Treatment options for glossopharyngeal neuralgia

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Glossopharyngeal neuralgia is a rare but painful affliction creating pain in the distribution of the nerve. It is an intermittent pain often provoked by non-noxious stimulation such as talking, swallowing or head movement. Glossopharyngeal neuralgia is best evaluated with a careful history, appropriate imaging and treated with pharmacologic approaches as well as surgery.

Glossopharyngeal neuralgia (GPN) is a rare condition involving throat and neck pain. The International Association for the Study of Pain (IASP) defines the condition as sudden, severe, brief, recurrent pains in the distribution of the glossopharyngeal nerve [1]. The International Headache Society recently classified GPN into classic and symptomatic [2]. They provided diagnostic criteria under the major group of cranial neuralgias and central causes of facial pain. Classic GPN is described as a severe transient stabbing pain experienced in the ear, base of the tongue, tonsilar fossa or beneath the angle of the jaw. The pain is felt in the distributions of the auricular and pharyngeal branches of the vagus and glossopharyngeal nerves. It is commonly provoked by swallowing, talking or coughing. It may remit for varying periods. Symptomatic GPN presents with the added presence of an aching pain that may persist between attacks. The diagnostic criteria are reviewed in Table 1.

Often, GPN occurs in episodes such as trigeminal neuralgia (TN), with each episode lasting for weeks to months, but in some patients can be unremitting. GPN is most often unilateral. The right side is affected more often with GPN than with TN. Bilateralism was noted less often in TN than in GPN cases [3]. Mechanical stimulation of the face by swallowing, talking and coughing often triggers the pain of GPN [4].

Keywords: glossopharyngeal nerve, glossopharyngeal neuralgia, temporomandibular disorder, temporomandibular joint, trigeminal neuralgia

Epidemiology

Katusic and colleagues published a 39-year retrospective study on the population of Rochester (MN, USA) [5]. A review of cases from 1945 until 1984 was carried out. It was found that the incidence rate of GPN in this population was 0.7 out of 100,000 for both sexes combined. There were no significant differences between the sexes. They concluded that GPN was generally a mild disease, since mild attacks were not uncommon, with only 3.6% of GPN sufferers having a second annual recurrence. Only 25% had to have surgery for symptom relief. In addition, 25% had bilateral symptoms.

Rushton and colleagues published a study in 1981 that examined GPN patients at the Mayo Clinic from 1922 to 1977 [6]. The authors reviewed 217 cases – a total of 57% were over than 50-years of age and 43% were between the ages of 18 and 50 years. A total of 161 patients had spontaneous remissions, 37 experienced no relief and 12% had bilateral pain. Syncope, which has been reported to occur with GPN, was rarely seen. A total of 25 patients experienced GPN and TN concurrently. Carbemazepine was the drug of choice and a total of 110 patients experienced good relief from pain with surgical intervention.

Patel and colleagues published a retrospective study of over 200 patients with GPN who underwent microvascular decompression (MVD) surgery at their institution over a 20-year period [7]. They found that 66.8% were female and 33.2% male. Mean age was 50.2 years, with a mean duration of pain of 5.7 years. The most common symptoms were throat and ear pain and throat pain alone. There was not a sided predilection with 54.8% of patients having left-sided symptoms and 45.2% having right-sided symptoms. Kondo and colleagues concluded in their study that GPN is rare, being seen 100-times less often then TN [8]. In summary, GPN is a relatively rare, usually unilateral painful condition that tends to be more common in middle-aged females.

Differential diagnosis

Differential diagnoses for GPN include, but are not limited to, TN, temporomandibular disorders (TMDs), Eagle's syndrome, certain short-lasting headaches and local pathology.

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Table 1. Integrated Health Service (IHS) criteria for glossopharyngeal neuralgia.

Criteria	Symptoms
Classical glossopharyngeal neuralgia	
A	Paroxysmal attacks of facial pain lasting from a fraction of a second to 2 minutes and fulfilling criteria B and C
B	Pain has all of the following characteristics:
1	Unilateral location
2	Distribution within the posterior part of the tongue, tonsilar fossa, pharynx or beneath the angle of the lower jaw and/or in the ear
3	Sharp, stabbing and severe
4	Precipitated by swallowing, chewing, talking, coughing or yawning
C	Attacks are stereotyped in the individual patient
D	There is no clinically evident neurologic deficit
E	Not attributable to another disorder
Symptomatic glossopharyngeal neuralgia	
A	Paroxysmal attacks of facial pain lasting from a fraction of a second to 2 minutes, with or without persistence of aching between paroxysms, and fulfilling criteria B and C
B	Pain has all of the following characteristics:
1	Unilateral location
2	Distribution within the posterior part of the tongue, tonsilar fossa, pharynx or beneath the angle of the lower jaw and/or in the ear
3	Sharp, stabbing and severe
4	Precipitated by swallowing, chewing, talking, coughing and/or yawning
C	Attacks are stereotyped in the individual patient
D	A causative lesion has been demonstrated by special investigations or surgery

TN is a unilateral disorder characterized by brief electric shock-like pains, abrupt in onset and termination, limited to the distribution of one or more divisions of the trigeminal nerve [9]. TN is the most similar condition to GPN and is mistaken for GPN more than any other condition.

TMDs can be unilateral or bilateral, painful or non-painful, and are located in and around the temporomandibular joint (TMJ). Painful TMD include arthridities of the TMJ, derangements of the joint, sprain of the joint, myalgia, myofascial pain, tendonitis, trismus, and spasm. These TMD pains can be similar to GPN when TMD is unilateral and intermittent. The best manner of differentiating these disorders is to ascertain the presence of temporomandibular joint noise, limited range of jaw motion (less than 40mm measured between the incisors), pain on joint palpation and worsening of the pain with chewing. If there is the presence of three out of four of the above it is more likely the pain is attributed to the jaw than GPN.

Eagle's Syndrome occurs from the elongation of the styloid process. The pain associated with this condition is often unilateral and can occur in the throat and ear, as in GPN. The symptoms from Eagle's Syndrome can occur from compression from the elongated styloid process of the glossopharyngeal, vagus, and/or trigeminal nerves. Bilateral elongation is common, but bilateral symptoms are less common [10]. Eagle's Syndrome is the most important cause of symptomatic GPN. Stylectomy is the treatment for Eagle's Syndrome and should be considered prior to surgical treatment for GPN [11].

Pathophysiology

In most cases it is thought that the primary cause of GPN is from compression of the nerve by a blood vessel. The vessel is usually the posterior inferior cerebellar artery [12]. This is supported from success of microvascular decompression surgery [13] as well as from electron microscopic observations of nerve injury of the glossopharyngeal nerve in patients with GPN [14]. This pathophysiology of vascular nerve compression is also supported by imaging that shows the compression. A study by Karibe and colleagues demonstrated that using magnetic resonance imaging (MRI) with a sequence of constructive interference in steady-state (CISS) they were able to accurately delineate microvascular compression to the glossopharyngeal nerve at its cisternal portion. They confirmed the results of the MRI during a microvascular decompression surgery [15].

Secondary causes of GPN

Compression of the nerve can also be caused by cerebellopontine angle tumors. Multiple sclerosis (MS) may also be associated with GPN. A report by Minagar and colleagues found that four of their multiple sclerosis patients had GPN. Three of the four patients responded to carbamazepine and the fourth responded to other treatment. They concluded that GPN may be associated with MS and responds to carbamazepine [16].

Other secondary causes of GPN include: Eagle's Syndrome, malignancies of the neck, Paget's disease, Arnold Chiari malformation [17,18], and neck trauma with foreign-body impaction [19].