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CASE REPORT

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The potential role of IgG subclass in NF155-associated nodopathy for targeting therapy: a pediatric case report highlighting the efficacy of pulsed oral corticosteroids

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Abstract

Background The autoimmune nodopathies are a distinct subgroup of immune-mediated polyneuropathies with characteristic clinical, pathophysiological, and electrophysiological features. IgG4 is the most frequently identified subclass and is typically associated with poor responsiveness to conventional therapies such as intravenous immunoglobulin and corticosteroids. However, emerging evidence highlights the relevance of non-IgG4 subclasses (e.g., IgG1), which may confer differential treatment responsiveness and prognostic implications.

Case presentation We report a 15-year-old boy with subacute, progressive distal sensorimotor polyneuropathy who initially fulfilled the clinical and electrophysiological criteria for the distal variant of chronic inflammatory demyelinating polyneuropathy. Despite multiple courses of IVIG, the patient's symptoms worsened, prompting further investigation for nodal/paranodal antibodies. Serum testing revealed high-titre anti-NF155 antibodies of the IgG1 subclass, the IgG4 subclass was not detected. Due to the negative results of IgG4 we have decided not to start treatment with rituximab but with high-dose pulsed oral dexamethasone which resulted in marked clinical and electrophysiological improvement over a 36-month period, with excellent tolerance and no adverse effects.

Conclusion Pediatric patients presenting with demyelinating polyneuropathies unresponsive to intravenous immunoglobulins should be evaluated for autoimmune nodo-paranodopathies, including subclass-specific antibody profiling. In IgG1-NF155 associated autoimmune nodopathy, pulsed oral corticosteroids may represent an effective therapeutic option, whereas the response to intravenous immunoglobulin can be suboptimal.

Keywords Nodo-paranodopathy, CIDP in children, Neurofascin-155, IgG subclasses, Immunotherapy, Pediatric neurology

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Introduction

Autoimmune nodo-paranodopathies have recently been recognized as a unique subset of immune-mediated neuropathies characterized by antibodies targeting cell-adhesion molecules at the nodes of Ranvier, such as neurofascin-155 (NF155), neurofascin-186 (NF186), contactin-1 (CNTN1), and contactin-associated protein 1 (CASPR1) [1–3]. Patients with these antibodies often present with a subacute onset, distal-dominant weakness, tremor, sensory ataxia, and significant disability, frequently accompanied by a poor response to intravenous immunoglobulin (IVIG), a clinical phenotype distinct from classic Guillain–Barré syndrome (GBS) or chronic inflammatory demyelinating polyneuropathy (CIDP) [1, 4–7] Paranodal dissection without classical macrophage-mediated demyelination is the characteristic feature of patients with autoantibodies to paranodal axo–glial junctional molecules [3].

While most cases of NF155 nodopathy are associated with IgG4 subclass antibodies and respond poorly to intravenous immunoglobulin (IVIG), recent studies have identified patients with exclusively non-IgG4 subclass (IgG1–3) antibodies, who may respond better to conventional immunotherapies [8–14]. Given the mechanistic and clinical heterogeneity associated with different IgG subclasses, their identification may play a potential role for treatment stratification [15].

In pediatric populations, nodo-paranodopathies remain underdiagnosed, and evidence guiding their management is limited [13, 14, 16–21]. Most therapeutic strategies are extrapolated from adult studies and traditional CIDP protocols, despite the unique immune and clinical profiles of these patients. Here, we present the case of an adolescent boy with IgG1 NF155-associated nodopathy who experienced significant and sustained clinical recovery following pulsed oral corticosteroid therapy.

Case report

A 15-year-old boy was admitted for evaluation of progressive lower limb weakness and gait disturbance. The symptoms had evolved over three months, beginning with lower back pain radiating to the left leg. He was initially managed for a presumed pseudoradicular syndrome, including rehabilitation. While the back pain resolved, new-onset distal lower limb weakness and paraesthesias progressed. He had a history of autoimmune thyroiditis, which was stable during follow-up, and had no family history of neuromuscular disorders.

Neurological examination revealed normal cognition, speech, and cranial nerve function. Upper limb strength was preserved proximally (Medical Research Council score - (MRC) 5) but mildly reduced distally (MRC 4), with discrete postural tremors and hyporeflexia. The lower limb findings included distal weakness (MRC 3), atrophy of the intrinsic foot and calf muscles, pes cavus, hammer toes, areflexia, impaired vibration sense, and a positive Romberg sign. Mild gait ataxia was also observed.

Routine blood tests, including autoimmune, infectious, metabolic, and vitamin panels, were unremarkable. CSF analysis revealed marked hyperproteinorachia (2.9 g/L) with a normal cell count. Electrophysiological studies were consistent with a symmetric, primary demyelinating sensorimotor polyneuropathy, fulfilling the EAN/PNS 2021 criteria [6] for distal variant CIDP (prolonged distal motor latencies, slowed conduction velocities, and delayed F-wave latencies).

Initial treatment with IVIG (2 g/kg followed by maintenance dosing of 1 g/kg every 4 weeks) yielded minimal improvement. After six months, distal muscle strength remained impaired (MRC 3), with a modified Rankin scale (mRS) score of 3, and electrophysiological parameters worsened, axonal involvement in lower limbs was also present. Given the lack of treatment response, hereditary neuropathy was considered; however, extensive genetic testing was negative.

Owing to the clinical course—subacute onset, pronounced distal involvement, gait ataxia, electrophysiological findings of severe primary demyelinating polyneuropathy with significantly prolonged distal latency, slow conduction velocity, prolonged minimal latency of F waves, severe axonal damage in lower limbs, significant CSF protein elevation, and poor IVIG response—serological testing for nodo/paranodal antibodies was performed. A cell-based assay confirmed anti-NF155 antibodies (endpoint titre of 1:200), with subclass analysis identifying IgG1. Other subclasses (IgG2,3,4) were not detected. No antibodies against NF186, CNTN1, or CASPR1 were detected (Fig. 1).

Treatment and outcomes

Owing to the lack of clinical response to IVIG and presence of IgG1 subclass of NF155 antibodies we decided to treat patient with corticosteroids. Pulsed oral corticosteroid therapy was initiated with dexamethasone 40 mg daily for four consecutive days every four weeks, alongside gastroprotective and bone health measures [22, 23]. After three months, the patient exhibited notable improvements in muscle strength, gait, and sensory symptoms.

The patient was monitored clinically and electrophysiologically every 3–6 months over the course of a 3-year follow-up. During this period, both his clinical status according to MRC sum score scale [24], upper and lower limb disability scale using the modified ONLS scale (ONLS - The Overall Neuropathy Limitations Scale, [25]), mRS - modified Rankin scale [26] and

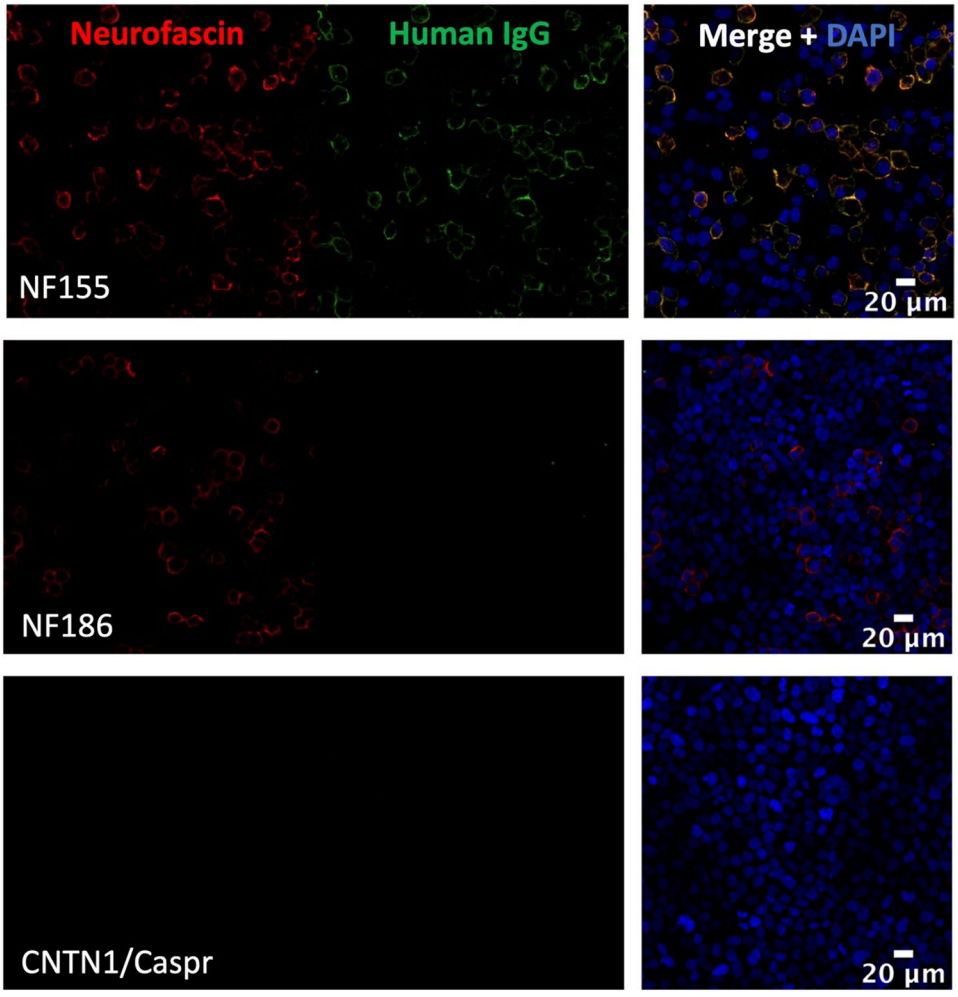

Fig. 1 Cell-based assays for nodal/paranodal antibodies HEK293 cells are transfected with plasmid vectors encoding the different isoforms of neurofascin, or co-transfected with plasmids encoding CNTN1 and Caspr1, which are subsequently expressed as proteins within the cell membrane. Incubation with patient sera allows antibodies (immunoglobulin IgG – green), if present, to bind to their target antigen. Colocalization with a commercial antibody that also targets neurofascin (red) confirmed this as the target antigen. The cell nuclei were stained with 4,6-diamidino-2-phenylindole (DAPI) (blue). Patients have antibodies targeting neurofascin isoform 155. Some nonspecific nuclear and perinuclear IgG staining can also be observed

electrophysiological findings demonstrated consistent improvement, as shown in the corresponding tables. In parallel, we gradually reduced the dose of dexamethasone in a stepwise manner, guided by the clinical response and tolerance, as detailed in the treatment timeline (Table 1).

At the 12-month follow-up, his paraesthesia had resolved, his upper limb strength was fully restored and his lower limb strength improved significantly (MRC sum score 58). The patient resumed athletic activities, including high-impact sports, and had an mRS score of 1. At the 24-month follow-up he was without clinical difficulties (ONLS score 0, MRC sum score 60, mRS 0).

Electrophysiological summary

Serial nerve conduction studies demonstrated significant improvements in distal latencies and conduction velocities (Table 2). Treatment was well tolerated, with no adverse effects reported. The regimen was tapered slowly down over a period of 3 years, with continued improvement observed.

Over the course of 36 months of dexamethasone therapy, the patient demonstrated substantial and sustained improvements in motor and sensory conduction parameters across multiple nerves. The CMAP amplitudes and conduction velocities improved progressively, whereas the distal latencies and F-wave latencies normalized. Sensory responses (particularly SNAPs) also showed marked recovery, reflecting both remyelination and a reduction in conduction block.

After 3 years of treatment, the serum IgG1 NF155 antibody titre was no longer detected. These findings paralleled clinical improvements and suggest IVIG resistance and the sustained remission following corticosteroids can be observed in IgG1 anti-NF155 autoimmune nodopathy.

Table 1 Summary of muscle strength, height, weight and disability scales before and during treatment

Timepoint (month)	Dexamethasone Dose (mg/day/4days)	Height/Weight (cm/kg)	Clinical Status (ONLS Score)	MRC Sum Score	mRS
Baseline (0)	40	182/83	5	52	3
3	40	183/83	4	54	3
6	30	183/83	3	58	2
9	20	185/84	2	58	2
12	10	188/85	2	58	1
18	8	192/85	1	60	1
24	4	194/87	0	60	0
30	2	195/87	0	60	0
36	2	195/87	0	60	0

Discussion

Autoimmune nodopathies associated with antibodies to paranodal proteins, particularly neurofascin-155, represent a distinct subgroup of immune-mediated polyneuropathies with unique clinical, pathophysiological, and therapeutic implications. While most described cases involve IgG4 NF155 antibodies, an increasing number of reports have identified non-IgG4 subclasses (IgG1, IgG2, IgG3, and IgM), highlighting significant heterogeneity in disease mechanisms and treatment responses [8–10, 13, 14, 27–33].

Epidemiologically, NF155 nodopathy is most often reported in young adult males; however, pediatric cases are increasingly recognized, often with more aggressive onset and rapid progression, sometimes misdiagnosed as Guillain–Barré syndrome (GBS) or chronic inflammatory demyelinating polyneuropathy (CIDP) [16–19, 34]. Children frequently present with ataxia, tremor, and early gait impairment, while cranial neuropathy, papilledema, or cerebellar ataxia are less common compared to adults [16, 17]. Supporting paraclinical findings such as markedly elevated CSF protein, CIDP-like NCS abnormalities, and lumbosacral root enlargement on MRI further aid diagnosis and overlap with adult findings [16, 17].

Our patient initially met the diagnostic and electrophysiological criteria for a distal variant of chronic inflammatory demyelinating polyneuropathy [6], therefore he was treated with IVIG in therapeutic dose (2 g/kg followed by maintenance dosing of 1 g/kg every 4 weeks). Given the lack of treatment response, and clinical and electrophysiological finding of a distal sensory motor polyneuropathy with foot deformity, hereditary neuropathy was also considered; however, extensive genetic testing was negative. The similar findings resembling hereditary neuropathy in patients with NF 155 autoimmune nodopathy have also been described in another study [13].

However, a significantly high level of protein in the cerebrospinal fluid is an important red flag for considering autoimmune-mediated nodopathy. Since this is a treatable polyneuropathy, early testing for antibodies to nodopathy in children with neuropathies and significant proteinorrachia might help to prevent irreversible nerve damage. A cell-based assay confirmed in our patient high-titre anti-NF155 antibodies (endpoint titre 1:200), with subclass analysis identifying IgG1 subtype. Neither other subtypes (IgG2,3,4 or IgM), nor antibodies against NF186, CNTN1, CASPR1 were detected. Indeed, our patient showed marked and sustained improvement with pulsed oral dexamethasone with normalization of antibody titers and no relapse occurring during the five years follow-up.

By contrast, non-IgG4 NF155 nodopathies (IgG1, IgG2, IgG3, or IgM) show more heterogeneous clinical and therapeutic outcomes [33]. Several mechanisms may explain this variability:

a) IgG1 and IgG3 antibodies are strong activators of the classical complement pathway and can mediate antibody-dependent cellular cytotoxicity (ADCC). This makes them pathogenic through inflammatory injury at the paranodes, causing conduction block and structural disruption [11, 23, 31, 35]. Consequently, patients with IgG1/3 antibodies may respond to therapies that suppress inflammation and complement activation, including corticosteroids, IVIG, or plasma exchange [8, 36, 37].
b) IgG2 antibodies are less efficient at complement activation, and their role is less well defined. Interestingly, Harris et al. described pediatric patients with IgG2-dominant NF antibodies who exhibited variable responses to IVIG, with some responding better to corticosteroids [14]. This suggests possible subclass–phenotype correlations that could guide individualized therapy.
c) IgM antibodies, although less common, have also been reported in nodopathies, sometimes associated with tremor. Their pathogenic role remains unclear, but they may indicate early or transient immune activation [33, 38]. These facts suggest the importance of IgG subclass determination in therapeutic planning.

Table 2 Electrophysiology of NF155 IgG1 before and after treatment with dexamethasone

Nerve	Parameter	1/2021 Before DEX	5/2021 3 mo DEX	5/2022 12 mo DEX	5/2023 24 mo DEX
Median	ML w/e/E (ms)	9.5/14.9/19.8	7.4/12.3	6.7/11.4/17.1	6.1/11.1
	CMAP w/e/E (mV)	8.0/7.4/3.2	9.5/9.0	9.6/9.6/3.6	9.9/9.1
	MCV w-e/e-E (m/s)	46.3/53.1	47.3/51.4	51.4/55.7	51.8
	FW lat (ms)	42.6	35.9	35.3	35.3
	DSL (ms)	ND	4.46	4.0	3.75
	SNAP (μV)	ND	0.48	4.3	5.5
	SCV (m/s)	ND	36.4	40.5	43.2
Ulnar	ML w/be/ab/E (ms)	7.4/12.6/14.5/18.7	5.3/10.4	4.8/9.5/11.2	4.4/8.7/10.6
	CMAP w/be/ab/E (mV)	2.3/3.7/3.6/3.2	3.1/3.8	4.5/4.4/4.3	5.6/5.4/5.2
	MCV w-e/be-ab/ab-E (m/s)	46.0/52.6/59.5	47.0	55.4/54.5	55.4/57.5
	FW lat (ms)	53.8	43.8	37.2	35.4
	DSL (ms)	ND	4.13	3.96	3.75
	SNAP (μV)	ND	5.2	17.3	22.1
	SCV (m/s)	40.5	40.5	43.2	–
Tibial	ML a/p (ms)	12.9/25.5	10.9/22.3	10.9/21.3	8.1/18.2
	CMAP a/p (mV)	0.48/0.66	0.94/0.78	2.2/1.5	3.8/3.6
	MCV a-p (m/s)	33.3	35.5	36.8	42.7
	FW lat (ms)	79.4	66.5	66.6	66.8
Peroneal	ML d/p/a (ms)	14.9/26.1/29.9	11.5/23.7/26.9	10.2/20.8/23.3	8.8/17.0/20.1
	CMAP d/p/a (mV)	0.83/0.9/0.9	1.5/1.2/0.98	3.7/3.4/2.7	4.4/4.1/3.3
	MCV d/b/a (m/s)	28.5/26.3	31.3/29.4	35.5/36.4	41.0/37.9
	FW lat (ms)	ND	74.5	69.2	68.9
Sural	SCV (m/s)	ND	40.5	40.5	42.3
	SNAP (μV)	ND	0.43	5.3	5.5
	DSL (ms)	ND	3.82	3.86	3.75

Abbreviations:CMAP Compound Muscle Action Potential, *DEX* Dexamethasone, *DSL* Distal Sensory Latency, *FW lat* F-wave Latency, *MCV* Motor Conduction Velocity, *ML* Motor Latency, *ND* Not Determined, *SCV* Sensory Conduction Velocity, *SNAP* Sensory Nerve Action Potential, *a/p* ankle/popliteal fossa, *d/b/a* distal/below/above fibular head, *w/e* wrist/elbow, *w/e/E* wrist/elbow/Erb's point

In pediatric CIDP more broadly, pulsed high-dose oral dexamethasone and other steroid regimens have shown favorable efficacy and safety, with reduced cumulative toxicity compared to daily administration [22, 23, 39]. Long-term steroid use in children carries well-established toxicities, including growth suppression, osteoporosis, weight gain, and behavioral effects [39, 40]. Furthermore, starting with corticosteroids provides rapid immunosuppression while allowing time to complete subclass testing and update vaccinations, which is critical before B-cell–depleting therapy given the risk of prolonged vaccine response blunting [41, 42]. The optimal treatment strategy for paediatric NF155 nodopathy remains uncertain, but we supposed that subclass-specific immunopathology provides a rationale for initiating therapy with corticosteroids and reserving rituximab for refractory or IgG4-mediated disease. Plasma exchange can provide rapid symptomatic improvement by removing pathogenic antibodies, but its effects are typically short-lived without maintenance therapy.

Conclusion

Conclusion Taken together, the evidence suggests that the IgG subclass profile in NF155 nodopathy plays an important role in determining the clinical course, prognosis and response to therapy. Our patient with IgG1 NF155 nodopathy experienced a poor therapeutic effect from IVIG, but a good response to corticosteroids with long-lasting remission. Therefore, subclass-specific antibody testing could provide more information for complex diagnostic evaluations, especially in pediatric patients with atypical CIDP presentations and/or recorded IVIG therapeutic resistance. Although NF155-IgG4 is frequently and predominantly elevated in patients with NF155-IgG, evaluating only NF155-IgG4 may result in overlooking patients who possess NF155-IgG1, IgG2, IgG3 or NF155-IgM antibodies. Standard subclass NF155-IgG definition in selected patients may be in the near future an additional tool for optimising the management of CIDP-like NF155 nodopathies.

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Authors' contributions
MTK designed the case report, collected and analyzed the data. ET, VM, SR assisted in datacollection. EK, SR assisted in data interpretation and revision. All authors reviewed andapproved the final version of the manuscript.

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Data availability
All data generated or analysed during the study are included in this published article.

Declarations

Ethics approval and consent to participate
All procedures performed in this study involving human participants were conducted in accordance with the ethical standards of the institutional and/or national research committee, and with the 1964 Helsinki Declaration and its later amendments or comparable ethical standards. Written informed consent was obtained from parents of the patient of this Case report.

Consent for publication
Written informed consent for publication of clinical details was obtained from the parents of the patient of this Case report. A copy of the written consent is available for review by the Editor of this journal.

Competing interests
The authors declare no competing interests.

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