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Glossopharyngeal Neuralgia: An Approach to Diagnosis and Management

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Abstract

Glossopharyngeal neuralgia is an uncommon facial syndrome with significant deleterious effect on the quality of life. The glossopharyngeal nerve is predominantly a sensory nerve with a limited number of motor and autonomic fibers. The central causes of glossopharyngeal neuralgia are commonly treated with neurosurgical interventions. Medical therapy and nerve blocks are predominantly used to treat peripheral causes of glossopharyngeal neuralgia. In this review article, we present clinical vignettes and describe practical aspects of intra-oral, extra-oral, and peripheral techniques to block the glossopharyngeal nerve. The glossopharyngeal nerve blocks should be performed in a monitored setting due to the potential for adverse complications.

Introduction

Facial pain syndromes are commonly associated with a significant effect on a patient's quality of life[1]. Glossopharyngeal neuralgia (GN) refers to a pain syndrome affecting mainly structures in the sensory distribution of the cranial nerve IX [2]. Patients with GN have episodic sharp, stabbing, severe unilateral pain in the posterior region of the tongue, tonsillar fossa, pharynx, angle of the jaw, or occasionally the inner ear. The symptoms often arise due to activities resulting in sensory stimulation in the distribution of the Glossopharyngeal nerve (GNv). The most common activities are chewing, swallowing, coughing, talking, and yawing [3]. Rarely patients can present with arrhythmias and syncope[4]. In this review, we describe the anatomy, clinical features, diagnostic work, and non-surgical therapies for the management of GN. In particular, we will focus on the management of post-tonsillectomy pain and Eagle's syndrome.

Anatomy

The GNv is predominantly a sensory nerve with some preganglionic parasympathetic fibers and motors fibers **(Figure 1)**.

Sensory Component: The afferent or sensory fibers of the GN transmit information from areas of the mouth and pharynx, middle ear, and posterior one-third of the tongue and end in the trigeminal sensory nucleus. Special visceral afferent fibers (lingual nerve) carry signals from the taste buds of the posterior third of the tongue, which terminate in the nucleus solitarius. The sensory information mediates the swallowing and gag reflex in communication with the nucleus ambiguous and hypoglossal nucleus. These sensory fibers (nerve of Hering) also carry information from the chemoreceptors in the carotid body and baroreceptors in the carotid sinus. GNv also carries

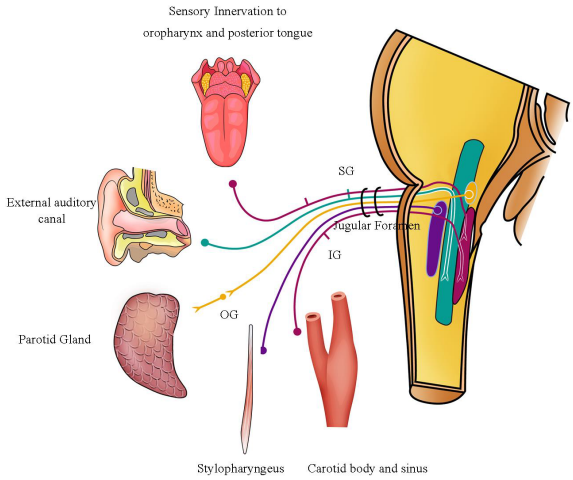

Figure 1. Anatomy of the Glossopharyngeal nerve. The nerve has predominantly sensory fibers that convey information to the brain stem. The GNv also supplies motor innervation (purple) to the stylopharyngeus muscle with fibers arising from the nucleus ambiguous. Sensory information from the posterior tongue, pharynx, and the carotid body is transmitted to nucleus solitarious. Sensory information from the external auditory canal is carried to the spinal trigeminal tract and nucleus (teal blue). The nerve also supplies parasympathetic innervation (yellow) to the parotid gland with synapses in the otic ganglion (OG). The GNv has additional connections with the superior (SG) and inferior ganglion (IG)

sensory information from the middle ear, mastoid through the tympanic branch, or Jacobson's nerve[5].

Motor Component: The GNv has limited motor fibers arising from the nucleus ambiguous, which innervate only the stylopharyngeus muscle involved in swallowing.

Autonomic Component: Preganglionic parasympathetic fibers of the GNv start in the inferior salivatory nucleus and synapse with the postganglionic fibers in the Otic ganglion which provide secretomotor fibers to the parotid gland.

Anatomical Course: GNv originates lateral to the olive in the rostral medulla as small rootlets, and then courses forward and laterally until it exits through the jugular foramen along with the vagus and spinal accessory nerves where it passes in-between the internal jugular vein (IJV) and the internal carotid artery (ICA). It descends anterior to the ICA and courses medially behind the styloid process where is starts to course away from the vagus nerve as it continues into its terminal branches. This extracranial course is along the posterior surface of the stylopharyngeus muscle. The nerve courses to the lateral surface eventually exiting through the gap between the superior and middle constrictors reaching the posterior surface of the tongue in the proximity of the palatine tonsil5. The carotid sinus nerve branches off the GNv as it exits the jugular foramen. Through its course, it has complex communication with the vagus nerve and sympathetic nerves.

Epidemiology

GN is a rare neuralgia syndrome, and it accounts for approximately 1% of all cranial neuralgias. The population-level incidence is around 0.7/100000 individuals [6]. The prevalence is similar among males and females, and the incidence increases with age. Most patients have unilateral pain (88%) and have spontaneous remission (76%) during follow up. GN is more common on the left side, while trigeminal neuralgia is more common on the right side [7].

Anatomical Sites Associated With GN

The GNv can be affected by lesions affecting the neuronal output from the brain before its origin in the brain stem, lesions affecting the nerve itself, extrinsic pressure from structures along the course, and by disease processes in the nearby structures **(Table 1)**. Multiple sclerosis is one of the causes that may affect the GNv prior to its origin in the brain.

Table 1. Causes of Glossopharyngeal Neuralgia

1. Idiopathic- No identifiable structural cause
2. Secondary
    a. Compression
        i. Tumors of the skull base
        ii. Paget's disease
        iii. Posterior fossa changes
        iv. Cervical malformation
        v. Eagle's syndrome
        vi. Vascular compression
            1. Arteriovenous malformations
            2. Persistent hypoglossal artery
            3. Dissection and aneurysm of the vertebral system
        vii. Oral or pharyngeal tumors
    b. Intracerebral changes
        i. Multiple sclerosis
        ii. Medullary Tumors
    c. Infectious and post inflammatory states
        i. Post tonsillectomy syndromes
        ii. Post neck dissection or radiation therapy
        iii. Sjogren's syndrome
        iv. Intraoral and skull base infections
        v. Post Herpetic neuralgia

Vascular compression, intracranial space-occupying lesions, posterior fossa and cervical malformations, and Eagle's syndrome; are examples of disease states affecting the nerve due to extrinsic pressure. Sjogren and other autoimmune diseases can lead to neuritis and GN. Peritonsillar, intra-oral infections, and other cancers of the mouth are other peripheral causes of GN. While the cause in most patients may remain unknown (idiopathic), most patients are treated with medical therapy, and neurosurgical intervention may be offered to patients with intrinsic causes of pressure prior to the exit at the jugular foramen.

Pathophysiology

The anatomical course of the nerve and its relationships explains symptoms in secondary causes of GN. Most cases do not have an identifiable cause of neuropathy and are termed idiopathic GN. Any inflammation, infection, or compression from the nearby structures may result in hyperexcitability of the nerve resulting in symptoms. Vascular compression of the nerve roots can lead to demyelination and ephaptic communication. The abnormal impulses due to repeated activation of the afferent neurons modulate hyperexcitability in the central nuclei [8].

Differential Diagnosis

Hyperactivity associated with other cranial nerves and myofascial Pain Dysfunction Syndrome are the main disorders that can be confused with GN [9]. Various hyperactive cranial nerve syndromes include trigeminal neuralgia (CN V), superior laryngeal neuralgia (CN X), and intermediate nerve neuralgia (CN VII) [2]. The diagnosis of GN is confirmed when symptoms improve with nerve block at the jugular foramen or with topical anesthesia of the pharynx.

Management

GN should be suspected based on clinical history and characteristics of pain associated with the episodes. A focused physical exam evaluating the potential causes of GN may provide additional clues. High-resolution Computed Tomography (CT) or Magnetic Resonance imaging of the brainstem

may be needed to identify compressive causes and demyelinating lesions, leading to GN. Radiography or high-resolution CT imaging may reveal an elongated styloid process suggestive of Eagle's syndrome as a cause for GN. Patient education, along with pharmacotherapy, is the mainstay of treatment for GN. In selected cases, nerve blocks and additional surgical interventions are warranted **(Table 2)**.

In the paper below, we focus on recent advances in the management of GN due to Eagles' syndrome, peritonsillar, and intra-oral lesions among patients who failed conservative therapy.

Table 2. Treatment Strategies for Glossopharyngeal Neuralgia

3. Patient education
4. Pharmacotherapy
    a. Carbamazepine
    b. Gabapentin
    c. Pregabalin
    d. Selective serotonin reuptake inhibitors
5. Nerve blocks
    a. Intraoral
    b. Extraoral
6. Interventional approaches
    a. Microvascular decompression
    b. Rhizotomy or Neurotomy
        i. Surgical
        ii. Percutaneous radiofrequency
    c. Stereotactic radiosurgery
    d. Pulsed radiofrequency neurolysis
    e. Gamma knife surgery
    f. Stylectomy
    g. Neuromodulation

Clinical Vignette

A 56-years-old female complained of chronic left-sided throat pain that was initially started in the recovery room after a tonsillectomy surgery under general anesthesia five years prior to this presentation. The pain was localized to the left base of the tongue, left tonsil area, and the soft palate with a sensation of swelling of these structures. She woke up from surgery with burning pain and a raw sensation in the interior of her throat that has not changed over the past five years. Mostly her pain increased with eating, especially spicy foods, and swallowing movements in her oropharynx. Her physical examination was within normal limits with evidence of scarring of the tonsillar fossa as expected after tonsillectomy surgery. She had allodynia in the areas shown in **Figure 2A**. After her tonsillectomy surgery, the patient had a prolonged empiric course of antibiotics under the guidance of an infectious disease specialist. She could not tolerate gabapentin because of side effects. She had tried pregabalin, duloxetine, baclofen, and tricyclic antidepressants without success.

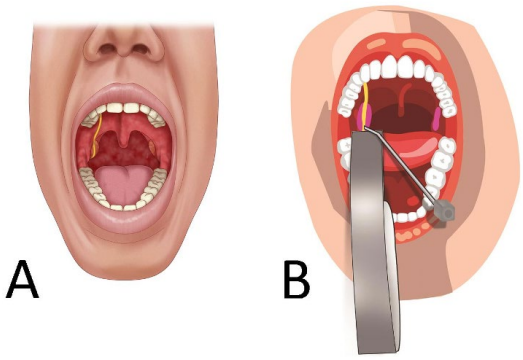

Figure 2. Panel A shows the relationship of the GNv to the left tonsillar pillar. The dotted outline shows the area of allodynia experienced by the patient. Panel B shows an approach to the Intraoral GNv block. A tongue depressor or retractor is used to expose the tonsillar surface, and local anesthetic is injected near the tonsillar pillars in the submucosal region after confirmation of negative blood aspiration.

GN was suspected, and a GNv block was suggested to confirm the diagnosis. She was interested and extremely anxious about the invasive approach to the glossopharyngeal nerve, especially in the light that she had not had much success with any previous treatments. A non-invasive intra-oral approach to GNv block was attempted for diagnostic purposes. After informed consent, the patient was placed in a recliner chair with monitors attached. A cotton pledget soaked in 4% lidocaine was held with angled-forceps ensuring the pledges extended beyond the metal tip to avoid injury. A string was wound around the pledges to avoid dislodgement. The soaked pledges were applied to the left side base of the tongue at the inferior portion of the tonsillar fossa in the area where the glossopharyngeal nerve crosses the posterior and anterior tonsillar pillars and is superficial. The soaked pledges were applied for 1-2 minutes, after which the patient described a numb sensation in the left side of the throat and reduction in pain in the recovery area. On follow up after three weeks, the patient reported a significant reduction in pain. She reported an overall 40% reduction in pain in the left throat. She continued to have reduced pain for about one month, at which time a spicy meal triggered her pain. She similarly underwent a second nerve block and continued to have improved pain control for over three months.

Intra-oral GNv Block: GN is a rare diagnosis that can occur uncommonly post-tonsillectomy, as in our patient. A GNv block can be challenging for pain physicians as it is not commonly performed because of the rarity of this neuralgia. Noninvasive intra-oral GNv block may be included as the first step in the diagnosis. GNv block using cotton pledges soaked in local anesthetics has been used by anesthesiologists for the management of difficult airways to limit gag reflex. The GNv can be blocked intra-orally in the vicinity of the tonsillar pillars (intra-oral/ premolar approach) and is considered safe. The patient is in a seated or supine position with a wide-open mouth, and the tongue is retracted medially and downward using a tongue depressor or a laryngoscope blade [10]. A bent needle or a 22-25G straight needle is used to aspirate and subsequently inject 1-2ml local anesthetic medication in the submucosal plane near the tonsillar pillar **(Figure 2 B)**. Relevant intraoral anatomy is shown in **Figure 3**. The needle should be rotated 90 degrees, and respiration attempted to minimize the risk of vascular injection [11]. Awake fiberoptic intubation requires a block of the GNv, superior laryngeal nerve, and translaryngeal nerve blocks. Recent reports of ultrasound guidance for nerve block along with local anesthetic spray in the oropharynx demonstrated successful nerve block of the airway even in an acidic environment like infected tissue [12]. Intraoral GNv block using lidocaine spray with video laryngoscopy has been described for pharyngeal anesthesia in a patient with severe nasal bleeding [13].

Clinical Vignette

A 45-year-old male presented with sharp neck pain with radiation to the ear and throat. He had a foreign body sensation and difficulty swallowing in the throat for the past year. His past medical history was otherwise unremarkable. A high-resolution CT scan showed an elongated styloid bone confirming the diagnosis of Eagle's syndrome. After an unsuccessful trial of gabapentin, the patient opted

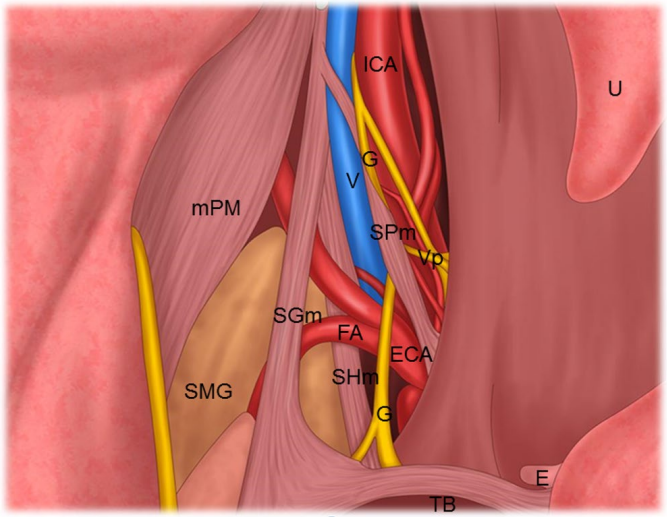

Figure 3. Intraoral submucosal course of GNv and its relationship to surrounding structures:
E: epiglottis, ECA: external carotid artery, FA: facial artery, G: glossopharyngeal nerve, ICA: internal carotid artery, mPM: medial pterygoid muscle, SGm: styloglossus muscle, SHm: stylohyoid muscle, SMG: submandibular gland, SPm: stylopharyngeus muscle, TB: tongue base, U: uvula, V: internal jugular vein, Vp: pharyngeal branch of vagus.

to undergo a fluoroscopic-guided GNv block. The patient had improvement in clinical symptoms and underwent a repeat injection at three months.

Extra-oral GNv Block: The percutaneous extra-oral GNv block is most often performed using imaging guidance. Approaches using fluoroscopy and ultrasound guidance have previously been described [14]. For the fluoroscopic approach, the patient is placed supine, and the head is turned to the unaffected side. Lateral fluoroscopic view is obtained to view the angle of the jaw and the mastoid process **(Figure 4)**. The midpoint of an imaginary line between the two points is an anatomic landmark for the styloid process. The skin is prepped with an antiseptic solution and is anesthetized with lidocaine. A 25G long needle is advanced until the styloid process is contacted. Once the styloid process is touched, the needle is walked-off posteriorly. Negative aspiration is confirmed before the injection of real-time contrast to rule out an intravascular spread. Then 2-3ml of local anesthetic with or without non-particulate steroids is injected.

Ultrasound approaches use a linear probe placed below the patient's ear lobe between the angle of the mandible and the mastoid process **(Figure 5)**. The patient's neck is prepared in a sterile fashion and turned to the unaffected site. The skin is anesthetized with local anesthetic solution. The styloid bone is visualized as a hyperechoic area medial to the mastoid process. When the styloid process is not identified, the internal carotid artery can be followed up to the ear lobe. The internal carotid artery is used as a landmark with a probe at the level of the ear lobe. The needle is directed posteriorly, and after a negative aspiration test, 2-3ml of local anesthetic with or without steroids is injected [15]. Newer techniques of GNv block more distally near the pharyngeal wall have been described. The pharyngeal wall and the horn of the hyoid bone are identified, and the local anesthetic is injected in the parapharyngeal space[16] **(Figure 6)**. This technique allows real-time visualization and

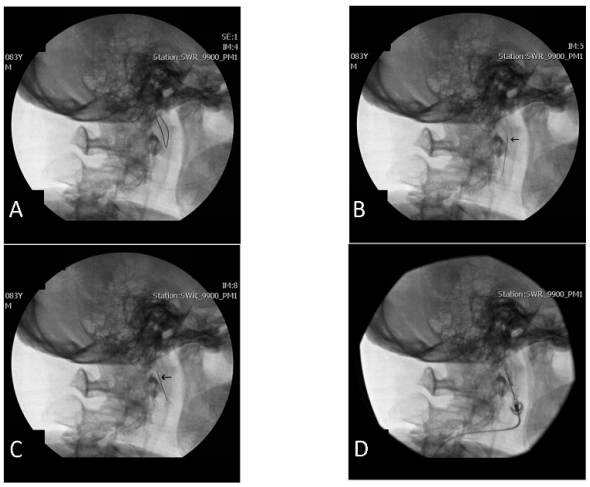

Figure 4. Fluoroscopic approach to extraoral GNv block:
Panel A shows a lateral view with the highlighted styloid process between the jaw-bone and the mastoid process. Panel B shows needle advancement to the styloid process. Panel C shows the needle walked off the posterior margin of the styloid process. Panel D shows contrast injection to confirm the absence of vascular spread prior to local anesthetic injection. Images reproduced with permission [19].

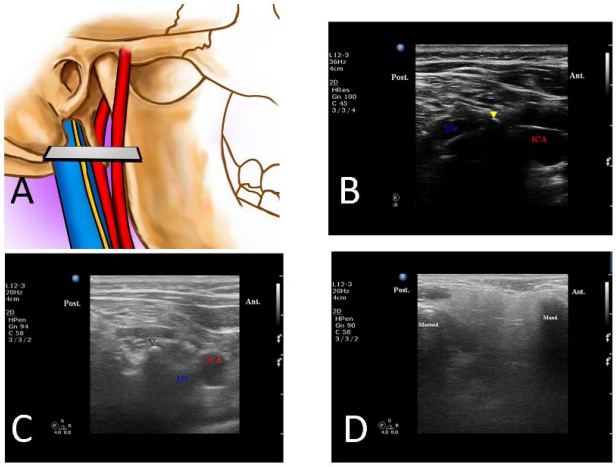

Figure 5. Ultrasound-guided extraoral proximal GNv block. Panel A illustrates the probe position and the resulting image plane below the earlobe and in between the mandible and mastoid process. Panel B illustrates styloid process (yellow arrow) and the relationship between the internal carotid artery (ICA) and Internal Jugular vein (IJV). Panel C illustrates a variable relationship with vessels based on the level of imaging. Panel D illustrates poor acoustic window in a patient. Images reproduced with permission [19].

at the same time avoids damage to the vascular structures and other nerve blockade.

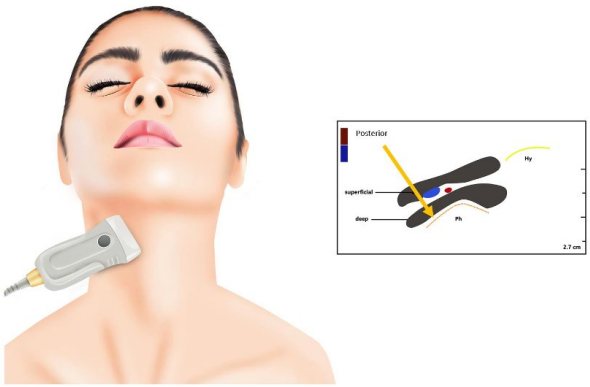

Figure 6. The left panel shows the position of the ultrasound probe for distal GNv block. The right panel shows a line diagram of an ultrasound image. The tissue plane of local anesthetic medication delivery is indicated by yellow arrow. There are two muscle layers seen in the figure. Facial artery and vein are seen traversing between the two muscle layers. Hy: hyoid bone, Ph: Pharynx. The depth of the imaging plane is optimized to visualize structures within 3 cm.

Precautions and Procedural Complications

Due to the close proximity of the GNv to major vascular structures and other cranial nerves, any interventions should be performed in a monitored setting with access to resuscitative equipment. Intravascular injections can lead to seizures and hemodynamic collapse. Local vascular injury in the neck can lead to hematoma formation and compression of the nearby structures. The risk of infection should be weighed in patients with oral infections undergoing intra-oral block or immunocompromised patients undergoing extra-oral GNv block. Patients with prior neck radiation therapy or neck surgery will require additional precautions to avoid complications. Intra-oral GNv block has a potential for injection into the tonsillar artery. Dysphagia and dryness may occur due to interruption of the motor supply to stylopharyngeus muscle. Dysphonia can result from recurrent laryngeal nerve palsy. Temporary weakness of the tongue and shoulder may occur due to block of the hypoglossal and spinal accessory nerves. Vagal nerve blockade can result in reflex tachycardia, hypertension, arrhythmias, and may be associated with hemodynamic instability.

Outcomes

Most of the GNv block reports are either single cases or case series. Several randomized trials show the safety and efficacy of GNv block for patients undergoing tonsillectomy. [17,18]

Conclusion

Glossopharyngeal neuralgia is a distinct syndrome associated with irritation or entrapment of the glossopharyngeal nerve (CN IX). Most cases do not have an identifiable cause. Patients are treated with medical therapy, interventional blocks in refractory cases, or with surgical treatment and neuromodulation. Both intraoral and extraoral GNbv block can be safely and effectively performed with a proper understanding of regional anatomy. Recent advances in ultrasound-guided distal block in the retropharyngeal space has the potential to minimize adverse events.

Contribution Statement

Vasudha Goel and Samer Narouze designed the review, drafted, and revised the paper. Vasudha Goel is a guarantor.

Disclosures

Dr. Samer Narouze is the current EIC for Annals of Headache Medicine. Dr Dmitri Souza was the acting EIC handling this manuscript.

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