

THE GLOSSOPHARYNGEAL NERVE (IX CRANIAL NERVE PAIR): ANATOMICAL CONSIDERATIONS AND NEURALGIA - LITERATURE REVIEW

O NERVO GLOSSOFARINGEO (IX PAR DE NERVO CRANIANO): CONSIDERAÇÕES ANATÔMICAS E NEURALGIA - REVISÃO DA LITERATURA

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ABSTRACT

The glossopharyngeal nerve (IX cranial nerve) is a mixed nerve, with both motor and sensory function. This relates to the tongue and pharynx. Glossopharyngeal neuralgia is a rare nervous neuropathy, with paroxysmal, lancinating and paritary crises, usually unilateral. The aim of the study was to review the literature on glossopharyngeal neuralgia of the nerve (IX cranial nerve), highlighting the anatomical aspects of this nerve and the possible causes and complications of neuralgia as well as forms of treatment. A literature review was carried out in the international Pubmed database. The literature review included 72 articles from 2015 to 2021. The keywords used were: "anatomy of glossopharyngeal neuralgia". Of the 72 articles, 7 were used for this literature review. Uncommon as nervous/glossopharyngeal etiologies and pathologies are neurological abnormalities/neurovascular and pathologies are neurovascular/neurovascular lesions. Pharmacological treatment approaches mentioned in the literature were therapy with antiepileptics and antidepressants such as carbamazepine and gabapentin; a microvascular decompression; and gamma knife radiosurgery.

UNITERMS: Glossopharyngeal nerve. Anatomy. Neuralgia.

INTRODUCTION

Glossopharyngeal neuralgia is a rare neuropathy, being reported about 12 cases per 100,000 individuals, and it is characterized by painful, lancinating and paroxysmal crises, usually unilateral. It was described by the International Headache Society (IH) as an acute, transient, and severe pain that occurs at the base of the tongue, tonsillar fossa or below the angle of the mandible. Pain episodes last from several seconds to minutes, with intervals between paroxysms lasting from minutes to hours. The intervals between attacks are irregular, but can vary from days to years, with patients being asymptomatic between episodes. Patients often have difficulty in identifying pain triggers, probably because it involves the deep structures of the mouth, pharynx, and ear. But

swallowing often triggers the paroxysms of pain, although chewing, yawning, and talking have also been implicated. Most cases of glossopharyngeal neuralgia are idiopathic and are usually caused by compression of the nerve, so the possible causes are compression by tumors or abscesses, compression by the elongated styloid process (Eagle's syndrome), compression by the stylohyoid ligament or calcified stylomandibular ligament, among other possible causes¹.

Neuralgia associated with cardiac syncope is even rarer and may then be called vagoglossopharyngeal neuralgia. In these cases, neuralgia may be associated with episodes of bradycardia, severe hypotension, and even cardiac arrest leading to syncope².

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Although certain surgical treatments have shown more permanent benefits to the patient, surgery is still reserved for those with medically refractory neuralgia. Neuralgia usually responds, at least initially, to pharmacotherapy, especially antiepileptics. It has been suggested that application of local anesthetics to the pharyngeal wall and tonsil may prevent attacks for a few hours¹.

The diagnosis of glossopharyngeal neuralgia is strictly clinical. The International Classification of Headache Disorders has developed diagnostic criteria that include the following: A: At least three episodes of unilateral pain, fulfilling criteria B and C; B: Pain located in the back of the tongue, tonsil fossa, pharynx, near the angle of the mandible and/or in the ear; C: Pain that has at least three of the following characteristics: pain recurring in paroxysmal attacks, lasting from seconds to 2 minutes; severe pain; lurching, stabbing, cutting pain; pain precipitated by swallowing, coughing, talking or yawning; D: No obvious clinical neurological deficits; E: Not better explained by another diagnosis of ICDH-3 beta³.

It has been recommended that all patients with glossopharyngeal neuralgia be referred to an otolaryngologist to investigate a possible structural cause. Brainstem imaging to identify tumors, or possible vascular compression, is recommended. Also, a high-resolution CT scan of the neck can identify an elongated styloid process. Although it is a rare condition, there are multiple etiologies that lead to the symptoms of glossopharyngeal neuralgia. Many cases are caused by variations in anatomy, which may be from the neurovascular structures that surround the glossopharyngeal nerve, the elongated styloid process (Eagle's syndrome), the stylohyoid ligament or a calcified stylomandibular ligament. Thus, anatomico-clinical knowledge is essential since glossopharyngeal neuralgia is caused by multiple etiologies and the diagnosis is strictly clinical¹.

Thus, the aim of the present study was to carry out a literature review on glossopharyngeal neuralgia (IX cranial nerve), highlighting the anatomical aspects of this nerve and the possible causes and complications of neuralgia as well as forms of treatment.

METHOD

A literature review was carried out in the international Pubmed database. The literature review included 79 articles from 2015 to 2022. The keywords used were: "glossopharyngeal neuralgia anatomy". The articles were searched on August 1st, 2022.

Of the 79 articles, 12 were eligible for this literature review. Among the 12 articles that were selected, 2 did not present the available full text. Thus, 10 articles were used for this literature review.

All types of articles were considered (4 literature reviews, 4 case reports and 2 original articles).

LITERATURE REVIEW

GLOSSOPHARYNGEAL NERVE ANATOMY (IX CRANIAL NERVE PAIR):

The glossopharyngeal nerve provides general sensory innervation to the oropharynx and special sensory innervation (taste) to the tongue base. Its path takes place laterally from the medulla to the jugular foramen⁴.

In general, the articles evaluated did not describe purely the anatomy of the glossopharyngeal nerve. Some of the works used anatomy to explain the neuralgia.

Saccomanno et al.⁵ described the case of a 60-year-old woman who presented with a severe unilateral trigeminal and glossopharyngeal neuralgia due to Eagle's syndrome and to explain the case related, the authors discussed about the craniofacial pain resembles glossopharyngeal neuralgia and it is secondary to the irritation of the surrounding neurovascular and muscular anatomical structures (carotid artery, cranial nerves, and muscles).

Shereen et al.¹ illustrated about the branches of the glossopharyngeal nerve to explain about the neuralgia. The authors related that the pain can essentially travel along any of the sensory segments for these branches. They described about the proximity of the styloid process as well as the internal carotid (branches have been removed for visualization of the glossopharyngeal nerve) which reflects the possibility of these structures compressing the nerve and causing the symptoms of glossopharyngeal nerve neuralgia.

Krugger et al.⁶ characterized the anatomy of the vagus nerve by evaluating intraoperative electrophysiological data to differentiate between motor and sensory fibers of the vagus nerve rootlets and define a surgical strategy to reduce the risk of undesired side effects for patients requiring rhizotomy for glossopharyngeal nerve neuralgia. Then, they described anatomical and electrophysiological findings on the vagus and glossopharyngeal nerves.

ETIOLOGIES OF GLOSSOPHARYNGEAL NERVE NEURALGIA:

Saccomanno et al.⁵ related that the Eagle's syndrome resembles glossopharyngeal neuralgia, and it is secondary to the irritation of the surrounding neurovascular and muscular anatomical structures (carotid artery, cranial nerves, and muscles). The authors explained that the pain deriving from the elongated styloid process may be due to compression of the glossopharyngeal nerve as it passes over the superior constrictor muscle.

Shereen et al.¹ reported that there are multiple etiologies that lead to the symptoms of glossopharyngeal neuralgia and Eagle's syndrome, such as anatomic variation of the neurovascular structures around the glossopharyngeal nerve, elongated styloid process and calcified stylohyoid or stylomandibular ligaments. Essentially, anything that

compresses or damages CN IX and/or its branches can cause symptoms of glossopharyngeal neuralgia. Often, the cause is unknown. According to Sherren et al.¹, of the known causes, the most common include neurovascular compression/vascular variations and pathologies (i.e., vertebral artery dissection), tumors, infection, infarction, and trauma.

Nagata et al.² reported a case of glossopharyngeal neuralgia with repeated syncope caused by the recurrence of esophageal carcinoma.

Li and Zhang⁷, Szmyd et al.⁸ and Lerner et al.⁹ attributed the neuralgia to neurovascular compression of the cranial nerve IX. When performing Magnetic Resonance in these cases, displacement and atrophy of the glossopharyngeal nerve caused by an adjacent arterial or venous structure can be found^{4,9}.

Kim and Lee¹⁰ reported a case of glossopharyngeal neuralgia caused by a posterior condylar canal dural arteriovenous fistula.

TREATMENT OF GLOSSOPHARYNGEAL NERVE NEURALGIA:

Glossopharyngeal neuralgia may generate tongue, throat, and floor of the mouth pain⁹. Also, glossopharyngeal disorders can provoke dental pain¹¹. Patients that present this condition may look for evaluation and treatment by their dentist¹¹.

Saccomanno et al.⁵ described the case of a 60-year-old woman who presented with a severe unilateral trigeminal and glossopharyngeal neuralgia due to Eagle's syndrome. The woman was subjected to conservative therapy for four months, consisting of 8% lidocaine spray on the oropharyngeal region corresponding to the tonsillar branches of the glossopharyngeal nerve, corticosteroids and NSAIDs per os. Although, the authors relate that it did not cause any symptoms relief. The patient's symptoms were intensifying and were not responding to painkillers. Then she was urgently subjected to surgical treatment, which was the surgical resection of the left stylomastoid process, via a transcervical approach and under general anesthesia. The procedure was effective, as it was observed sudden remission of symptoms even without using any anti-inflammatory drugs. Saccomanno et al.⁵ concluded that the surgical approach in Eagle's syndrome patient's is unquestionable such surgical styloidectomy shows a cure rate of 80%.

A literature review performed by Shereen et al.¹ found 7 articles with respect to pediatric glossopharyngeal neuralgia. The authors verified that this condition has different etiologies. Then, it was concluded that the treatment's approach choice is dependent on clinical judgment. Among the options found, the following can be mentioned: pharmacological therapy with antiepileptics and antidepressants, as carbamazepine and gabapentin; microvascular decompression; gamma knife radiosurgery. When treating Eagle's syndrome, the

surgical approach is more directed to the removal of the styloid process¹.

Nagata et al.² reported a case of a 78-year-old Japanese man that presented repeated syncopal attacks that might have been induced by a glossopharyngeal neuralgia caused by the recurrence of esophageal carcinoma. The authors claim that performing a surgical resection was not indicated because it was too risky due to the location of the tumor in the present case. Treatment with sympathomimetic drug was not effective, but the reduction of the tumor size could help on relieving symptoms. Therefore, concurrent chemoradiation therapy (cCRT) was performed, the tumor regressed and the episodes of glossopharyngeal neuralgia with syncope were totally solved. According to Nagata et al.², carbamazepine is a proposed drug for cases of glossopharyngeal neuralgia, but patients can become tolerant to it after prolonged use and thus the treatment is ineffective. Also, this drug may provoke some adverse effects, such as eruption, drowsiness, dizziness, nausea, and vomiting. Neurosurgical treatment of glossopharyngeal neuralgia is performed through microvascular decompression or nerve rhizotomy⁶.

A study performed by Krüger et al.⁶ aimed to define a surgical strategy to attenuate the occurrence of side effects for patients requiring rhizotomy for glossopharyngeal neuralgia. Twelve patients who underwent posterior fossa craniotomy with intraoperative electrophysiological monitoring of the vagus nerve rootlets were included in this study, and 7 of them had glossopharyngeal neuralgia. The surgical approach was defined by the pattern of vagus nerve the patients presented (Type A or Type B). If the patient showed Type A pattern (pure sensory innervation of the rostral rootlet), it was possible to perform decompression or section of those rootlets without any undesired complication. If the patient showed Type B pattern (motor innervation in the rostral vagal rootlet), that rootlet was decompressed but no vagus nerve rootlets were sacrificed, avoiding a motor complication due to the procedure. According to the authors, none of the patients with glossopharyngeal neuralgia developed any permanent neurological deficits with this surgical strategy⁶.

Boeddinghaus and Whyte⁴ stated that treatment of glossopharyngeal neuralgia can happen through ablative procedures and neurosurgical microvascular decompression.

Li and Zhang⁷ reported a case of a 57-year-old woman who was diagnosed with glossopharyngeal neuralgia due to neurovascular compression of this cranial nerve. For symptom relief, oxcarbazepine, pregabalin, and mecobalamin were tried but it was insufficient. Then, the authors performed microvascular decompression and patient related complete disappearance of symptoms immediately after the procedure.

Kim and Lee¹⁰ treated the glossopharyngeal neuralgia caused by a posterior condylar canal dural arteriovenous fistula using transvenous coil embolization. The authors reported that after the procedure, the patient's symptoms were totally resolved.

CONCLUSION

In conclusion, according to the literature reviewed, the glossopharyngeal nerve neuralgia is unusual, and the most common etiologies are neurovascular compression/vascular variations and pathologies and trauma. The treatment's approach mentioned area pharmacological therapy with antiepileptics and antidepressants, as carbamazepine and gabapentin; microvascular decompression; gamma knife radiosurgery.

RESUMO

O nervo glossofaríngeo (IX par de nervo craniano) é um nervo misto, contendo função tanto motora como sensitiva. Este nervo relaciona-se com a língua e com a faringe. A neuralgia do nervo glossofaríngeo é uma neuropatia rara, sendo caracterizada por crises dolorosas, lancinantes e paroxísticas, geralmente unilaterais. O objetivo do estudo foi realizar uma revisão de literatura sobre a neuralgia do nervo glossofaríngeo (IX par de nervo craniano), destacando os aspectos anatômicos deste nervo e as possíveis causas e complicações da neuralgia bem como formas de tratamento. Foi realizada uma revisão da literatura na base de dados internacional Pubmed. A revisão da literatura incluiu 72 artigos no período de 2015 a 2021. As palavras-chave utilizadas foram: "anatomia da neuralgia do glossofaríngeo". Dos 72 artigos, 7 foram utilizados para esta revisão de literatura. Verificou-se que a neuralgia do nervo glossofaríngeo é incomum e as etiologias mais encontradas foram compressão neurovascular/variações vasculares, patologias e traumas. As abordagens dos tratamentos mencionadas na literatura foram a terapia farmacológica da área com antiepilépticos e antidepressivos, como carbamazepina e gabapentina; a descompressão microvascular; e radiocirurgia com faca gama.

UNITERMOS: Nervo glossofaríngeo. Anatomia. Neuralgia.

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