Glossopharyngeal Neuralgia

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Key Words

Glossopharyngeal neuralgia · Theodore Weisenburg · Wilfred Harris · Microvascular decompression

Abstract

Glossopharyngeal neuralgia is a distinctive syndrome, named by Wilfred Harris. Investigation must exclude multiple sclerosis, and local compression, especially by tumours which require treatment. Dandy deserves credit for first indicating vascular compression of cranial nerve roots as a cause of cranial neuralgias, and Jannetta for establishing neurovascular decompression. Vascular compression is a common and treatable cause but does not account for all previously designated idiopathic cases.

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Glossopharyngeal neuralgia is a rare condition, occurring with a frequency of about 1% of that of trigeminal neuralgia. Its reported incidence is approximately 0.8 per 100,000 people.

Historical Note

The first description of glossopharyngeal neuralgia is credited to Theodore H. Weisenburg [1] in 1910. Weisenburg practised in Philadelphia and became editor of *A.M.A. Archives of Neurology and Psychiatry*. His patient

presented with the classical symptoms of lancinating pain in the ear and neck. Not until 6 years later when the patient died was an autopsy performed that revealed the underlying cerebellopontine angle tumour. The tumour compressed the trigeminal nerve and stretched the glossopharyngeal nerve.

Ten years later, Sicard and Robineau [2] described 3 patients who had ‘algie velo-pharyngee essentielle’ (pain in the distribution of the glossopharyngeal nerve of unknown cause). Treatment with sedatives and physical agents failed; the patients became suicidal. Sectioning the glossopharyngeal nerve through the neck was, however, successful in relieving the pain in all 3 patients.

A year later, Wilfred Harris [3] coined the term ‘glossopharyngeal neuralgia’ (fig. 1) describing in 7 cases (3 with malignancy in the tonsillar region) how it ‘resembles trigeminal neuralgia major precisely in the severity and suddenness of onset, in their brevity, and duration ... Repeated coughing or clearing the throat during the pain is suggestive ... paroxysms are started by certain movements, especially swallowing and yawning, eating and talking.’

Harris could cite only two reports, that of Sicard and Robineau, and one of Doyle [4] of the Mayo Clinic (4 cases). At that time, he was unaware of Weisenburg’s work, but in his later book in 1937 [5], he referred to Weisenburg’s report. Harris then advocated avulsion of the nerve and its two ganglia, exposed behind the angle of the jaw, which Sir Geoffrey Jefferson performed successfully (and reported in 1931); this spared the patient an intracranial operation.

Fig. 1. Wilfred Harris' *Neuritis and Neuralgia*, 1926.

Although Weisenburg's priority has been debated [6, 7], his study established glossopharyngeal neuralgia as a clinical entity, distinct from trigeminal neuralgia, with which it was often confused. Two so-called variants have been described: an otitic form with pain mainly deep in or near the ear, and an oropharyngeal form with the pain in the pharynx, tonsil, soft palate, and back of the tongue [8]. Like trigeminal neuralgia, symptomatic relief is obtained in many instances by carbamazepine and other anticonvulsants [9]. Harris first differentiated a symptomatic form secondary to carcinomata and a primary, idiopathic form. He reported attacks of unconsciousness in paroxysms in a young barrister. Rushton in 1981 [8]

reported four instances of syncope in 217 patients at the Mayo Clinic. George Bruyn [6] analysed 304 reported such instances [10], observing: 'a cardiovascular type of the neuralgia, with bradycardia, or asystole and convulsions or coma.'

Anatomy and Pathogenesis

The glossopharyngeal nerve has been called 'the neglected cranial nerve', because it is small, lies deep within the neck and is often unnoticed in surgical dissections. It is the nerve of the third branchial arch. It exits the medulla laterally, just rostral to the vagus nerve. The glossopharyngeal nerve receives sensory fibres from the posterior two thirds of the tongue, including taste, and afferents from the carotid bodies that enter the nucleus of tractus solitarius. Visceral pain passes to the spinal nucleus of V. It supplies parasympathetic fibres to the parotid gland via the otic ganglion. Motor fibres supply the stylopharyngeus muscle (from nucleus ambiguus) and the upper pharyngeal muscles.

Familial cases, and association with multiple sclerosis (MS), trauma and medullary tumours have each been occasionally implicated, as have laryngeal and nasopharyngeal tumours, Paget's disease and diverse tumours of the skull base. Oppenheim [11] described demyelinating plaques at the root entry zone as the potential cause of the closely related trigeminal neuralgia in MS patients. The characteristic finding in MS is demyelination of intrapontine trigeminal fibres, shown in T₂-weighted MRI.

Vascular Compression

Dandy [10], who introduced partial section of the sensory root, first incriminated vascular arterial contact with the dorsal root of the trigeminal nerve in 1929; he said:

'In many instances the nerve is grooved or bent in an angle by the artery. This I believe is the cause of tic douloureux.'

Vascular compression of the nerve root entry zone is thought to cause demyelination and ephaptic transmission. Alternatively, it may cause repetitive activation of primary afferents in the nerve causing hyperexcitability in central neurons. Activation of N-methyl-D-aspartic acid receptor has been invoked as a possible factor. The conventional view in the analogous trigeminal neuralgia is of a peripheral irritative lesion of the nerve causing

pain. A mixed central-peripheral mechanism has recently been postulated in which abnormal impulses derived from demyelinated axons (MS, vascular compression and other possible causes of demyelination along the central and the peripheral course of Gasserian ganglion fibres) modulate the nuclear activity [12].

It was not until the 1950s that Gardner and Miklos [13] and Taarnhøj [14] reported the beneficial effects of decompressing the trigeminal nerve for tic douloureux. This hypothesis was the rationale for microvascular decompression. Similar vascular compression of the seventh cranial nerve in hemifacial spasm was reported in 1947. Despite these seminal observations, microvascular decompression was not an accepted treatment for cranial nerve syndromes until the late 1960s. It is likely that root entry zone lesions of the glossopharyngeal nerve are associated with ephaptic conduction and underlie the pathogenesis of glossopharyngeal neuralgia.

Clinically, it is therefore important to exclude secondary causes: compression or irritation to the glossopharyngeal nerve that may result in neural hyperexcitability and neuralgia. One possible cause is Eagle's syndrome [15] or stylalgia in which the glossopharyngeal nerve is compressed by an elongated styloid process (>25 mm) or calcification of the stylohyoid ligament that occurs, however, in 4–10% of asymptomatic subjects; thickening or ossification of those structures may be more important as an occasional cause of glossopharyngeal pain, often treated by stylectomy.

Treatment

Medical treatment [16] of glossopharyngeal neuralgia with carbamazepine or gabapentin can be effective in suppressing painful paroxysms. Although spontaneous remissions are common, the relapses may become refractory to drug therapy. Surgical methods include nerve section, tractotomy [17] or microvascular decompression [18]. Intracranial root section has been the most often employed and is generally effective but additional section of the upper vagal rootlets is considered necessary in some cases [8].

Microvascular decompression introduced by Jannetta can afford complete relief of pain in 76% and substantial improvement in a further 16% as reported in a large series, with a mean follow-up of 48 months [19]. More recently, endoscopy has been employed as the sole imaging modality in glossopharyngeal nerve decompression. In the analogous trigeminal neuralgia, there is 93% success

3 years after endoscopic surgery [20], but an annual recurrence rate of 3.5% [21].

Sophisticated three-dimensional MRI shows that contact or contiguity between blood vessels and neural tissue contact is not visibly different in the symptomatic and asymptomatic sides [22], thus discounting any consistent causal relationship to pain. However, neural grooving, distortion or deviation are more likely to cause neuralgia. As with trigeminal neuralgia, precisely how pain is caused and relieved is an incompletely resolved question.

Conclusions

Glossopharyngeal neuralgia is a rare but distinctive syndrome, named by Wilfred Harris. It has many causes. Investigation must exclude MS, and local compression, especially by tumours which require treatment in their own right. Dandy deserves credit for first indicating vascular compression of cranial nerve roots as a cause of cranial neuralgias, and Jannetta for establishing neurovascular decompression. Vascular compression is a common and treatable cause but does not account for all previously designated idiopathic cases. The mechanism involves hyperexcitability of the nerve and ephaptic conduction, but is not yet fully understood.

References

- 1 Weisenburg T: Cerebello-pontine tumor diagnosis for six years as tic douloureux: the symptoms of irritation of the ninth and twelfth cranial nerves. *JAMA* 1910;54:1600–1604.
- 2 Sicard R, Robineau J: Algie vélo-pharyngée-essentielle. Traitement chirurgical. *Revue. Neurologique* 1920;36:256–257.
- 3 Harris W: Neuritis and Neuralgia. London, Oxford University Press, 1926, pp 327–335.
- 4 Doyle JB: A study of four cases of glossopharyngeal neuralgia. *Arch Neurol Psychiatry* 1923;9:34–36.
- 5 Harris W: The Facial Neuralgias. London, Oxford University Press, 1937, pp 100–102.
- 6 Bruyn GW: Glossopharyngeal neuralgia. *Cephalalgia* 1983;3:143–157.
- 7 Katusic S, Williams DB, Beard CM, et al: Incidence and clinical features of glossopharyngeal neuralgia, Rochester, Minnesota, 1945–1984. *Neuroepidemiology* 1991;10:266–275.
- 8 Rushton JG, Stevens C, Miller RH: Glossopharyngeal (vagoglossopharyngeal) neuralgia. A study of 217 cases. *Arch Neurol* 1981;38:201–205.
- 9 Ekblom KA, Westerberg CE: Carbamazepine in glossopharyngeal neuralgia. *Arch Neurol* 1966;14:595–596.
- 10 Dandy WE: An operation for the cure of tic douloureux. Partial section of the sensory root at the pons. *Arch Surg* 1929;18:687–734.
- 11 Oppenheim H: Textbook of Nervous Disease. 5th ed. New York, Stechert and Co., 1911.
- 12 Ferroli P, Franzini A, Farina L, La Mantia L, Broggi G: Does the presence of a pontine trigeminal lesion represent an absolute contraindication for microvascular decompression in drug resistant trigeminal neuralgia? *J Neurol Neurosurg Psychiatry* 2002;72:122–123.
- 13 Gardner WJ, Miklos MV: Response of trigeminal neuralgia to ‘decompression’ of the sensory root. Discussion of cause of trigeminal neuralgia. *JAMA* 1959;170:1773–1776.
- 14 Taarnhøj P: Decompression of the trigeminal root and the posterior part of the ganglion as treatment in trigeminal neuralgia. Preliminary communication. *J Neurosurg* 1952;9:288–290.
- 15 Soh KBK: The glossopharyngeal nerve, glossopharyngeal neuralgia and the Eagle’s syndrome – Current concepts and management. *Singapore Med J* 1999;40:659–665.
- 16 Rozen TD: Trigeminal neuralgia and glossopharyngeal neuralgia. *Neurol Clin* 2004;22:185–206.
- 17 Kunc Z: Treatment of essential neuralgia of the 9th nerve by selective tractotomy. *J Neurosurg* 1965;23:494–500.
- 18 Laha RK, Jannetta PJ: Glossopharyngeal neuralgia. *J Neurosurg* 1977;47:316–320.
- 19 Resnick DK, Jannetta PJ, Bissonnette D, et al: Microvascular decompression for glossopharyngeal neuralgia. *Neurosurgery* 1995;36:64–69.
- 20 Kabil MS, Eby JB, Shahinian HK: Endoscopic vascular decompression versus microvascular decompression of the trigeminal nerve. *Minim Invasive Neurosurg* 2005;48:207–212.
- 21 Cheshire WP: Trigeminal neuralgia: diagnosis and treatment. *Curr Neurol Neurosci Rep* 2005;5:79–85.
- 22 Lang E, Naraghi R, Tanrikulu L, et al: Neurovascular relationship at the trigeminal root entry zone in persistent idiopathic facial pain: findings from MRI 3D visualisation. *J Neurol Neurosurg Psychiatry* 2005;76:1506–1509.