

Treatment of Glossopharyngeal Neuralgia by Gamma Knife Radiosurgery

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Glossopharyngeal neuralgia (GPN) is a rare facial pain syndrome affecting the sensory distribution of the glossopharyngeal (IX) and sometimes vagus (X) cranial nerves. It is characterized by severe paroxysmal pain typically on one side of the throat, ear, base of the tongue, and angle of jaw. GPN can be associated with bradycardia and syncopal episodes that rarely can cause life-threatening hemodynamic instability. Pain attacks may be elicited by triggering stimuli, such as swallowing, coughing, talking or chewing. The majority of cases of GPN are idiopathic and, like trigeminal neuralgia (TN), can be caused by microvascular compression of the nerve roots. Initial management of GPN consist of anticonvulsant medications. For refractory cases, microvascular decompression (MVD) is an option with good rates of pain relief,¹ however, significant morbidity and mortality can occur with open surgery. GKRS is a well-accepted treatment for TN, but its use for GPN remains controversial. There have been few cases reported in the literature to date.²⁻⁸ To add to this, we present the first case in Canada of a medically refractory patient in which GPN was successfully treated with GKRS.

CASE REPORT

A 43-year-old man presented with an 8-year history of electric shocklike pain in the left neck from the angle of the mandible and the throat, which was elicited by triggering stimuli like swallowing and talking. He did not have associated hemodynamic instability. The painful episodes were increasing in intensity and frequency in recent months despite optimal medical management which had included carbamazepine, baclofen, pregabalin, and gabapentin. There was no associated objective neurological deficit. MRI demonstrated possible contact between the left glossopharyngeal nerve and the left vertebral artery. The patient was offered MVD, but declined and was referred to the Gamma Knife Clinic of the Centre Hospitalier Universitaire de Sherbrooke. Detailed information was given concerning expected risks and benefits for treatment of GPN. Informed consent was obtained for Gamma Knife radiosurgery (GKRS).

The Gamma Knife procedure was performed in a standard manner. Briefly, light sedation was administered to the patient and a Leksell G stereotactic frame was then placed under local anesthesia. Stereotactic images were then obtained, including high-resolution CT scan of the skull and volumetric MRI sequences (constructive interference steady-state and contrast-enhanced magnetization-prepared rapid gradient-echo). The Leksell GammaPlan software (Elekta AB, Atlanta, GA) was used for dose planning. The glossopharyngeal nerve was targeted at the glossopharyngeal meatus of

the jugular foramen using a single 4-mm isocentre (Figure 1). A maximum dose of 80 Gy was given. The procedure was performed using Leksell Gamma Knife Perfexion model (Elekta AB).

The patient had gradual, complete relief of pain starting a few days after the procedure. At the last follow-up, 44 months after treatment, the patient had no pain recurrence and remains off medication. No complications were noted. At the time of follow-up, neurologic examination was performed and no signs of glossopharyngeal or vagus nerve dysfunction were noted.

DISCUSSION

GPN is a rare disorder for which limited data are available in the literature. The treatment of choice remains anticonvulsant medication. In patients refractory to medical treatment, neurosurgical intervention, such as microvascular decompression, may be considered. According to the literature, short- and long-term success rates approximate those seen for TN, but severe morbidity and mortality remains a possibility.¹ Rey-Dios et al reported, in a literature review, a rate of pain freedom of 85%, with mean follow-up durations of 4.9 years. They also found a mean rate of permanent cranial nerve X deficit of 5.5% in MVD alone and 19.1% with rhizotomy.¹ Stereotactic radiosurgery is an alternative for patients who do not want or who are poor candidates for open surgery. Its use is well-established for the treatment of TN with low morbidity and high efficacy.

Based on data available for TN, some centers proposed gamma knife treatment for GPN (Table 1). The majority of cases reported have shown good response to radiosurgery. The maximum dose received was 80 Gy or more in 18 of 22 patients. Only two of those 18 patients reported no change in pain and had subsequent MVD, which was also unsuccessful. Among patients who received a maximum dose of 80 Gy or more, 16 patients (89%) had significant or complete reported pain relief, and three had eventual pain recurrence that was controlled with medication. In comparison, three of the four patients who received maximum doses lower than 80 Gy required new surgery for failure or pain recurrence.³ No major side effects were noted in all 22 patients treated with a Gamma Knife. Martinez-Alvarez et al used a

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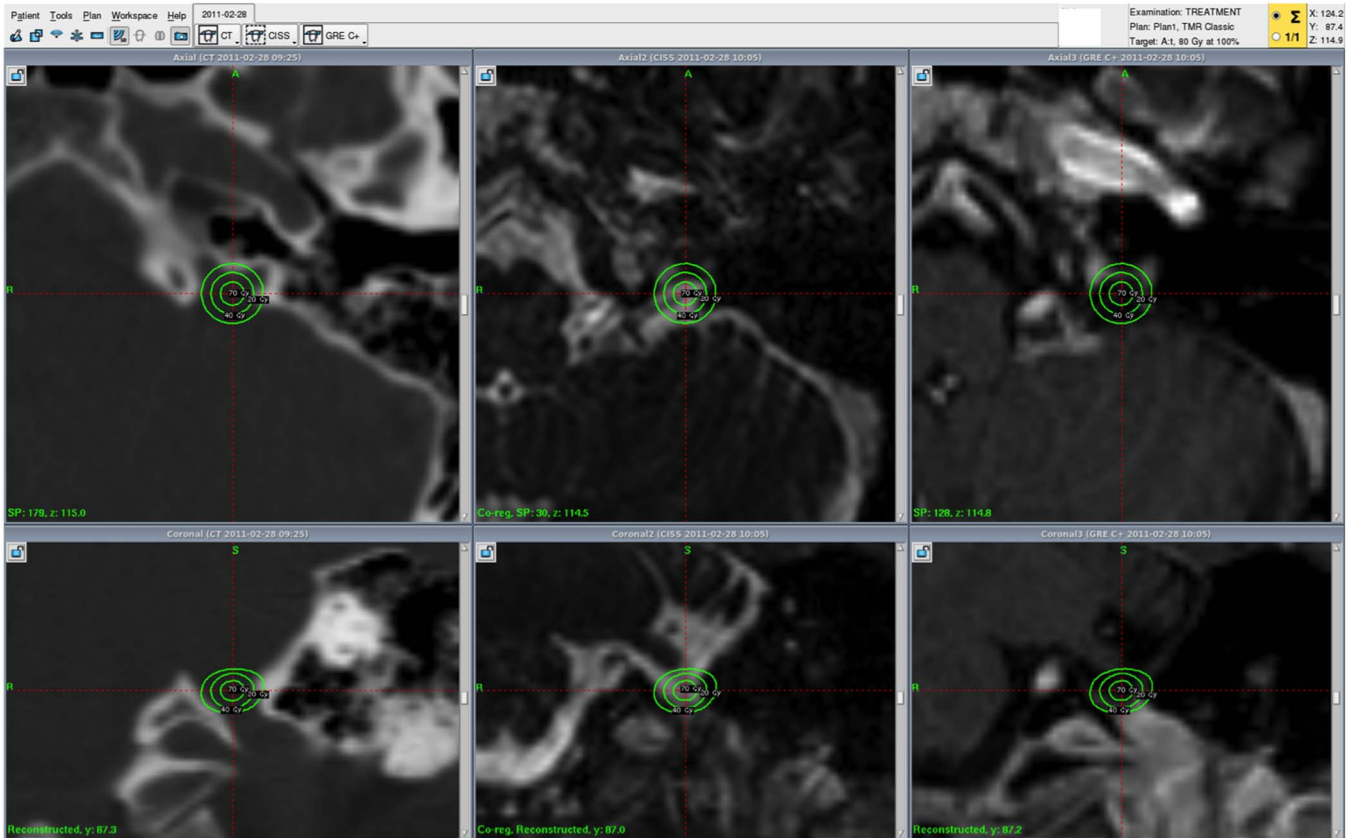


Figure 1: Leksell GammaPlan snapshot of the treatment plan. The treatment isocenter is shown at the glossopharyngeal meatus. Upper row shows the axial plane; lower row, the coronal plane. Left column shows CT images; middle column, constructive interference steady-state MIs; right column, contrast-enhanced magnetization-prepared rapid gradient-echo MRIs.

maximum dose of 90 Gy in four of their patients without side effects observed. As reported in TN studies, higher radiation doses provide better pain relief but seems to be associated with an increased risk of sensory side effects.⁹ It remains to be seen whether these data can be extrapolated to GPN patients. The treatment of GPN using a maximum dose of 80 Gy seems effective and so far the effect appears to be maintained over time. Using a dose below 75 Gy appears to be less effective,³ but it might be premature to draw firm conclusions because of the limited number of patients and short follow-up times reported.

Medically refractory glossopharyngeal neuralgia remains an uncommon condition. Because of this, successful treatment with GKRS might prompt authors to publish their case series, whereas failed cases might not be reported, leading to potential publication bias. Nevertheless, we believe the growing evidence of treatment success supports the use of GKRS in this setting. Prospective studies on a larger scale would be of value, but are difficult to conduct because of the rarity of the pathology.

CONCLUSIONS

Based on our patient evolution and cases found in the literature, Gamma Knife radiosurgery appears to be a safe and effective treatment option for medically refractory glossopharyngeal neuralgia. Targeting the nerve the glossopharyngeal meatus using a maximum radiation dose of 80 Gy is recommended.

DISCLOSURES

None.

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Table 1: Summary of reported cases of glossopharyngeal neuralgia treated with gamma knife radiosurgery

Authors	Age- and gender	Target	Previous surgery	Dose (Gy)	Follow-up (mo)	Pain relief	Pain recurrence	Subsequent surgery
Stieber, 2005	NS, F	GPM	None	80	6	Complete	Yes	None
Williams, 2010	47, F	GPM	None	80	11	Complete	No	None
Levêque, 2011								
Case 1	83, F	GPM	None	60	17	Complete	Yes	Repeat GKRS, thermocoagulation
Case 2	62, M	Cistern	None	70	24	More than 50%	Yes	Cortical stimulation
Case 3	66, M	Cistern	None	70	24	More than 50%	Yes	MVD
Case 4	49, M	GPM	None	75	32	Complete	No	None
Case 5	71, M	GPM	None	80	13	Complete	Yes	None
Case 6	36, F	GPM	None	80	10	Complete	No	None
Case 7	65, M	GPM	None	80	8	Complete	Yes	None
Pollock, 2011								
Case 1	Median patient age 61 (34-83)	GPM	None	80	19	Complete	No	None
Case 2		GPM	None	80	16	Complete	No	None
Case 3		GPM	None	80	13	Complete	No	none
Case 4		GPM	None	80	5	No change	No change	MVD
Case 5		GPM	None	80	2	No change	No change	MVD
O'Connor, 2013	99, F	GPM	None	80	16	Complete	No	none
Stanic, 2012	51, M	Cistern	MVD	80	12	Complete	No	none
Martinez-Alvarez, 2014								
Case 1	56, F	GPM	None	80	83	Complete	No	None
Case 2	73, F	GPM	Rhizotomy	90	71	Complete	No	None
Case 3	62, F	GPM	MVD	90	31	Complete	No	None
Case 4	66, M	GPM	None	90	19	Complete	No	None
Case 5	37, F	GPM	MVD	90	14	Complete	No	None
Current report	43, M	GPM	None	80	44	Complete	No	None

F = female; GKRS = Gamma Knife radiosurgery; GPM: = glossopharyngeal meatus; M = male, MVD: microvascular decompression