Section Editors: Clare Fraser, MD
Susan Mollan, MD

# Vision Loss as a Presenting Feature of Chronic Inflammatory Demyelinating Polyneuropathy: A Case Series

Adam M. Kruszewski, MD, Zujaja Tauqeer, MD, DPhil, Elana A. Meer, BA, Sana Ali Bautista, MD, Neena R. Cherayil, MD, Marcus E. Cimino, MD, Sami L. Khella, MD, Eric Lancaster, MD, PhD, Shawn J. Bird, MD, Steven S. Scherer, MD, PhD, Karen E. Revere, MD, Ali G. Hamedani, MD, MHS, Grant T. Liu, MD, Madhura A. Tamhankar, MD



**Background:** Chronic inflammatory demyelinating polyneuropathy (CIDP) is an acquired, immune-mediated, and clinically heterogeneous demyelinating disease affecting the nerve roots and peripheral nerves. We report a series of 4 patients who presented with early and progressive vision loss in the context of new-onset CIDP: 3 due to papilledema and 1 due to optic neuropathy without papilledema.

**Methods:** This was a retrospective case series of 4 patients with vision loss as a presenting feature of CIDP evaluated at the Hospital of the University of Pennsylvania from January 2016 to August 2021. Demographic, clinical, diagnostic, and treatment data were collected via retrospective medical record review.

**Results:** Case 1 was a 51-year-old man with 2 months of progressive bilateral papilledema associated with reduced visual acuity (count fingers at 1 foot in each eye) and severely constricted visual fields. Case 2 was a 36-year-old man with 4 months of worsening headaches, reduced visual acuity (count fingers at 1 foot in each eye), severely constricted visual fields, and papilledema. Case 3 was a 39-year-old man with papilledema causing progressive vision loss (20/80 in both eyes), headaches, and relapsing limb sensorimotor deficits. Case 4 was a 19-year-old man with 3 months of progressive bilateral visual decline (20/400 in the right eye, 20/600 in the left eye), central scotoma, and optic disc pallor consistent with optic neuropathy without papilledema. All 4 patients met clinical and electrodiagnostic criteria of CIDP. Cases 3 and 4 each tested positive for serum neurofascin-155 IgG4 antibodies. All patients were managed with immunomodulatory therapy. Cases 1 and 2 also each required surgical intervention with bilateral optic nerve sheath fenestration and cerebrospinal fluid (CSF) shunting procedures.

**Conclusion:** Vision loss from optic neuropathy with or without papilledema has rarely been reported in CIDP, and typically has been described in the context of longstanding disease. Our cases highlight how CIDP can present with early vision loss that may be profound and challenging to manage if diagnosis is delayed. CIDP should be considered in any patient with new progressive vision loss when associated with peripheral sensorimotor symptoms and elevated CSF protein. The small subgroup of CIDP patients with neurofascin-155 antibodies may be at particular risk of optic nerve involvement.

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Departments of Ophthalmology (AMK, ZT, SAB, GTL, MAT), and Neurology (AMK, MEC, SLK, EL, SJB, SSS, AGH, GTL, MAT), University of Pennsylvania, Philadelphia, PA; Division of Ophthalmology (ZT, SAB, KER, GTL), The Children's Hospital of Philadelphia, Philadelphia, PA; The Perelman School of Medicine (EAM, SLK, EL, SJB, SSS, KER, AGH, GTL, MAT), University of Pennsylvania, Philadelphia, PA; and Departments of Neurology and Ophthalmology (NRC), Feinberg School of Medicine, Northwestern Medicine, Chicago, IL.

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A. M. Kruszewski and Z. Tauqeer contributed equally to this article as co-first authors.

Address correspondence to Madhura A. Tamhankar, MD, Division of Neuro-Ophthalmology, Scheie Eye Institute, University of Pennsylvania, 51 N. 39th Street, Philadelphia, PA 19104; E-mail: madhura.tamhankar@pennmedicine.upenn.edu

Chronic inflammatory demyelinating polyneuropathy (CIDP) is an immune-mediated demyelinating neuropathy that predominantly affects nerve roots and peripheral nerves. CIDP diagnosis is based on at least 2 months of progressive or relapsing motor and/or sensory dysfunction, absent limb reflexes, elevated cerebrospinal fluid (CSF) protein without pleocytosis, and electrodiagnostic (or histopathologic) evidence of demyelination (1).

Although CIDP is a peripheral neuropathy, central nervous system (CNS) involvement can occur. Electrophysiologic studies have detected optic nerve dysfunction in

patients with CIDP, even in the absence of overt visual symptoms (2). Patients with known CIDP have been reported to develop papilledema, optic neuropathy without papilledema, or ophthalmoparesis (3,4). Although the occurrence of vision loss with CIDP has been reported in the literature, it is rare and mostly described in the context of longstanding disease (3,4).

## METHODS

This was a retrospective case series of 4 patients with vision loss as a presenting feature of CIDP evaluated at the hospital of the University of Pennsylvania from January 2016 to August 2021. Data on patient demographics, history, examination, diagnostic testing, and treatment were collected via retrospective medical record review and reported in a deidentified manner. Each patient met clinical and electrodiagnostic criteria for CIDP (1).

## RESULTS

### *Case 1*

A 51-year-old obese man (body mass index [BMI], 43.2 kg/m$^2$) was found to have bilateral optic disc edema (Fig. 1A, B) with normal afferent visual function on routine ophthalmologic examination. He was initially managed outside of our institution without neuro-ophthalmology referral. MRI brain was normal. Per records, initial diagnosis was presumed pseudotumor cerebri syndrome (PTCS) given presence of bilateral optic disc edema, obesity, and normal MRI brain in the absence of other neurologic symptoms. He was placed on acetazolamide 500 mg twice daily. Lumbar puncture (LP) was not completed at symptom onset. Over the subsequent 2 months, he developed progressive bilateral visual decline and mild distal limb sensorimotor dysfunction. He was then referred to neuro-ophthalmology emergently. Examination revealed reduced visual acuity (VA) of 20/200 in the right eye and 20/250 in the left eye, 0/11 color plates in both eyes, constricted visual fields in both eyes, and sluggish pupil constriction to light in both eyes without relative afferent pupillary defect (rAPD). Fundus examination revealed bilateral grade 4 optic disc edema with disc hemorrhages and macular edema (Fig. 1C, D). He underwent urgent LP, which showed elevated opening pressure of 47 cm $H_2O$ and elevated CSF protein of 255 mg/dL (normal 15–45 mg/dL).

Because of profound visual decline, he was started on intravenous (IV) acetazolamide 1,000 mg twice daily and IV methylprednisolone 250 mg 4 times daily. Neurologic examination revealed proximal-predominant limb weakness, distal limb sensory loss, absent distal limb reflexes, and gait dysfunction. Nerve conduction studies (NCS) and electromyography (EMG) showed an acquired demyelinating motor-sensory polyneuropathy. Repeat MRI and MR venogram (MRV) of the head revealed signs of intracranial hypertension without mass or venous sinus thrombosis (Fig. 2). He met diagnostic criteria for

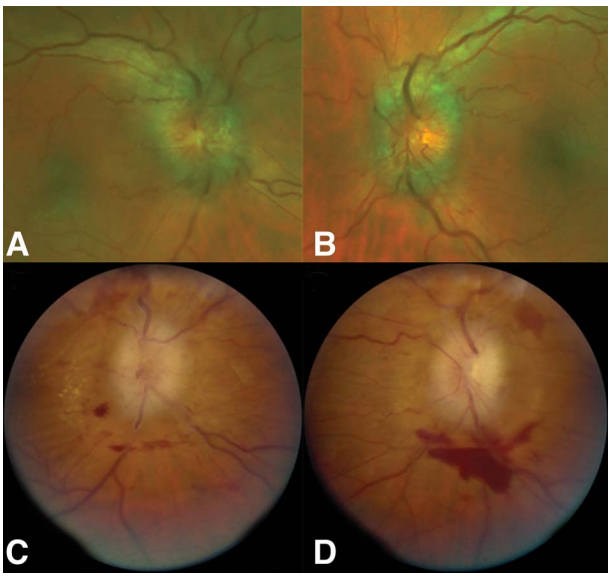


**FIG. 1.** Case 1. Initial Optos fundus images of the right (**A**) and left (**B**) optic nerve showing 360° optic disc elevation with partial obscuration of vessels. Fundus photographs of the right (**C**) and left (**D**) optic nerve showing worsened grade IV papilledema with 360° swelling, peripapillary retinal and preretinal hemorrhages, and partial obscuration of vessels.

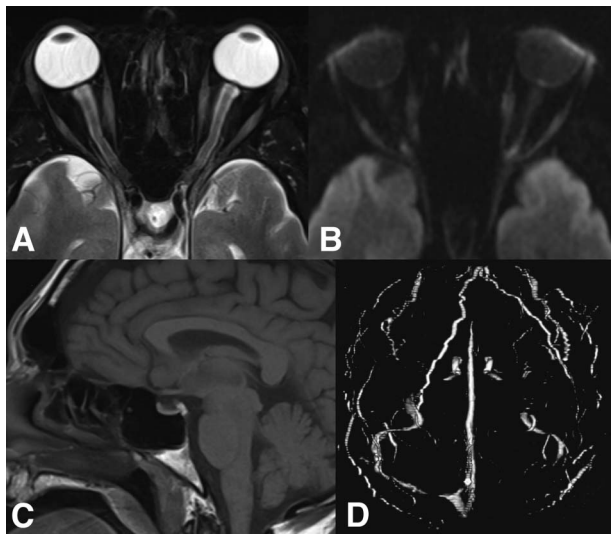


**FIG. 2.** Case 1. MRI/MRV brain with signs of intracranial hypertension. (**A**) Axial MRI T2 image showing mild prominence of the CSF surrounding the optic nerves with protrusion into the posterior sclera, flattening of the posterior globes, and mild tortuosity of each optic nerve. (**B**) Axial MRI DWI image with faint diffusion restriction of the optic discs. (**C**) Sagittal MRI T1 image showing partially empty sella, with concavity of the superior pituitary margin. (**D**) Axial MR Venography three-dimensional reconstruction showing stenosis of the left > right transverse venous sinuses without evidence of venous sinus thrombosis. CSF, cerebrospinal fluid; MRV, MR venogram.

CIDP. Treatment with plasmapheresis followed by intravenous immunoglobulin (IVIG) was initiated. Because of vision threatening papilledema, he underwent emergent surgical CSF diversion via external ventricular drain placement. Despite these measures, visual acuity declined to counting fingers at 1 foot in each eye. He then underwent bilateral optic nerve sheath fenestration (ONSF) to salvage vision. A significant egress of fluid was observed intraoperatively on decompression of each optic nerve sheath. Postoperatively, despite improved papilledema, there was optic atrophy and hand motion visual acuity in both eyes. Following IVIG treatment, polyneuropathy symptoms gradually improved over weeks to months.

### *Case 2*

A 36-year-old obese man (BMI 45.5 kg/m$^2$) presented with 4 months of progressively worsening bilateral vision loss, headaches, distal leg numbness, and gait instability. Neuro-ophthalmologic evaluation revealed reduced visual acuity (20/30 in the right eye and 20/25 in the left eye), 5/8 color plates in both eyes, and dense nasal greater than temporal visual field constriction bilaterally. Fundus examination revealed severe bilateral optic disc edema (Fig. 3A, B). Static perimetry testing showed severe generalized constriction with relative temporal and paracentral sparing in both eyes (Fig. 4A, B). Neurologic examination demonstrated distal leg sensory loss and absent ankle reflexes. LP showed elevated opening pressure of 38 cm $H_2O$ and elevated CSF protein of 531 mg/dL. MRI brain showed signs of increased intracranial pressure (Fig. 4C, D). EMG/NCS demonstrated an acquired demyelinating motor-sensory polyneuropathy. He was diagnosed with CIDP and treated with IVIG. Because of vision threatening bilateral papilledema, he was treated with IV acetazolamide, IV methylprednisolone, and underwent bilateral ONSF. A significant egress of fluid was noted on decompression of each optic nerve sheath. Fundus examination subsequently showed improvement in optic nerve swelling (Fig. 3C, D). He was continued on an oral prednisone taper, oral acetazolamide 1,000 mg 3 times daily, and IVIG every 3 weeks. After immune therapy, polyneuropathy symptoms improved.

One month later, he developed worsening vision and headaches. Neuro-ophthalmic examination showed VA of 20/400 in both eyes, markedly constricted visual fields, 0/8 color plates in both eyes, and optic disc edema with pallor in both eyes. Repeat LP revealed elevated opening pressure at 30 cm $H_2O$. Vision deteriorated over days to counting fingers at 1 foot in both eyes, and an emergent ventriculoperitoneal shunt was placed. On follow-up, VA improved to 20/50 right eye and 20/500 left eye, visual field constriction slightly improved in both eyes, and fundus examination showed severe bilateral optic disc pallor.

### *Case 3*

A 39-year-old man developed limb numbness, weakness, headaches, and mild vision loss 1 to 2 weeks after an upper respiratory infection. These symptoms resolved without intervention over several weeks, except for persistent mild subjective vision blurriness. Two months after initial symptom onset, gait dysfunction worsened, and he was

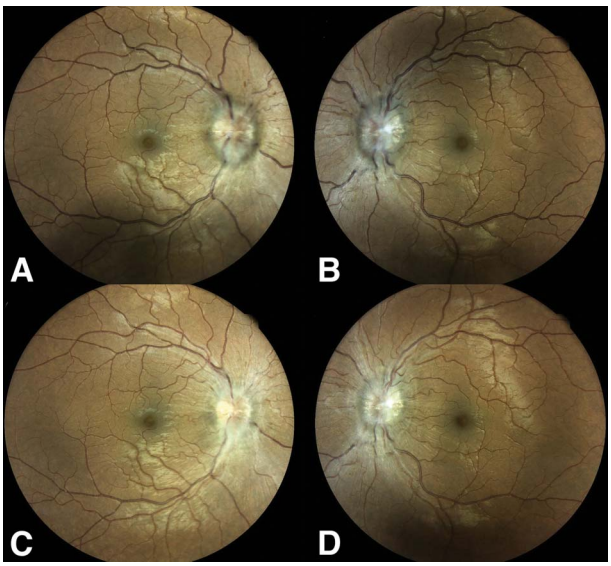


**FIG. 3.** Case 2. Fundus photographs of the right (**A**) and left (**B**) optic nerve showing severe papilledema with 360° swelling and partial obscuration of vessels. Fundus photographs of the right (**C**) and left (**D**) optic nerve showing improved papilledema after bilateral optic nerve sheath fenestration.

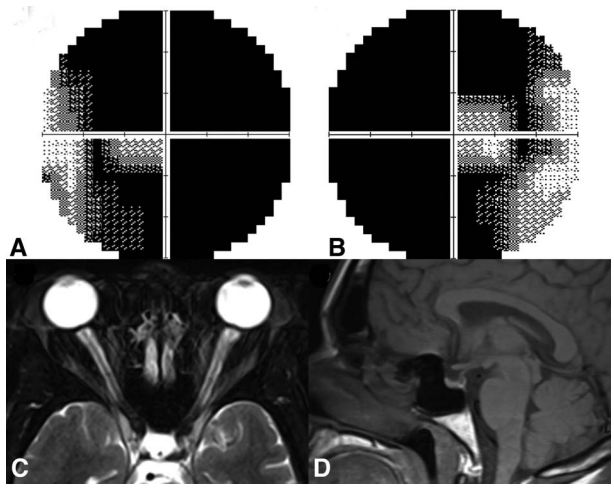


**FIG. 4.** Case 2. Humphrey visual field 30-2 demonstrating severe constriction of the left (**A**) and right (**B**) visual fields. (**C**) T2 axial MRI image showing enlargement of the CSF spaces in the optic nerve sheaths and flattening of the posterior globes. (**D**) T1 sagittal MRI image showing partially empty sella, with mild concavity of the superior pituitary gland. CSF, cerebrospinal fluid.

evaluated by neurology. Examination revealed moderate limb weakness, numbness, and absent leg reflexes. EMG/NCS revealed an acquired demyelinating motor-sensory polyneuropathy. MRI brain was unremarkable. He was diagnosed with CIDP 4 months after symptom onset. Treatment with monthly IVIG was initiated; however, gait dysfunction did not improve.

At 10 months after initial symptom onset, he developed acute on chronic blurred vision with worsened headaches and pulsatile tinnitus. Ophthalmologic examination revealed reduced visual acuity of 20/50 in the right eye and 20/40 in the left eye, mildly reduced color vision in both eyes, full visual fields to confrontation, sluggish pupillary reaction to light in both eyes, and bilateral 360° optic disc edema. He was referred to neuro-ophthalmology. Over weeks, visual acuity worsened to 20/80 in both eyes and automated perimetry showed enlarged blind spots in both eyes. Urgent MRI/MRV of the head was negative for structural abnormality. LP opening pressure was 23 cm $H_2O$, with significantly elevated CSF protein of 550 mg/dL. He was treated with plasmapheresis, oral acetazolamide 500 mg twice daily, and oral prednisone 80 mg daily with slow taper. Visual symptoms and headaches improved following treatment. Within a month, visual acuity improved to 20/30 in the right eye and 20/25 in the left eye, color vision normalized, and papilledema significantly improved. However, polyneuropathy symptoms continued to gradually worsen despite plasmapheresis, ongoing IVIG, and oral prednisone treatments. Extensive work-up was negative including serum protein electrophoresis (SPEP), vascular endothelial growth factor (VEGF), myelin-associated glycoprotein (MAG) antibodies, IL-6 titer, human herpes virus 8 DNA quantification, skeletal survey, and body CT. He underwent a trial of monthly IV cyclophosphamide for 6 months without benefit. He then tested positive for serum neurofascin-155 IgG4 antibodies. He was subsequently initiated on rituximab therapy. He showed improvements in limb strength, sensation, and gait after 1 dose of rituximab. Within several months after treatment, he was able to walk unassisted.

### *Case 4*

A 19-year-old man presented to the ophthalmology clinic with 3 months of progressive bilateral vision loss, gait imbalance, and distal limb numbness and weakness. Initial examination noted reduced visual acuity (20/200 in the right eye, count fingers in the left eye), dyschromatopsia, and normal appearing optic discs and maculae on fundus examination. Static perimetry demonstrated central scotomas breaking out temporally in the right eye and infratemporally and supranasally in the left eye (Fig. 5A, B). He was referred to neurology and examination revealed mildly reduced strength and sensation in the bilateral distal limbs, unsteady

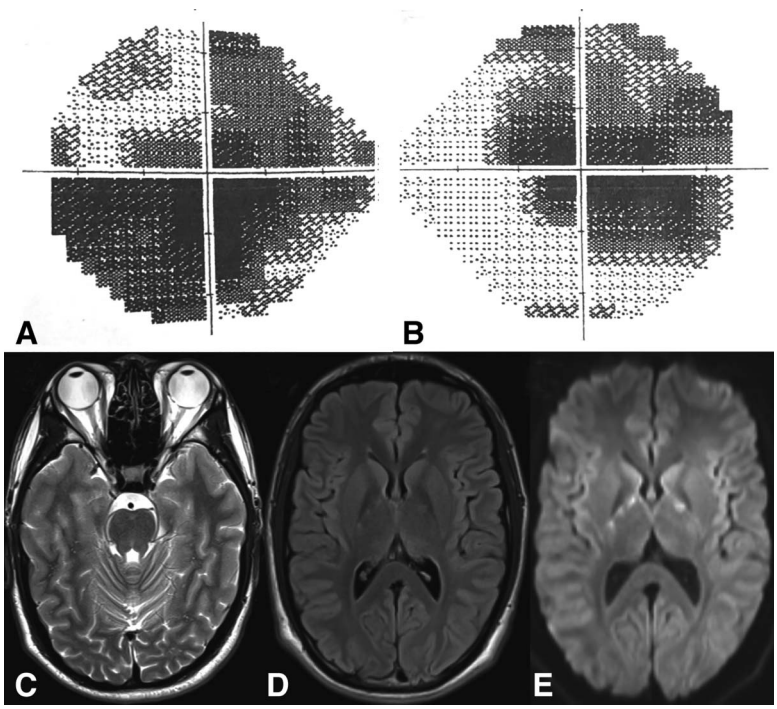


**FIG. 5.** Case 4. Humphrey visual field 24-2 demonstrating central scotomas breaking out infratemporally and supranasally in the left eye (**A**) and temporally in the right eye (**B**). (**C**) T2 axial MRI image showing normal appearing orbits. (**D**) Axial MRI FLAIR image showing foci of hyperintensity in the bilateral posterior limb of the internal capsule. This was associated with increased signal on isotropic diffusion-weighted imaging (**E**) confirmed by decreased signal on apparent diffusion coefficient imaging, consistent with abnormal diffusion restriction in this region.

gait, and absent limb reflexes. EMG/NCS revealed an acquired demyelinating motor-sensory polyneuropathy. LP opening pressure was normal at 14 cm $H_2O$ with elevated CSF protein of 396 mg/dL. MRI brain showed a few foci of nonenhancing T2/FLAIR hyperintensity in the bilateral posterior limb of the internal capsule associated with restricted diffusion (Fig. 5C, D, E). The optic nerves and visual pathways appeared normal on MRI. Extensive work-up was unrevealing with normal vitamin B12, folate, vitamin B1, creatine kinase, VEGF, SPEP, as well as negative Lyme, Sjogren, antinuclear, double-stranded DNA, and MAG antibodies. The patient was diagnosed with CIDP and started on monthly IVIG, but his symptoms did not improve.

Neuro-ophthalmologic evaluation revealed visual acuity of 20/400 in the right eye and 20/600 in the left eye. Color plates were 0/8 in both eyes. Pupils sluggishly constricted to light in each eye without rAPD. Visual field testing revealed persistent bilateral central scotomas. There was trace temporal optic disc pallor with sharp disc margins in both eyes on fundus examination. Visual evoked potential studies revealed absent P100 responses in each eye. Vision loss was attributed to chronic optic neuropathy in association with CIDP. He was found to have IgG4 antibodies against neurofascin-155. Monthly treatments of high-dose IV methylprednisolone were initiated while awaiting insurance approval for rituximab. After 4 months of steroid treatment, he showed improvement in polyneuropathy symptoms. Visual acuity also improved to 20/60 in the right eye and 20/50 in the left eye with improvements in color vision. Rituximab was initiated along with a prolonged oral prednisone taper, and polyneuropathy symptoms continued to improve. Three months after rituximab initiation, there was significant improvement in visual acuity (20/25 in both eyes) and color vision (11/11 color plates in both eyes), with persistent trace right rAPD.

## CONCLUSIONS

We report a series of 4 patients who presented with early and progressive vision loss in the context of new onset CIDP: 3 due to papilledema and 1 due to optic neuropathy without papilledema. Furthermore, 2 of these cases had IgG4 antibodies against neurofascin-155, which have been rarely identified in patients with CIDP (5,6).

### *Discussion*

All 4 cases exhibited moderate-to-severe vision loss as a presenting feature of CIDP and were treated with immunomodulatory therapy. The 3 cases with papilledema received acetazolamide. Cases 1 and 2 had severe papilledema from CIDP, requiring surgery with CSF shunting and ONSF procedures. ONSF has been rarely performed in CIDP-associated intracranial hypertension, with favorable visual outcome (7). Although vision improved following early surgical decompression in Case 2, vision did not improve in Case 1. Cases 3 and 4 showed significant vision improvement with medical therapy alone, and both had antibodies to neurofascin-155.

Visual dysfunction from optic neuropathy with or without papilledema in CIDP is rare. Papilledema associated with acute polyneuritis was first described by Gilpin in 1936 (8). A review of the literature found papilledema reported in 8 of 613 (1.3%) CIDP patients from 8 large case series (3,9–15). However, these studies did not comment on the presence, timing, degree, or mechanism of visual impairment in affected patients. Other reports describe papilledema occurring long after limb polyneuropathy onset in CIDP (4). Very few cases report optic nerve dysfunction as a presenting feature in new-onset CIDP (4,16), and none clearly describe papilledema preceding polyneuropathy. Acute and chronic optic neuropathy in patients with known CIDP have been rarely described, and very few report acute optic neuritis preceding limb neuropathy (2,17). Visual evoked potential studies have revealed electrophysiologic evidence of subclinical optic nerve dysfunction in those with long-standing CIDP (2,4). A recent optical coherence tomography study found subtle retinal ganglion cell/inner plexiform layer neurodegeneration in patients with CIDP compared with healthy controls (18). The precise mechanism of optic nerve dysfunction in CIDP remains unclear. One theory is that elevated CSF protein causes obstruction of arachnoid granulations, thereby increasing CSF pressure leading to papilledema, perhaps in combination with an immune-mediated or infiltrative optic neuropathy (4).

Given optic neuropathy in CIDP is rare, other differential diagnostic considerations of optic disc edema must be considered. These include more common conditions such as PTCS, inflammatory, infiltrative, infectious, or neoplastic causes. PTCS is a common cause of papilledema in overweight individuals and was a diagnostic consideration in Cases 1 and 2 until CSF analysis revealed elevated CSF protein, warranting evaluation for alternative etiologies. In Case 1, it was atypical that mild papilledema preceded vision loss and polyneuropathy symptoms for 2 months. Notably, both patients with severe papilledema (Cases 1 and 2) were also obese. One could question whether obesity predisposed these patients to abnormal CSF flow dynamics that were exacerbated by CIDP, leading to a more fulminant increase in intracranial pressure compared with nonobese patients, although this remains speculative and warrants further study. Despite multiple medical and surgical attempts to lower intracranial pressure in Case 1, vision did not improve. This led us to hypothesize that a CIDP-associated immune-mediated or infiltrative optic neuropathy could have contributed to severe visual loss. POEMS syndrome (polyneuropathy, organomegaly, endocrinopathy, monoclonal protein, and skin changes) is

another diagnostic consideration, because it is often associated with papilledema and demyelinating features on EMG/NCS (4). No patient in this series met diagnostic criteria for POEMS syndrome.

Antibodies to neurofascin-155 have been found in a small subset of patients with CIDP who tend to be younger in age, present with tremor, have poor response to IVIG therapy, and have better response to corticosteroids and rituximab (5,6). Neurofascin-155 is part of a transmembrane cell adhesion protein complex that stabilizes paranodal axo-glial junctions flanking the nodes of Ranvier in central and peripheral nervous system myelin (19). Neurofascin-155 IgG4 antibodies disrupt axo-glial junction integrity, causing slowed nerve conduction (6,19). One case of bilateral optic neuritis occurring 18 months after limb neuropathy and 4 cases of papilledema (without other ophthalmologic details) have been reported in patients with neurofascin-155 antibody-positive CIDP (6,20). We describe 2 patients with neurofascin-155 IgG4 seropositive CIDP that presented with bilateral vision loss because of optic neuropathy, 1 with papilledema (Case 3), and 1 without papilledema (Case 4). Both were younger at symptom onset (ages 39 and 19), responded poorly to IVIG, and responded well to corticosteroids and rituximab. In Case 3, CSF opening pressure was 23 cm $H_2O$. Given initial LP was performed 6 months after starting monthly IVIG, it is possible intracranial pressure improved following IVIG therapy. Alternatively, vision loss could have been driven by an immune-mediated mechanism more than intracranial hypertension, and may partly explain why he had better visual outcome with immunosuppressive treatment alone compared with Cases 1 and 2 who had severe papilledema. In Case 4, abnormal white matter signal associated with restricted diffusion on MRI brain (Fig. 5D, E) could possibly represent ischemia or active CNS demyelination suggesting combined central and peripheral demyelination, which has been rarely reported in neurofascin-155 antibody-positive CIDP (6). The presence of neurofascin-155 IgG4 antibodies in CIDP is rare and reported in about 3% to 4% of CIDP cases (5,6). Thus, presence of IgG4 antibodies is overrepresented in our cohort (50%) and may indicate that patients with CIDP and neurofascin-155 IgG4 antibodies could have an increased risk for developing optic neuropathy.

In conclusion, we describe vision loss as a rare presenting clinical feature in new-onset CIDP. This series is unique in providing clinical information regarding presence, timing, and degree of visual impairment that has heretofore been lacking in the CIDP literature. Moreover, it alerts the clinician that CIDP-associated visual decline can be profoundly progressive but potentially reversible with early diagnosis and expedited therapeutic intervention.

STATEMENT OF AUTHORSHIP
Conception and design: A. M. Kruszewski, Z. Tauqeer, E. A. Meer, M. A. Tamhankar; Acquisition of data: A. M. Kruszewski, Zujaja Tauqeer, E. A. Meer, S. A. Bautista, M. A. Tamhankar; Analysis and interpretation of data: A. M. Kruszewski, Z. Tauqeer, E. A. Meer, S. A. Bautista, N. R. Cherayil, M. E. Cimino, S. L. Khella, E. L., S. J. Bird, S. S. Scherer, K. E. Revere, A. G. Hamedani, Grant T. Liu, M. A. Tamhankar. Drafting the manuscript: A. M. Kruszewski, Z. Tauqeer, E. A. Meer, M. A. Tamhankar. Revising the manuscript for intellectual content: A. M. Kruszewski, Z. Tauqeer, E. A. Meer, S. A. Bautista, N. R. Cherayil, M. E. Cimino, S. L. Khella, E. Lancaster, S. J. Bird, S. S. Scherer, K. E. Revere, A. G. Hamedani, G. T. Liu, M. A. Tamhanka. Final approval of the completed manuscript: A. M. Kruszewski, Z. Tauqeer, E. A. Meer, S. A. Bautista, N. R. Cherayil, M. E. Cimino, S. L. Khella, E. Lancaster, S. J. Bird, S. S. Scherer, K. E. Revere, A. G. Hamedani, G. T. Liu, M. A. Tamhankar.

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