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Before CIDP and Sjögren's Syndrome are Causally Linked, the Two Diagnoses Must Be Confirmed

***Josef Finstere*[1], *and Sounira Mehri*[2]**

[1]*MD, PhD , Neurology & Neurophysiology Center, Vienna, Austria, ORCID: 0000-0003-2839-7305*

[2]*MD, Biochemistry Laboratory, LR12ES05 "Nutrition-Functional Foods and Vascular Health", Faculty of Medicine, Monastir, Tunisia, ORCID 0000-0002-2221-7193*

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LETTER TO THE EDITOR

We read with interest the article by Süt, *et al*. [Süt, N. Y. *et al*., 2023] who reported on a 13 year-old male diagnosed with typical chronic inflammatory demyelinating polyneuropathy (CIPD) according to the revised EAN criteria [Van den Bergh, P. Y. K . *et al*., 2021]. Since the patient was also diagnosed with subclinical Sjögren's syndrome, a causal relation between CIDP and Sjögren's syndrome was suspected [Süt, N. Y. *et al*., 2023]. The patient benefited from monthly intravenous immunoglobulins and daily steroids for six months [Süt, N. Y. *et al*., 2023]. The study is compelling but has limitations that should be discussed.

We disagree with the diagnosis CIDP. CIDP is characterised by motor and sensory disturbances. The patient did not complain of any loss of sensitivity and the sensory tests in the clinical neurologic exam were normal. There was normal pin-prick testing on the clinical neurologic examination [Süt, N. Y. *et al*., 2023]. How can the discrepancy between absent sensory nerve action potentials in the lower limbs and the absence of sensory disturbances in the index patient be explained? There was no MRI with contrast medium that showed enhancing nerve roots.

We also disagree with diagnosis of Sjögren's syndrome. Anti-nuclear antibodies (ANA) were only slightly increased to 1:80 [Süt, N. Y. *et al*., 2023]. A single anti-Ro test combining anti-Ro-60 and anti-Ro-52 antibodies in solid-phase immunoassays was negative in the index patient [Süt, N. Y. *et al*., 2023]. Anti-Ro/SSA and anti-Ro/SSB were also negative [Süt, N. Y. *et al*., 2023]. Ro60 antibodies were not determined. Therefore, the index patient's autoimmune pathophysiology was insufficiently supported by the data provided.

We disagree with the assumption that CIDP was due to Sjögren's syndrome [Süt, N. Y. *et al*., 2023]. Several arguments can be made against Sjögren's syndrome as the trigger of CIDP. First, Sjögren's syndrome was subclinical, while the neurological manifestations were very pronounced. Second, differential triggers of CIDP have not been adequately ruled out. Given that CIDP appears to have evolved during the pandemic, it is crucial that SARS-CoV-2 infection has been sufficiently ruled out as a trigger for CIDP. Has the past history been positive for COVID-19? Has the patient ever tested positive for SARS-CoV-2? It is also not mentioned whether the patient had received an anti-SARS-CoV-2 vaccination or not [Süt, N. Y. *et al*., 2023]. CIDP has been reported as a complication of COVID-19 vaccination [Fotiadou, A. *et al*., 2022]. Contactin antibody titre results are also lacking to assess whether the index patient has nodopathy due to contactin antibodies. CIDP, in particular nodopathies, may be associated with antibodies against contactin [Dong, M. *et al*., 2022]. It is therefore important that not only antibodies against neurofascin but also against contactin are determined. There is no mention whether malignancy could be ruled out with certainty. The results of the virus panel in serum and CSF were not provided. Was the index patient HIV negative?

There was evident uncertainty as to the diagnosis of CIDP. Why else was the patent genetically tested for Charcot Marie Tooth disease? Was hereditary neuropathy suspected? Which genes were tested? How was GBS diagnosed in the cousin? Was there evidence for neuropathy in other first-degree relatives, particularly the index patient's parents or sisters and brothers?

In summary, the interesting study has limitations that put the results and their interpretation into perspective. Addressing these issues would

***Corresponding Author:** Josef Finsterer

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strengthen the conclusions and could improve the status of the study. CIDP patients usually complain about sensory disturbances and Sjögren's syndrome is usually diagnosed when both Ro/SSA and Ro/SSB antibodies are positive.

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