

# Expert Opinion

CME

## Glossopharyngeal Neuralgia

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In 1910, Weisenberg<sup>1</sup> described the first symptomatic case of glossopharyngeal neuralgia due to a cerebellopontine angle mass. Harris described the idiopathic type and coined the term “glossopharyngeal neuralgia” in 1921.<sup>2</sup>

### CASE

This 60-year-old woman presented with an 11-year history of severe throat pain recurring approximately every couple of years lasting for about 3 months. The current episode had been present for 5 weeks, the prior episode occurred over 1 year previously. She described an electrical pain with an intensity of 10 of 10 felt in the left throat around the tonsillar area sometimes shooting to the left ear lasting about 5 seconds occurring 30 times or more per day. The paroxysms could be triggered by swallowing cold water, opening her mouth, talking a lot, brushing her teeth, and eating.

She had previously seen 2 other neurologists. She was initially on phenytoin with mild help. Gabapentin, topiramate, and clonazepam were of no benefit. Oxcarbazepine titrated up to 1200 mg twice a day reduced

the number of paroxysms to 3 to 4 per day, but made her feel unsteady and “spaced out.” Baclofen had just been added on a slowly increasing dose without benefit thus far. An MRI scan of the brain and skull base with and without contrast was normal. There was no history of syncope or seizures. Past medical history was negative. Neurological examination was normal.

**Questions.**— What is the diagnosis? Are her triggers typical? What other medications might be of benefit if baclofen did not help? What is the efficacy of the various medication and surgical treatments? What is the natural history?

**Expert Opinion.**—Since the age of 49, the patient has been complaining of periodic pain in the throat. The location, unilaterality and nature of pain, the intensity and duration of attacks, and the presence of specific triggers suggest a form of glossopharyngeal neuralgia (GN), according to the diagnostic criteria of the International Classification of Headache Disorders.<sup>3</sup> In addition, the absence of clinically evident neurological deficits and of neuroimaging alterations indicate a diagnosis of *classical* neuralgia, ruling out secondary neuralgia.

GN is an uncommon disease. According to a study conducted in Rochester, USA, by reviewing health records for the 4 decades between 1945 and 1984, the crude incidence rate of GN is 0.7/100,000/year (0.9 and 0.5 in men and in women, respectively) and disease

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frequency increases slightly with age.<sup>4</sup> GN incidence is 1 of 70 to 1 of 100 that of trigeminal neuralgia.<sup>5</sup> This neuralgia does not seem to have a different distribution in males as opposed to females and it usually occurs after age 50. Data about its seasonal recurrence are conflicting.

The disorder is characterized by clusters of unilateral attacks of sharp, stabbing, and shooting pain localized in the throat radiating to the ear or vice versa. The distribution of pain is diagnostic: the pain shoots from the pharynx, tonsil, and posterior tongue base upwards to the Eustachian tube and inner ear or to the mandibular angle. The paroxysms usually last for seconds to a few minutes. The intervals between the paroxysms range from a few minutes to a few hours and usually the attacks occur during the day. GN may have a periodic course, the mean duration of clusters being about 4 weeks.

The trigger factors reported by the patient are typical. GN paroxysms are usually triggered by swallowing, and the act of taking cold liquids seems especially likely to induce pain. It is the passage of food or of liquid brushing the area rather than the act of swallowing that precipitates the attacks. Other trigger factors often mentioned include the act of sneezing, chewing, coughing, talking, cleaning the throat, and touching the gums or oral mucosa. In some cases, even sudden movements of the head, raising the arm on the side of the pain, and the lateral movement of the jaw precipitated the pain. Finally, several patients found that touching the external auditory canal, the side of the neck, and the skin anterior to the ear triggered the pain on the same side.<sup>6</sup>

As regards the natural history of the disease, we know that GN patients commonly experience a remission of pain, even though clusters have an irregular course. In Rushton et al's case series,<sup>5</sup> comprising 217 patients with GN, about one-fourth of the cases had pain-free periods lasting less than 6 months, while 30% reported they had been pain free for more than 2 years. In this connection, some authors<sup>4</sup> believe that GN is not a severe condition, because (a) the average annual recurrence rate for a second episode is low (3.6%); (b) two-thirds of the cases had only 1 episode; and (c) only one-fourth of the cases had to have surgery for relief of symptoms.

GN may be associated with trigeminal neuralgia, accounting for 8 to 11% of reported cases; like trigeminal neuralgia, GN too, may be significantly associated with MS.<sup>7</sup>

Cases were reported in which GN paroxysms were associated with bradycardia and asystolia, possibly inducing syncopal symptoms or convulsive phenomena.<sup>8</sup> The most acceptable mechanism underlying the association between syncope and GN is the close connection between the vagus nerve and the glossopharyngeal nerve, which may favor the creation of a vago-glossopharyngeal reflex arch. Afferent nerve impulses from irritative or ischemic lesions arising from the glossopharyngeal nerve may reach the nucleus of the tractus solitarius of the midbrain and via collaterals reach the dorsal motor nucleus of the vagus nerve resulting in reflex bradycardia or asystolia. Convulsive movements, limb clonus, automatic smacking movements of the lips, and upward turning of the eyes are signs of cerebral hypoxia induced by the bradycardia. The electroencephalogram recorded during attacks shows slowing of background rhythms (delta activity) or even spike activity.

The pathogenesis of GN is unknown, but probably involves both central and peripheral mechanisms. GN has been attributed to vascular compression of the nerve root entry zone, resulting in demyelination and ephaptic transmission. Alternatively, vascular compression of a nerve has been hypothesized to induce repetitive activation of primary afferents in the nerve and has led to hyperactivity and hyperexcitability of the central neurones. Activation of *N*-methyl-D-aspartic acid receptors has been invoked as another possible explanation.<sup>9</sup>

Both medical and surgical treatments may be used to treat GN. The patient described here tried the medication usually recommended for GN, except for carbamazepine (CBZ), without any substantial benefits. If baclofen did not help, other drugs, such as lamotrigine, valproic acid, and topiramate at the smallest possible pain-relieving doses, could be used that have proved effective in cases of trigeminal neuralgia, even though to date there are no reports in the literature about their use in GN except for a single case report of lamotrigine.<sup>10</sup> CBZ, which is a first-choice medication for GN, is not indicated in this patient

given the therapeutic failure of oxcarbazepine and its high rate of side effects. Finally, for patients refractory to medical therapy, surgery should be considered as a treatment option. The best established surgical treatments are rhizotomy of the glossopharyngeal and upper vagal nerve roots, and microvascular decompression. Intracranial root section has been the most often employed treatment and is generally regarded as curative. In the largest case series reporting the results of treatment<sup>5</sup> and in a smaller series with long-term follow-up,<sup>11</sup> there were no recurrences after preganglionic section of the ninth and upper tenth nerve roots. More recently, microvascular decompression has been employed with complete relief of pain in 76% of the cases and substantial improvement in a further 16% in the largest series.<sup>12</sup> A follow-up study showed that most GN patients treated with microvascular decompression were still free from pain after more than 10 years.<sup>13</sup> Operative methods should be chosen depending on the 2 following circumstances: presence or absence of compression of nerve roots by affected vessels; and the condition of compression. Recently, case reports have been published in which patients benefited from pulsed radiofrequency neurolysis and gamma knife surgery.

*Conflict of Interest:* None Declared

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