

BACKGROUND

This case report describes a patient presenting with autoimmune nodopathy related to anti-Contactin-1 IgG4 antibodies, alongside membranous glomerulopathy with NELL1 IHC positivity. The patient initially presented with sudden onset back pain and progressed to severe sensorimotor polyneuropathy, consistent with Guillain-Barré syndrome."

CASE REPORT

This is a 79 yo patient with PMH of HTN, T2DM, who initially presented with back pain thought to be associated with pyelonephritis. Later in patient clinical course, patient was diagnosed with membranous glomerulopathy associated with NELL1 IHC positivity. He had progressive weakness with the development of sensorimotor polyneuropathy leading to quadriplegia, and the initial diagnosis of Guillain-Barré syndrome with features suggesting autoimmune nodopathy. EMG/NCS revealed a combination of demyelinating and axonal loss patterns, further supported by serological findings of anti-Contactin-1 IgG4. This patient had worsening of symptoms and clinical deterioration resulting in death, despite plasmapheresis (PLEX) and supportive care.

Target Antigens in Membranous Nephropathy

Malignancy	Autoimmune	Infection	Medications	Pediatric
NELL-1 THSD7A PCHD7	EXT1/2 TGFB3 NCAM1 PCHD7 CNTN1 (CDIP) FAT1 (HSC or transplant)	NDNF-3 (syphilis)	PCSK6 (NSAID) NELL-1 (lipoic acid) (mercury) Cationic BSA (milk allergen)	SEMA3B Neutral endopeptidase (antenatal)

DISCUSSION

There are several documented case reports illustrating concomitant CIDP with membranous nephropathy. According to prior studies, Contactin-1 IgG4 antibody is a pathological link between immune mediated neuropathies and nephrotic syndrome caused by membranous glomerulonephritis. This particular case demonstrates an unclear association between concurrent diagnosis of NELL 1IHC positive membranous nephropathy with anti Contactin-1 IgG4 autoimmune nodopathy.

CONCLUSION

In this patient, as seen in prior studies, the onset of nephrotic syndrome coincided with the development of severe ascending sensorimotor neuropathy. While in prior studies the Contactin-1 IgG4 antibody seemed to be the link between the nephropathy/nodopathy, this study illustrates the possibility of a NELL 1 IHC positive MN alongside a Contactin-1 IgG4 autoimmune nodopathy.

References

- Fehmi, Janev et al. "Contactin-1 links autoimmune neuropathy and membranous glomerulonephritis." *PLoS one* vol. 18,3 e0281156. 9 Mar. 2023, doi:10.1371/journal.pone.0281156
- Progress in diagnosis and treatment of chronic inflammatory demyelinating polyradiculoneuropathy
Bunschoten, Carina et al. *The Lancet Neurology*, Volume 18, Issue 8, 784 - 794

CIDP Subtypes

