

Non-Invasive Treatment of Pterygopalatine Fossa Schwannoma with Gamma Knife Radiosurgery

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Abstract

Background: Surgical resection for pterygopalatine fossa schwannomas (PFS) can be challenging due to the complex anatomy and potential morbidity. Gamma Knife radiosurgery (GKRS) offers a minimally invasive alternative with precise targeting and minimal damage to surrounding structures.

Case report: A 21-year-old female patient who had a history of progressive left-sided facial numbness over the past year and was diagnosed with progressive PFS underwent GKRS. The radiological and clinical outcomes of the patient were evaluated over a period of 15 years, with the patient remaining symptom-free and experiencing no adverse effects from the treatment.

Conclusion: The patient's positive outcome, with significant tumor reduction and symptom relief, underscores the potential of this non-invasive technique as a primary treatment modality for schwannomas in challenging anatomical locations.

Keywords

gamma knife radiosurgery, numbness, pterygopalatine fossa, schwannoma, skull base

Introduction

The pterygopalatine fossa (PPF) is a pyramidal cavity between the maxillary, sphenoid, and palatine bones. Tumors that originate in or affect the PPF are extremely rare, making up just 0.5% of all head and neck tumors.¹ Pterygopalatine fossa schwannoma (PPFS) is a rare primary benign tumor that typically presents with nonspecific symptoms such as facial pain, numbness, and nasal congestion, which can be challenging for clinicians to diagnose. Despite the availability of advanced radiological tests, some patients still present with a large PPFS.

The traditional treatment approach for PPFS involves surgical resection. However, the intricate and complex anatomy of the PPF poses significant challenges during these procedures.² To address these difficulties, surgeons began utilizing endoscopic techniques, which offer improved visualization and access with less invasiveness.³ Despite these advantages, endoscopic techniques require extensive training and still present certain complications⁴⁻⁸, necessitating alternative minimally invasive treatment modalities, such as Gamma Knife radiosurgery (GKRS), for treating such challenging-to-reach lesions.⁹⁻¹² Patients may also need GKRS as salvage therapy in cases where subtotal resection is performed.⁵

Herein, we present the first case report of a PPFS patient treated with GKRS and followed in our clinic for 15 years after the treatment.

Case report

A 21-year-old female presented with a history of progressive left-sided facial numbness over the past year. Initially diagnosed with a PPFS by a referring clinic and followed for one year, the patient showed tumor progression on further magnetic resonance imaging (MRI), leading to referral to our clinic for further treatment as the patient did not want to undergo surgery. The patient had no significant medical history and was not on any regular medications. She reported no history of trauma, infections, or surgeries in the affected region. On physical examination, no masses were detectable intraorally or on facial inspection. Neurological examination was largely unremarkable, with no deficits in cranial nerve functions. The patient's visual acuity and ocular movements were normal. The initial workup included a contrast-enhanced MRI, which revealed a well-defined, homogeneously enhancing mass in the left pterygopalatine fossa, exhibiting characteristics consistent with a schwannoma.

After a multidisciplinary discussion, the patient was offered GKRS as the primary treatment modality, considering the tumor's location and the patient's preference to avoid open surgery. The radiosurgical procedure was conducted under local anesthesia with the patient in a stereotactic frame to ensure precise targeting. High-resolution thin-slice MRI was used to delineate the tumor margins. A marginal dose of 14 Gy was delivered to the 50% isodose line, encompassing the entire tumor volume in a single session (Figure 1). The procedure was completed without complications, and the patient was monitored in the recovery area before being discharged the same day.

The patient was monitored with regular follow-up visits and imaging. At the 6-month follow-up, the patient reported significant improvement in facial numbness. Neurological examination remained normal. At this time, MRI showed stable tumor size with no evidence of progression. The 12-month follow-up MRI showed further tumor shrinkage, with a total reduction of 35% in size from the initial measurement. The patient remained symptom-free, and no adverse effects of the treatment were noted. At the 15-year follow-up, the MRI confirmed further tumor shrinkage and stable findings with continued symptom relief (Figure 1).

Discussion

PPFSs are exceedingly rare, accounting for a small fraction of all schwannomas. Their rarity, coupled with the complex anatomical region they inhabit, presents unique challenges in diagnosis and management. As in other schwannomas, this case highlights the efficacy and safety of GKRS in treating PPFSs. The precise delivery of high-dose radiation minimizes damage to adjacent critical structures, offering a favorable outcome.

Traditional management of schwannomas involves surgical resection, especially for tumors causing significant mass effects or in cases where a definitive histopathological diagnosis is required. Tumors in this region have been excised using an open lateral or anterior surgical corridor. However, these techniques are highly invasive, requiring extensive facial incisions, craniotomies, or osteotomies. Endoscopic approaches with image guidance have become integral to the skill set of skull base surgeons, offering improved cosmetic outcomes and quality of life. On the other hand, these techniques require extensive training and are considered an advanced procedure that should be performed in specialized referral centers. Despite this, complications can still occur as surgical access is technically challenging in the PPF due to the region's intricate anatomy. The PPF is filled with adipose tissue, specifically the pterygopalatine extension of the buccal fat pad. It contains the maxillary nerve and its branches, the pterygopalatine ganglion, the nerve of the pterygoid canal, and the pterygopalatine segment of the maxillary artery, along with its branches and corresponding veins.² Thus, surgical approaches, whether open or endoscopic, may lead to significant morbidity.¹³ Post-operative cerebrospinal fluid (CSF) leak is the most common complication following endoscopic approaches and can lead to complications such as meningitis, pneumocephalus, or acute subdural hemorrhage.¹⁴ Additionally, CSF leaks can result in increased hospital stays and a higher risk of readmission. Injury to the internal carotid artery is a rare but serious complication of the endoscopic endonasal approach, with an incidence ranging up to 1% and a mortality rate of up to 10%.^{15, 16} In addition to major complications, decreased olfaction, sensory changes, nasal crusting, nasal obstruction, rhinosinusitis, and mucocele formation can also be observed.¹⁷ Dry eye, V2 hypesthesia, and trismus are also site-specific complications.¹⁸

GKRS has emerged as a valuable alternative for treating benign tumors like schwannomas, particularly in complex anatomical regions like the PPF or those with contraindications to surgery. The procedure is minimally invasive, typically performed on an outpatient basis, and

associated with minimal recovery time and morbidity. The efficacy of GKRS in treating schwannomas, including vestibular and non-vestibular types, has been well-documented.¹⁹⁻²¹ Studies have shown high rates of tumor control and symptom relief with no to minimal complications. Although this is the only case report of PPFS managed with GKRS, Niranjana et al.⁹ reported an overall tumor control rate of 94.5% in a cohort of 309 patients with trigeminal schwannomas, with only 9% experiencing perilesional edema. The authors reported symptom improvement in 45% of the patients at the last follow-up. In this case report, the patient experienced significant symptomatic relief, substantial tumor reduction, continued symptom relief, and no new neurological deficits at the 15-year follow-up.

Conclusion(s)

In conclusion, this case report is the first in the literature to document the successful treatment of a PPFS using GKRS. The patient's positive outcome, with significant tumor reduction and symptom relief, underscores the potential of this non-invasive technique as a primary treatment modality for schwannomas in challenging anatomical locations. Further studies are needed to validate these findings and establish standardized treatment protocols.

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Bullet Point Summary

- This paper documents the first successful treatment of PPFS with GKRS.
- GKRS is an effective and safe minimally invasive treatment for PPFS.
- GKRS has long-term success in local tumor control and symptom relief.

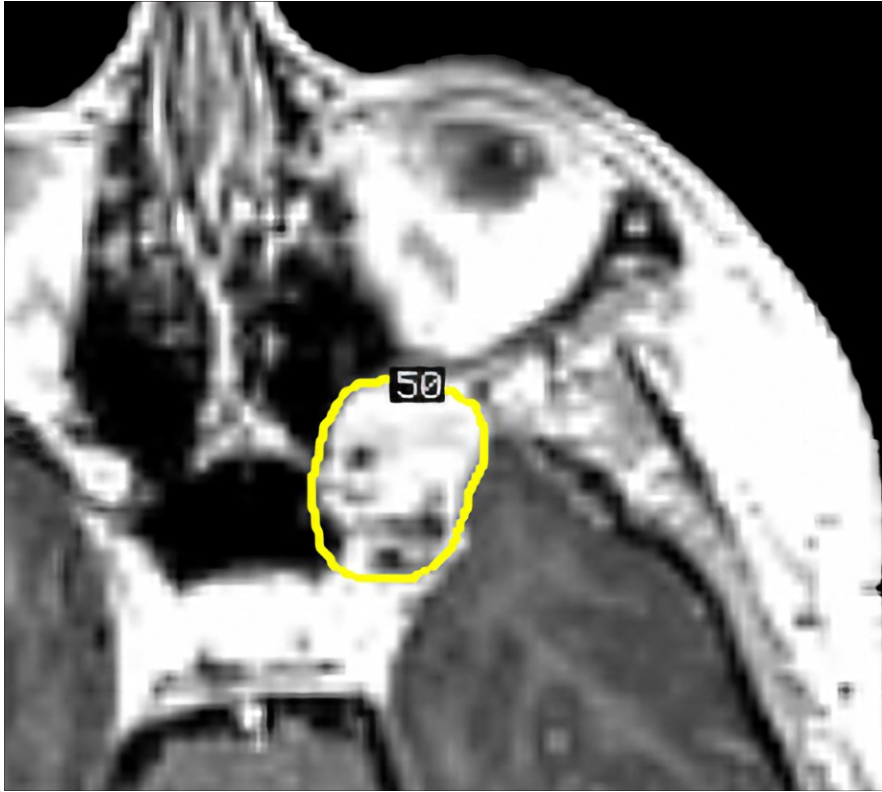


Figure 1

