

# Concurrent Glossopharyngeal Neuralgia and Hemi-Laryngopharyngeal Spasm (HeLPS): A Case Report and a Review of the Literature

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**BACKGROUND AND IMPORTANCE:** Hemi-laryngopharyngeal spasm (HeLPS) has been recently described but is not yet widely recognized. Patients describe intermittent coughing and choking and can be cured following microvascular decompression of their Xth cranial nerve. This case report and literature review highlight that HeLPS can co-occur with glossopharyngeal neuralgia (GN) and has been previously described (but not recognized) in the neurosurgical literature.

**CLINICAL PRESENTATION:** A patient with GN and additional symptoms compatible with HeLPS is presented. The patient reported left-sided, intermittent, swallow-induced, severe electrical pain radiating from her ear to her throat (GN). She also reported intermittent severe coughing, throat contractions causing a sense of suffocation, and dysphonia (HeLPS). All her symptoms resolved following a left microvascular decompression of a loop of the posterior inferior cerebellar artery that was pulsating against both the IXth and Xth cranial nerves. A review of the senior author's database revealed another patient with this combination of symptoms. An international literature review found 27 patients have been previously described with symptoms of GN and the additional (but not recognized at the time) symptoms of HeLPS.

**CONCLUSION:** This review highlights that patients with symptoms compatible with HeLPS have been reported since 1926 in at least 4 languages. This additional evidence supports the growing recognition that HeLPS is another neurovascular compression syndrome. Patients with HeLPS continue to be misdiagnosed as conversion disorder. The increased recognition of this new medical condition will require neurosurgical treatment and should alleviate the suffering of these patients.

**KEY WORDS:** Cough, Glossopharyngeal neuralgia, Hemi-laryngopharyngeal spasm, Laryngospasm, Microvascular decompression, Neurovascular compression

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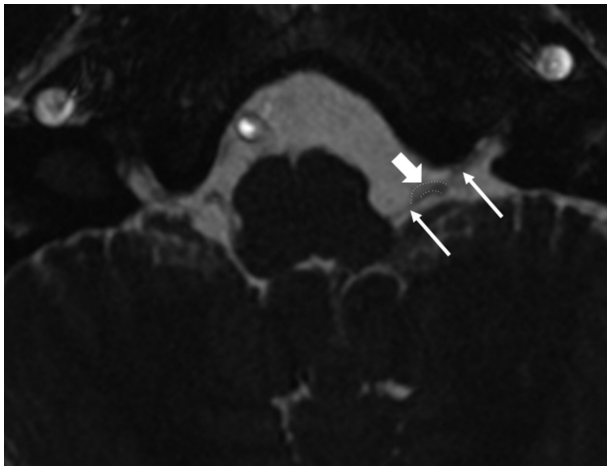
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**G**lossopharyngeal neuralgia (GN) is a well-recognized intermittent, unilateral throat pain that can be due to a vascular compression of the IXth cranial nerve. Hemi-laryngopharyngeal spasm (HeLPS) is a recently described, but not yet well known, surgically curable condition due to unilateral vascular compression of the Xth cranial nerve.<sup>1,2</sup> Symptoms include progressive, intermittent

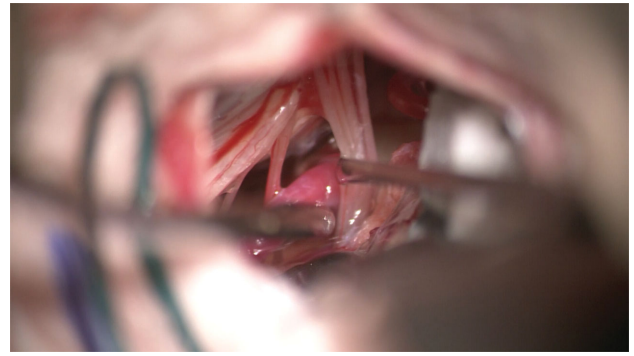
throat contractions and cough without pain. Patients can also report intermittent dysphonia. With a similar etiology and the close proximity of these two cranial nerves, one would expect that these two conditions would occasionally be found concurrently in the same patient.

We present a case report of a patient with concurrent GN and HeLPS and review their symptomology, imaging, intraoperative findings, and outcome following microvascular decompression. We reviewed our neurosurgical database and the international literature on GN and found previously described, but unrecognized, cases of concurrent GN and HeLPS. These cases, described in 4 different languages, reported patients from 8 different countries.

**ABBREVIATIONS:** GN, glossopharyngeal neuralgia; HeLPS, Hemi-laryngopharyngeal spasm; MRI, magnetic resonance imaging



**FIGURE 1.** MRI preoperative imaging. Axial section with constructive interference in steady state sequence (1.5 T MRI) shows the left posterior inferior cerebellar artery (thick arrow pointing to dotted lines) abutting and posteriorly distorting the IXth cranial nerve (thin arrows) as it traverses from the brainstem to the jugular foramen.



**VIDEO.** Intraoperative video microscopy during MVD for the index patient. A loop of the posterior inferior cerebellar artery is pulsating posteriorly against, and distorting, the IXth and Xth cranial nerves.

## CLINICAL PRESENTATION

Presentation of this case and the data obtained from our reviews followed approval from our institution's Clinical Research Ethics Board (H17-00830) and consent from the patient. A 48-yr-old woman presented with a 5-yr history of left-sided GN with intense, sharp, and sometimes electric-like pain radiating from deep to her left ear down the left side of her throat. The symptoms were intermittent and could be triggered by swallowing, laughing, talking, and anything that increased her heart rate. She had spent the previous year sleeping on the ground floor of her home for fear of triggering the pain by the exertion of climbing the stairs to her bedroom. She also presented with symptoms compatible with HeLPS. She described intermittent coughing and choking. The coughing was triggered by a tickling sensation deep to her suprasternal notch and was severe enough to cause vomiting and bladder incontinence. The choking was described as a circumferential tightness of her throat that caused a sensation of suffocation. It was triggered by loud, prolonged talking, or drinking a cold liquid. It could also follow a coughing episode. She described an additional symptom: anything that increased her heart rate could cause dysphonia (hoarseness of her voice). A trial of carbamazepine incompletely reduced her neuralgia but had no effect on her HeLPS. The medication was discontinued because of rash. A trial with gabapentin was discontinued because of lethargy. Neurological examination between these episodes was normal. A magnetic resonance imaging (MRI) demonstrated her left posterior inferior cerebellar artery (PICA) was curving into and posteriorly distorting her left lower cranial nerves (Figure 1).

A left microvascular decompression was performed (Video). The left glossopharyngeal nerve and vagus nerve were distorted posteriorly by a loop of the PICA. This vessel was surgically displaced anteroinferiorly and maintained in position with Teflon between the vessel and brainstem.

Following the surgery, she had a 3-wk exacerbation of her GN, which then settled and completely resolved by 6 wk postoperative. She has not had further GN during the last 2 yr. Her hemi-laryngopharyngeal symptoms also improved but at a different rate. There was no improvement until approximately 3 mo postoperatively. She then noticed a slow, but continuous, improvement with complete resolution of her coughing, choking, and voice changes with exercise by 1-yr postoperative. At the 2-yr follow-up, she had no symptoms of GN or HeLPS.

## Database Review

The senior author's neurosurgical database was reviewed for patients with a diagnosis of GN. All patients were contacted and asked if they had any additional symptoms potentially related to concurrent HeLPS, such as coughing, choking, and/or dysphonia. Fourteen patients were identified. Eight were female, and the average age at surgery was 56 yr (range 41-81). The average postoperative follow-up was 7 yr (range 1-12 yr). One additional patient described symptoms compatible with HeLPS (intermittent coughing and choking) that completely resolved following microvascular decompression.

## Literature Search

A PubMed literature search for "glossopharyngeal neuralgia" identified 1,162 articles. Abstracts were screened for articles describing the patients' symptoms in either English, German, Spanish, Russian, Polish, or Dutch. Cases secondary to neoplasia, review papers, and papers not describing the symptoms were excluded. One hundred and sixty-three articles were identified. Each article was then reviewed for the description of the patients' symptoms. Patients with GN and the additional

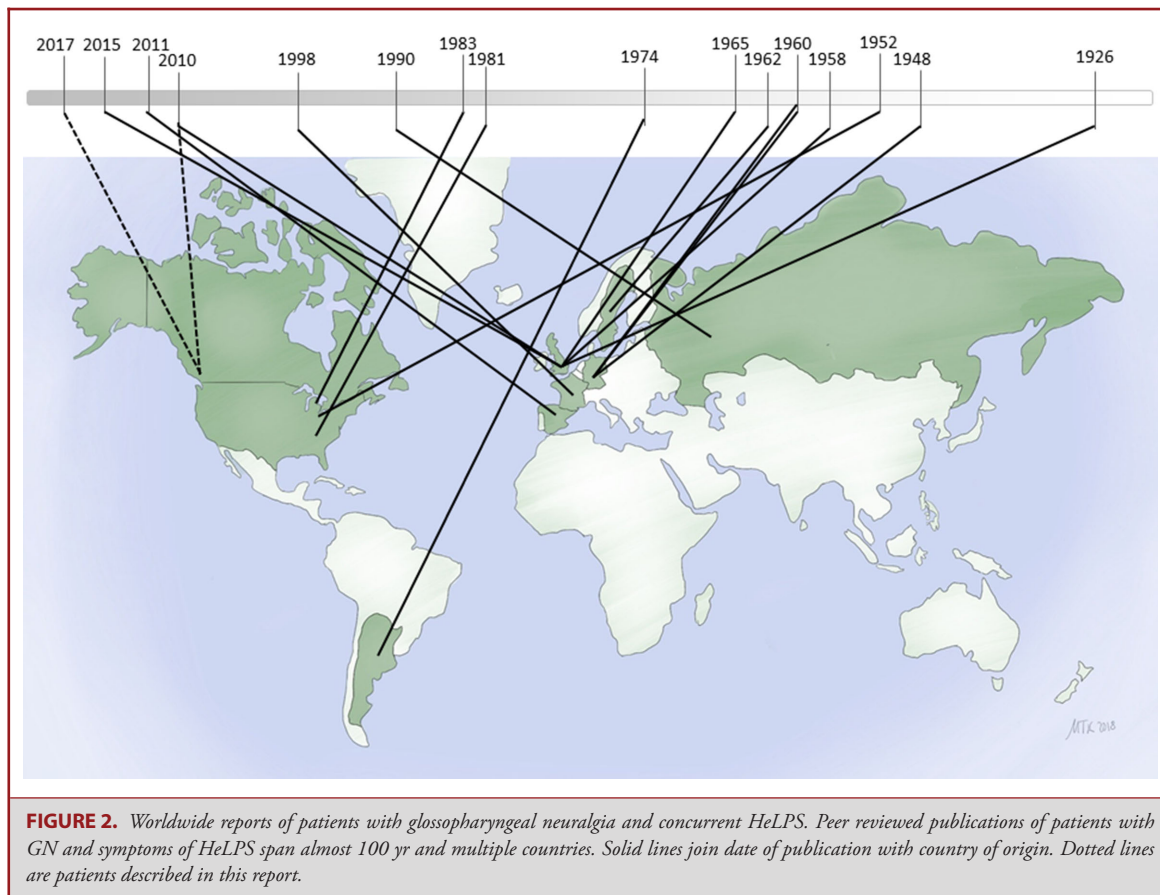
**TABLE. Previously Published Descriptions of Patients With Glossopharyngeal Neuralgia and Additional Symptoms Compatible With HeLPS**

No.	Year	Country (language)	Gender, age number in series	GN	Additional symptoms	Treatment and outcome	Reference
1	1926	England (English)	N/A	Yes	Coughing	No surgical treatment	Harris <sup>6</sup>
2	1948	Germany (German)	F, 70 yr	Left	Coughing	Section of IX GN and coughing resolved	Okonek <sup>7</sup>
3	1952	United States (English)	F, 48 yr	Left	Coughing	Minimal pain relief after cutting IX Later, complete pain relief after cutting upper two rootlets of X. No mention of coughing outcome.	Stowell <sup>8</sup>
4	1958	England (English)	M, 26 yr	Right	Coughing (Tingling and taste disturbance)	No surgical treatment	Laskiewicz <sup>9</sup>
5	1960	Germany (German)	M, 41 yr	Right	Coughing (Taste disturbance)	Medical treatment symptoms improved	Jankoff <sup>10</sup>
6	1960	Germany (German)	M, 70 yr	Yes	Coughing (Tearing)	Section of IX pain relief but no mention of coughing outcome	Usbeck <sup>11</sup>
7	1962	Sweden (English)	2/18	Both left	(1) Coughing and hoarseness (2) Coughing and throat irritation	(1) No details of surgery (2) Pain remained after section of IX; resolution after cutting upper X	Bohm <sup>12</sup>
8	1965	England (English)	F, 65 yr	Left	Coughing	"Tympanic neurectomy" Pain relief but no mention about cough outcome	Kersley <sup>13</sup>
9	1974	Argentina (Spanish)	N/A	Yes	Coughing	Medical treatment Resolution of pain and cough	Borello <sup>14</sup>
10	1981	United States (English)	21/217	all	Coughing (18) Stridor (1) Hoarseness (2) (Syncope (4))	Surgery for 129 of the 217 patients but no outcome reported for the patients with HeLPS	Rushton <sup>15</sup>
11	1983	United States (English)	M, 65 y	Left	Tightness in throat with stridor (Mild syncope)	Section of IX and 3 rootlets of X Relieve of "all" symptoms but no specific mention about stridor	Marshall <sup>16</sup>
12	1990	Russia (Russian)	N/A	Yes	Coughing	Unclear (MVD?) Outcome not stated	Puzin <sup>17</sup>
13	1998	France (English)	F, 60 y 1/2	Left	Coughing	MVD of IX Relieve of pain but no mention about cough	Boch <sup>18</sup>
14	2010	England (English)	2/21	Yes	1) Coughing with syncope 2) Coughing and TN	1) MVD (IX and X) 2) MVD V, IX and X Outcomes unclear	Kandan <sup>19</sup>
15	2010	United States (English)	F, 47 y	Left	Choking	Gamma knife of IX No mention of choking outcome	Williams <sup>20</sup>
16	2011	Spain (English)	1/14	Yes	Aphonia	MVD or rhizotomy No mention of outcome	Martínez-González <sup>3</sup>
17	2015	England (English)	F, 37 y	Left	Coughing	Rhizotomy IX and upper X, then MVD IX and X after recurrence No mention of coughing outcome	Price <sup>21</sup>

hemi-laryngopharyngeal symptoms of intermittent coughing, choking, or vocal changes were identified and described (Table). Seventeen articles described 27 patients with one or more of these additional symptoms compatible with HeLPS. These articles were in 4 different languages and from 8 different countries (Figure 2). A few of these reports also documented that the symptoms of HeLPS resolved following the surgical treatment for GN.

## DISCUSSION

The coexistence of symptoms caused by pathology of adjacent cranial nerves is rare but well described. Patients can present with simultaneous trigeminal neuralgia and hemifacial spasm or concurrent hemifacial spasm and GN or trigeminal neuralgia and GN.<sup>3-5</sup> It should, therefore, not be surprising to find patients who



report symptoms related to compression of their IXth (GN) and Xth (HeLPS) cranial nerves.<sup>6-21</sup>

Although the first descriptions of patients with HeLPS were only recently reported,<sup>1,2</sup> there must have been patients with this condition since antiquity. We postulate that the patients described in Table had HeLPS (as well as GN). We can, therefore, trace the earliest descriptions of HeLPS back to 1926. In fact, it has been described in 4 different languages and from multiple countries.

A common cause of GN is a vascular compression of the IXth cranial nerve and, occasionally, the upper rootlets of the Xth cranial nerve. The proposed pathophysiology of HeLPS is also a vascular compression of the Xth cranial nerve because microvascular decompression of that nerve has cured the symptoms.<sup>1,2</sup> A vessel compressing the IXth cranial nerve is more likely to compress the rostral than the caudal rootlets of the adjacent Xth cranial nerve. This may explain why the associated symptoms are far more likely to be coughing than choking. Perhaps the sensory fibers of the Xth cranial nerve, which trigger a tickling sensation deep to the suprasternal notch that can lead to coughing, are located more rostrally in the nerve. Another possible explanation for the higher association of coughing than choking (both symptoms of HeLPS) with GN may be that the sensory fibers are more susceptible to compression than the motor fibers.

Our index case had a common cause for both their GN and HeLPS: a vascular compression of the respective cranial nerves. The different time course of symptom resolution of GN and HeLPS in this case may have been due to different degrees of neuronal demyelination or healing capacities of the nerves.

### Limitations

There are limitations of this work. The review of our database found 1 patient (in addition to the index case) who retrospectively endorsed symptoms of coughing and throat contractions during a structured phone interview. Their medical records did not mention these symptoms and were either not mentioned or not documented because of assumed irrelevance to their painful neuralgia. Our literature review may have underestimated the co-occurrence of these conditions, because the authors were focusing on GN and may not have asked about or reported any symptoms compatible with HeLPS. A better estimate of the incidence of concurrent GN and HeLPS would require a prospective database.

### CONCLUSION

HeLPS has been recently described, but the condition is not yet widely recognized, and patients continue to be misdiagnosed as

psychogenic.<sup>1,2</sup> This report highlights that the etiology of HeLPS is likely the same as GN—a neurovascular compression—because both conditions respond to the same microvascular decompression. The literature review also highlights that HeLPS is not new. Its symptoms have been described in multiple countries (but not recognized) for almost a century.

## Disclosures

The authors have no personal, financial, or institutional interest in any of the drugs, materials, or devices described in this article.

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## COMMENT

I applaud the authors for this contribution. Because I have never encountered a patient with hemi-laryngopharyngeal spasm, however, I will restrict my comments to the diagnosis and management of glossopharyngeal neuralgia.

This patient's fear of "climbing the stairs" to avoid triggering an exacerbation of glossopharyngeal neuralgia symptoms is unusual. More importantly, in Figure 1, the neurovascular compression (NVC) of the posterior inferior cerebellar artery and glossopharyngeal nerve is far distal to the centrally myelinated portion of the glossopharyngeal nerve. In my experience, the NVC of glossopharyngeal neuralgia is always along the first few millimeters of the glossopharyngeal and vagus nerves. Finally, patients with glossopharyngeal neuralgia who undergo an appropriate decompression of the glossopharyngeal and vagus nerves, respectively, awaken with immediate and complete cessation of symptoms. It is as if one has turned off a light switch. Unlike trigeminal neuralgia, recurrences of glossopharyngeal symptoms following microvascular decompression are distinctly uncommon. This patient required three weeks to become pain free. This is also unusual.

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