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The Glossopharyngeal Nerve, Glossopharyngeal Neuralgia and the Eagle's Syndrome – Current Concepts and Management

K B K Soh

ABSTRACT

Glossopharyngeal neuralgia is not just a painful condition. At times, it may be life-threatening as a result of associated cardiovascular consequences. Even in the absence of life-threatening consequences, it can be a severe debilitating disease with depression, suicidal tendencies, fear of swallowing, loss of weight and under-nutrition.

The treatment for glossopharyngeal neuralgia and Eagle's syndrome has evolved over time.

This review summarises the scientific evidence and philosophy about current management and therapy.

Emphasis is placed on the importance of excluding secondary causes of glossopharyngeal neuralgia before embarking on nerve section through the posterior cranial fossa approach. The Eagle's syndrome due to an elongated styloid process is the most important cause of secondary glossopharyngeal neuralgia. Stylectomy is effective and should be considered before embarking on any neurosurgical procedure. Peripheral cervical and trans-tonsillar appoaches to the glossopharyngeal nerve are also discussed.

Keywords: glossopharyngeal, neuralgia, Eagle's syndrome, cardiovascular, transtonsillar

INTRODUCTION

Amongst the lower 6 cranial nerves, the glossopharyngeal nerve is the smallest in terms of nerve diameter, importance and clinical significance. When compared with the facial, vestibulococchlear, vagus, accessory and hypoglossal nerves, the glossopharyngeal nerve appears to dwarf in comparison. Otolaryngologists and other clinicians are consciously aware of the presence of the facial, vagus, accessory and hypoglossal. These are commonly encountered in neck surgery. Inadvertent surgical injury to these nerves result in clinically obvious problems like facial palsy, vocal cord palsy, shoulder dysfunction from denervation of the trapezius muscle, and speech problems arising from tongue deviation. The vestibulococchlear nerve subserves the important sense of hearing and balance. The science of audiology has evolved specifically to test and probe the functions of this nerve.

Department of Otolaryngology
National University Hospital
5 Lower Kent Ridge Road
Singapore 119074

K B K Soh, FRCS (Edin), FRCS (Glas), FAMS (ORL), MBA
Consultant Otolaryngologist

Correspondence to:
Dr K B K Soh

However the glossopharyngeal nerve remains the neglected cranial nerve. This is because the nerve is small and lies deep within the neck, and surgeons often do not encounter the nerve even with deep dissections of the neck. The nerve is not commonly identified or visualised even when performing a major neck operation, for example a radical neck dissection. A more important reason is because the glossopharyngeal nerve supplies important structures in the head and neck region only in the company of another cranial nerve. It does not supply an important structure in isolation and has no monopoly in the innervation of any critical organ. Mother Nature had not entrusted the glossopharyngeal nerve an important vital function in the same way that she had given roles of importance to the other cranial nerves.

Surgical anatomy of the glossopharyngeal nerve[1-3]

The glossopharyngeal nerve is the nerve of the third pharyngeal arch. The third pharyngeal arch in embryonic life goes on to form the lower part of the body and the greater horn of the hyoid bone. Unlike the first, second, fourth and sixth pharyngeal arches which play major roles in head and neck development, the contribution by the third arch is small. The fifth arch only makes a transitory appearance in embryonic life, and thereafter disappears, and can therefore be considered to be even less important than the third arch in head and neck development.

The glossopharyngeal nerve has both sensory and motor components. It receives somatic sensory fibers from the oro-pharynx, posterior third of the tongue, Eustachian tube, middle ear and mastoid. The sensory supply to the middle ear and mastoid passes along the tympanic branch or Jacobson's nerve. The glossopharyngeal nerve also receives special sensory fibers for taste in the posterior third of the tongue as well as chemoreceptor and baroreceptor afferent inputs from the carotid body and carotid sinuses respectively. The cell bodies of these sensory cells lie in the petrous ganglion. Their central processes pass to the nucleus of tractus solitarius for taste, and to the nucleus of spinal tract of trigeminal for somatic sensory. The afferent fibers for chemoreceptors and baroreceptors

pass on to the dorsal nucleus of the vagus, which is the main autonomic nucleus, and from there to the respiratory and vasomotor centers.

The motor component supplies the striated muscle stylopharyngeus and secretomotor parasympathetic fibers to the parotid gland. The supply to the stylopharyngeus begins in the nucleus ambiguus which receives bilateral supranuclear innervation from corticobulbar fibers. The parotid supply begins in the inferior salivatory nucleus. The fibers pass along the Jacobson's nerve to the tympanic plexus and lesser superficial petrosal nerve to relay in the otic ganglion. The postganglionic parasympathetic fibers then reach the parotid gland by way of the auriculotemporal nerve.

The tympanic branch or Jacobson's nerve is a very important branch of the glossopharyngeal nerve. It carries somatic sensory fibers which receive pain and touch from the middle ear and mastoid, and secretomotor parasympathetic fibers to the parotid gland. It does not supply the external ear canal or pinna. The Jacobson's nerve emanates from the petrous ganglion of the glossopharyngeal nerve at or above the level of the jugular foramen. It runs to the tympanic plexus lying on the promontory on the medial wall of the middle ear.

The other important branch is the carotid sinus nerve (nerve of Hering). This nerve supplies the carotid body and carotid sinus. It conveys chemoreceptor and stretch baroreceptor information centrally for respiratory and circulatory reflex function. It branches from the glossopharyngeal nerve at the level of the carotid bifurcation.

The long credentials of the glossopharyngeal nerve appear quite impressive. Yet nerve sacrifice for the treatment of glossopharyngeal neuralgia seldom ever result in significant untoward effects. The loss of the sensation of taste to the posterior tongue is unlikely to be missed because the posterior tongue is a midline structure with a bilateral nerve supply and a great deal of overlap. Also the supply to the anterior tongue by the chorda tympani more than compensates for any loss of taste sensation in a small part of the posterior tongue. The loss of somatic sense of pain and touch to the middle ear and mastoid will not be perceived by the patient, although a hyperactivity syndrome like glossopharyngeal neuralgia can cause excruciating pain in the ear. The stylopharyngeus muscle is only one of three muscles that originates from the styloid process. Isolated loss of stylopharyngeus muscle function will not cause a perceptible problem with swallowing or speech. The isolated loss of secretomotor function to the parotid will not cause a remarkable reduction in the volume or quality of saliva being produced. The presence of the opposite parotid gland, both submandibular and sublingual glands, and the numerous minor salivary gland in the rest of the aerodigestive tract will more than compensate for the loss. On the contrary, abnormal secretory function of the secretomotor fibers, as seen in the Frey's syndrome after parotid resection, can be an important cause of discomfort and distress.

Thus we have seen that hypofunction of the glossopharyngeal nerve is unlikely to be a significant cause of morbidity. This is because the nerve, and the territory that it supplies, namely the structures derived from the third pharyngeal arch, are of minor significance when compared to the other cranial nerves. In contrast however, a hyperfunctional irritative lesion of the glossopharyngeal nerve can be a very important cause of distress and suffering for the patient. In some instances, an irritated glossopharyngeal nerve can also be potentially deadly and life-threatening.

Glossopharyngeal neuralgia

Glossopharyngeal neuralgia was first described by Weisenburg[4] in 1910 as "tic doloureux". His patient had presented with the classical symptoms of lancinating pain in the ear and neck. It was only discovered 6 years later when the patient died and an autopsy was performed, that the patient had a cerebellopontine angle tumour. The tumour was noted to be compressing the trigeminal nerve and stretching the glossopharyngeal nerve at autopsy.

Ten years later in 1920, Sicard and Robineau[5] described three patients who had "algie velo-pharyngee essentielle" ie pain in the distribution of the glossopharyngeal nerve without any known cause. Their patients developed suicidal tendencies after treatment with sedatives or physical agents did not work. However, sectioning of the glossopharyngeal nerves through the cervical approach was successful in relieving the pain in all three of their patients.

A year later, Harris[6] coined the term "glossopharyngeal neuralgia", describing it as a painful syndrome characterised by paroxysms of unilateral and severe lancinating pain occurring in the distribution of the nerve, and which may be elicited by stimulation of trigger points in regions supplied by the nerve. The pain may be spontaneous or precipitated by a variety of actions that stimulate the region supplied by the glossopharyngeal nerve namely yawning, coughing, swallowing and talking.

Glossopharyngeal neuralgia has often been compared with trigeminal neuralgia. As both are neuralgic pain syndromes associated with cranial nerves, there is a tendency to assume that similar pathogenetic processes are operational in both syndromes. It is reasonable to suggest that successful treatment strategies for trigeminal neuralgia may be extrapolated for use in treating glossopharyngeal neuralgia.

However, there is also evidence that the two pain syndromes are not so similar after all. There is a difference in the overall age- and sex-adjusted annual incidence rates which demonstrate that trigeminal neuralgia is about six[7] to a hundred[8,9] times more common than glossopharyngeal neuralgia. Glossopharyngeal neuralgia is a milder disease than trigeminal neuralgia based on the number of episodes, treatment and characterisation of pain[7]. However, glossopharyngeal neuralgias are more resistant to treatment with carbamazepine.

Glossopharyngeal neuralgia has been divided into two clinical types[8] based on the distribution of pain: tympanic type which affects mainly the ear, and the oropharyngeal type which affects mainly the oropharyngeal area. The presence of pain in the ear is attributed to the somatic sensory supply of the tympanic membrane, middle ear and mastoid by the Jacobson's nerve, which branches off the glossopharyngeal nerve at the petrous ganglion.

The importance of differentiating between the two clinical types lie in making the choice between a low or high approach to sectioning the glossopharyngeal nerve. A low approach avulses the nerve distal to the petrous ganglion, thus leaving the Jaconson's nerve intact. A high approach avulses the nerve at or proximal

to the petrous ganglion, or even at the nerve root entry zone at the brainstem.

If the trigger area is in the pharyngeal area and otologic symptoms are minimal, then a simple trans-tonsillar pharyngeal approach[10] or a cervical approach may be utilised. However, if otologic symptoms are predominant, then the Jacobson nerve has to be included in the resection, and a high cervical or retrosigmoid, posterior fossa intracranial operation is the surgical treatment of choice.

The symptoms induced by glossopharyngeal neuralgia is usually described as severe and paroxysmal. However, an equal number may also have a constant dull ache which may exist in isolation or be accompanied by short attacks of severe paroxysmal pain. Although there is no apparent difference in these two types of pain patterns in terms of relief of otalgia after nerve section[11], it is not known if there is any difference in terms of severity of pain, prognosis and response to carbamazepine.

Pathogenesis

Glossopharyngeal neuralgia usually occurs without any obvious cause. A thorough physical examination usually does not reveal any abnormality other than the identification of trigger points. Radiological examination including CT scans, MRIs, and angiograms will be normal.

Why then does the nerve go into a state of hyperexcitability causing "idiopathic glossopharyngeal neuralgia"?

When good results were reported for microvascular decompression of the glossopharyngeal nerve[12,13] in 1977, it became apparent that most of these cases of "idiopathic" glossopharyngeal neuralgia could be caused by vascular compression of the glossopharyngeal nerve at the nerve root entry zone, causing a hyperactive rhizopathy. The implicating vessel is usually the posterior inferior cerebellar artery (PICA)[14] which frequently also compresses on the rootlets of the vagus nerve. It is impossible to image radiologically, and to determine pre-operatively, the presence of this compressive relationship, which can only be discovered at the time of explorative surgery through the posterior fossa approach.

This concept of nerve hyper-excitability induced by vascular compression is not new. Although it has only recently been thought to play a part in causing idiopathic glossopharyngeal neuralgia, it is a well recognised and documented pathologic mechanism in other cranial nerve syndromes. It has been known for some time that trigeminal neuralgia, hemifacial spasm, spastic dysphonia, hearing loss[12] and vertigo from vestibular paroxysmia[15,16] and spasmodic torticollis can be a form of vascular compression neuropathy. These syndromes are quite effectively treated by a microvascular decompression procedure of the relevant vessels.

Before embarking on an intracranial explorative procedure, it is important to exclude secondary causes of glossopharyngeal neuralgia. Any sort of compression or irritation to the glossopharyngeal nerve can result in neural hyper-excitability and neuralgia. The most common secondary cause of neuralgia is the Eagle's syndrome[17-20] or styalgia. It is a glossopharyngeal nerve hyper-excitability syndrome caused by compression of the nerve against an elongated or fractured[21] styloid process or a calcified stylo-hyoid ligament[22].

Other causes of secondary glossopharyngeal neuralgia include: cerebellopontine angle tumours[4,23], parapharyngeal space lesions[24], metastasis to petrous temporal bone from breast carcinoma[25], post-tonsillectomy[26], local infection[27,28], carcinoma of the parapharyngeal space[28,29] carcinoma of the pharynx[6,30], nasopharyngeal carcinoma[31], posterior fossa arterio-venous malformation[32].

Atypical glossopharyngeal pain

But pain from glossopharyngeal neuralgia may not be localised to the area supplied by glossopharyngeal nerve. Pain may radiate to atypical sites in the face, forehead, hypopharynx, larynx, external ear canal and pinna, areas which are not supplied by the glossopharyngeal nerve.

There are two possibilities for this. The atypical features may be due to either a multicranial neuropathy, or it may be due to cross-talk between fibers in adjacent cranial nerves. The occurrence of multi-cranial neuropathy can be most easily explained using the concept of intracranial vascular compression or arachnoidal adhesions[33]. The PICA commonly sits on and compresses the glossopharyngeal and vagus nerves, while the AICA does the same for the vestibulo-cochlear nerve and nervus intermedius[34]. The vascular loops of AICA and PICA can compress several cranial nerves simultaneously leading to neuralgic syndromes that involve areas that are more extensive than would be expected of a single cranial nerve. Sometimes, neuralgic syndromes may also be associated with hearing loss[35]. It is therefore important, though difficult, to distinguish the various pain syndromes namely geniculate neuralgia, nervus intermedius syndrome, vagal neuralgia, and trigeminal neuralgia. These neuralgic syndromes can occur in isolation or in combination.

The concept of multiple ipsilateral cranial rhizopathy due to compression by intracranial vessels or arachnoidal adhesions on several nerves simultaneously accounts for why decompression or avulsion of the glossopharyngeal nerve alone may not always induce pain relief until other nerves are also decompressed or avulsed. The variable contribution by the vagus, nervus intermedius, glossopharyngeal, and trigeminal nerves gives rise to an atypical facial pain syndrome that will not respond adequately to treatment of a single cranial nerve.

Cross-talk is a theoretical concept where hyperactive impulses originating from the glossopharyngeal nerve spreads, diffuses or contaminates other cranial nerves. It is presumed to be the result of the fact that the trigeminal, glossopharyngeal, vagal, and upper cervical roots all contribute to the descending spinal nucleus of the trigeminal nerve[36]. It helps to explain why pain from glossopharyngeal neuralgia can sometimes extend beyond the apparent distribution of the glossopharyngeal nerve.

Alternatively, we have to presume that the glossopharyngeal nerve supplies different structures in different individuals thus accounting for the different distribution of pain in different patients. However this is not a likely possibility.

Glossopharyngeal neuralgia can be life-threatening

Glossopharyngeal neuralgia can be extremely disabling. Some patients, in whom swallowing is a trigger factor, can suffer from weight loss and under-nutrition from fear of swallowing[37,38]. The depression from debilitating pain

may become so severe that there have been cases of neuralgic patients who have attempted suicide[5].

Even worse, the effects associated with glossopharyngeal neuralgia can be life threatening[39]. In 1921, Harris[6] reported that glossopharyngeal neuralgia can be associated with cardiac dysrhythmia and instability. This relationship is a well accepted one, having been documented by many authors, subsequently. The various reports and case studies has been compiled and summarised by Ferrante et al[40].

Intense irritability and hyper-stimulation of the glossopharyngeal nerve feedback onto the vasomotor center in the brainstem, giving rise to a heightened vagal response. This results in cardiac dysrhythmia, bradycardia, hypotension, and even asystole and subsequent syncope. This effect is similar to that seen in carotid sinus massage for the treatment of supraventricular tachycardias. Massaging the carotid sinus causes a hyper-stimulation of the glossopharyngeal afferent pathway, resulting in an exaggerated parasympathetic vagal efferent response. In the case of glossopharyngeal neuralgia, the hyper-stimulation is induced by either an intrinsic irritability of the nerve, or compression of the nerve by blood vessels or styloid process.

Alternatively, the enhanced vagal response may be due to direct compression by the PICA in the posterior fossa. However, this is thought to be a less common mechanism compared to mediation by the glossopharyngeal nerve.

Treatment for the cardiovascular effects of glossopharyngeal neuralgia with conventional cardiologic management, eg. cardiogenic drugs or electrical pacing, have not been demonstrated to yield satisfactory or long-lasting results[24,29,39,40]. Fortunately, treatment which is directed at relieving the irritability of the nerve is usually a more feasible option. Carbamazepine or surgical approaches like avulsion of the glossopharyngeal nerve or microvascular decompression have been very effective in the management of both the neuralgic pain and its associated cardiac effects.

There is a subset of patients with demonstrable cardiologic manifestations without the typical neuralgic symptoms, who have responded very well to glossopharyngeal nerve avulsion or microvascular decompression of the nerve rootlets. Though these patients have cardiac dysrhythmias that have been refractory to the usual cardiologic manipulations, good outcomes have resulted from treatments that have been directed at reducing glossopharyngeal nerve irritability. Such syndromes have been termed non-neuralgic glossopharyngeal neuralgia[41], in recognition of the fact that glossopharyngeal nerve irritability may not always give rise to a pain syndrome.

Cardiovascular events and syncope have also been associated with parapharyngeal space lesions[24]. There are several possibilities for this. It may be due to compression of the carotid sheath by the mass, involvement of the vagus nerve or sympathetic trunk, or stretching and compression of the glossopharyngeal nerve. It is likely that glossopharyngeal nerve compression and irritability is the most likely pathophysiologic mechanism that can account for cardiovascular instability. The reason for making such an assumption is twofold. Firstly, the attacks of bradycardia and syncope occur in a paroxysmal and episodic fashion which simulates the cardiovascular events that is seen to accompany true glossopharyngeal neuralgia. Secondly, microvascular decompression of the glossopharyngeal nerve has been shown to cause a dramatic and long-lasting improvement in cardiac instability. Cicogna et al[24] reported 11 cases of recurrent and severe vaso-vagal attacks associated with parapharyngeal masses that had responded very well to intracranial resection of the glossopharyngeal nerve.

Cicogna et al[24] also discussed the three types of reflex cardiovascular syndromes linked to the glossopharyngeal nerve. These are the carotid sinus syndrome, the glossopharyngeal neuralgia-asystole syndrome, and the parapharyngeal space lesions syncope syndrome.

Further evidence is provided by Sobol et al[28] who reported an interesting case of cardiac syncope syndrome associated with a parapharyngeal abscess which they thought was mediated by the glossopharyngeal nerve pathway. The cardiac abnormalities disappeared when the abscess was drained.

It is therefore important when dealing with neck masses to exclude abnormalities of cardiac rhythm that is brought about by glossopharyngeal afferent-vagal efferent hyperexcitability. When these masses are excised or abscesses drained under general anesthesia, close hemodynamic monitoring of blood pressure and cardiac rhythm is mandatory.

In spite of the wealth of data implicating primary glossopharyngeal neuralgia as a cause of cardiovascular instability, no similar associations are reported for the Eagle's syndrome or other causes of secondary glossopharyngeal neuralgia. Why this is so remains poorly understood.

One possibility could be that the easy diagnosis of Eagle's syndrome and other secondary causes of neuralgia results in quick and expedient treatment, thereby halting the progress of nerve irritability and the development of cardiovascular consequences.

A second possibility is that secondary causes of neuralgia, with the exception of parapharyngeal masses, tend to give rise to less stretching and compression of the nerve.

A third possibility is that the Eagle's syndrome may have been underdiagnosed. The Eagle's syndrome is a well known entity among otolaryngologist. However, neurosurgeons may not be familiar of the existence of this condition, and may not realise that an elongated styloid process can cause glossopharyngeal neuralgia. Failure to identify this more peripheral cause of glossopharyngeal neuralgia may result in subjecting patients to unnecessary neurosurgery when the matter can be dealt with through a safer and simpler intra-oral approach to resection of the styloid process.

DIAGNOSIS

The first priority is to ascertain the diagnosis of neuralgia, and to exclude other causes of pain due to inflammation and neoplasia. The description of the pain will help. Neuralgic pain is severe, episodic, lancinating, and of short duration, which may be associated with intervening periods of a low grade dull ache. In contrast, inflammatory or neoplastic pain is more constant, of longer duration, and has a deep-seated boring quality.

Next, the distribution of the pain has to be mapped out. This is important for two reasons. Firstly, there is a need to know if the neuralgic pain is typically glossopharyngeal, or if it involves other cranial nerves, namely the trigeminal nerve or nervus intermedius. Secondly, if the pain has a typical glossopharyngeal distribution, whether it is predominantly tympanic or oro-pharyngeal in distribution.

It is important to determine the site of any trigger points. Is the trigger point in the oropharyngeal area or is it in the ear? Is the neuralgic pain precipitated by oral activities eg. swallowing, talking, yawning, or is it brought about by hearing activities eg. pain on exposure to loud sounds? Are there any otologic symptoms? Is the neuralgia predominantly tympanic or oro-pharyngeal in its distribution? This is an important point to consider when trying to evaluate the chances of success with low glossopharyngeal nerve section via either the cervical or pharyngeal trans-tonsillar approach[10].

Determine if it is possible to relieve the neuralgic pain by anesthesising the trigger point. Coccaine or lignocaine 10% pledget may be applied to the tonsil or pharynx to see if the neuralgic pain disappears. If the pain is precipitated by certain movements, then see if the pain induced by these movements disappear. Alternatively, if the patient does not have pain at that point in time, but anticipates that he would have it later in the day, the trigger point may be injected with lignocaine 2% or marcaine 0.5% to see if it can avert another attack of pain.

If the symptoms are primarily otologic, inject lignocaine 2% or marcaine 0.5% into the external auditory meatus to see if it abolishes the pain that is present at that time or whether it will avert a subsequent attack.

Lastly, determine if this is an idiopathic type of glossopharyngeal neuralgia, or whether there is a secondary cause. The most important cause of a secondary glossopharyngeal neuralgia is the Eagle's syndrome due to either an elongated styloid process or calcification of the stylohyoid ligament. However, it is important to search for other causes of secondary glossopharyngeal neuralgia in the neck and ENT area, like malignant neck masses and nasopharyngeal carcinomas. Always remember to check the nasopharynx as nasopharyngeal carcinoma is known to be a secondary cause of glossopharyngeal neuralgia[31]. Check for dental causes of neuralgic pain[42].

In the absence of any obvious signs on clinical examination, it is important to exclude intracranial lesions[4] which may cause secondary glossopharyngeal neuralgia. A CT scan or MRI of the brain and temporal bone will detect any cerebellopontine angle tumours that may press upon, stretch and irritate the nerve.

Treatment

Management will depend on whether it is an idiopathic or secondary type of neuralgia. Obviously, if the neuralgia is due to a mass in the cerebellopontine angle or the neck, then it has to be removed for diagnostic purposes and to alleviate pressure on the glossopharyngeal nerve. In the case of a parapharyngeal abscess, drainage will alleviate pain from inflammation, from pressure buildup in the abscess cavity, and from stimulation of the glossopharyngeal nerve.

Cases of idiopathic neuralgia or Eagle's syndrome should be given a trial of anti-convulsant medical therapy with carbamazepine[9,43]. Cardiac abnormalities associated with glossopharyngeal neuralgia will also respond to therapy with carbamazepine[43]. They should be started at low doses and build up until relief of neuralgia is obtained or if side effects such as drowsiness or giddiness start to appear. Some authors feel that the response to medical treatment in glossopharyngeal neuralgia may not be as effective as in the case of trigeminal neuralgia. There is also the question of how long should treatment with carbamazepine continue. No answers are as yet available as studies have not been done to address this issue. Also the benefit from anti-convulsant therapy tends to decrease with time. Recurrences of neuralgia are usually resistant to further medical treatment. Furthermore there is also the risk from long-term anti-convulsant treatment. However, it is generally agreed that a course of medical treatment is still worthwhile.

The next step will be surgical treatment. If an elongated styloid process is present, then resection of the process will give good results. The styloid process can be approached either externally through the neck or through the tonsillar fossa after a preliminary tonsillectomy has been done. The preferred approach is through the tonsil fossa as it will not leave any scars on the neck and is a simple procedure. The parapharyngeal space is entered when the superior constrictors are separated to access the styloid process. The patient should therefore be on prophylactic antibiotics during the procedure to prevent contamination of the parapharyngeal space by intra-oral contents.

If the styloid is not elongated, and the symptoms predominantly oro-pharyngeal in distribution, serious consideration should be given to avulsing the glossopharyngeal nerve low in its course through the neck. The glossopharyngeal nerve can be approached through an external neck incision or the pharyngeal trans-tonsillar approach. The external approach is difficult[44] as the glossopharyngeal nerve is small and lies deep within the neck. The pharyngeal approach as proposed by Wilson and McAlpine[10] is a much simpler approach as the glossopharyngeal nerve can be found just lateral to the superior constrictor muscle which forms the bed of the tonsil fossa. Avulsion of the nerve at the level of the tonsils have been reported to give good results in patients with the oropharyngeal type of glossopharyngeal neuralgia[10,45].

The only caution with the pharyngeal approach is that symptom control is inadequate if the distribution of pain is in the ear. In the tympanic type of neuralgia, the hypersensitivity and irritability of the Jacobson's nerve is a major contributor to symptomatology. The pharyngeal approach to resection of the glossopharyngeal nerve, by itself, is likely to fail. It should be combined with a tympanotomy and avulsion of the nerves of the tympanic plexus to deal with the contribution by the Jacobson's nerve.

Alternatively, the glossopharyngeal nerve has to be divided proximal to where the Jacobson's nerve branch out from the petrous ganglion at the level of the jugular foramen. This would require either a high cervical approach or a retrosigmoid posterior fossa approach to the glossopharyngeal nerve. The high cervical

approach is a hazardous procedure with a high risk of inadvertent damage to the sympathetic chain, vagus and accessory nerves as they exit from the jugular foramen. Adson[46] describes the high cervical approach as a "highly formidable procedure", that he recommends intracranial division of the glossopharyngeal nerve. Thus, in the presence of significant tympanic neuralgia, the posterior cranial fossa approach should be used.

Glossopharyngeal nerve resection through the posterior fossa approach was first used by Dandy[23] in 1927. He reported very good results from the procedure. However, there was a subset of patients who failed to improve with the Dandy procedure. This group responded very well with a second procedure to resect the upper vagal rootlets through the posterior fossa approach. The resection of the upper vagal rootlets was based on the assumption that patients who did not respond well to isolated glossopharyngeal resection probably has a contributory vagal neuralgia. Based on the experience of several authors[8,47,48], it was recommended that the upper vagal rootlets as well as the glossopharyngeal nerve should be divided at the nerve root entry zone in patients with symptoms of glossopharyngeal neuralgia.

Laha and Jannetta[13] in 1977 first reported the technique of microvascular decompression of the glossopharyngeal and upper vagal rootlets for the treatment of glossopharyngeal neuralgia.

The posterior fossa approach enables the surgeon to inspect the blood vessels in the posterior cranial fossa to determine if the neuralgic symptoms are attributable to a vascular compressive effect on the glossopharyngeal nerve[33,36]. Any offending vessel, in particular the posterior inferior cerebellar artery (PICA)[36], can be dealt with by mobilising and separating it away from the glossopharyngeal and upper vagal rootlets. Also, arachnoid adhesions which can be responsible for causing neuralgia can be lysed and divided. Such non-ablative manoevers are effective in relieving glossopharyngeal neuralgia, and is therefore preferable to performing nerve resection and sacrifice[33]. It is only in the event that no identifiable cause of nerve compression is found, that the glossopharyngeal nerve should be resected at the root entry zone. The experience documented in the literature with glossopharyngeal nerve resection has been favourable, with very minimal and imperceptible post-operative neurologic sequelae.

Some authors recommend a combined middle fossa - posterior fossa approach[11,49]. This allows the nervus intermedius, glossopharyngeal and vagus nerves to be accessed via the posterior fossa, while the geniculate ganglion can be reached via the middle fossa. The purpose of performing a geniculate ganglionectomy is that it enables a more complete resection of the sensory fibres of the nervus intermedius, and therefore better resolution of facial pain in cases where the nervus intermedius plays a contributory role. Also, extradural geniculate ganglionectomy, performed through the middle fossa, is facilitated by first doing an intradural dissection of the posterior fossa[49].

In spite of the popularity of microvascular decompression surgery, critics abound who claim that the good result from decompression surgery arises more from nerve damage during surgery and a reduction in nerve irritability, rather than an actual benefit from decompression[50]. Nerve trauma which is induced while dissecting the vessel away from the nerve, result in an effect similar to partial neurectomy, and thus effecting a clinical cure. Several authors claim that vessels are commonly in contact with nerve rootlets even in patients who do not have neuralgic symptoms[14,50,51]. Yet others theorise that vascular contact at the root entry zone is an epiphenomenon created by viral neuritis[15]. They suggest that a preceding event, like inflammation or focal neuritis, causes the formation of arachnoid adhesions between vessel and nerve, resulting in contact.

Despite the criticisms, microvascular decompression of the glossopharyngeal and upper vagal rootlets through the posterior fossa approach still remains a very popular treatment option.

Why Eagle's syndrome should be considered

There is a predominance of literary contributions which deal with the management of glossopharyngeal neuralgia through intracranial procedures or high cervical approaches. Unfortunately, many fail to mention or consider the possibility of Eagle's syndrome as an important cause of neuralgic symptoms.

An elongated styloid process or calcification of the stylohyoid ligament is a fairly common occurrence in normal asymptomatic subjects. It does not always give rise to symptoms. However, when pain in the glossopharyngeal distribution is associated with an elongated styloid process, dramatic results are obtained by performing a stylectomy through the trans-tonsillar approach. This is a very safe and simple procedure that carries very little morbidity.

The Eagle's syndrome is frequently missed because an elongation of the styloid process has not been sought out through careful clinical examination and palpation of the tonsil fossa, or radiological investigations. That being so, many patients are probably being subjected to unnecessary neurosurgery, when all they would require is a simple transtonsillar operation to resect the styloid process or avulse the glossopharyngeal nerve.

Microvascular decompression surgery carries with it a morbidity that is associated with trauma to the important vessels in the posterior fossa. The complex anatomy of the posterior fossa, and the tortuosity of the vessels in the area, predisposes to injury of the PICA, resulting in cerebellar infarction and necrosis. The operation is being performed in the vicinity of the important respiratory and cardiovascular control centers in the brainstem. Close monitoring during anaesthesia is of utmost importance. Deaths have occurred as a result of hemodynamic instability, acute intra-operative hypertension[33], and profound bradycardia and hypotension[52]. Furthermore, the need to open the dura increases the risk of post-operative meningitis. The operation takes several hours, and hospitalisation is prolonged. On the contrary, patients can often be discharged the day after a trans-tonsillar operation. Often the procedure takes only half an hour, and may be performed as day surgery. It is therefore obvious that the risk-benefit profile for trans-tonsillar procedures is far superior to that of an intracranial operation. If an equitable amount of pain relief is achievable using either procedure, then obviously a trans-tonsillar approach is the surgical option of choice.

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