Microvascular decompression for glossopharyngeal neuralgia through a microasterional approach: A case series

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Abstract

Background: Glossopharyngeal neuralgia (GPN) is an uncommon craniofacial pain syndrome. It is characterized by a sudden onset lancinating pain usually localized in the sensory distribution of the IX cranial nerve associated with excessive vagal outflow, which leads to bradycardia, hypotension, syncope, or cardiac arrest. This study aims to review our surgical experience performing microvascular decompression (MVD) in patients with GPN.

Methods: Over the last 20 years, 14 consecutive cases were diagnosed with GPN. MVD using a microasterional approach was performed in all patients. Demographic data, clinical presentation, surgical findings, clinical outcome, complications, and long-term follow-up were reviewed.

Results: The median age of onset was 58.7 years. The mean time from onset of symptoms to treatment was 8.8 years. Glossopharyngeal and vagus nerve compression was from the posterior inferior cerebellar artery in eleven cases (78.5%), vertebral artery in two cases (14.2%), and choroid plexus in one case (7.1%). Postoperative mean follow-up was 26 months (3–180 months). Pain analysis demonstrated long-term pain improvement of 114 ± 27.1 months and pain remission in 13 patients (92.9%) (P = 0.0001) two complications were documented, one patient had a cerebrospinal fluid leak, and another had bacterial meningitis. There was no surgical mortality.

Conclusions: GPN is a rare entity, and secondary causes should be discarded. MVD through a retractorless microasterional approach is a safe and effective technique. Our series demonstrated an excellent clinical outcome with pain remission in 92.9%.

Key Words: Glossopharyngeal nerve, microvascular decompression, neuralgia, neurovascular compression, vagus nerve
INTRODUCTION

Glossopharyngeal neuralgia (GPN) is an uncommon craniofacial pain syndrome, representing 0.2–1.3% of facial pain syndromes, with an annual incidence of 0.7 cases per 100,000 inhabitants per year according to a population-based study. It is characterized by a sudden onset of lancinating acute pain, lasting seconds to minutes, usually in the sensory distribution of the auricular and pharyngeal branches of the IX and X cranial nerve. The pain is felt in the pharynx, tongue, tonsillar fossa, internal ear, and mandible angle. In some cases, it is associated with excessive vagal outflow; which leads to bradycardia, hypotension, syncope, or cardiac arrest.

The first GPN description is attributed to Theodore H. Weisenburg in 1910. Dandy elucidated the pathophysiology of trigeminal neuralgia and proposed vascular compression as the main etiology, causing demyelization, and ephaptic transmission; which is the same pathophysiology of GPN. First line medical treatment, including carbamazepine and gabapentin, may sometimes improve pain paroxysms. However, in cases with refractory GPN various surgical approaches have been attempted. In 1920, Sicard and Robineau proposed sectioning the glossopharyngeal nerve through the neck as a definitive treatment; which evolved to intracranial rhizotomy of the glossopharyngeal nerve performed by Dandy. Later on, Sweet introduced percutaneous compression at the middle fossa and finally Jannetta, popularized microvascular decompression (MVD) as a definitive surgical treatment for this pathology. MVD series have reported good outcomes in 90–98%, long-term pain improvement have been observed in 64% with a low mortality ranging from 0% to 5.8%.

This study aims to review our surgical experience performing MVD using a microasterional approach in patients with GPN.

METHODS

Patients

This study is a consecutive case series of 14 patients, who underwent MVD for the treatment of idiopathic GPN at the National Institute of Neurology and Neurosurgery “Manuel Velasco Suárez”, in Mexico City, between 1994 and 2014. The senior author (Rogelio Revuelta-Gutiérrez) performed all the surgeries. A retrospective analysis of the clinical charts was performed. Patient data including gender, the age of onset, symptoms, previous medical management, operative findings, complications, and clinical outcome were collected. Pain intensity was graded according a three-grade scale: (1) No pain, no need for medication; (2) pain controlled with medical management; (3) pain not controlled with medication. All patients were previously managed with conservative treatment including carbamazepine, gabapentin, and pregabalin. No pain improvement for at least 6 months before surgical procedure was documented. Diagnosis work-up included a 3T magnetic resonance imaging (MRI). T1, T2, gadolinium-enhanced and FIESTA sequences were assessed to discard a secondary cause of the symptoms and identify vascular compression.

A statistical analysis was performed using SPSS Version 20 (IBM SPSS Statistics, New York, USA). Categorical variables were expressed as proportions and continuous variables were expressed using means and standard deviations. Clinical outcome was evaluated according to the surgical management, use of medications, pain recurrence, and postoperative complications. Descriptive statistics was performed for the patient data and the grade of pain preoperatively and postoperatively was analyzed using Wilcoxon signed-rank test. P < 0.05 was considered statistically significant.

Surgical technique

Under general anesthesia patients were placed in park bench position with the head fixed in a Mayfield skull clamp. The upper shoulder was retracted, and the head was rotated 60° to the opposite side of the exposure with slight cervical lateral tilting (10°) toward the floor. A 5 cm retrosigmoid incision centered over the asterion was performed and a keyhole (2.5–3 cm) asterional craniectomy exposed the angle of the transverse and sigmoid sinuses [Figures 1 and 2a]. Curvilinear durotomy was performed under microscope magnification and intradural dissection started toward the dural angle between the tentorium and petrous surface [Figure 2b]. Cerebrospinal fluid (CSF) was released through arachnoid dissection without using cerebellar retractors. The dissection was directed caudally, and the lower vascular nervous complex involving the glossopharyngeal...
nerve was exposed, identifying its exit through the jugular foramen. Once the identification of the vascular element compressing the glossopharyngeal nerve was observed [Figure 2c], blunt dissection was done, and a small piece or multiple pieces of Teflon were placed between the glossopharyngeal nerve and the compressing vessels (arterial or venous) [Figure 2d, Video 1].

RESULTS

Patient demographics
A total of 14 patients were diagnosed with GPN and were surgically treated [Table 1]. The median age of onset was 58.7 ± 11 years, with a male to female ratio of (1:1.8). The mean time duration from symptom onset to surgery was 8.8 years. Pain trigger was described when swallowing in seven cases, talking in four cases and without previous stimuli in three cases. Carbamazepine was the most used medication (78%), followed by gabapentin and pregabalin; 64.2% patients were on more than one drug. All patients from this study had no clinical improvement with full dose carbamazepine, gabapentin, pregabalin, and daily analgesic medication. Three patients were misdiagnosed before they were referred to our institution; stiloidectomy was performed in two patients (14.3%) and previous dental surgery in one patient (7.1%). Mean time from diagnosis to surgery was 106.3 ± 95.7 months (males 86.4 ± 78.4 months and females 117.3 ± 106.9 months; \(P = 0.58\)).

The pain was more common on the left side (78.6%) compared to the right (21.4%). The primary location of the pain was pharyngeal in 13 cases (92.9%) and preauricular in one case (7.1%). Pain irradiation was referred in 6 cases (42.9%), 5 of them to the preauricular area and one to the pharynx. One patient (7.1%) presented with syncope and another one had an intraoperative vasovagal reflex during decompression.

Neuroradiological and operative findings
MRI showed vascular compression from the posterior inferior cerebellar artery (PICA) [Figure 3] in three patients (21.4%), vertebral-basilar arteries in three patients (21.4%), and an inflammatory process in one patient (7.1%). Seven patients were reported as normal on MRI scan (49.7%). At the time of the surgery, all 14 patients were found to have compression of the vagal and glossopharyngeal nerve roots. Vascular compression was from PICA in 11 cases (78.5%), vertebral artery in two cases (14.2%), and compression from the choroid plexus in one case (7.1%).

Clinical outcome
All 14 patients were contacted for long-term follow-up. Postoperative mean follow-up of was 26 months (3–180 months). All patients referred initial pain relief, and 13 were pain-free with no need of medication in the long-term follow-up. Only one patient referred pain 1 month after surgery and was treated with carbamazepine with complete relief of the pain and no further surgery was required. Pain analysis demonstrated long-term pain improvement of 114 ± 27.1 months and pain remission in 13 patients (92.9%) (\(P = 0.0001\)) [Table 1].

Complications
Two patients presented complications related to surgical treatment. One patient presented with CSF leak, which resolved with lumbar drainage and acetazolamide 500 mg TID for 5 days without any complications. The second patient presented with meningitis and was treated with intravenous vancomycin 1 g. BID for 5 days recovering completely without clinical sequelae [Table 1]. There was no surgical mortality in this case series.

DISCUSSION

Wilfred Harris applied the term GPN when he described an entity similar to trigeminal neuralgia. At his initial report in 1937, Harris described two types of pathologies: Primary or idiopathic and secondary to carcinoma. Idiopathic GPN is explained due to nerve compression by a vessel, as it exits the medulla oblongata.\[24\] This theory is supported by the success of MVD in the treatment of this pathology.\[12\] The main symptom of the GPN is a lancinating pain lasting seconds to minutes. However, some cases have reported the presence of pain associated to syncope.\[7\] In this regard, Gardner associated the

| Table 1: Clinical data and outcome of patients with glossopharyngeal neuralgia |
|----------------------------------|
| n (%)                      |
| **Pain localization**          |
| Pharyngeal                  | 13 (92.9) |
| Preauricular                | 1 (7.1)  |
| **Pain side**                |
| Left                        | 11 (78.6) |
| Right                       | 3 (21.4)  |
| **Pain trigger**             |
| Abrupt onset                | 3 (21.4)  |
| Swallowing                  | 7 (50)    |
| Talking                     | 4 (28.6)  |
| **Pain irradiation**         |
| Yes                         | 6 (42.9)  |
| No                          | 8 (57.1)  |
| **Preoperative pain**        |
| Grade 3                     | 14 (100)  |
| **Long-term follow-up postoperative pain** |
| Grade 1                     | 13 (92.9) |
| Grade 2                     | 1 (7.1)   |
| Grade 3                     | 0 (0)     |
| **Complications**            |
| Cerebrospinal fluid leak     | 1 (7.1)   |
| Meningitis                  | 1 (7.1)   |
proximity of the glossopharyngeal nucleus to the vagal nucleus. The activation of the nucleus produces activation of the vagal nerve, which results in bradycardia and hypotension secondary to a decrease of the peripheral vascular resistance. Another theory explains the vascular resistance impairment secondary to inhibition of vasomotor centers.

Traditionally, a lateral suboccipital approach provides adequate exposure to the trigeminal, facial, and lower cranial nerves. Kawashima et al. proposed a transcondylar fossa approach advocating the wide operative view of the cerebellomedullary cistern, smaller retraction of the cerebellum, less risk of cranial nerve injury, and enough space to perform the sling retraction technique. However, we believed that a minimally invasive technique as an asterional approach described previously by the senior author is enough for adequate exposure of PICA, vertebral artery, and the relationship with the glossopharyngeal nerve. There is no need of retractors, and after the CSF is released with adequate and careful arachnoid dissection, the cerebellum is out of the way, and there is enough space for working without the necessity of removing the jugular tubercle.

Jannetta popularized the MVD using a suboccipital craniotomy. After years of experience, the approach was modified according to the surgical goal. Initially, it is important to focus bone exposure to the junction of the transverse and sigmoid sinuses. A smaller tailored craniectomy according to the cranial nerve approach can be performed. For lower cranial nerve exposure; McLaughlin et al. recommended a triangular craniectomy with the apex at the edge of the jugular bulb. In our experience, our circular microsurgical craniectomy [Figure 2a] at the edge of the transverse and sigmoid sinuses gives enough bone exposure to access the trigeminal, facial, and glossopharyngeal nerves.

In the MVD series, the overall surgical mortality is 1.1%. The rate of long-term pain remission is 84.7% with recurrence in 7%. Transient X cranial nerve dysfunction occurred in 13.2% and permanent deficits in 5.5%. In our case series, we did not have any mortality, and no permanent deficits occurred after the surgery. We did not have cerebellar lesions or hearing the loss in this case series; it is explained because we do not use retractors over the cerebellum, the surgical route place minimal traction on the VII–VIII nerve complex and we perform a careful microsurgical vascular dissection with minimal bipolar coagulation. However, we had two complications; a CSF leak and a case of meningitis that was successfully treated.

Rey-Dios and Cohen-Gadol demonstrated in his analyses that the most effective surgical procedure to treat GPN is the MVD. Several studies used rhizotomy as the preferred procedure, but a 3-fold increase in the risk of permanent postoperative vagus dysfunction is objectionable in comparison to MVD. It is also well demonstrated that the rate of pain control is slightly better with rhizotomy (95%) than with MVD (86%). However, in our series we had 92.9% pain remission with 3–180 months (mean 26 months) of follow-up; only one case had pain recurrence that was treated with carbamazepine. GPN is a rare condition in which the clinical findings are not always typical. The mean duration from symptom onset to surgery is 5–8 years.
case series, we had a mean time for diagnosis of 8.8 years, however, despite the time for diagnosing GPN the clinical outcome of our patients is similar to the reported in the literature.\cite{27}

It is important to rule out secondary causes such as neoplasms,\cite{13} infections,\cite{19} trauma,\cite{19,40} vascular malformations,\cite{10} Chiari malformation,\cite{41} choroid plexus overgrowth,\cite{22} Tornwaldt's cyst,\cite{31} Eagle syndrome,\cite{6} pontine lesions,\cite{19} multiple sclerosis,\cite{21} and previous surgical interventions (vagal nerve stimulator).\cite{5} It is essential to have a careful selection and an accurate diagnosis of idiopathic GPN to avoid negative exploratory operations. Two of our patients were previously diagnosed as Eagle syndrome, in both of them stilopectomy was performed without pain improvement, and one of them had a tooth extraction before referral to our Institution. During the diagnosis workup, we ruled out secondary causes and confirmed an idiopathic GPN in all patients and MVD was performed.

As Lister et al.\cite{18} previously described in a microsurgical anatomic study, PICA has the most variable course of the cerebellar arteries, but most of the time it passes under the glossopharyngeal nerve. In most of the recent clinical series\cite{13,15,30} PICA is the most common vessel compressing the glossopharyngeal nerve. In our series, during dissection we found PICP compression in eleven cases (78.1%), in all of them we did the transposition of the vessel and apply Teflon in between the nerve and the vessel.

In refractory cases to MVD, we believe that sectioning the glossopharyngeal nerve and the upper roots of the vagus nerve involved an unaccepted high morbidity. We advocate for compression of the glossopharyngeal and upper roots of the vagus nerve as a last option for pain recurrence as previously demonstrated for trigeminal neuralgia.\cite{26} Other noninvasive treatment options have been described: Percutaneous radiofrequency neurolysis\cite{1} is an alternative in cases who failed medical treatment or in which they cannot undergo intracranial surgery. Gamma Knife radiosurgery is also a potential option to relieve the pain without reported side effects but a high early recurrence risk.\cite{14,42}

**CONCLUSION**

Glossopharyngeal MVD through a retractorless microasterional approach is a safe technique in which surgical anatomical knowledge is essential to obtain good results with minimal morbidity. Our series demonstrate an excellent clinical outcome (pain remission - 92.9%) following MVD for GPN.

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**Conflicts of interest**

There are no conflicts of interest.

**REFERENCES**

1. Arbit E, Krol G. Percutaneous radiofrequency neurolysis guided by computed tomography for the treatment of glossopharyngeal neuralgia. Neurosurgery 1991;29:580-2.

2. Ceylan S, Karakus A, Duru S, Baykal S, Koca O. Glossopharyngeal neuralgia: A study of 6 cases. Neurosurg Rev 1997;20:196-200.

3. Chawla JC, Falconer MA. Glossopharyngeal and vagal neuralgia. Br Med J 1967;3:529-31.

4. Dandy W. Glossopharyngeal neuralgia (tic doloreux). Its diagnosis and treatment. Arch Surg 1927;15:198-214.

5. Duhaime AC, Melamed S, Clancy RR. Tonsillar pain mimicking glossopharyngeal neuralgia as a complication of vagus nerve stimulation. Case report. Epilepsia 2000;41:903-5.

6. Eagle WW. Symptomatic elongated styloid process: report of two cases of styloid process-carotid artery syndrome with operation. Arch Otolaryngol 1949;49:490-503.

7. Esaki T, Osada H, Nakao Y, Yamamoto T, Maeda M, Miyazaki T, et al. Surgical management for glossopharyngeal neuralgia associated with cardiac syncpe: Two case reports. Br J Neurosurg 2007;21:599-602.

8. Fraioli B, Esposito V, Ferrante L, Trubiani L, Lunardi P. Microsurgical treatment of glossopharyngeal neuralgia: Case reports. Neurosurgery 1989;25:630-2.

9. Fraioli B, Esposito V, Guidetti B, Cruccu G, Manfredi M. Treatment of trigeminal neuralgia by thermocoagulation, glyceralization, and percutaneous compression of the gasserian ganglion and/or retrogasserian rootlets: Long-term results and therapeutic protocol. Neurosurgery 1989;24:239-45.

10. Galetta SL, Raps EC, Hurst RW, Flamm ES. Glossopharyngeal neuralgia from a posterior fossa arteriovenous malformation: Resolution following embolization. Neurology 1993;43:1854-5.

11. Greene KA, Karahalios DG, Speetzler RF. Glossopharyngeal neuralgia associated with vascular compression and choroid plexus papilloma. Br J Neurosurg 1995;9:809-14.

12. Jannetta PJ. Observations on the etiology of trigeminal neuralgia, hemifacial spasm, acoustic nerve dysfunction and glossopharyngeal neuralgia. Definitive microsurgical treatment and results in 117 patients. Neurochirurgia (Stuttg) 1977;20:145-54.

13. Kandan SR, Khan S, Jeayaraen DS, Lhatoo S, Patel NK, Coakham HB. Neuralgia of the glossopharyngeal and vagal nerves: Long-term outcome following surgical treatment and literature review. Br J Neurosurg 2010;24:441-6.

14. Katusic S, Williams DB, Beard CM, Bergstrahl EJ, Kurland LT. Epidemiology and clinical features of idiopathic trigeminal neuralgia and glossopharyngeal neuralgia: Similarities and differences, Rochester, Minnesota, 1945-1984. Neuroepidemiology 1991;10:276-81.

15. Kawashima M, Matsushima T, Inoue T, Mineta T, Masuoka J, Hirakawa N. Microvascular decompression for glossopharyngeal neuralgia through the transcondylar fossa (supracondylar transjugular tubercle) approach. Neurosurgery 2010;66:275-80.

16. Korkes H, de Oliveira EM, Brollo L, Hachul DT, Andrade JC, Peres MF, et al. Cardiac syncope induced by glossopharyngeal “neuralgia”: A rare presentation. Arq Bras Cardiol 2006;87:e189-91.

17. Laha RK, Jannetta PJ. Glossopharyngeal neuralgia. J Neurosurg 1977;47:316-20.

18. Lister JR, Rhoton AL Jr., Matsushima T, Peace DA. Microsurgical anatomy of the posterior inferior cerebellar artery. Neurosurgery 1982;10:170-99.

19. McCarroll MO, Bone I. Glossopharyngeal neuralgia referred from a pontine lesion. Cephalalgia 1999;19:115-7.

20. McLaughlin MR, Jannetta PJ, Clyde BL, Subach BR, Comey CH, Resnick DK. Microvascular decompression of cranial nerves: Lessons learned after 4400 operations. J Neurosurg 1999;90:1-8.

21. Minagar A, Shereemata WA. Glossopharyngeal neuralgia and MS. Neurology 2000;54:1368-70.

22. Occhiogrosso M, De Tommasi A, Vailati G. Choroid plexus compression
of glossopharyngeal nerve in patients with glossopharyngeal neuralgia. J Neurosurg Sci 1996;40:37-41.

23. Patel A, Kasama A, Horowitz M, Chang YF. Microvascular decompression in the management of glossopharyngeal neuralgia: Analysis of 217 cases. Neurosurgery 2002;50:705-10.

24. Pearce JM. Glossopharyngeal neuralgia. Eur Neurol 2006;55:49-52.

25. Revuelta-Gutierrez R, Beltran-Rochin J, Escobedo-Rios F, Flores-Orozco J. Microcraniectomy asterional: Una opción quirúrgica para la patología del ángulo ponto-cerebeloso. Rev Ecuat Neurol 1999;8:6-10.

26. Revuelta-Gutierrez R, Martinez-Anda JJ, Coll JB, Campos-Romo A, Perez-Peña N. Efficacy and safety of root compression of trigeminal nerve for trigeminal neuralgia without evidence of vascular compression. World Neurosurg 2013;80:385-9.

27. Rey-Dios R, Cohen-Gadol AA. Current neurosurgical management of glossopharyngeal neuralgia and technical nuances for microvascular decompression surgery. Neurosurg Focus 2013;34:E8.

28. Rozen TD. Trigeminal neuralgia and glossopharyngeal neuralgia. Neurol Clin 2004;22:185-206.

29. Rushton JG, Stevens JC, Miller RH. Glossopharyngeal (vagoglossopharyngeal) neuralgia: A study of 217 cases. Arch Neurol 1981;38:201-5.

30. Sampson JH, Grossi PM, Asaoka K, Fukushima T. Microvascular decompression for glossopharyngeal neuralgia: Long-term effectiveness and complication avoidance. Neurosurgery 2004;54:884-9.

31. Sicard R, Robineau J. Algie vélo-pharyngée-es-sentielle. Traitement chirurgical. Rev Neurol 1920;36:256-7.

32. Slavin KV. Glossopharyngeal neuralgia. Semin Neurosurg 2004;15:71-9.

33. Stern LZ, Hall SW. Tornwaldt’s disease. Onset as symptomatic (secondary) glossopharyngeal neuralgia. Neurology 1972;22:1182-5.

34. Stieber VW, Bourland JD, Ellis TL. Glossopharyngeal neuralgia treated with gamma knife surgery: Treatment outcome and failure analysis. Case report. J Neurosurg 2005;102 Suppl:155-7.

35. Sweet WH. Percutaneous methods for the treatment of trigeminal neuralgia and other faciocephalic pain; comparison with microvascular decompression. Semin Neurol 1988;8:272-9.

36. Taha JM, Tew JM Jr. Long-term results of surgical treatment of idiopathic neuralgias of the glossopharyngeal and vagal nerves. Neurosurgery 1995;36:926-30.

37. Teixeira M., de Siqueira SR, Bor-Seng-Shu E. Glossopharyngeal neuralgia: Neurosurgical treatment and differential diagnosis. Acta Neurochir (Wien) 2008;150:471-5.

38. Thomson JL. Glossopharyngeal neuralgia accompanied by unconsciousness. J Neurosurg 1954;11:511-4.

39. Waga S, Kojima T. Glossopharyngeal neuralgia of traumatic origin. Surg Neurol 1982;17:77-9.

40. Webb CJ, Makura ZG, McCormick MS. Glossopharyngeal neuralgia following foreign body impaction in the neck. J Laryngol Otol 2000;114:70-2.

41. Yglesias A, Narbona J, Vanaclocha V, Artieda J. Chiari type I malformation, glossopharyngeal neuralgia and central sleep apnoea in a child. Dev Med Child Neurol 1996;38:1126-30.

42. Yomo S, Arkha Y, Donnet A, RéGIS J. Gamma knife surgery for glossopharyngeal neuralgia. J Neurosurg 2009;110:559-63.