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LETTER TO THE EDITOR

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Author response to "Absence of proximal muscle weakness, dysarthria, and facial diplegia suggest Guillain–Barre syndrome rather than CIDP"

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Abstract

In many instances, the differential diagnosis between Guillain–Barre syndrome and chronic inflammatory demyelinating polyneuropathy (CIDP) may be challenging. The aim of this letter to the editor is to elucidate comments and concerns raised, regarding our latest published article dealing with two patients that developed acute-onset CIDP after SARS-CoV-2 infection and Ad26.COV2.S vaccination, respectively.

Keywords SARS-CoV-2, COVID-19, Vaccination, CIDP, Guillain–Barre syndrome

Background

The aim of this letter to the editor is to clarify the comments and concerns raised by Professor Josef Finsterer [1] with regard to our latest published manuscript in the *Egyptian Journal of Neurology, Psychiatry and Neurosurgery*, presenting two patients who developed acute-onset chronic inflammatory demyelinating polyneuropathy (CIDP) after infection with SARS-CoV-2 and vaccination with an mRNA-based anti-SARS-CoV-2 vaccine (Ad26.COV2.S), respectively [2].

Main text

With regard to our patient 2 and the argument that "bilateral prosopoplegia is only rarely reported in patients with CIDP" [1], we agree that cranial nerve involvement is more frequent in Guillain–Barre syndrome (GBS) than CIDP and that facial diplegia is uncommon as a first manifestation of CIDP and have also commented on this within our manuscript ("it is proposed that patients with A-CIDP generally exhibit no cranial nerve dysfunction"). However, facial nerve involvement is estimated to occur in 63% of CIDP patients [3, 4]. Moreover, Bagella and colleagues [4] and de Souza and colleagues [5] have also reported cases of acute-onset CIDP (A-CIDP) presenting with bilateral facial palsy after COVID-19 vaccination. Thus, it seems that "patients with A-CIDP following COVID-19 vaccination present with bifacial paralysis and generally a more severe clinical phenotype at initial presentation that may mimic GBS, rendering early clinical distinction between COVID-19 vaccination related GBS from A-CIDP practically impossible", as mentioned in our text.

With respect to the second argument against CIDP, that in our second case "the disease course was not progressive during 8 weeks as requested by the EFNS criteria for CIDP" [1], we disagree with the notion that clinical evolution is solely progressive in CIDP. According to the European Academy of Neurology/Peripheral Nerve Society (EAN/PNS) clinical diagnostic criteria, CIDP evolution can also be relapsing–remitting, as in our case. Moreover, as the clinical nadir was reached beyond the

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8th week after disease onset, our diagnosis is compatible with A-CIDP [6].

With regard to the third argument that "acute-onset CIDP (A-CIDP) requires the presence of proximal and distal muscle weakness" [1], we agree that according to EAN/PNS guidelines, proximal and distal muscle weakness is present in typical CIDP. However, the same guidelines include atypical CIDP variants, such as the distal CIDP variant that involves distal sensory loss, muscle weakness predominantly in lower limbs and gait instability, and the sensory-predominant CIDP variant that is characterized by gait ataxia, impairment of vibration and position sense and changes in cutaneous sensation, as in our case [6]. Myelin-associated glycoprotein (MAG) peripheral neuropathy is usually associated with distal CIDP. Testing for anti-MAG was performed but was negative.

With respect to the notion that the author "disagrees with the description in Table 2 that involvement of respiratory muscles is absent in CIDP" [1], we should first mention that Tables 1 and 2, differentiating GBS with treatment-related fluctuations (GBS-TRF) from A-CIDP, were modified from van Doorn [7] and not constructed by us. In any case, we agree with their included information, as according to EAN/PNS guidelines respiratory involvement is exceptional in CIDP [6] and only a few cases of respiratory failure due to phrenic nerve involvement are mentioned in the literature [8].

We should clarify that dysarthria in patient 2 was due to bilateral facial palsy solely. On the other hand, there was no evidence of cranial nerve IX and X involvement or altered sensorium that would have been suggestive of Bickerstaff brainstem encephalitis (BBE). Thus, from a therapeutic standpoint, we did not consider that cranial magnetic resonance imaging (MRI) with contrast medium would have added significant information, as the constellation of clinical, cerebrospinal fluid (CSF) and neurophysiological findings were already consistent with the diagnosis of primarily demyelinating immune-mediated neuropathy.

With regard to the comments raised for patient 1, we agree that the latency between COVID-19 infection and CIDP onset is long for a causal relationship to be established. Nevertheless, in reported cases of late-onset GBS after COVID infection latency ranges from 53 to 100 days [9–11].

We also agree that the determination of cytokines, chemokines, and glial fibrillary acidic protein GFAP would have offered additional information. However, their testing is not included in best practice guidelines for GBS [12] or CIDP [6], thus was not ordered. Serum and CSF testing for antiganglioside antibodies was performed but was negative.

Finally, we agree that in nodopathies cranial nerve involvement is a common feature and also considered this possibility. However, testing for nodopathies, including neurofascin and contactin antibodies, was negative in both patients.

Conclusion

As evidenced by our cases and the concerns posed by Professor Finsterer, the differential diagnosis between GBS and A-CIDP is not always straightforward, and perhaps our diagnoses might have been wrong. In any case, we should state that both patients being treated as A-CIDP exhibit remarkable clinical recovery, so far.

Abbreviations

BBE	Bickerstaff brainstem encephalitis;
CIDP	Chronic inflammatory demyelinating polyneuropathy
A-CIDP	Acute-onset CIDP
CSF	Cerebrospinal fluid
GBS	Guillain–Barre syndrome
GBS-TRF	GBS with treatment-related fluctuations
MRI	Magnetic resonance imaging
SARS-CoV-2	Severe acute respiratory syndrome coronavirus 2

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Author contributions

AF: data collection and drafting the manuscript. DT: basic idea, data collection, drafting and revising the manuscript. SKa: data collection and drafting the manuscript. SKi: revising the manuscript. Il: critical comments during drafting and manuscript revision. The authors read and approved the final manuscript.

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Availability of data and materials

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Declarations

Ethics approval and consent to participate

Not applicable.

Consent for publication

Not applicable.

Competing interests

The authors declare no competing interests.

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