

# Anti-GFAP-antibody positive postinfectious acute cerebellar ataxia and myoclonus after COVID-19: a case report

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**Abstract:** We present a case of acute cerebellar ataxia and myoclonus with detected anti-GFAP-antibodies in a patient recently recovered from COVID-19. Main symptoms consisted of acute gait and limb ataxia and myoclonus. The patient improved considerably upon treatment with high-dose intravenous (IV) steroids. While cerebrospinal fluid (CSF) and magnetic resonance imaging (MRI) findings were unremarkable, anti-GFAP-antibodies were detected in the patient's serum and disappeared upon clinical remission at a 3-month follow-up. This case suggests that anti-GFAP-antibodies might be associated with some of the increasingly observed cases of postinfectious acute cerebellar ataxias in COVID-19 patients and aid in the diagnosis of this autoimmune complication. We recommend searching for these antibodies in serum and CSF in suspected cases. Early steroid treatment may prove beneficial for these patients.

**Keywords:** anti-GFAP, ataxia, cerebellitis, COVID-19, myoclonus, steroids

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## Case description

A 44-year-old previously healthy patient presented to the emergency department with rapidly progressive cerebellar syndrome 14 days after diagnosis of COVID-19. Initial COVID-19-related symptoms consisted of joint pain, fatigue, diarrhea, and temporary loss of smell and had already ceased. On presentation, he complained of a debilitating impairment of speech, an unsteady gait and limb clumsiness. Clinical examination showed severe generalized action myoclonus also affecting his speech, generalized ataxia, and dysgraphia (see Supplemental video). The patient was admitted to the Neuro-COVID-ward in Essen University Hospital for further diagnosis and treatment.

Blood lab work was normal. SARS-CoV-2-RNA was still detectable in the nasopharyngeal swab specimen. Magnetic resonance imaging (MRI) of the brain showed no related abnormalities, particularly no cerebellar or brainstem pathology or contrast agent accumulation. Electroneurography

yielded no pathological findings. Investigation of cerebrospinal fluid (CSF) revealed normal cell count with 2 leukocytes/ $\mu$ l and borderline protein levels (51 mg/dl); SARS-CoV-2 as well as other viral agents (HSV 1&2, CMV, EBV, VZV, Adeno-, Entero-, Parecho-, Parvovirus, HHV 6&7) were negative by polymerase chain reaction (PCR) testing in CSF.

Suspecting parainfectious autoimmune pathogenesis, we initiated anti-inflammatory treatment with a 5-day course of 1 g intravenous (IV) methylprednisone daily. Symptoms significantly improved in the days following treatment initiation. Results of CSF and serum testing for autoimmune antibodies revealed positive anti-GFAP-IgG antibodies in the patient's serum (Glial fibrillary acidic protein; titer of 1:1000, determined by indirect immunofluorescence assay), but not in CSF. No other coexisting neural autoantibodies were found in serum or CSF (including anti-Hu, anti-Yo, anti-Ri, anti-AMPA-R, anti-NMDA-R, anti-ZIC4, anti-GAD65, anti-LGI1, anti-AQP-4).

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After discharge, the patient experienced further steady improvement. Three months later, he showed no residual neurological symptoms. A recent control MRI of the brain was unchanged to the previous scan. No structure suspicious of neoplasms were detected in a computed tomography (CT) of the thorax and abdomen. At this time, GFAP-Serum antibodies had decreased to undetectable levels ( $< 1:100$ ).

### Discussion

Various neurological complications of COVID-19 have been observed during the acute infection,<sup>1</sup> but as the pandemic continues, there is now also rising evidence for associated neurologic diseases that emerge in the postinfectious phase with some time delay after convalescence from the virus. Here, we present a case of SARS-CoV-2-triggered, GFAP-antibody positive autoimmune acute cerebellar ataxia with myoclonus. This diagnosis is supported by the time course of the disease, positive response to steroid treatment, and the detection as well as the kinetics of the neural autoantibody.

Neural autoreactivity is proposed to be underlying a pattern of observed neurological symptoms after COVID-19. These often manifest after the acute viral replicative phase and are also, if not preferred, present in young, otherwise healthy patients with a competent immune system and only mild prior course of viral disease.

Within the last months, several reports of COVID-19-patients with rapidly developing cerebellar ataxia, often also accompanied by myoclonus, emerged in the literature.<sup>2-5</sup> Predominantly, adult men exhibited neurologic symptoms days to a few weeks after recovery from a mild course of COVID-19. As in our case, MRI of the brain of these patients was normal, and CSF revealed no or only very discrete signs of inflammatory reactivity and was negative for SARS-CoV-2 RNA. Treatment regimen consisted of high-dose IV steroids, some received IV immunoglobulin (IVIg)-therapy and supportive administration of anticonvulsants. The overall course was benign in the majority of cases and patients completely recovered over the period of weeks. No disease relapses have been reported so far, indicating a monophasic course of disease. In our patient, symptoms stayed in complete remission without the need for sustained immunosuppressive therapy during close follow-up monitoring.

As a hint to the underlying pathomechanism in our patient, we detected a high titer of serum-anti-GFAP-antibodies during the early phase after onset of neurological symptoms. No defined autoimmune antibodies were found in serum or CSF in previous comparable cases,<sup>2-6</sup> albeit none of them explicitly stated that testing included anti-GFAP-antibodies. The decrease of antibody titer below the detection limit paralleled the complete medical recovery in our case, suggesting a close link to the acute pathomechanism in this autoimmune disease.

In 2016, Fang *et al.*<sup>7</sup> defined the novel entity of steroid-responsive meningoencephalomyelitis with autoimmune GFAP astrocytopathy by IgG antibody detection in serum or CSF. The presence of the antibody in the serum was shown to be highly specific for this condition, which exhibited varying clinical manifestations, cerebellar ataxia and postural tremor being among them. A paraneoplastic occurrence has been described in 38% of cases. CT-based tumor screening revealed no masses suspicious for malignancy in our patient.

### Conclusion

Here, we report of GFAP-antibody-associated acute cerebellar ataxia with myoclonus in a patient with COVID-19. Symptoms consisted of gait and limb ataxia and myoclonus including the vocal folds and responded well to high-dose corticosteroid treatment. This case presents an expansion to the spectrum of neurologic diseases associated with SARS-CoV-2. Clinicians should be aware of this possible complication which can affect young patients with only mild respiratory symptoms. Clinical management entails ruling out other potential causes, antibody testing, CSF investigation, and MRI of the brain. Early steroid treatment may be beneficial.

### Author contributions

LA collected clinical data, wrote the manuscript, and edited the video. SK provided conceptual input for symptomatology and clinical classification. CK supervised diagnosis and treatment. MS led inpatient care and provided conceptual input. MK guided diagnosis and treatment and provided conceptual input. All authors read, reviewed, and approved the final manuscript.

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**Conflict of interest statement**

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**Patient consent**

The patient gave written informed consent to publication of medical information and the video.

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**Supplemental material**

Supplemental material for this article is available online.

**References**

1. Fleischer M, Kohrmann M, Dolff S, *et al.* Observational cohort study of neurological involvement among patients with SARS-CoV-2 infection. *Ther Adv Neurol Disord* 2021; 14: 1–14.
2. Dijkstra F, Van den Bossche T, Willekens B, *et al.* Myoclonus and cerebellar ataxia following Coronavirus Disease 2019 (COVID-19). *Mov Disord Clin Pract* 2020; 7: 974–976.
3. Schellekens MMI, Bleeker-Rovers CP, Keurlings PAJ, *et al.* Reversible myoclonus-ataxia as a postinfectious manifestation of COVID-19. *Mov Disord Clin Pract* 2020; 7: 977–979.
4. Shah PB and Desai SD. Opsoclonus myoclonus ataxia syndrome in the setting of COVID-19 infection. *Neurology* 2021; 96: 33.
5. Foucard C, San-Galli A, Tarrano C, *et al.* Acute cerebellar ataxia and myoclonus with or without opsoclonus: a parainfectious syndrome associated with COVID-19. *Eur J Neurol* 2021; 28: 3533–3536.
6. Heming M, Li X, Räuber S, *et al.* Neurological manifestations of COVID-19 feature T cell exhaustion and dedifferentiated monocytes in cerebrospinal fluid. *Immunity* 2021; 54: 164–175. e6.
7. Fang B, McKeon A, Hinson SR, *et al.* Autoimmune glial fibrillary acidic protein astrocytopathy: a novel meningoencephalomyelitis. *JAMA Neurol* 2016; 73: 1297–1307.

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