Glossopharyngeal Neuralgia: A Comprehensive Update

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Abstract

Glossopharyngeal neuralgia (GN) is an unusualclinical condition where severe paroxysmal pain of the glossopharyngeal nerve. GN pain should be differentiated from trigeminal neuralgia pain, as the two neuralgias share several similar features. Thorough medical history coupled with a detailed clinical examination and hematologic and radiographic investigations is necessary for early and accurate diagnosis. Anticonvulsants form the cornerstone of the medical management, and surgical interventions (microvascular decompression and gamma knife surgery) is needed in cases resistant to pharmacotherapy.

Keywords: *Carbamazepine, glossopharyngeal neuralgia, microvascular decompression, neuralgia.*

Introduction

Neurologica non-nociceptive pain or pain which does not relate to the activation of pain receptor cells in the body. It's a type of pain due to a lesion or any disorder of the somatosensory system.[1]

As per ICHD-3 (International Classification of Headache Disease-3) classification, glossopharyngeal neuralgia (GN) is a disorder characterized by a brief periodic unilateral pain, with sharp and piercing in character, with abrupt onset and cessation, in the glossopharyngeal nerve distribution (angle of the jaw, ear, tonsillar fossa and the tongue base). It falls under the International Classification of Diseases (ICD) category as ICD-10-CM-G52.[1 2]

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The International Association for the Study of Pain (IASP) defines it as sudden, severe, brief, recurrent pain along the glossopharyngeal nerve distribution.[3]

GN is an extremely uncommon occurrence and accounts for only 0.2%–1.3% of the cases with facial pain.[4,5] GN usually affects the male individuals above 50 year-age, and the reported incidence of GN is roughly 0.8 per 100,000 persons per year.[4,6-11]

It must be asserted that GN is not as rare as reported in the literature due to non-familiarity with the disease, diagnostic obstacles, and more so with the escalating number of patients with Eagle's syndrome.[12]

Historical Background: Theodore H. Weisenburg (1910) was the first to provide the classic description of GN in a patient who reported with a lancinating ear and neck. Autopsy reports revealed that the pain was due to a cerebellopontine angle tumor, which presses the trigeminal nerve and stretches the glossopharyngeal nerve.[13]

After 10 yrs in 1920, Sicard and Robineau (1920) reported 3 patients with "Algie Velo-pharyngee essentielle" i.e. Pain along the distribution of glossopharyngeal nerve idiopathically[10,13]

The term "glossopharyngeal neuralgia" was framed by Wilfred Harris. He described GN as a painful condition characterized by episodes of severe lancinating, unilateral pain along the nerve division, which may be triggered by trigger point stimulation in areas supplied by the nerve.[10,13]

Two noteworthy advancements were reported for the efficient management of glossopharyngeal neuralgia in 1927. J. Doyle published a case series of 18 GN patients, and W. Dandy performed the first successful intracranial resection for glossopharyngeal neuralgia.[14]

Riley et al. (1942) reported an uncommon association of GN with life-threatening cardiovascular manifestations- asystole, transient/persistent bradycardia, syncopal episodes, and even seizures.[8]

Classification:

1. Based on the International Headache Society (IHS) Classification, GP is classified as (1) classical GPN (occasional or episodic pain) and (2) symptomatic GPN (persistent pain; persisting between the episodes of peaks and troughs of pain.).[15]
2. Based on the pain distribution, GN is classified as- (a) Otitic pain: Pain in (or around) the auricle/ ear lobe (b) Oropharyngeal pain: Pain of neck and maxillofacial regions. Nevertheless, these regions show considerable overlap inthe cranial nerve-supplied areas.[16]
3. Based on the causal factors, GN is classified as (a) Idiopathic/essential GN- The majority of GN cases are idiopathic, where no apparent lesion is noticed.[17] GN is caused by neurovascular compression of nerve IX by the postero-inferior cerebellar artery (PICA),[18] which was first observed postmortem by Brihaye et al.[19] (b) Secondary GN- where a demonstrable lesion is seen. Eagle's syndrome/ styalgia accounts for the most common secondary cause of GN .[13]

Eagle's syndrome is an unusual disorder, clinically alarger in size of the styloid process or stylohyoid ligament complex is mineralized.

Clinical Features: Glossopharyngeal neuralgia is a rare disease entity, characterized by pain paroxysms in the regions supplied by the glossopharyngeal nerve (auricular and pharyngeal). The GN pain is typically characterized as brief episodes of unilateral severe, piercing/lancinating pain, confined primarily of the ear, ventral surface of tongue, throat, tonsillar region, or beneath the angle of the jaw.[21]

The GN pain distribution is diagnostic: The pain generally commences from the pharyngeal, tonsillar, and posterior base of tongue and includes the eustachian tube and inner ear or spreads to the mandibular angle. Exacerbating factor in GN is generally deglutition, and cold liquids mainly evoke the pain.[21]

GN pain is transient and excruciating to begin with (lasts for 30sec), although it may recur after days, weeks/ months and persists in the same areas.[22,23] Swallowing, chewing, sneezing, talking, cleaning the throat, and light touch to the gums/oral mucosa, abrupt head movements, pain on lifting of the arm of the effected site, and the lateral jaw movement may also precipitate the pain.[24]

Vagoglossopharyngeal neuralgia refers to an uncommon linkage between GN and syncope,[2] where the affected individual presents with episodes of bradycardia, severe hypotension, syncope, and even cardiac arrest.[25] The probable reason for these life-threatening manifestations in GN may be attributed to the anatomical vicinity of glossopharyngeal nerve with the vagus and nervus intermedius.[26]

It is important to distinguish GN from trigeminal neuralgia (TGN), that shares some features with GN and can combined form in 10% of patients.[27] Spontaneous remission is seen in both GN and TGN.[8]

GN is an uncommon clinical entity as compared to TGN (Reported prevalence rate of GN and TGN is approximately 0.8/100,000 population and 4.7/100,00 population respectively).[9] The left side of the face is usually affected in GN in contrast to the right side in TGN.[28]

The two neuralgias can also be differentiated based on the site affected and triggering factors. GN has the predilection for the throat and tonsillar region, and TGN primarily affects check, ala of nose, vermillion border of lips (facial region along the trigeminal nerve division). GN pain is triggered by chewing and swallowing, whereas, a light facial touch while washing/brushing the teeth exacerbates the TGN pain.[16]

A dull ache usually persists between pain episodes in GN,[29] in contrast to symptom-free periods between pain paroxysms in TGN. Any individual experiencing a dull ache between pain paroxysm does not fulfill TGN diagnostic criteria.[30]

GN pain occurs mostly during the daytime, but the pain may disturb the sleep and the patient may wake up in the night. However, there is no pain during sleep in TGN.[29] Topical anesthetic application on pharyngeal mucosa may relieve GN pain, thus, distinguishing it from other neuralgias. Vasoglossopharyngeal neuralgia may be associated with life-threatening cardiovascular features- syncope, hypotension, cardiac arrhythmias, in contrast to trigeminal neuralgia.[29]

Diagnostic Aids: Characteristic clinical manifestations usually suffice for the diagnosis of GN, and definitive investigative tests do not exist. GN patients are usually recommended for an ear, nose, and throat (ENT) consultation to rule out a possible anatomical lesion.[2] The distinctive unilateral, severely piercing, episodic pain in the throat, triggered on swallowing provides confirmation for GN pain, and differentiates it from the pain of inflammatory origin (constant pain, lasts for minutes). Glossopharyngeal nerve involvement/ association with other cranial nerves may be evaluated by assessing the pain distribution. Ototic pain may be detected by injecting an anesthetic solution (2% Lignocaine or 0.5% bupivacaine) into the region with a close approximation to the trigger point.[11] Potential etiologies such as vascular compression/neoplasms may be demonstrated by radiographic evaluation of the brain stem. A high-resolution neck CT scan may demonstrate Eagle's syndrome.[2]

Treatment: GN may be treated with either Pharmacologic/non-pharmacologic interventions.[31]

Carbamazepine is regarded as the drug of choice in GN. The drug has an inhibitory action on the brain stem reflex activity.[32] Also, the drug has a possible action on preventing bradycardia.[33,34]

Gabapentin, pregabalin, and lamotrigine are the other pharmacotherapeutic agents used in GN. Gabapentin and pregabalin act on the α2δ subunit of calcium channels, and diminish the release of excitatory neuro-transmitters. Anticholinergic therapy, β2-adrenoceptor agonists (atropine, isoprenaline) and temporary pacing may be used to manage the bradyarrhythmia in GN.[35] Published literature has also elucidated the efficacy of selective serotonin reuptake inhibitors (20–50mg/day paroxetine and 50–200mg/day sertraline) and vitamin B12 in the management of GN.[36]

Only a few cases of GN have shown response to non-steroidal anti-inflammatory drugs (due to their probable action on unidentified acute inflammation) and opioids (as an ancillary agent to the anti-convulsants).[17]

Glossopharyngeal nerve block provides quick pain amelioration and acts as an excellent ancillary aid in the medical management of GN. It will bedone with either (local anesthetic agents) non-neurolytic agents with or without additions of (steroid, ketamine, etc.) or with(phenol, alcohol, glycerol, etc). neurolytic agents [37]

Surgical management is usually adopted in young aged, recalcitrant cases and is associated with high morbidity.[17]

Microvascular decompression (MVD) is the most preferred surgical technique associated with the highest success rates.[28] MVD is a more efficient treatment than Gamma knife surgery (GKS) due to both initial and long-term pain remission, lower relapse rates, and maintaining a pain-free status.[38]

Conclusion

Glossopharyngeal neuralgia is an uncommon neuropathic pain that occurs along the distribution of the glossopharyngeal nerve. GN can be classified based on the nature of pain, site predilection, and etiologic factors. This neuralgic pain mimics trigeminal neuralgia; hence, an early and accurate diagnosis is mandatory. The condition is seldom associated with life-threatening cardiovascular manifestations like bradycardia, hypotension, syncope and rarely cardiac arrest. Pharmacotherapy with anti-convulsants is usually sufficient, although surgical intervention may be required in resistant cases.

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