

## The Joint Occurrence of Autoimmune Diseases Requires Special Diagnostic Care

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### LETTER TO THE EDITOR

We read with interest the article by Segura-Chavez *et al.* on a 38 year-old male who developed progressive quadraparesis, facial diplegia, and limb paresthesias 10 days after an intestinal infection presumably with *Campylobacter jejuni* [Segura-Chávez, D. *et al.*, 2023]. Due to affection of respiratory muscles and muscular respiratory failure he required intubation and mechanical ventilation but made a complete recovery of limb weakness after administration of intravenous immunoglobulins (IVIGs) [Segura-Chávez, D. *et al.*, 2023]. He was additionally diagnosed with primary biliary cholangitis (PBC) upon jaundice and generalised pruritus, elevated liver transaminases, and antibodies against mitochondria and centromere-B, which incompletely resolved upon steroids and azathioprine [Segura-Chávez, D. *et al.*, 2023]. The study is compelling but has limitations that should be discussed.

We disagree with the diagnosis PBC [Segura-Chávez, D. *et al.*, 2023]. PBC is diagnosed not only upon the clinical presentation, elevated liver transaminases, cholestasis parameters, and elevated anti-mitochondrial and anti-centromere-B antibodies, but also by magnetic resonance (MR) cholangio-pancreatography (MRCP) [Yacoub, H. *et al.*, 2023]. There is also a role for liver biopsy in the diagnosis of PBC, which was not performed in the index patient [Manzo-Francisco, L. A. *et al.*, 2023]. There is also no mention that MRCP had been carried out. Therefore, the diagnosis PBC is unsupported and questionable.

We also disagree with the diagnosis acute, motor, axonal neuropathy (AMAN) [Segura-Chávez, D. *et al.*, 2023]. Although no compound muscle action potentials could be elicited when stimulating motor fibers of the median, ulnar, peroneal, and tibial nerves on the initial electrophysiological study, the patient also complained about sensory disturbances. However, no NCSs of sensory nerves

were reported at onset of the symptoms. Only nerve conduction velocity of sural nerve after treatment was listed in table 2, which was normal

There is a discrepancy between the description of symptoms and NCVs. The patient reported paresthesias of the lower limbs but NCSs did not reveal any sensory disturbances. Since there were sensory and motor symptoms, the condition should not be classified as AMAN but rather as AMSAN. It would be necessary to know whether sensory NCSs had been performed prior to onset of IVIG. AMAN can be diagnosed only in the absence of sensory symptoms and normal sensory nerve conduction.

There is a discrepancy between the modified Rankin scale (mRS) of 2 and the statement that the patient recovered completely his limb strength [Segura-Chávez, D. *et al.*, 2023]. If the patient recovered completely one would expect a mRS of 1 or 0. Did facial weakness also recover under IVIG?

A limitation of the study is that the drugs the patient received during hospitalisation were not reported. It is crucial to know the current medication to assess whether hepatopathy and cholestasis could have been also due to the liver toxic effect of any of the applied drugs. Furthermore, a detailed account of the immunomodulatory regimen is essential, as advances in treatment strategies continue to evolve our approach to complex autoimmune presentations [Venna, P.R. and Gannina, B., 2021]. Elevation of anti-mitochondrial and anti-centromere-B antibodies is a non-specific finding. They can be elevated also in inflammatory myopathies, systemic sclerosis, and cardiomyopathy [Kainaga, M. *et al.*, 2023].

Another limitation of the study is that antibodies against *Campylobacter jejuni* were not reported. Since GBS was causally related to the 2019 *Campylobacter jejuni* outbreak in Peru, it is critical

to determine these antibodies. Another cause of GBS that needs to be ruled out is Zika. Since SARS-CoV-2 started already at least in 12/19 it is conceivable that the patient was even infected by SARS-CoV-2, which should have been ruled out by a negative PCR.

There is no explanation for hepatomegaly. There is usually no hepatomegaly in case of PBC. Was there tricuspid insufficiency or right ventricular heart failure?

Overall, the interesting study has limitations that put the results and their interpretation into perspective. Addressing these issues would strengthen the conclusions and could improve the status of the study. AMAN should not be diagnosed if there are sensory disturbances and if NCSs reveal sensory neuropathy. To detect the trigger of GBS it could be helpful to determine antibodies against *Campylobacter jejuni* and against Zika, particularly in areas where these infections are endemic.

## REFERENCES

1. Segura-Chávez, D., Tagle-Lostaunau, I., Sifuentes-Monge, J. & Aquino-Peña, F. "Concurrence of Guillain-Barré syndrome and primary biliary cholangitis not related to

SARS-CoV-2: Case report." *Medwave*, 23.3 (2023). English, Spanish.

2. Yacoub, H., Ben Azouz, S., Hassine, H., Debbabi, H., Cherif, D., Ghayeb, F., Boukriba, S., Kchir, H. & Maamouri, N. "Overlap syndrome of primary biliary cholangitis and primary sclerosing cholangitis: two case reports." *J Med Case Rep*, 17.1 (2023): 169.
3. Venna, P.R. and Gannina, B. "Advances in Immunomodulatory Treatments for Autoimmune Disorders: A Roadmap for Future Therapeutic Strategies." *Afr. J. Biomed. Res.* 24.3 (2021): 486-492
4. Manzo-Francisco, L. A., Aquino-Matus, J., Vidaña-Pérez, D., Uribe, M. & Chavez-Tapia, N. "Systematic review and meta-analysis: Transient elastography compared to liver biopsy for staging of liver fibrosis in primary biliary cholangitis." *Ann Hepatol* (2023): 101107.
5. Kainaga, M., Sasaki, T., Kitamura, M., Nakayama, T., Masuda, K., Kakuta, Y., Nishino, I. & Imafuku, I. "Inflammatory Myopathy Associated with Anti-mitochondrial Antibody-negative Primary Biliary Cholangitis Diagnosed by a Liver Biopsy." *Intern Med*, 62.5 (2023): 797-802.

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