Cranial Nerve IX and X Neurectomy for Glossopharyngeal Neuralgia: Case Report and Operative Video

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BACKGROUND AND IMPORTANCE: Glossopharyngeal neuralgia (GPN) is a rare condition that is often misdiagnosed as trigeminal neuralgia. The condition is characterized by intermittent, severe pain in the distribution of the glossopharyngeal nerve. We present an illustrative case of GPN with an operative video detailing neurectomy of the glossopharyngeal nerve and the upper rootlets of the vagus nerve for treatment of idiopathic GPN in a patient with a history of squamous cell carcinoma.

CLINICAL PRESENTATION: A 62-year-old man with a history of left mandibular alveolar squamous cell carcinoma status postresection presented with left-sided severe, paroxysmal pain in the posterior one-third of his tongue refractory to medical treatment and without evidence of recurrent malignancy or vascular compression on imaging studies. After he failed medical management, glossopharyngeal neurectomy was performed through a left suboccipital craniotomy during which cranial nerves IV, V, VI, VII/VII, IX, X, and XI were visually inspected for malignant recurrence, and the glossopharyngeal nerve and the upper 2 to 3 nerve rootlets of the vagus nerve were severed. The patient had immediate, complete, and durable resolution of his symptoms without any new neurological deficits.

CONCLUSION: Glossopharyngeal neurectomy has been shown to be an efficacious surgical treatment for GPN, as first described by Walter Dandy in 1920. In this report, we describe the workup and treatment of GPN with important diagnostic considerations and present a detailed video demonstrating technical and anatomic considerations when performing glossopharyngeal neurectomy.

KEY WORDS: Glossopharyngeal neuralgia, Glossopharyngeal neurectomy

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Glossopharyngeal neuralgia (GPN) is a rare syndrome characterized by severe pain in the distribution of the glossopharyngeal nerve (CN IX) and the pharyngeal and auricular branches of the vagus nerve (CN X). The prevalence of GPN is 153 per 1 000 000 people, and the incidence is 4.3 new cases per 100 000 people annually.[1,2] Patients typically experience recurrent, paroxysmal attacks of unilateral pain in the distribution of the glossopharyngeal nerve. Idiopathic GPN, the most common form, is thought to arise from vascular compression of CN IX or X at or near the root entry zone by the vertebral artery, posterior inferior cerebellar artery, or other nearby blood vessels. Secondary GPN may be caused by intracranial or perineural tumors, peritonsillar abscess, demyelinating lesions, and Eagle syndrome.[3-6]

ABBREVIATION: GPN, glossopharyngeal neuralgia.

Although its name implies involvement of only CN IX, involvement of the upper rootlets of CN X in GPN also contributes to the pain associated with swallowing and phonation.[7] Indeed, in a closely related condition termed vago-glossopharyngeal neuralgia, patients experience GPN symptoms and symptoms related to excessive stimulation of CN X during episodes of pain such as bradycardia, hypotension, syncope, seizures, or cardiac arrest.[8]

Medical management options for GPN are similar to trigeminal neuralgia and may include a combination of gabapentin, carbamazepine, lamotrigine, other anticonvulsants, or topical anesthetics.[9-12] Surgical options for medically refractory GPN include microvascular decompression of CN IX or CN X, neurectomy of CN IX and the upper rootlets of CN X, or gamma knife radiosurgery.[13-15] Surgical considerations include severity of symptoms, distribution of pain, and evidence of vascular compression on MRI.[13-16] Herein, we describe an illustrative case of GPN treated

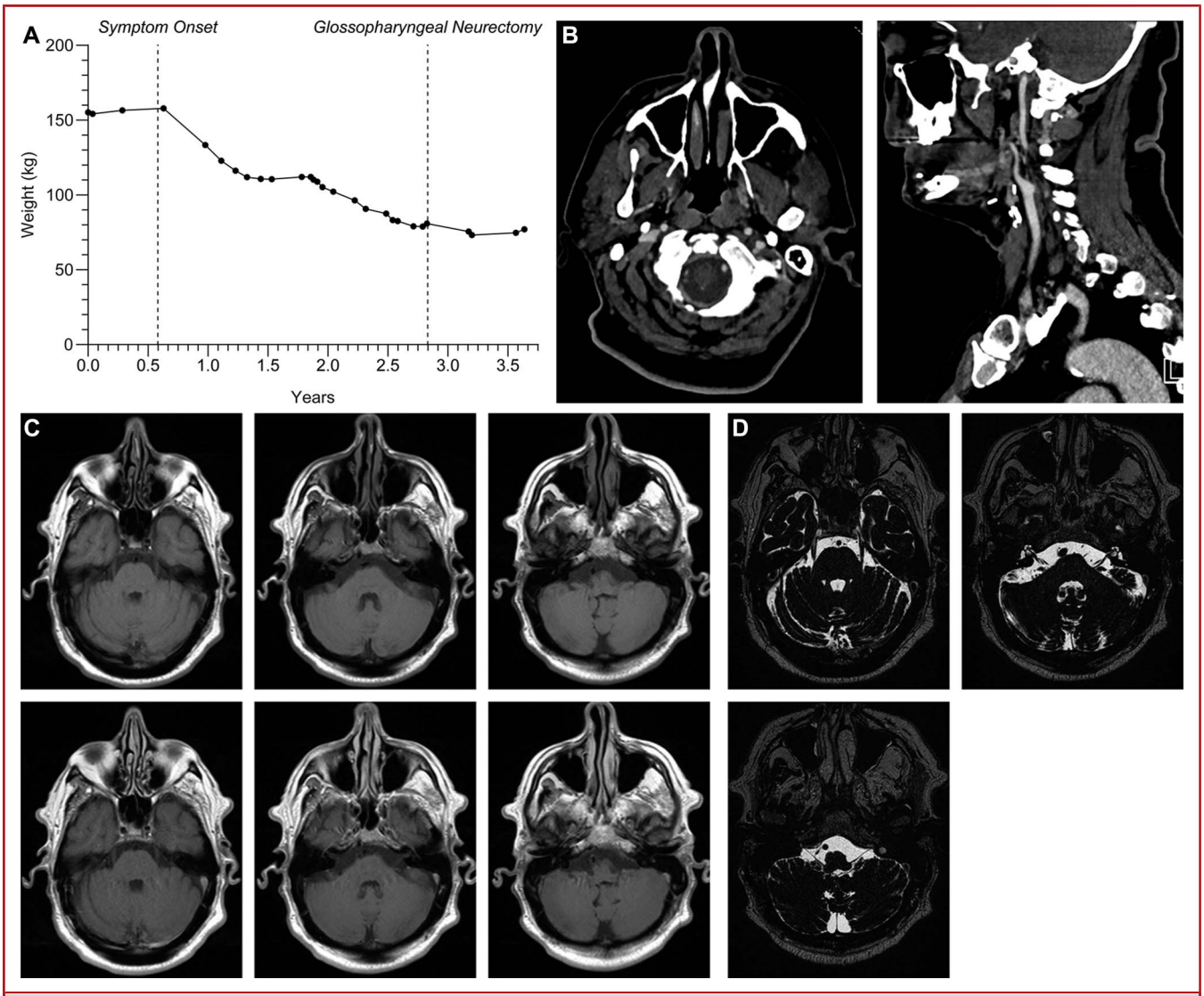

FIGURE 1. **A**, *Precipitous weight loss depicted in relation to symptom onset.* **B**, *Computed tomography with contrast showing absence of recurrent malignancy.* **C**, *T1-weighted MRI with/without contrast showing no neoplastic lesion or contrast enhancement of the trigeminal, facial/vestibulocochlear, and glossopharyngeal nerves.* **D**, *Constructive interference in steady state MRI showing no distortion or compression of the trigeminal, facial/vestibulocochlear, and glossopharyngeal nerves.*

with glossopharyngeal neurectomy with an accompanying operative video.

CLINICAL PRESENTATION

A 62-year-old man with left mandibular alveolar squamous cell carcinoma was treated with mandibulectomy, buccal resection, and selective neck dissection (American Academy of Otolaryngology–Head and Neck Surgery levels 1A, 1B, 2A, 2B, and 3) 2.5 years before neurosurgical evaluation.[17] He began experiencing numbness in the anterior tongue and left lower lip immediately after his neck surgery and left-sided posterior lancinating tongue pain which began several months after his surgery but was also initially thought to be related to his surgery. His tongue pain radiated toward the left ear, first occurring approximately once per month but ultimately progressing to daily episodes triggered by swallowing and leading to significant weight loss (Figure 1A).

Diagnostic Workup

Computed tomography of the neck with contrast, laryngoscopy, and biopsy of the prior surgical site confirmed no recurrence of his previously treated carcinoma (Figure 1B). Cranial nerve examination was notable only for loss of sensation in the left lower lip (mental nerve distribution, a branch of CN V3) as well as the

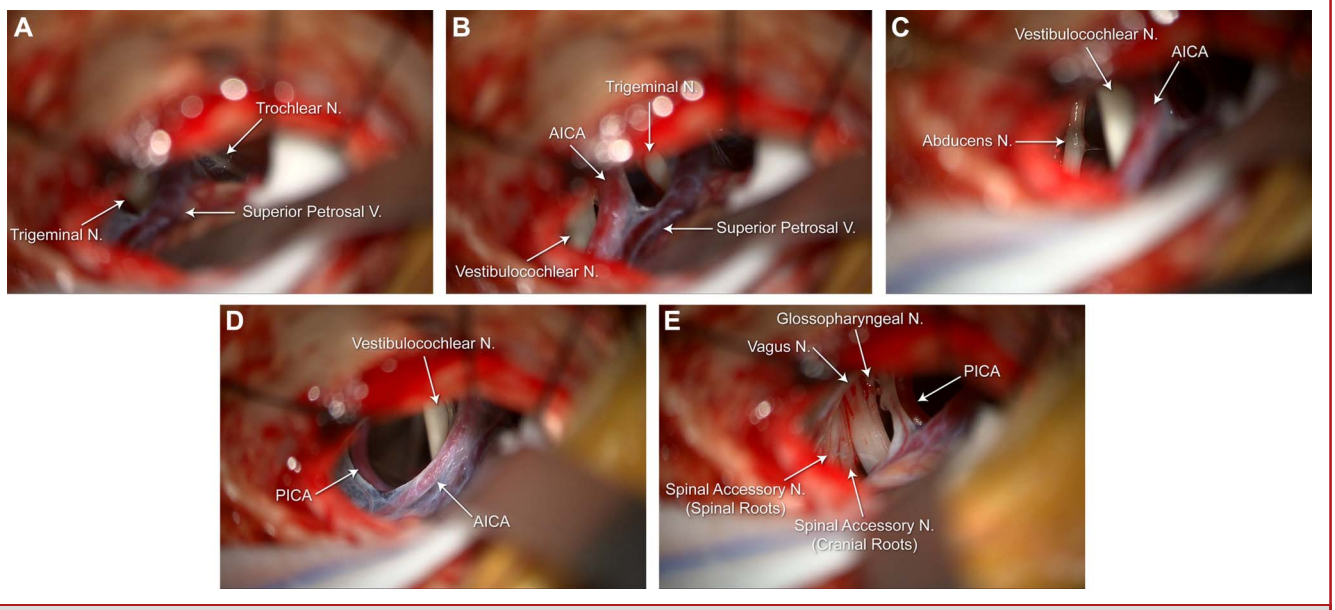

FIGURE 2. A-E, *Intraoperative views of the left cerebellopontine angle progressing sequentially from rostral to caudal, grossly normal appearing cranial nerves labeled. AICA, anterior inferior cerebellar artery; PICA, posterior inferior cerebellar artery.*

anterior two-thirds of the tongue (lingual nerve distribution, a branch of CN V3), both expected findings given the patient's otolaryngological procedure. Sensation was intact in the posterior one-third of the tongue, where the patient was experiencing pain. Importantly, no dissection in the region of the jugular foramen or along the course of the extracranial glossopharyngeal nerve was performed during the patient's otolaryngological surgery. Based on the history, neurological examination, and exclusion of neoplastic process with MRI scan (Figure 1C), a diagnosis of left-sided GPN was made. High-resolution MRI of the cranial nerves showed no distortion of CN IX and no vascular compression at CN IX root entry zone (Figure 1D). After failing to respond to treatment with carbamazepine, gabapentin, oxycodone, acetaminophen, and cetacaine, the patient underwent surgical treatment with glossopharyngeal neurectomy, as described below.

Operative Details

Informed consent was not required by our Institutional Review Board for this case report. After providing informed consent for the procedure, the patient was taken to the operating room and positioned supine with his head secured in a Mayfield head clamp and turned to the right to expose the left suboccipital region. Monitoring devices were attached for intraoperative auditory brainstem response and facial electromyography monitoring. A curvilinear incision was created posterior to the ear, and a retractor was used to reflect the scalp and muscle. A left suboccipital craniectomy was created to expose the transverse-sigmoid junction and then expanded inferiorly along the sigmoid sinus to the tip of the mastoid. The dura was opened 5 mm from the edge of the sinuses. The arachnoid was dissected under microscopic visualization and cerebrospinal fluid was aspirated for brain relaxation to maximize visualization. Further arachnoid dissection was performed to identify cranial nerves IV, V, VI, VII/VII, IX, X, and XI, with each nerve being visually inspected for any evidence of tumor invasion given the patient's history of carcinoma (Figure 2 and Video). Consistent with the preoperative MRI, no blood vessels were compressing CN IX or X, including at their root entry zones. Next, CN IX was separated from CN X with a dissector and severed using microscissors. The upper rootlets of CN X were then separated from the rest of the nerve and severed using microscissors. The dura was closed with suture, and wax was applied to bone edges for hemostasis and to occlude mastoid air cells. The craniectomy defect was filled with methyl-methacrylate and the muscle and skin reapproximated with sutures.

Immediately after surgery, the patient experienced complete resolution of tongue pain without any new neurological deficits. One year postoperatively, the patient continued to have a durable response, with no recurrence of posterior tongue or throat pain and no further weight loss with normal oral intake (Figure 1A).

DISCUSSION

GPN is a rare, debilitating condition. In a recently published analysis of 97 patients with GPN treated with microvascular decompression, 92% of patients experienced immediate and complete pain relief postoperatively, with long-term probability of being pain free at 1, 3, 5, and 10 years being 89.7%, 74.1%, 68.3%, and 66.3%, respectively. Notably, in this series, 9% of patients experienced permanent neurological deficit such as dysphagia, hoarseness, and

facial paresis, and 7 patients underwent reoperation, which included partial rhizotomy, after the initial surgery failed to provide relief.[18]

Here, we discuss a case of GPN with successful surgical treatment including an operative video depicting sectioning of the glossopharyngeal nerve and the upper rootlets of the vagus nerve. As illustrated, diagnosing GPN can be challenging, especially in the setting of red herrings such as prior head and neck surgery. Care must be taken to obtain a detailed history with detailed description of symptomatology from the patient. This information must then be evaluated in the context of a thorough neurological examination, close inspection of imaging studies, and thoughtful consideration of the anatomy of the glossopharyngeal and other cranial nerves, particularly the trigeminal nerve and its branches, to arrive at the correct diagnosis.

CONCLUSION

This report detailing treatment of a patient with GPN includes an intraoperative video of intracranial glossopharyngeal neurectomy for GPN, highlighting sectioning of the glossopharyngeal nerve as well as the upper nerve rootlets of the vagus nerve. As illustrated in this case, the diagnosis of GPN can be challenging, with patients often suffering from symptoms for long periods before a correct diagnosis is made. In this case, after failing to respond to medical management, the patient had complete and durable symptom relief after glossopharyngeal neurectomy.

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VIDEO. This video presentation begins with a brief narrative of the patient's presentation and workup as detailed in the manuscript and accompanying figures. We then provide the surgeon's perspective from the operating microscope pausing briefly at critical moments to label important blood vessels and nerves that must be carefully preserved. Toward the end of the video, we demonstrate sectioning of the glossopharyngeal nerve, followed by dissection and sectioning of the upper rootlets of the vagus nerve. Sectioning of both nerves is critical to ensure success of the surgery.