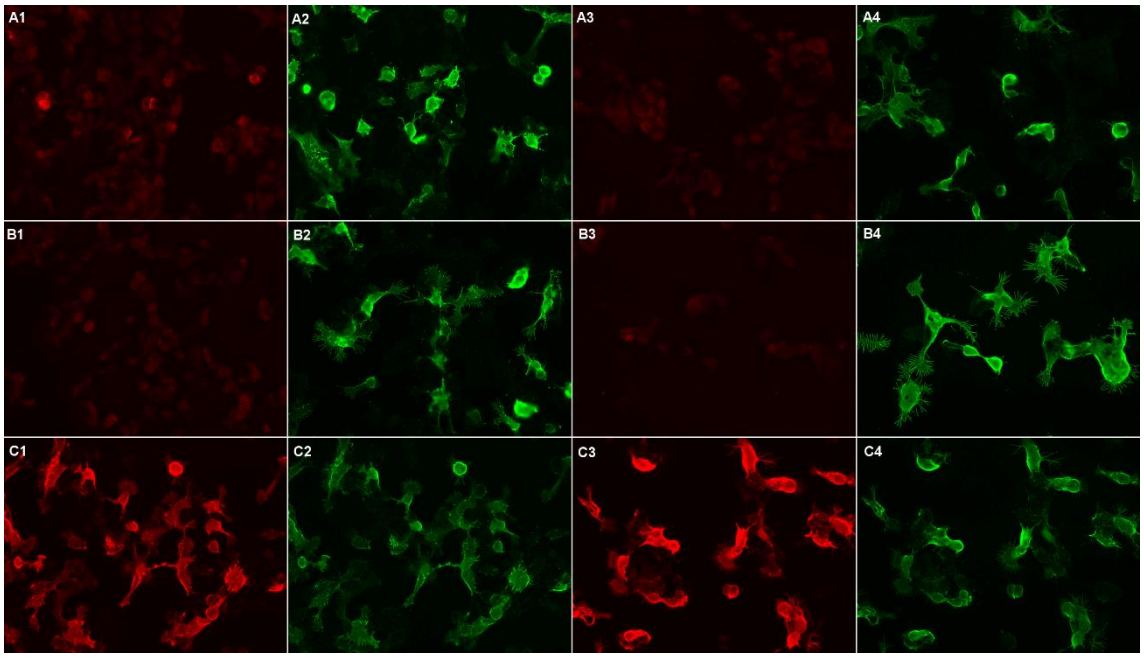
SUPPLEMENTARY RESULTS:

One patient was diagnosed post-mortem. This patient was initially diagnosed as a cervical myelopathy because the symptoms started after an accident. At that moment an electromyography, without nerve conduction studies, was performed showing the presence of spontaneous activity. The MRI did not show a cervical myelopathy and the clinical suspicion of CIDP was then considered but the EMG was not repeated. The patient was treated with corticosteroids and IVIg without response, so nodo/paranodal antibodies were then tested. The patient was tetraplegic, on a ventilator, and after careful medical-ethic deliberation, it was decided with the family to withdraw supportive therapy. Anti-NF155 positivity was received afterwards. A systemic autopsy was performed, revealing no additional organ damage of any relevance, but nerve histopathology could not be performed.

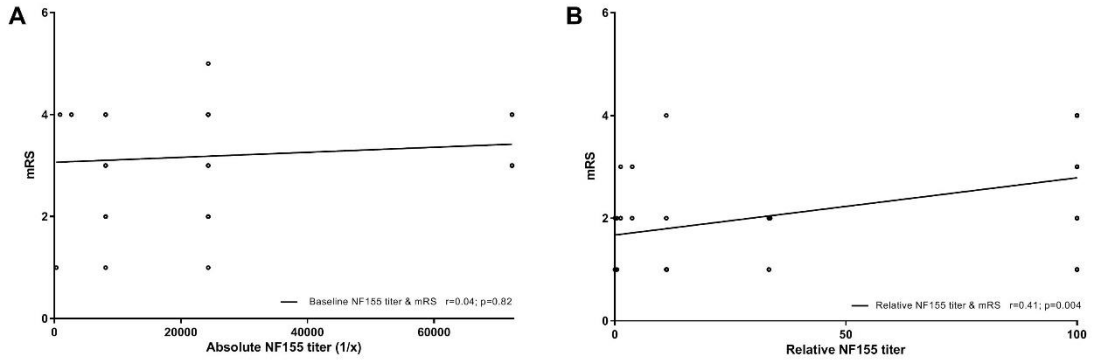
SUPPLEMENTARY FIGURES:

Figure e-1. CBA results for false positive patients for NF155.



The first row (A) corresponds to a patient with a false positive result, the second (B) corresponds to a negative control and the third (C) to a patient with anti-NF155 + AN. We show the sera reactivity in red and the HEK293 transfected cells with NF155 in green. Numbers 1 and 2 correspond to transfected cells with the NF155 plasmid containing the DDK-myc tag and numbers 3 and 4 to transfected cells with the untagged plasmid. In the false positive patient, we can observe a cross-reactivity (A1) that partially colocalized with the NF155 (A2) and that disappeared in the transfected cells with the untagged-neurofascin-155 plasmid (A3).

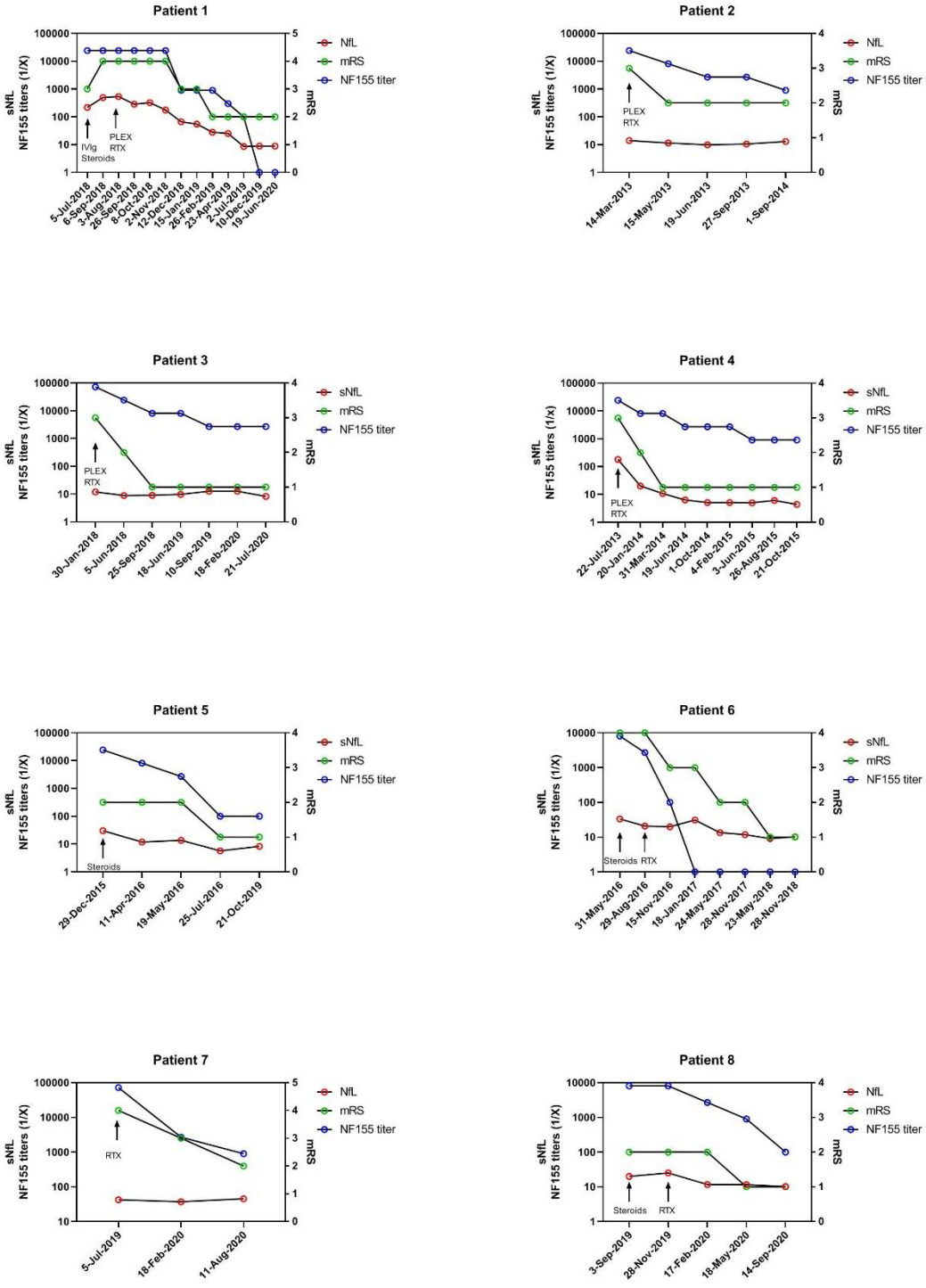
Figure e-2. A. Correlation between absolute NF155 titers at baseline and mRS. B. Correlation between relative NF155 titers and mRS.



The lines represent the correlation between NF155 titers and mRS.

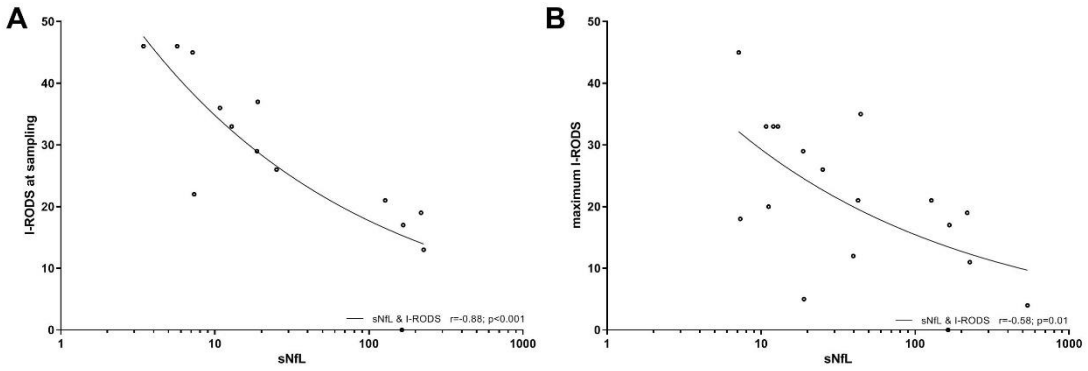
mRS= modified ranking scale, NF155= neurofascin 155

Figure e-3. Clinical status, sNfL and autoantibody titers in anti-NF155 AN patients.



IVIg= intravenous immunoglobulin; mRS= modified ranking scale; NF155= neurofascin-155; NfL= neurofilament light chain; PLEX= plasma exchange, RTX= rituximab

Figure e-4. A. Correlation between sNfL levels and I-RODS at sampling. B. Correlation between sNfL levels and maximum I-RODS.



The lines represent the correlation between sNfL levels and I-RODS. I-RODS= Inflammatory Rasch-built Overall Disability Scale; sNfL= serum neurofilament light chain.

SUPPLEMENTARY TABLES:

Table e-1. Correlation between sNfL, CMAP amplitudes and presence of spontaneous activity.

	Patients (n, %)	Rho Spearman	p
Peroneal CMAP	31 (77.5%)	-0.066	0.73
Tibial CMAP	27 (67.5%)	-0.042	0.84
Median CMAP	28 (70%)	-0.34	0.09
Ulnar CMAP	25 (62.5%)	-0.012	0.96
Sum CMAP	24 (60%)	-0.24	0.26
Spontaneous activity	24 (60%)	-	0.17

CMAP= compound muscle action potential. The sum of CMAP is reported in all patients in whom 4 nerves were evaluated in nerve conduction studies (peroneal, tibial, median, and ulnar).

Table e-2: Relationship between follow-up time and clinical improvement in rituximab-treated patients

Improvement in mRS after rituximab treatment	Number of patients (n, %)	Follow-up time (months; median, IQR)	p
No change	4 (17.4%)	9.5 [6-39.25]	0.43
1 point	6 (26%)	20.5 [7.25-31.5]	
2 points	8 (34.8%)	23.5 [17.25-65.5]	
3 points	5 (21.7%)	26.5 [12.5-46.5]	

mRS= modified ranking scale; IQR= interquartile range

Table e-3. Patients with and without facial diplegia, clinical status and sNfL.

	Patients with facial diplegia (n=7)	Patients without facial diplegia (n=33)	p
Maximum mRS (n=39) (median, IQR)	4 [3-4]	3 [2-4]	0.12
Final mRS (n=39) (median, IQR)	3 [2-3]	2 [1-2]	0.022
Maximum I-RODS (n=17) (median, IQR)	22 [4-30]	47 [38-57]	0.003
Final I-RODS (n=22) (median, IQR)	43	61 [55-88]	0.035
sNfL levels (median, IQR)	136.49 [9.5-51.04]	25.12 [9.53-51.04]	0.036

I-RODS: Inflammatory Rasch-built Overall Disability Scale; IQR= interquartile range; mRS= modified ranking scale.