Case report

Opsoclonus-Myoclonus Syndrome in a COVID-19 positive patient

A. GIORGETTI, E. CORENGIA, S. LEVA, M. SERVIDA, L. POLITINI, A. MAZZONE, P. CLERICI, P. FOCIANI, M. CORBELLINO*, A. PRELLE

Aziende Socio Sanitarie Territoriali (ASST) Ovest Milanese, Legnano (Milano)
** ASST "Fatebenefratelli Sacco", Milano*

SUMMARY: *Opsoclonus-myoclonus syndrome is a rare immune-mediated disease in adults. However the pathogenic role is not well understood. We assessed wether there was an association between Opsoclonus-myoclonus syndrome and the positivity for Severe Acute Respiratory Syndrome CoronaVirus-2 (SARS-CoV-2) infection. A 58 years old man with symptomatology suggestive of opsoclonus-myoclonus syndrome was admitted to our hospital; during the hospitalization he developed a typical radiological pattern for COVID-19 pneumoniae. He was studied for possible immuno-mediated causes of the disease or correlation to SARS-CoV-2 infectio. MR imaging, electroencephalography and cerebrospinal fluid examination was normal as the detection of antibodies to neuronal surface antigens. A total body CT scan revealed a typical pattern of ground glass opacities in both lungs. Naso-pharyngeal swab was repeated and revealed a positivity for SARS-CoV-2. Reverse transcriptase-polymerase chain reaction for Sars-CoV-2 RNA in the cerebrospinal fluid was negative. Patient was transferred to a COVID-19 Department. In the following days, a progressive improvement was observed. Number of patients with COVID-19 experiencing different neurological signs and symptoms is rapidly growing in the literature. Opsoclonus-myoclonus syndrome should be added to the spectrum of clinical manifestations associated with this new disorder.*

KEY WORDS: *COVID-19, Myoclonus Syndrome, Opsoclonus.*

INTRODUCTION

Opsoclonus-myoclonus syndrome is a rare immune-mediated disease in adults. The main symptoms include opsoclonus, myoclonus, ataxia, cognitive and behavioural disorders. Opsoclonus is characterized by involuntary, multidirectional saccades with horizontal, vertical and torsional components. OMS is classified as an idiopathic, post-infectious or paraneoplastic syndrome. In adults, small-cell lung cancer, breast and ovarian cancer are the main underlying tumors. Post-infectious brainstem encephalitis, toxic-metabolic disorders and other conditions should also be considered as potential causes. In OMS, several autoantibodies directed against a variety of antigens were found, but diagnostic immunological markers are yet to be discovered[1,2].

CASE REPORT

A 58 years old man presented to emerging department with opsoclonus, myoclonus, ataxia, psychomotor slowing, progressive gait instability and

Correspondence: Dr. Alessandro Praelle, UOC di Neurologia, ASST Ovest Milanese, Ospedale Nuovo, 20025 Legnano (MI), ph. +39-(0)331-449558, e-mail: alessandro.prelle@asst-ovestmi.it
Progress in Neurosciences 2020; 5 (1-4): 59-60 *ISSN: 2240-5127.*

LIST OF ACRONYMS AND ABBREVIATIONS: **5-OH** = 5-hydroxy; **AMPA** = α-Amino-3-hydroxy-5-Methyl-4-isoxazolePropionic Acid receptor; **CASPR** =Contactin-ASsociated PRotein; **COVID-19** = COronaVIrus Disease-19; **CV2** = CrossVeinless 2; **CSF** = Cerebrospinal Fluid Examination; **DPPX** = DiPeptidyl-Peptidase-like protein 6; **ED** = Emerging Department; **GABA-B2** = Gamma-AminoButyric Acid B receptor 1; **HIV** = Human Immunodeficiency Virus; **HNK1** = Human Natural Killer 1; **HSV-1** = Herpes Simplex Virus 1; **HSV-2** = Herpes Simplex Virus 2; **MRI** = Magnetic Resonance Imaging; **NMDAR** = N-Methyl-D-Aspartate Receptor; **NPS** = Naso-Pharyngeal Swab; **OMS** = Opsoclonus-Myoclonus Syndrome; **PNMA2** = ParaNeoplastic antigen Ma2; **RT-PCR** = Reverse Transcriptase-Polymerase Chain Reaction; **SARS-CoV-2** = Severe Acute Respiratory Syndrome CoronaVirus-2; **VZV** = Virus Varicella Zoster.

behavioural impairment. Fifteen days earlier, patient developed fever up to 38 °C for 3 days.

His clinical history was unremarkable, no current therapy. In ED NPS for SARS-CoV-2 RNA was negative by real time RT-PCR. He was admitted to our COVID-19 free Department of Neurology.

MRI, performed with contrast fluid, was normal; electroencephalography showed no slowing activity or epileptiform discharges.

CSF showed normal proteins and cells; PCR testing for the DNA of HSV-1, HSV-2, VZV and cytomegalovirus was negative. Cytopathologic examination revealed a mild non-specific inflammation with sediment consisting of some red blood cells, some polymorphs, rare lymphocytes and macrophages.

Blood tests results were normal and excluded Epstein-Barr virus and HIV infections, Lyme-disease and auto-immune disorders; antineuronal cerebellar antibodies (anti-Hu, anti-Yo, anti-Ri, anti-PNMA2, anti-CV2 and anti-amphiphysin) were not detected.

Detection of antibodies to neuronal surface antigens reported in association in OMS: anti-NMDAR, anti-GABA B1, anti-DPPX, anti-HNK1, anti-CASPR 2, anti-AMPA 1 AMPA 2 were all normal as well as the dosing of the protein tau and 14.3.3 in the CSF.

A total body CT scan revealed a typical pattern of ground glass opacities in both lungs, which were highly suggestive for COVID-19 pneumonia.

NPS was repeated and revealed a positivity for SARS-CoV-2.

Patient was transferred to a COVID-19 Department where he received supportive care, and 5-OH Chloroquine.

A second MRI and lumbar puncture were, respectively, repeated 7 and 11 days after admission and were both unrevealing; RT-PCR for SARS-CoV-2 RNA in the CSF was negative.

In the following days, a progressive improvement in both cognitive and behavioural symptoms as well as myoclonus-opsoclonus, trunk and gait ataxia was observed.

□ DISCUSSION

Our patient presented with a prominent neurological syndrome, in the absence of the typical COVID respiratory symptoms .

Although we could not detect SARS-CoV RNA in the CSF, this seems to be the case for most of the reports published to date[3]. Number of patients with COVID-19 experiencing different neurological signs and symptoms is rapidly growing in the literature[3]. OMS should be added to the spectrum of clinical manifestations associated with this new disorder. Further studies are warranted to unravel the mechanisms underlying the pathogenesis of the neurologic manifestations in COVID-19.

□ REFERENCES

1. Oh S-Y, Kim J-S, Dieterich M. Update on opsoclonus-myoclonus syndrome in adults. J Neurol 2019; 266 (6): 1541-1548.
2. Kurian M, Lalive PH, Dalmau JO, Horvath J. Opsoclonus-myoclonus syndrome in anti-N-methyl-D-aspartate receptor encephalitis. Arch Neurol 2010; 67 (1): 118-121.
3. Mao L, Jin H, Wang M et al. Neurologic manifestations of hospitalized patients with coronavirus disease 2019 in Wuhan, China. JAMA Neurol 2020; 77 (6): 683-690.