TWO RARE CASE REPORTS OF AUTOIMMUNE NODOPATHY

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INTRODUCTION

- Autoimmune nodopathy are diseases where antibodies targeting peptide structures of the node (NF186/140, Gliomedin) and paranode (CNTN1, Caspr1, NF 155) were identified in patients with clinical features of Guillain–Barré syndrome (GBS) and chronic inflammatory demyelinating polyradiculoneuropathy (CIDP).
- Patients with positive anti-NF186/140 IgG are rare(<3%) and we highlight two such cases.

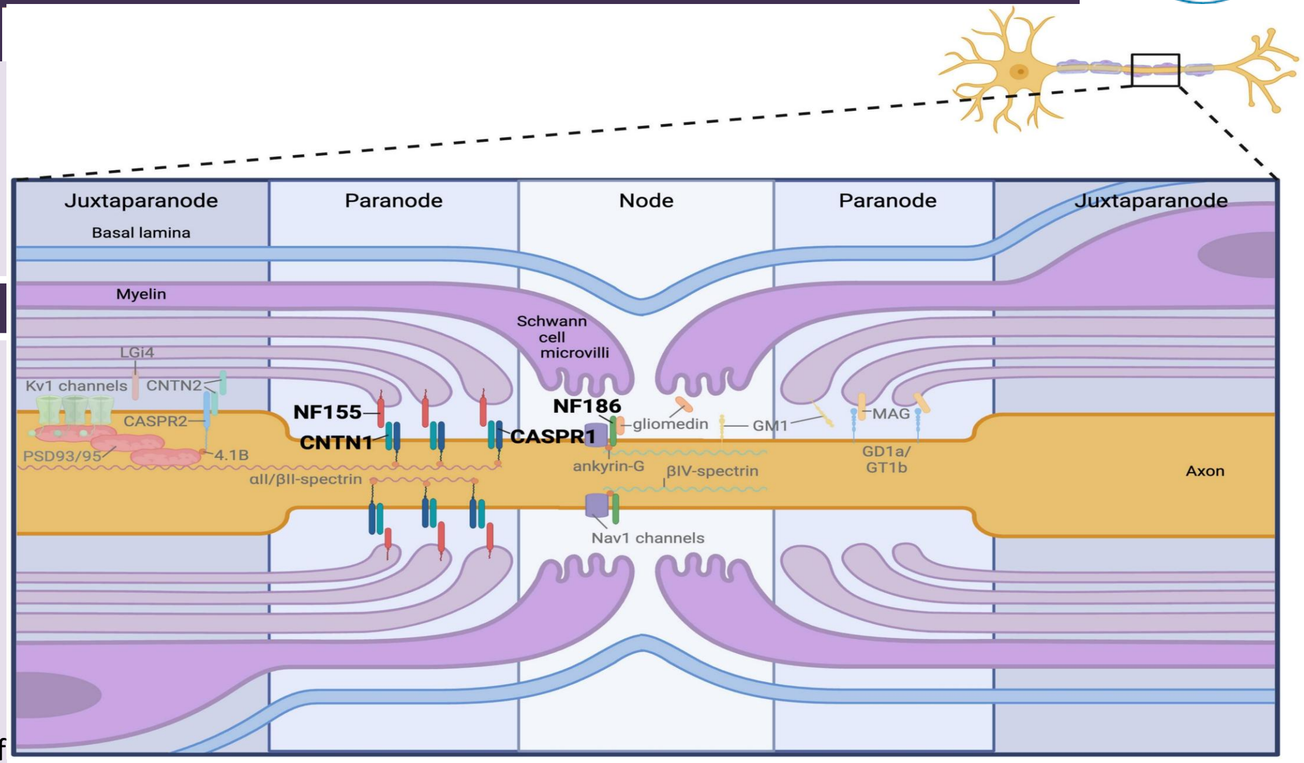
CASE REPORTS

A 39 year old man presented with distal paresthesia since 4 days and quadriparesis since one day. Examination revealed flaccid quadriplegia with autonomic dysfunction and respiratory failure. NCS showed severe motor axonal polyneuropathy. He was treated with IVIG following which his respiratory failure improved. Autoimmune nodopathy panel came positive for **NF 140** antibody. Though he received IV Methylprednisolone and Rituximab, there was no further improvement.

A 46 year old woman presented with progressive sensory ataxia of four months. Examination showed severe large fibre sensory loss. Electrophysiology disclosed pure sensory axonal polyneuropathy. Etiological workup in terms of autoimmune, paraneoplastic, vasculitic, and serological markers were normal. **NF 186** IgG antibody was positive. Patient received 5 days of IV pulse Methyl prednisolone and 1st cycle of Rituximab. She improved from a completely bedbound state to being able to walk with minimal support.

DISCUSSION

Though these patients presented as AMAN variant of GBS and sensory variant of CIDP both of them did not respond to IVIG. Nodo-paranodopathy should be suspected in patients presenting with acute to subacute onset predominantly distal motor neuropathy with rapid progression and severe nadir disability; associated features- severe sensory ataxia, tremor, autonomic dysfunction or respiratory insufficiency, associated disorders- Nephrotic syndrome; less responsive to IVIG and responsive to PLEX, rituximab, corticosteroids.



Anatomy and molecular organization of node

CONCLUSION

Patients with specific antibodies to NF186/NF 140 are rare.
Their prompt recognition and institution of appropriate therapy can lead to improved outcomes and help in prognostication and conserve costly resources.