patient received initially, symptomatic treatment with clonazepam and valproic acid, with poor response. Then, she received treatment with 1000 mg intravenous metylprednisolone during 5 days, with posterior maintenance dose, with excellent response without sequealae.



Case 1

Case Two

60 year/old man who, a week after a flu-like syndrome, started suddenly with generalized spontaneous abnormal brief movements and unsteady gait.

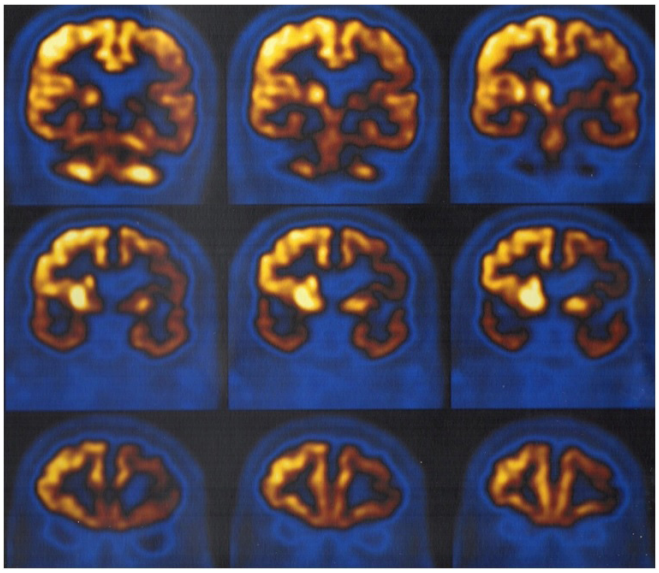
On examination, he presented severe generalized spontaneous and associated with movement myoclonus, including velopalatine myoclonus, which was responsible of his eating difficulty. He had a severe ataxia. He was unable to remain seated.

He had a severe limitation of his daily activities such as walking, sitting, or eating.

We performed a blood test, with rheumatology test; PCR for Epstein Barr, Herpes Simplex, and Varicella Zoster virus; serology for HIV; onconeural antibodies; cerebrospinal fluid analysis and Brain MRI. All of these tests were normal.

We performed a whole body PET scan, which showed a left cortical hypometabolism.

He received treatment with 1000 mg intravenous of Metylprednisolone during 3 days with excellent outcome and full recovery within 2 months.



Case 2

Discussion

Adult's OMAS is a rare neurological disorder in which more than fifty percent of the cases are related to paraneoplastic disease (small cell lung, breast and ovarian cancers). While, idiopathic, parainfectious disease, toxic or metabolic etiologies account for the other half [3,4].

It is essential to confirm the etiology as soon as possible because it is close related with the outcome.

Idiopathic OMAS has been related with an excellent outcome with immunotherapy. On the contrary, when the etiology is paraneoplastic disease, the treatment should be directed to the tumor, because it will define the outcome [5].

In idiopathic cases, the brain MRI or CT are usually normal. Blood tests and CSF analysis are necessary to exclude an infection. In the same way, PET scan is essential to exclude the possibility of hidden tumor [6,7]. However, we didn't find reports about brain metabolic changes in idiopathic OMAS.

In the patients we describe, PET scans showed cortical unilateral hypometabolism without cerebellar involvement which, to our knowledge, has never been described before.

Another important point in our patients is the presence of unspecific viral disease, within two weeks before the symptoms started. This has been described by other authors as a characteristic in idiopathic etiology.

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