

## Opsoclonus-Myoclonus Syndrome Associated with Covid-19

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### Abstract

Since the emergence of the SARS-Cov-2, we have learned a lot about the neurological manifestations of this disease. We will introduce a case of opsoclonus-myoclonus-ataxia associated with an acute infection caused by SARS-Cov-2, and we will make a brief review of what has been published regarding this topic.

**Keywords:** SARS-Cov-2; Opsoclonus-Myoclonus Syndrome; Covid-19

### Introduction

The opsoclonus-myoclonus-ataxia syndrome (OMA) is a rare neurological disorder characterized by opsoclonus, an involuntary, abnormal eye saccadic movement (fast, eye movement that redirects the fovea from one object to another), that is characterized for being arrhythmic, chaotic, and with a horizontal, vertical and torsional component. If the oscillations are small, they are asymptomatic, but they can also produce blurred vision and/or oscillopsia (a feeling that objects have a slight oscillation). The syndrome occurs during fixation, eye tracking and convergence, and it continues during sleep and the palpebral closure. Such syndrome is associated with action myoclonus that affect the limbs, the trunk and the head. Myoclonus are arrhythmic, involuntary, sudden and short movements (< to 100 ms) that occur after a muscular contraction (positive myoclonus) or a muscular inhibition (negative myoclonus) that originate in the central nervous system. They have been described within the manifestations of the COVID-19 disease [1]. The OMA syndrome can also present with cerebellar dysfunction with dysarthria and truncal ataxia and, in some patients, with encephalopathy. It can be the manifestation of a paraneoplastic syndrome, but it also occurs in connection to several clinical conditions (infectious, postinfectious, toxic and metabolic conditions) and sometimes without a clear etiology [2].

Recently, cases of opsoclonus-myoclonus-ataxia syndrome associated with the infection caused by COVID-19 have been reported [3,4].

### Clinical Case

We present a 57-year-old female patient, with no known pathological habits nor history, who initially starts with headache, myalgia, fever and loss of smell. This required hospitalization 72 hours later due to a pneumonia that didn't need oxygen therapy. She had a positive PCR (polymerase chain reaction) result after a nasal swab and an IgM serology for COVID-19.

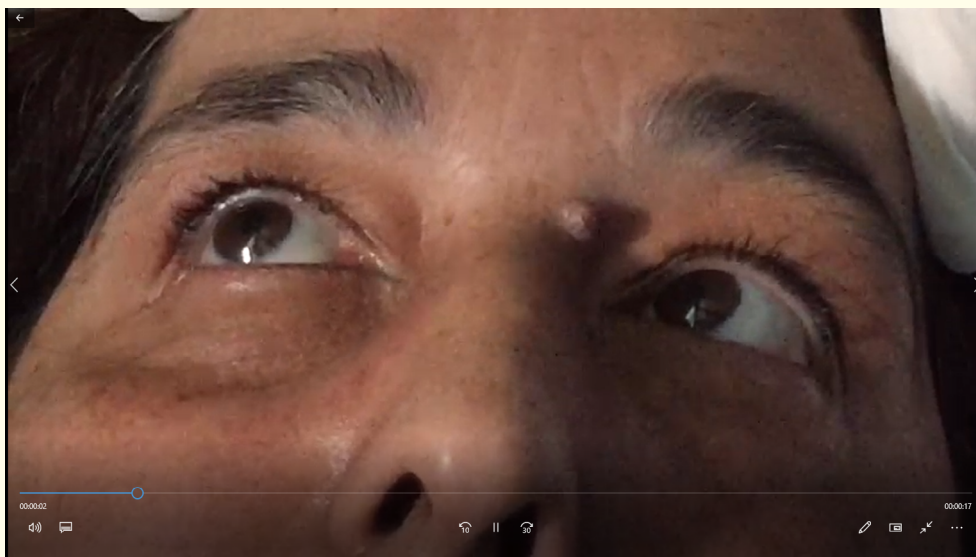
During the 4<sup>th</sup> day of hospitalization, the patient was referred to our Hospital because she presented with generalized myoclonus induced by voluntary movements, associated with a staggered gait: she had a marked instability related to myoclonus.

At the moment of admission to our hospital, the patient was haemodynamically stable and afebrile. Dysarthria with distraction and disorientation episodes were confirmed.

The neuro-ophthalmological exam showed conjugated, involuntary, quick, constant, chaotic, multidirectional eye movements, that were present during fixation and slow tracking (Video 1 and 2). Also, myoclonus of upper and lower limbs and trunk induced by action were observed (Video 3-5).



Video 1



Video 2



**Video 3**



**Video 4**

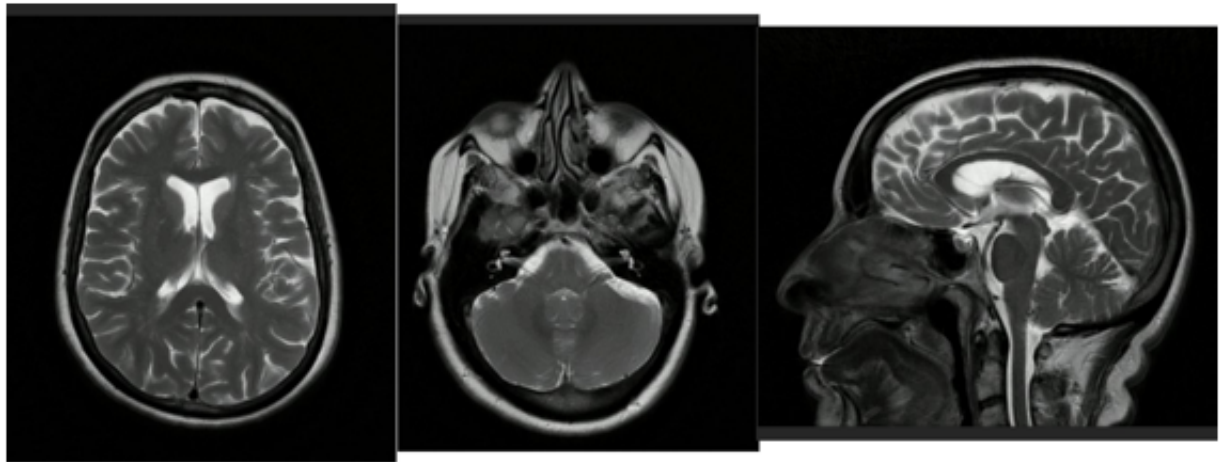


**Video 5**

The patient could not stand up nor wander due to her marked instability and her myoclonus.

#### Additional examinations

Brain MRI performed with a high field equipment and a follow-up 15 days later with normal contrast (See figure 1).



*Figure 1*

Chest CT compatible with pneumonia due to COVID-19.

Lab test without alterations.

We confirmed an opsoclonus-myoclonus diagnostic associated with COVID-19, so it was decided to administer pulse methylprednisolone 1g per day for 5 days and then continue with oral meprednisone with a descending withdrawal scheme, and levetiracetam 1000 mg every 12 hours to treat the myoclonus.

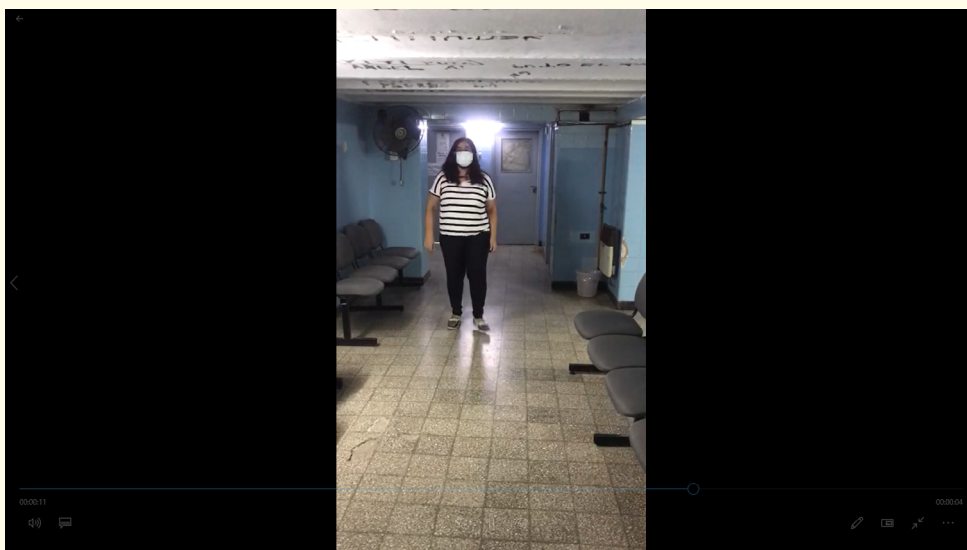
The patient's clinical condition significantly improved during hospitalization, including the disappearance of eye opsoclonus one week later (Video 6), a reduction of the myoclonus and a marked improvement of the gait. A clinical follow-up was done one month later, showing a disappearance of her myoclonus and a normal gait (Video 7 and 8).



*Video 6*



Video 7



Video 8

## Discussion

On 8<sup>th</sup> December 2019, the severe acute respiratory syndrome type 2 (SARS-CoV-2) was diagnosed for the first time, which caused the coronavirus disease 2019 (COVID-19) and it has been spread all over the world since then.

The new SARS-CoV-2 coronavirus (severe acute respiratory syndrome 2) is the cause of the COVID-19 pandemic [5]. The SARS-CoV-2 is similar to the SARS-CoV and the MERS-CoV in terms of the clinical manifestations and the pathological findings.

In fact, even though the most common symptoms of COVID-19 include fever, cough, shortness of breath and a pulmonary disease (from pneumonia to acute respiratory distress syndrome) the SARS-CoV-2 seems to affect other organs, including the nervous system. Apart

from the reported anosmia and the dysgeusia, other reported neurological manifestations include encephalopathies, para/post-infectious central nervous system (CNS) syndromes, cerebrovascular diseases (ischemic and hemorrhagic) and neurological autoimmune disease, like the Guillain-Barré syndrome [5].

The SARS-CoV-2 can damage the central nervous system through 3 ways: (1) as a consequence of the systemic and pulmonary involvement (for example, stroke and post-hypoxic encephalopathies), (2) through direct viral invasion of the CNS (through trans-synaptic or dissemination through the hematoencephalic barrier) and (3) postinfectiously (immune-mediated) [6].

The presence of the polymerase chain reaction of the SARS-CoV-2 in serum together with the pulmonary findings in the CT scan emphasize the probability of a parainfectious etiology of COVID-19 in this patient.

It is believed that the pathogenesis of parainfectious etiologies is directly linked to the viral invasion from tissues to the nervous system or to the postinfectious immune response. Brain MRIs do not often show an anatomic lesion on these parainfectious cases of COVID-19. The route of entry of COVID-19 is through the receptors of the angiotensin-converting enzyme 2, that are not only expressed on the lungs, but are also present on the glial cells and neurons [7].

Therefore, it is believed that this occurs because of a direct invasion of the brain stem and the cerebellar pathways, including the deep cerebellar nuclei or because of the susceptibility of these regions to hyperinflammation (“cytokine storm”) [8] associated to COVID-19 as a pathogenic mechanism in this syndrome.

The opsoclonus-myoclonus-ataxia syndrome associated with infectious processes has different characteristics; its course is usually self-limiting with a spontaneous recovery and the prognosis is usually positive [9], as the case we have presented.

### Conclusion

We presented case of opsoclonus-myoclonus-ataxia associated with an acute infection caused by SARS-Cov-2, an unusual neurological manifestation. Although there have been publications of similar cases since the beginning of the pandemic and there are several hypotheses about how the virus causes neurological disorders, there is little information about the pathophysiology, the management and the prognosis. The subsequent publications should study these topics in order to improve our understanding and the management of these patients.

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