

Facial diplegia, a possible atypical variant of Guillain-Barré Syndrome as a rare neurological complication of SARS-CoV-2



David Salomón Juliao Caamaño^{a,*}, Rubén Alonso Beato^b

^aMedical Resident at the Department of Medical Oncology, Hospital General Universitario Gregorio Marañón, Madrid, Spain

^bMedical Resident at the Department of Internal Medicine, Hospital General Universitario Gregorio Marañón, Madrid, Spain

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ABSTRACT

We present a case of facial diplegia after 10 days of SARS-CoV-2 confirmed infection symptoms in a 61 year old patient without prior clinically relevant background. There are few known cases of Guillain-Barré Syndrome (GBS) related to SARS-CoV-2 infection; we propose this case as a rare variant of GBS in COVID-19 infection context, due to its chronology, clinical manifestations and cerebrospinal fluid (CSF) findings.

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1. Introduction

Several typical and not so typical neurologic late manifestations of viral previous pandemics have been proposed throughout human history, for example, with never fully certain causality, there is encephalitis lethargica and its relationship with the influenza pandemic of 1918 [1]. SARS-CoV-2 pandemic is spreading in an era of scientific advances that offers medical community higher chances of establishing causal relationships and common efforts to fight disease and sequelae.

Patients with several neurological involvement due to SARS-CoV-2 infection have been described in the recent outbreak in China [2]; hitherto, there are few described cases of Guillain Barré and SARS-CoV-2 (Table 1). More cases are foreseen due to the virus pathogenesis and epidemiology [3].

Coronaviruses can cause nervous tissue injuries through several known mechanisms (direct infection injury, hypoxia, ACE2 receptors, immune injury [2]; immunomodulatory treatment and gut microbial translocation) [4]. In a series of 214 patients, Mao et al. reported dizziness, headache, hypogeusia and hyposmia as the most common CNS and PNS manifestations [5]. GB is not yet considered a common complication.

2. Case presentation

A 61 year old patient had fever and coughing without dyspnea on day 1 of the illness; after telephone contact with his primary care physician, chest plate and nasopharyngeal sampling RT-PCR, He was diagnosed and treated as a SARS-CoV-2 infection with pneumonia (hydroxychloroquine and lopinavir/Ritonavir for 14 days).

After one week, his symptoms disappeared and on day 10 of his illness, He noted liquid dripping on his right facial commissure and went to the ER. With a diagnosis of right peripheral facial nerve palsy, He was transferred to the low risk COVID19 dedicated hospital and the day after He was transferred back to our ward due to progression towards bilateral facial nerve palsy (Fig. 1) with

unresponsive blink reflex on both eyes. He had no other neurological findings at examination, including symmetrical and normal force, sensitivity, reflexes, ocular movements and a normal gait.

His chest plate showed significant improvement of pneumonia (Figs. 2 and 3) and He had no remarkable laboratory findings besides CSF data. Brain CT and MRI were performed without any acute pathological findings and an image guided lumbar puncture demonstrated mildly elevated levels of proteins (44 mg/dL), absent leukocytes and a negative RT-PCR for SARS-CoV-2 on CSF.

Our patient completed treatment for SARS-CoV-2 adequately and did not present life-threatening signs at any time; He was treated with low dose oral prednisone and after two weeks He started with a barely notable improvement on both sides.

3. Discussion

We regard the patient's neurological complication as directly related to SARS-CoV-2 infection, considering its clinical course and absence of other possible causes in later diagnostic testing. According to previously described GBS variants we propose it as a DP, clinically coherent with the diagnostic criteria for DP by Wakerly et al. (2015) [6]; similar features with the series of DP by Suzuki et al. (2009) [7] and resembling a single case from the recent series by Toscano et al. (2020) [8] related with SARS-CoV-2 infection.

GBS is a peripheral nervous system disorder that presents as a rapidly progressive, ascending, flaccid paralysis with diminished or absent reflexes. The disease is often triggered by infectious processes. *Campylobacter jejuni* infection is the most commonly identified precipitant of GBS. Some viruses like Cytomegalovirus, Epstein-Barr virus, human immunodeficiency virus (HIV), and Zika have also been associated with GBS [9].

GBS used to be considered a single entity characterized by lymphocytic inflammation and peripheral nervous system demyelination. Now, it is usually clinically defined as a more diverse disorder divisible into several patterns and with various clinical manifestations. One proposed clinical variant is facial diplegia [10].

According to diagnostic criteria proposed by Wakerly et al. (2015) [6] our case accomplishes bifacial symmetrical weakness, absence of limb, neck or ocular weakness, an antecedent of prior infectious disease in the previous 3 days to 6 weeks and albu-

* Corresponding author at: Department of Medical Oncology, Hospital General Universitario Gregorio Marañón, Calle del Dr. Esquerdo 46, Madrid, Spain.

E-mail address: davidsalomon.juliao@salud.madrid.org (D.S. Juliao Caamaño).

Table 1
Reported Guillain Barré Syndrome cases related to SARS-CoV-2 infection, demographic characteristics and country.

Sex and age [†]	F – 77 M – 23 M – 55 M – 76 M – 61	F – 76	M – 64	M – 54	M – 65	F – 61	M – 61
Country	Italy	Spain [11]	France [12]	USA [13]	Iran [14]	China [15]	Spain*

[†] F: Female, M: Male.

* Julio Caamaño, Alonso-Beato, in this publication.



Fig. 1. Facial Diplegia on admission. [Video on Appendix 1A.](#)



Fig. 2. Chestplate on admission showing bilateral frosted glass pneumonia.

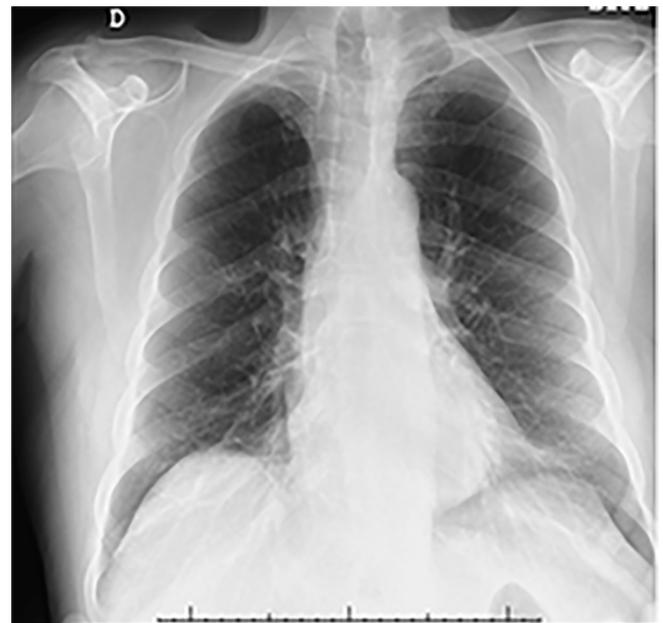


Fig. 3. Chestplate 10 days later with notable improvement.

minocytological dissociation in CSF. We hypothesize that SARS-CoV-2 infection might have triggered this atypical clinical variant of GBS in our patient.

Some other case reports (Table 1) have been published relating typical variants of GBS and SARS-CoV-2 infection so we suggest a possible association between them, but more cases are needed to support causality. The absence of anti ganglioside antibodies testing available data is a limiting factor in comparing these cases.

4. Conclusion

There is a clear emerging group of neurological manifestations during and after SARS-CoV-2 infection; some directly linked, others not so much. The relationship between viral infections and GBS with a consistent chronological sequence is widely accepted. As in previous human pandemics; We are facing unknown possible complications of an extensively expressed disease; according to clinical resemblance with other reports and coherence with diag-

nostic criteria, our case is a highly probable GBS DP variant. More similar cases are needed to establish confident causality and only time will let us know more about this phenomenon.

Declaration of Competing Interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

Appendix A. Supplementary data

Supplementary data to this article can be found online at <https://doi.org/10.1016/j.jocn.2020.05.016>.

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Long-term follow-up in infantile-onset SCAR18: A case report

Alessandro Iodice^a, Carlotta Spagnoli^{b,*}, Margherita Cangini^b, Luca Soliani^b, Susanna Rizzi^b, Grazia Gabriella Salerno^b, Daniele Frattini^b, Francesco Pisani^c, Carlo Fusco^{b,d}



^a Child Neurology and Psychiatry Unit, Santa Chiara Hospital, APSS, Trento, Italy

^b S.C. Neuropsichiatria Infantile, Arcispedale Santa Maria Nuova, AUSL – IRCCS di Reggio Emilia, Reggio Emilia, Italy

^c Child Neuropsychiatry Unit, Parma University, Parma, Italy

^d Laboratorio di Neurofisiologia dell'età evolutiva, S.C. Neuropsichiatria Infantile, Arcispedale Santa Maria Nuova, AUSL – IRCCS di Reggio Emilia, Reggio Emilia, Italy

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ABSTRACT

Autosomal recessive spinocerebellar ataxia type 18 (SCAR18) is caused by pathogenic variants in the Glutamate Receptor, Ionotropic, Delta-2 (*GRID2*) gene. We describe the long-term follow-up from 1 to 31 years of an Italian patient with congenital SCAR18 who is compound heterozygous for a maternally-inherited nonsense variant and a de novo microdeletion. To date, this is the longest follow-up in congenital SCAR18.

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1. Introduction

In 2013 homozygous deletions of Glutamate Receptor, Ionotropic, Delta-2 (*GRID2*) gene were first linked to a recessive syndrome featuring cerebellar ataxia and atrophy [1,2].

We present an adult Italian patient with long-term follow-up.

2. Case report

A male was born from Italian non-consanguineous parents, with unremarkable pre/perinatal and family history.

He presented shortly after birth due to nystagmus. He then developed hypotonia, early ataxia, motor delay (supported walking with unsteady gait: 18 months, independent walking: never acquired), and slight language delay with dysarthria. Over the years, ocular flutter, oculomotor apraxia and bilateral exophoria became apparent. His reflexes were brisk.

Brain MRIs revealed progressive cerebellar atrophy, more prominent in the vermis, stable since fourteen years of age (last at 30 years, Fig. 1). No evidence of peripheral polyneuropathy arose (last examined at 30 years).

At last neurological evaluation (31 years), he walks unsteadily with a walker, shows action tremor and sits independently with frequent truncal oscillations and has mild distal hypotrophy in his lower limbs, without pyramidal signs. He is dysarthric.

During follow-up, he underwent various genetic and neurometabolic investigations. We identified a compound heterozy-

* Corresponding author at: S.C. Neuropsichiatria Infantile, Arcispedale Santa Maria Nuova, AUSL – IRCCS di Reggio Emilia, viale Risorgimento, 80, 42100 Reggio Emilia, Italy.

E-mail address: carlotta.spagnoli@ausl.re.it (C. Spagnoli).