



Conductive hearing loss in chronic inflammatory demyelinating polyneuropathy (CIDP): A case report

Mark A. Fadel^{*,a}, Kevin Y. Zhan^b, Edward E. Dodson^b

^a University of Toledo College of Medicine & Life Sciences, 3000 Arlington Ave, Toledo, OH, 43614, USA

^b The Ohio State University Wexner Medical Center, Department of Otolaryngology- Head & Neck Surgery, 915 Olentangy River Rd, Columbus, OH, 43212, USA

ARTICLE INFO

Keywords:

Chronic inflammatory demyelinating polyneuropathy
Hearing loss
Ossicular erosion
Cranial neuropathy

ABSTRACT

Chronic inflammatory demyelinating polyneuropathy (CIDP) is a progressive autoimmune disorder that targets peripheral nerves. It commonly presents with motor-predominant dysfunction and enlargement of cranial nerves. With regards to hearing loss, a few cases of sensorineural loss have been described. We present a novel case of conductive hearing loss caused by a mass on the tympanic segment of the facial nerve in the setting of CIDP.

1. Introduction

CIDP was first described in the mid-1900s as a progressive motor-dominant disease causing severe weakness (Dyck and Tracy, 2018). It was later identified as an autoimmune disorder targeting myelin sheaths rather than axons themselves due to its lack of associated muscle atrophy (Dyck and Tracy, 2018). While CIDP triggers have not been identified, the pathogenesis results from segmental inflammatory infiltrates in the perivascular space of nerves (Peltier and Donofrio, 2012). Its prevalence ranges from 1 to 8.9/100,000, and is typically higher in the male population and can occur at any age (Baig et al., 2012; Hattori et al., 2001).

A wide range of symptoms exist, but common hallmarks of CIDP include relapsing-remitting, progressive motor and/or sensory loss of distal nerves (Dyck and Tracy, 2018; Peltier and Donofrio, 2012; European Federation of Ne, 2010). Progressive and ongoing proximal or distal motor weakness typically worsens over 8 weeks (Dyck and Tracy, 2018). Other symptoms with variable severity include sensory ataxia, areflexia, and decreased sensation (Dyck and Tracy, 2018; Peltier and Donofrio, 2012).

The variable presentation of CIDP has several shared diagnostic findings, including elevated cerebrospinal fluid (CSF) protein levels and slowed conduction velocities on electrophysiological testing (Peltier and Donofrio, 2012; European Federation of Ne, 2010). The most commonly affected nerves are typically large fibers with plentiful myelin, like the sural, superficial peroneal, or gracilis motor nerves

(Baig et al., 2012; European Federation of Ne, 2010). As with most inflammatory conditions, steroids and intravenous immunoglobulins (IVIg) are the mainstay of treatment (Dyck and Tracy, 2018; Baig et al., 2012; European Federation of Ne, 2010).

Central nervous system involvement in CIDP may mimic multiple sclerosis (MS) (Dyck and Tracy, 2018). However, MRI criteria supportive of MS classically involves periventricular white matter lesions (Prof et al., 2016; Polman et al., 2011). Whereas, MRI in CIDP demonstrates hypertrophy outside of the brain, most commonly the cauda equina and lumbosacral or cervical nerve roots (European Federation of Ne, 2010). Several reports of hypertrophic cranial nerves in CIDP as a result of blood-nerve barrier breakdown have been described (Duarte et al., 1999). These reports have involved a variety of cranial nerves, including the optic nerve (Stojkovic et al., 2000), oculomotor nerve (Alwan and Mejico, 2007; McCann et al., 1995), trigeminal nerve (Duarte et al., 1999; Alwan and Mejico, 2007; McCann et al., 1995; Lucchesi et al., 2015; Guedes and Cury, 2015; Okuzumi et al., 2014), and vestibulocochlear nerve (Stojkovic et al., 2000; Hengstman et al., 2004; Sivakumar and Fife, 2005; Orhan et al., 2016; Mowry and King, 2017; Frohman et al., 1996). CSF analyses also differentiate the two: MS is supported by 2 or more oligoclonal bands (Polman et al., 2011) and CIDP displays elevated protein and a leukocyte count less than 10/mm³ (Baig et al., 2012; European Federation of Ne, 2010). These distinguishing characteristics are essential to prescribe proper treatment for each since IVIg is not the mainstay of treatment for MS (Melzer and Meuth, 2013). This case recognizes a constellation of these findings in

* Corresponding author.

E-mail addresses: mark.fadel@rockets.utoledo.edu (M.A. Fadel), Edward.Dodson@osumc.edu (E.E. Dodson).

<https://doi.org/10.1016/j.joto.2018.10.001>

Received 3 September 2018; Received in revised form 13 September 2018; Accepted 9 October 2018

1672-2930/© 2018 PLA General Hospital Department of Otolaryngology Head and Neck Surgery. Production and hosting by Elsevier (Singapore) Pte Ltd This is an open access article under the CC BY-NC-ND license (<http://creativecommons.org/licenses/by-nc-nd/4.0/>).

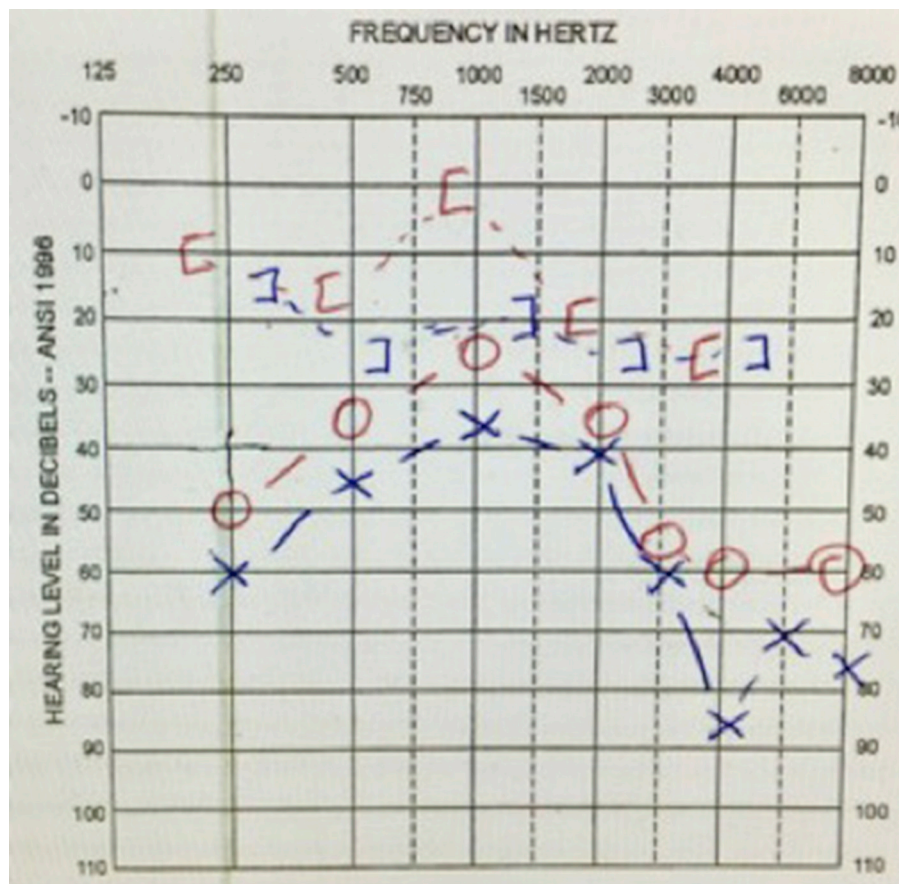


Fig. 1. Audiogram demonstrating conductive hearing loss. “O” = right unmasked air. “X” = left unmasked air. “[“ = right masked bone. “]“ = left masked bone.

the setting of CIDP: bilateral cranial polyneuropathy with hearing loss.

2. Case study

A 35-year-old male Iraqi immigrant was referred to our neurotology clinic for persistent conductive hearing loss despite multiple sets of tympanostomy tubes for serous otitis media. His hearing loss began seven years prior, with the left ear worse than right. He had been diagnosed with CIDP by a local neurologist three years prior based on fluctuating episodes of bilateral lower extremity weakness and numbness since age seven, elevated CSF protein levels, and slowed conduction velocities. He was actively receiving IVIg and steroids for the past two years.

He denied otorrhea, tinnitus, vertigo, or aural fullness. He had no history of frequent ear infections, previous ear surgery, head trauma, use of ototoxic medications or a family history of deafness. His only other surgery included bilateral lateral orbital wall decompression for Graves ophthalmopathy. On exam, he had bilateral ptosis, proptosis and myringosclerosis posteriorly with tympanostomy tubes in place. Audiogram revealed an air-bone gap at all frequencies, left worse than right (Fig. 1). The pure tone averages were 40 dB on the left and 32 dB on the right.

An MRI ordered by his neurologist demonstrated multiple enlarged cranial nerves, including the tympanic and mastoid segments of both facial nerves (Fig. 2; Fig. 2a; Fig. 2b). Other findings included bilateral enlargement of the oculomotor and trigeminal nerves, extraocular muscles, foramen ovale (Fig. 2a), foramen rotundum and stylomastoid foramen. His extraocular muscle and oculomotor nerve enlargement likely both contributed to his ptosis and proptosis.

The patient underwent left middle ear exploration with intraoperative facial nerve monitoring. After elevating a tympanomeatal

flap, a significant soft tissue mass was identified just medial to the chorda tympani nerve (Fig. 3), suspicious for a grossly enlarged facial nerve sheath. This was confirmed with a stimulus from the facial nerve monitor. There was erosion of the incus and partial erosion of the stapes superstructure. The hypertrophic nerve was gently manipulated in an attempt to identify the stapes footplate, but it could not be well visualized, and given these findings, further ossicular chain reconstruction was contraindicated and the procedure was terminated. It was recommended that he pursue bilateral amplification.

3. Discussion

CIDP is rarely associated with hearing loss and only a few cases of sensorineural loss have been described (Hengstman et al., 2004; Sivakumar and Fife, 2005; Orhan et al., 2016; Mowry and King, 2017). These reports describe the onset of hearing loss concurrent with the initial CIDP symptoms and were attributed to demyelination of segments of the vestibulocochlear nerve considering its proximity in presentation to CIDP onset (Hengstman et al., 2004; Sivakumar and Fife, 2005; Orhan et al., 2016; Mowry and King, 2017). None had other cranial neuropathies (Hengstman et al., 2004; Sivakumar and Fife, 2005; Orhan et al., 2016; Mowry and King, 2017). However, there is one report of primarily vestibular involvement, rather than cochlear (Frohman et al., 1996).

Two of the four reported cases completely recovered hearing with steroids and IVIg (Hengstman et al., 2004; Orhan et al., 2016). The other three patients described in the literature did not have improvement in hearing with steroids or IVIg (Sivakumar and Fife, 2005; Mowry and King, 2017). This paper describes a novel case of conductive hearing loss and bilateral cranial polyneuropathy. His hearing loss persisted despite two years of steroids and IVIg therapy. Initially, the

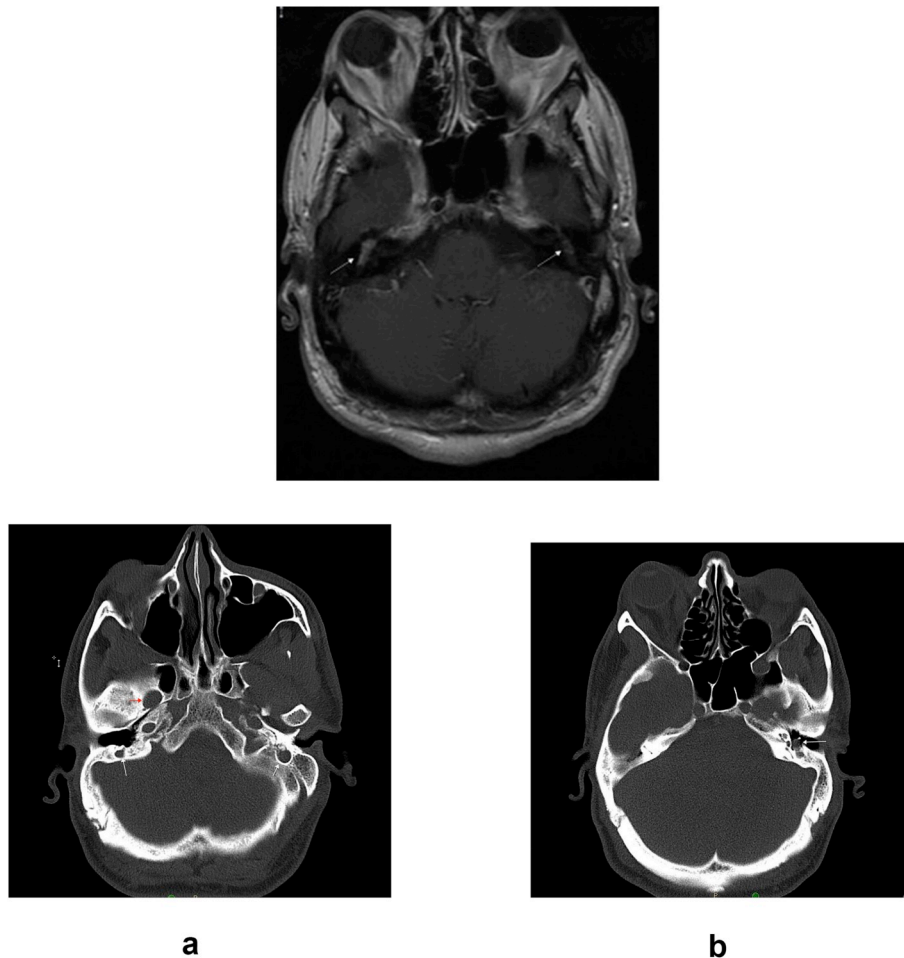


Fig. 2. Post contrast axial T1-weighted MRI demonstrating bilaterally thickened tympanic segments of the facial nerve (arrows). a: Non-contrast axial CT scan showing bilaterally enlarged fallopian canals in the mastoid segment (white arrows), as well as dilated foramen ovale (red arrow). b: Non-contrast axial CT scan showing ossicular erosion secondary to enlarged tympanic segment of the facial nerve.

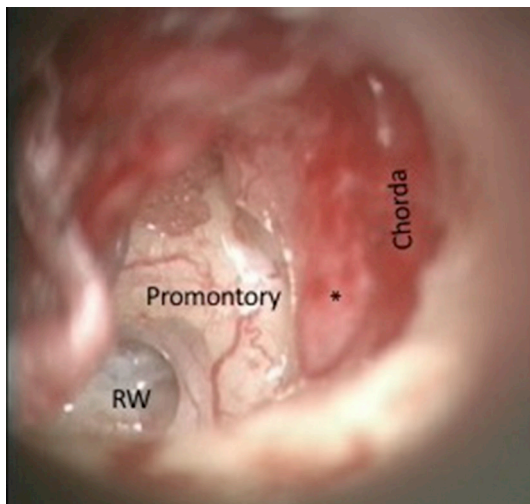


Fig. 3. Intraoperative finding of the soft tissue mass (*) just medial to the chorda tympani nerve (Chorda). The photo is in surgical position, with the left side of the photo as inferior and top half of the photo as anterior. There is a small piece of surgical packing anteroinferiorly. “RW” = round window. “Promontory” = cochlear promontory.

hearing loss was likely secondary to a mass effect on the ossicles with subsequent ossicular erosion from pressure necrosis. After further discussion with his neurology team, further treatment was not pursued and deemed unlikely to reduce facial nerve hypertrophy.

Facial nerve hypertrophy has rarely occurred in the setting of CIDP (Waddy et al., 1989). These findings of hypertrophy have been attributed to the frequent demyelination-remyelination cycles causing Schwann cell proliferation (Lucchesi et al., 2015). Our patient's symptoms had some similarities and differences compared to previous reports. He lacked facial paresthesias despite his thickened trigeminal nerves while another case reported mandibular hypesthesia (Lucchesi et al., 2015). However, he presented with ptosis and proptosis as described in previous case reports with oculomotor nerve or extraocular muscle thickening (Duarte et al., 1999; Alwan and Mejico, 2007; Lucchesi et al., 2015). Some of these cases also involved Graves ophthalmopathy which may suggest an association between CIDP and increased incidence of autoimmune disease (Alwan and Mejico, 2007; Lucchesi et al., 2015).

4. Conclusion

CIDP has a wide range of symptoms with varying courses and severity of disease that should be distinguished from other autoimmune demyelinating disorders like MS. The patient presented in this case report exhibits a unique cranial polyneuropathy with resultant hearing loss from mass effect secondary to facial nerve hypertrophy and subsequent ossicular erosion.

Declarations of interest

None.

Funding

None.

References

- Alwan, A.A., Mejico, L.J., 2007. Ophthalmoplegia, proptosis, and lid retraction caused by cranial nerve hypertrophy in chronic inflammatory demyelinating polyradiculoneuropathy. *J. Neuro Ophthalmol.* 27 (2), 99–103.
- Baig, F., Knopp, M., Rajabally, Y.A., 2012. Diagnosis, epidemiology and treatment of inflammatory neuropathies. *Br. J. Hosp. Med.* 73 (7), 380–385.
- Duarte, J., et al., 1999. Hypertrophy of multiple cranial nerves and spinal roots in chronic inflammatory demyelinating neuropathy. *J. Neurol. Neurosurg. Psychiatry* 67, 685–687.
- Dyck, J.B., Tracy, J.A., 2018. History, diagnosis, and management of chronic inflammatory demyelinating polyradiculoneuropathy. *Mayo Clin. Proc.* 93 (6), 777–793.
- European federation of neurological societies/peripheral nerve society guideline on management of chronic inflammatory demyelinating polyradiculoneuropathy: report of a joint task force of the european federation of neurological societies and the peripheral nerve society—first revision. *J. Peripher. Nerv. Syst.* 15 (1), 1–9.
- Frohman, E.M., et al., 1996. Vestibular dysfunction in chronic inflammatory demyelinating polyneuropathy. *Ann. Neurol.* 39, 529–535.
- Guedes, B.F., Cury, R.G., 2015. Trigeminal Nerve Thickening in Chronic Inflammatory Demyelinating Polyneuropathy. pp. 634–635 vol. 73(7).
- Hattori, N., et al., 2001. Age of onset influences clinical features of chronic inflammatory demyelinating polyneuropathy. *J. Neurol. Sci.* 184, 57–63.
- Hengstman, G., Schelhaas, H.J., Zwarts, M.J., 2004. Auditory dysfunction in chronic inflammatory demyelinating polyradiculopathy. *Neuro* 62 (8), 1446–1448.
- Lucchesi, C., et al., 2015. Chronic inflammatory demyelinating polyradiculoneuropathy with cranial nerves hypertrophy, thyroid-related orbitopathy and IgG monoclonal gammopathy: a case report. *Neurol. Sci.* 36, 1027–1029.
- McCann, E.L., et al., 1995. Severe cranial nerve involvement in longstanding demyelinating polyneuropathy: a clinicopathologic correlation. *Acta Neuropathol.* 91, 309–312.
- Melzer, N., Meuth, S.G., 2013. Disease-modifying therapy in multiple sclerosis and chronic inflammatory demyelinating polyradiculopathy: common and divergent current and future strategies. *Clin. Exp. Immunol.* 175, 359–372.
- Mowry, S.E., King, S., 2017. Cochlear implantation in chronic demyelinating inflammatory polyneuropathy. *Cochlear Implants Int.* 18 (2), 116–120.
- Okuzumi, A., et al., 2014. Ophthalmic nerve hypertrophy in chronic inflammatory demyelinating polyradiculoneuropathy. *Neuro* 82 (17), 1566–1567.
- Orhan, G., Bohm, F., Paulus, W., 2016. Treatment of Chronic inflammatory polyneuropathy may improve associated hearing loss: a case report and review of the literature. *Muscle Nerve* 53 (5), 828–829.
- Peltier, A.C., Donofrio, P.D., 2012. Chronic inflammatory demyelinating polyradiculoneuropathy: from bench to bedside. *Semin. Neurol.* 32 (3), 187–195.
- Polman, C.H., et al., 2011. Diagnostic criteria for multiple sclerosis: 2010 Revisions to the McDonald criteria. *Ann. Neurol.* 69 (2), 292.
- Prof, M.F., et al., 2016. MRI criteria for the diagnosis of multiple sclerosis: MAGNIMS consensus guidelines. *Lancet Neurol.* 15 (3).
- Sivakumar, K., Fife, T.D., 2005. Autoimmune hearing loss in chronic inflammatory demyelinating polyneuropathy. *Neuro* 65 (10).
- Stojkovic, T., et al., 2000. Visual evoked potentials study in chronic idiopathic inflammatory demyelinating polyneuropathy. *Clin. Neurophysiol.* 111 (12), 2285–2291.
- Waddy, H.M., et al., 1989. Focal cranial nerve involvement in chronic inflammatory demyelinating polyneuropathy: clinical and MRI evidence of peripheral and central lesions. *J. Neurol.* 236, 400–405.