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Review Article

Geniculate neuralgia successfully treated with microvascular decompression

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ABSTRACT

Background: First described by John Nottingham in 1857, geniculate neuralgia remains a rare condition associated with vascular compression of the nervus intermedius by the anterior inferior cerebellar artery (AICA), which results in paroxysmal unilateral periauricular pain. Furthermore, limited and controversial treatment options for symptom relief exist given the rarity of the condition and limited cases reported in the literature.
Case description: This is a case of a 37-year-old one-pack-per-day smoker with diabetes mellitus who presented to our clinic for evaluation of episodic lancinating pain localizing to the right periauricular region. The patients symptoms were attempted to be managed medically, however, remained refractory to medical management for a period greater than one year. The patient's exam demonstrated a trigger point slightly anterior and inferior to the right tragus, and the pain was reproducible when touched or tapped. The patient was otherwise neurologically intact. Magnetic resonance imaging (MRI) was performed and demonstrated a loop of the AICA in contact with the root entry zone of the facial nerve. This patient was offered an elective microvascular decompression (MVD) for treatment of geniculate neuralgia.
Conclusions: Surgical microvascular decompression is a safe and effective treatment option for patients suffering from neuralgia refractory to medical therapy. Furthermore, our case report demonstrates that MVD is an effective treatment option for patients suffering from geniculate neuralgia with imaging evidence of AICA compression of the nervus intermedius that is refractory to medical management.

1. Introduction

Geniculate neuralgia, also referred to as nervus intermedius neuralgia or Hunt's neuralgia, is a rare condition first described in 1857 by John Nottingham, with fewer than 150 cases reported in the literature from 1932 through 2012 [1–3]. Geniculate neuralgia is a condition most commonly described by its association with herpes zoster or vascular compression of the nervus intermedius, a small branch of the facial nerve, frequently by the anterior inferior cerebellar artery (AICA). The nervus intermedius, also referred to as the intermediate nerve of Wrisberg, was first identified in 1563 [1,2]. However, it was not documented in the literature until 1777 by Heinrich August Wrisberg at the German University of Göttingen, describing it as the "*portio media inter comunicantem faciei et nervum auditorium.*" [1,2] (Table 1).

John Nottingham first described the clinical features of geniculate neuralgia as paroxysmal flushing and pain of the ear in 1857, characterizing it as "tic douloureux of the ear" [1,2,4]. However, it was not until 1907 when James Ramsay Hunt identified the correlation between tic douloureux of the ear and the nervus intermedius from his work with herpes zoster oticus, coining the term geniculate neuralgia [1,2,4]. Currently, the International Headache Society describes the diagnostic criteria for geniculate neuralgia as paroxysmal unilateral periauricular shooting or stabbing pain that lasts a few seconds to minutes, is severe in intensity, and is exacerbated by a trigger point within the distribution of the nervus intermedius [5].

Anatomically and functionally, the nervus intermedius contains sensory fibers, parasympathetic secretory fibers, and special cutaneous afferent sensory fibers. The parasympathetic fibers are derived from the superior salivary nucleus of the facial nerve located in the dorsal pons, which travel to innervate the submandibular, sublingual, and lacrimal glands [2,6]. Somatosensory components of the nervus intermedius are branches of the facial nerve located within the geniculate ganglion, carrying cutaneous sensorium input from the external auditory meatus, the cymba, and cavum of the external ear [2,7]. The nervus intermedius is also composed of special afferent fibers from the gustatory nucleus, relaying taste from the anterior two-thirds of the tongue, the palate, and

Abbreviations: AICA, anterior inferior cerebellar artery; BAER, brainstem auditory evoked responses; FIESTA, fast imaging employing steady-state acquisition; MRI, Magnetic resonance imaging; MVD, microvascular decompression

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Table 1
Previously published studies reporting patients with geniculate neuralgia, treatment modality and outcomes.

Paper	Treatment Dates	Demographics	Symptoms	Number of Patients	Intervention	Outcome	Complications	Other
Pulec 2002	Between 1966 and 1996	Not stated	Not stated	64	Surgical excision of the nervus intermedius and geniculate ganglion via the middle cranial fossa approach	All had complete resolution of pain No CSF leak No hearing loss	11 with temporary facial paralysis 1 with prolonged facial paralysis that recovered	3 patients had selective retrolabyrinthine Vth nerve section after excision of the nervus intermedius and geniculate ganglion. 13 had previously undergone a selective retrolabyrinthine Vth nerve section
	1994	47-year-old Caucasian female	2 years of "ice pick to the ear" left ear pain. 2–3 episodes per day for 3 min duration	1	3-mm excision of nervus intermedius, excision of geniculate ganglion and 25% of the anterior portion of the motor part of the facial nerve at the internal genu. greater petrosal nerve was severed so that a 1-cm segment of this nerve was excised along with the geniculate ganglion	Complete resolution of pain immediately post-op and at 2-year follow-up	None reported	N/A
Peris-celda et al., 2018	2000–2017	Median age was 48 7 Female 4 Male	Not stated	11 (9 primary, 2 reoperations)	Retrosigmoid approach in all cases. 9 patient treated with nervus intermedius rhizotomy 2 patients treated with microvascular decompression (MVD) Trigeminal neuralgia treated by MVD in 4, rhizotomy in 1 Glossopharyngeal Neuralgia treated by MVD in 2, rhizotomy in 2	All patients initially had clinically significant pain relief. Complete resolution in 8 patients 5 patients with recurrence	2 recurrences with debilitating pain recurrence	N/A
Saers et al., 2011	2011	24 y/o Female	9-year history of otalgia	1	Microvascular decompression	Complete resolution 1-year post-operatively	None	N/A
Rupa et al., 1991	1976–1991	Ages 12–71 10 Male 8 Female	Otalgia	NI: 14 Geniculate Ganglion: 10	NI Rhizotomy (14 patients) Geniculate Ganglion Rhizotomy (10 patients) Ninth Nerve Rhizotomy (14 patients) Tenth Nerve Rhizotomy (11 patients) Tympanic Nerve Rhizotomy (4 patients) Chorda Tympani Rhizotomy (1 patient) MVD (9 patients)	72.2% successful pain relief with mean follow-up of 3.3 years	Decreased lacrimation, salvation, and taste related to NI sectioning. Transient Hoarseness and diminished gag related to 9th and 10th nerve sectioning Sensorineural hearing loss, vertigo, and transient facial nerve paresis in 4 patients 1 CSF leak 1 aseptic meningitis	N/A
Tubbs et al., 2013	2013	39 y/o Male	Otalgia	1	Retromastoid craniotomy and transection of NI, ("Arterial loop that was in close contact with the lower cranial nerves at the level of the brainstem was mobilized using a polytetrafluoroethylene (Teflon) implant" = microvascular decompression?)	Complete pain resolution after 16 months	None	N/A

floor of the oral cavity [2,6,7,8].

Definitive diagnosis of geniculate neuralgia remains complex due to contributions of the vagus, glossopharyngeal, trigeminal nerve, as well as the nervus intermedius [2,6,7]. Yet, multiple treatment approaches continue to be developed and refined in an effort to treat this rare condition. The following describes a case of geniculate neuralgia that was successfully treated with microvascular decompression (MVD) of the nervus intermedius.

2. Case report

2.1. History of presenting illness

A 37-year-old one-pack-per-day smoker with diabetes mellitus presented to our neurosurgical clinic for evaluation of episodic lancinating pain localizing to the right periauricular region. The patient reported experiencing approximately three attacks of ear pain per day that were refractory to medical management over a period greater than one year with failed trials of amitriptyline, carbamazepine, gabapentin, baclofen, meclizine, and meloxicam. The pain originated from the right tragus and radiated to the deep facial structures and the ipsilateral eye. The pain distribution did not fit that of classical trigeminal neuralgia.

2.2. Physical examination and diagnostic imaging

On the preoperative neurological exam, the patient was alert and oriented to person, place, and time. Pupils were three millimeters and reactive with intact extraocular movements. The patient's hearing was intact bilaterally, with no other cranial nerve deficits detected on exam. A trigger point slightly anterior and inferior to the right tragus would reproduce the pain when touched or tapped. No pronator drift was detected and strength and sensation were full and intact in all extremities. Magnetic resonance imaging (MRI) was performed and demonstrated a loop of the AICA in contact with the root entry zone of the facial nerve (Fig. 1). As the patient met the diagnostic criteria for geniculate neuralgia, he was offered elective intervention for symptomatic control, and was ultimately consented for MVD of the nervus intermedius via a right retrosigmoid cranial approach.

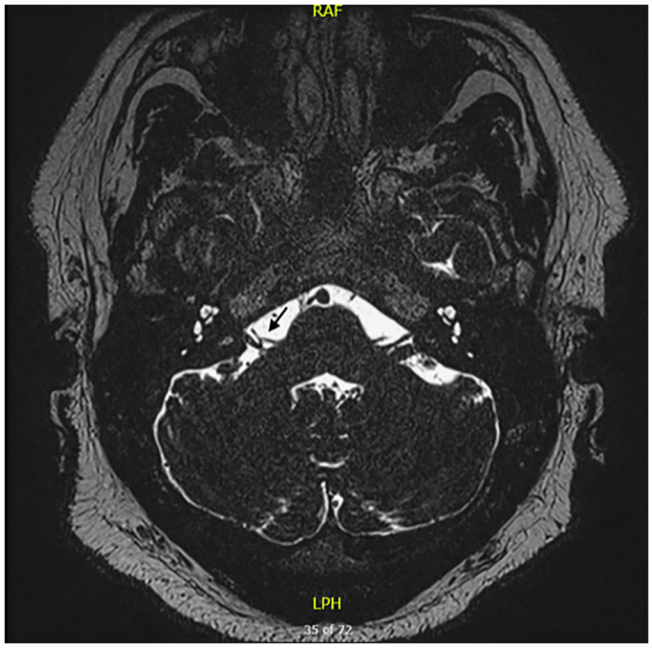

Fig. 1. Axial T2 weighted magnetic resonance image demonstrating AICA compression at the root entry zone of the facial nerve (arrow).

2.3. Operative details

Preoperatively, the patient was placed in the lateral decubitus position for lumbar drain insertion. The patient was then repositioned supine on a standard operative table and placed in the Mayfield clamp. An ipsilateral shoulder and hip bump were placed to facilitate the positioning of the right mastoid at the apex of the operating field. The patient's retrosigmoid scalp was shaved, and anatomical landmarks were used to delineate the junction of the transverse and sigmoid sinus. A standard linear incision was planned one-third above the transverse sinus and two-thirds below. The patient was then prepped and draped in the usual sterile fashion. Neuromonitoring was utilized throughout the procedure to monitor the function of the facial nerve, as well as brainstem auditory evoked responses (BAER).

A right retrosigmoid craniotomy was performed. The transverse and sigmoid sinuses were identified and a curvi-linear dural incision was made. Tack-up sutures were placed along the dural edge to increase the visual field. The lumbar drain was opened and 15 ml of cerebral spinal fluid (CSF) was evacuated for relaxation of the retrosigmoid corridor. Additionally, the patient was hyperventilated to a pCO2 of 25 mmHg and 100 g of intravenous mannitol was delivered.

Cottonoid® patties were placed over the cerebellar hemisphere as a retractor-free approach along the right retrosigmoid corridor and was conducted with the aid of microscissors to facilitate sharp dissection. The lower cranial nerves were identified and dissection superiorly revealed the vestibulocochlear and facial nerve complex. After meticulous dissection of the arachnoid, it became apparent that AICA did not form a classical loop. Instead, the offending artery passed between the facial and cochlear nerve and imbedded into the root entry zone. Electrical stimulation confirmed the identity of the facial nerve. A fixed retractor was placed with the Budde halo and gentle dissection provided a plane between the brainstem and AICA. A small Teflon pledget was placed in this space to protect the root entry zone. Further mobilization AICA from the nerves was attempted but limited because manipulation of the nerves began to corelate with increased latencies of the BAERs. The facial nerve was stimulated again and its function confirmed with a stimulus of < 0.05 mA. No effort was made to isolate and ligate the nervus intermedius. Finally, more Teflon pledgets were placed circumferentially around AICA and secured using fibrin sealant (Tisseel, Baxter International Inc., Deerfield, IL, USA).

Following hemostasis and irrigation, the dura was closed in a watertight fashion. A 2-inch $\times$ 2-inch synthetic dural repair material (DuraGen, Integra Lifesciences, Plainsboro, NJ, USA) was placed over the site of durotomy. The cranial flap was plated with titanium brackets and secured with screws. The nuchal muscles, dermis, and skin were then reapproximated using interrupted sutures in a multilayer fashion. The patient was awakened without event and observed overnight in the intensive care unit. The lumbar drain was left in place post-operatively, with intermittent drainage of 10 ml/h. It was clamped after 24 h and removed on post-operative day two.

3. Results

On post-operative day 1, the patient's facial pain had completely resolved. All cranial nerves were functioning at the pre-operative baseline activity. Touching or tapping the trigger point did not elicit any pain. Patient was seen in clinic 4 weeks post-operative and continued to be pain free. At 5 months post-operatively, patient continues to report 60% pain control compared to pre-operative pain levels. Follow-up MRI demonstrated separation of the vessel loop from the nervus intermedius (Fig. 2).

4. Discussion

Geniculate neuralgia remains a clinically challenging entity secondary to the rarity of the disorder, the complexity of diagnosis, the

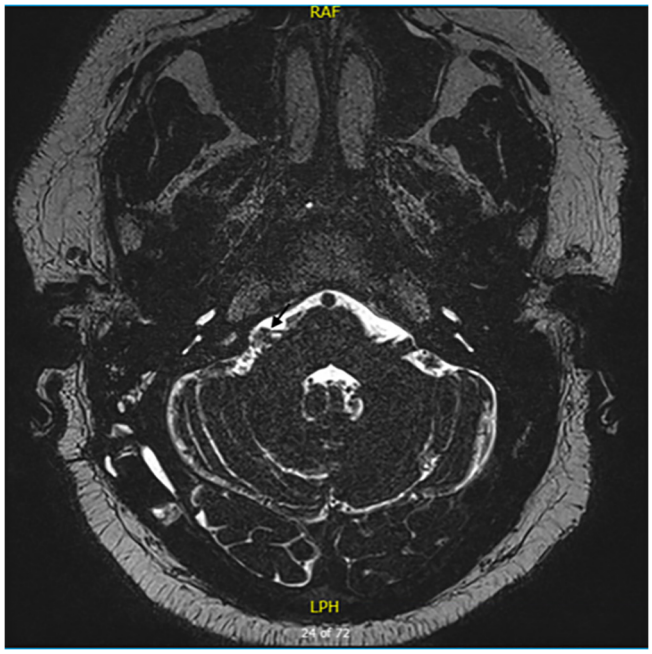

Fig. 2. Axial T2 weighted magnetic resonance image demonstrating separation of the vessel loop from the root entry zone of the facial nerve (arrow).

frequency of failed medical management, and the technical skill required to negotiate the operative nuance. However, continued reporting of these rare cases, in addition to further technological advances in MR imaging have made the diagnosis more reliable and familiar. Burmeister et al. reported a novel way of identifying the nervus intermedius in 2011 through high-field 3 T MRI that was not previously possible using CT or 1.5 T MRI [9]. The authors reportedly evaluated 54 nervus intermedi in 27 healthy volunteers and were able to identify bilateral facial nerve branches in 57.4% of cases, and at least one nervus intermedius in 70.4% of individuals [9].

There is a lack of evidence for a definitive management algorithm for geniculate neuralgia. Although it is generally accepted that medical management should be trialed prior to operative intervention, a superior first-line medication has not been identified. The most common techniques for surgical management of geniculate neuralgia remain nervus intermedius sectioning, geniculate ganglion excision, and microvascular decompression [2,3,6,8]. Accessing the nervus intermedius for operative intervention can be limited in up to 20% of cases as the intracisternal portion of the nerve remains adherent to the superior vestibular nerve and does not separate until within the internal acoustic meatus, making identification virtually impossible intraoperatively [8,10]. When identifiable, the nervus intermedius is often in close proximity to and between the facial nerve and superior portion of the vestibular nerve, making it challenging to surgically manipulate [8].

Pulec et al. described the outcomes of 64 patients that underwent sectioning of the nervus intermedius and excision of the geniculate ganglion through a middle cranial fossa approach [11]. The investigators determined that this technique was safe and effective for treatment of geniculate neuralgia refractory to conservative management. The report documented only a single case of facial paralysis that ultimately recovered [11].

Rupa et al. described 18 cases of primary otalgia over a 15-year period that were treated with four different operative procedures based on presumed etiology: tympanotomy for chorda tympani and tympanic nerve section, the technique described by Pulec utilizing a middle cranial fossa approach for geniculate ganglionectomy, posterior cranial fossa approach for nervus intermedius, ninth, and tenth cranial nerve sectioning, and a combination of the posterior and middle fossa approach for nervus intermedius and fifth, ninth, and tenth nerve exposure with geniculate ganglionectomy [12]. They demonstrated a total success rate of 72.2%, with increased pain relief in patients that had sectioning of the seventh nerve (geniculate ganglion, nervus intermedius, and chorda tympani), ninth, tympanic nerve, and the upper rootlets of the tenth nerve [12].

More recently, Peris-Celda et al. reported a retrospective review of 11 patients treated for geniculate neuralgia (nine primary, two re-operative) from 2000 to 2017 [13]. The authors describe nine patients who were treated with sectioning of the nervus intermedius, and two who underwent microvascular decompression [13]. Of the two patients treated with MVD, one had complete resolution of their pain post-operatively and no longer required neuropathic pain medications, whereas the second had improved, but experienced incomplete resolution of pain and remained on neuropathic pain medication [13]. Saers et al. reported a case of geniculate neuralgia in a 24-year-old female in 2011 who was successfully treated with MVD after determining neurovascular compression of the nervus intermedius by the AICA on fast imaging employing steady-state acquisition (FIESTA) MRI [14]. The authors reported complete resolution of pain immediately post-operatively and at one year follow-up without complication [14]. The goal of this report is to increase the awareness geniculate neuralgia and to demonstrate MVD is an effective technique to achieve symptomatic relief in patients refractory to medical management. To date, our patient continues to report satisfactory pain control.

5. Conclusion

Geniculate neuralgia is a rare disorder that is clinically challenging to diagnose. Definitive medical and surgical treatments have yet to be clearly elucidated. Furthermore, prospective studies to determine the outcomes and complications following nervus intermedius sectioning with geniculate ganglion excision versus microvascular decompression appear unlikely for this infrequent disease. This report demonstrates that nervus intermedius MVD is an effective technique for those patients with imaging evidence of AICA compression that is refractory to medical management.

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Declaration of Competing Interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

Appendix A. Supplementary data

Supplementary data to this article can be found online at https://doi.org/10.1016/j.inat.2019.100583.

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