Gamma Knife surgery for optic glioma

Report of 2 cases

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Optic pathway/hypothalamic gliomas represent approximately 2%–5% of brain tumors in children. Total excision, subtotal excision, subtotal excision followed by irradiation, radiation therapy alone, chemotherapy, and no treatment at all have been reported. In this article the authors discuss the results of Gamma Knife surgery (GKS) for optic gliomas in 2 children.

Two pediatric patients, a boy and a girl, underwent GKS for optic gliomas at our hospital between March 2005 and August 2005. The children’s ages were 10 and 16 years at presentation. The histological diagnosis was confirmed to be pilocytic astrocytoma in both cases. The tumor involved the optic chiasm in 1 patient and the right optic nerve in the other patient. Treatments were planned with the prescription of 11 Gy to the 50% isodose line for the optic chiasm glioma and 15 Gy to the 50% isodose line for the optic nerve glioma.

In both patients, GKS was well tolerated. The follow-up periods were 60 and 55 months. Complete response with near-total disappearance of the tumors was observed in both patients. During the follow-up period, neither of the patients developed any endocrine dysfunction.

Gamma Knife surgery permits treatment of optic glioma with good tumor control and no clinically relevant morbidity. With the ability to deliver a high dose to the tumor while sparing normal brain tissue, especially the optic nerve, optic chiasm, and pituitary gland, GKS should be the choice of treatment for optic gliomas. A larger number of patients and long-term follow-up are required for further evaluation of the efficacy and potential side effects of GKS. (DOI: 10.3171/2010.7.GKS10945)

KEY WORDS • Gamma Knife • stereotactic radiosurgery • optic glioma

Optic pathway gliomas are intrinsic low-grade neoplasms involving the optic nerve, chiasm, and tracts (the anterior visual pathway). They comprise 2%–5% of all intracranial tumors in children. Up to 60%–80% of these rare tumors occur in pediatric patients during the 1st decade of life. The most common histological entity in children is low-grade astrocytoma.

Approximately 10%–70% of these tumors are associated with neurofibromatosis. The disease course is variable and unpredictable, ranging from spontaneous regression to rapid progression and resulting in severe morbidity and death. The rarity of these tumors and their unpredictable course make assessment and standardization of treatment modalities difficult.

The treatment of optic gliomas remains controversial. Total excision, subtotal excision, subtotal excision followed by irradiation, RT alone, brachytherapy, chemotherapy, and no treatment at all have been reported. Chemotherapy was introduced in the 1990s to postpone or replace irradiation in the treatment of children with optic gliomas. In the past few decades, chemotherapy has gained significant recognition in the treatment of optic gliomas. It is expected to have fewer long-term side effects than radiation treatment. Radiation therapy has been recommended by many authors as the treatment of choice in patients for whom total resection of tumor is not possible with acceptable morbidity. Furthermore, RT is considered to be effective and recommended for patients who experience tumor recurrence after resection. Potential late toxicity of conventional RT, such as endocrine disorders and neurophysiological deficits, limits its general application, especially in very young children. Fractionated stereotactic radiotherapy and SRS reportedly have advantages over conventional RT because they can minimize the amount of normal tissue affected by high doses of radiation. We report the results of GKS for optic gliomas in 2 children.

Methods

In this study, we report on 2 pediatric patients, 1 boy and 1 girl, who underwent GKS for optic gliomas at our hospital between March 2005 and August 2005.
Gamma Knife surgery for optic glioma

Gamma Knife Surgery

Gamma Knife surgery was performed following the administration of a local anesthetic agent. After a Leksell model G stereotactic frame (Elekta AB) had been affixed to the head, each patient underwent stereotactic MR imaging to identify the tumor, optic nerve, and optic chiasm. Magnetic resonance imaging was performed in axial planes by using short repetition time sequences. Targeting was based on the axial images. The sequences were performed at 1-mm slice intervals. These imaging sequences provided graphic depiction of the optic nerve and tumor. Treatment planning was performed using the Leksell GammaPlan (version 5.32) for the model C Gamma Knife (Elekta AB). Target volumes were drawn in the presence of a neuroradiologist, a neurosurgeon, and a physicist. Each treatment plan was created using the same clinical criteria: a plan that is as conformal as possible without exceeding dose constraints for critical structures. The plug function was used to protect critical structures.

Case Reports

Case 1

This boy was 10 years old at the time of presentation for GKS for a 6.2-cm³ tumor in the optic chiasm. He had previously undergone craniotomy but no cytotoxic chemotherapy or RT. The lesion was confirmed to be a pilocytic astrocytoma by histological analysis.

Operation. The prescription dose used to treat the optic chiasm glioma was 11 Gy to the 50% isodose line. Treatment was completed and GKS was well tolerated in this patient.

Follow-Up. Local tumor control was obtained, and there was almost complete disappearance of the tumor by the time of the last follow-up at 55 months post-GKS (Fig. 2). Following GKS there was no improvement in the patient’s total blindness on the right side. Her visual acuity was 1.0 on the left side and remained stable throughout the follow-up period. Endocrine testing found no sign of endocrine dysfunction in this patient. She displayed good learning performance at school.

Discussion

The natural history of optic gliomas is unpredictable. Some tumors may exhibit little growth or remain quiescent for years; others enlarge rapidly and are associated with high rates of morbidity. Several authors have reported spontaneous regression of optic gliomas in patients with and without neurofibromatosis. Complete resection has proved successful in tumors confined to the optic nerve. For disease that has progressed into or beyond the optic chiasm, on the other hand, total resection is not practical. Partial resection may provide symptom relief and will provide a tissue diagnosis.

In the past few decades, chemotherapy has become the first-line treatment of choice. It delays RT or surgery until the disease has progressed. Nevertheless, although chemotherapy has emerged as a promising therapy, no regimen thus far has been universally accepted. Many studies clearly demonstrate that postoperative RT directed to the chiasm is much more effective than surgery alone, resulting in a reduced treatment failure rate. Jenkin et al. showed a significant difference in 20-year relapse-free survival rates: 75% for patients given RT compared with 41% for patients treated with primary subtotal resection alone. Kovalic et al. treated 33 patients with optic gliomas using conventional RT. They reported that 5 patients developed learning disabilities following RT. Endocrine dysfunction is a well-known late effect of irradiation of hypothalamic structures. Kovalic et al. reported a 10-year incidence of 63% in children younger than 10 years of age and a 10% incidence for older pa-
tic nerve glioma. The follow-up periods were 24 and 43 months. In both cases were 10 and 48 months. Kurt and colleagues described 2 patients with optic gliomas treated with GKS; follow-up MR imaging in these cases demonstrated a decreased tumor volume. The follow-up periods for the 2 cases were 10 and 48 months. Kurt and colleagues reported on 6 patients with optic pathway tumors who were treated with proton radiation therapy. At a mean follow-up period of 3.3 years, all 6 patients maintained useful vision or obtained improved visual status.

Few reports have demonstrated the effectiveness and application of SRS for optic gliomas. Lim and Leem reported on 2 patients with optic gliomas treated with GKS. The follow-up periods were 24 and 43 months. In both cases, follow-up MR imaging showed a marked decrease in tumor size, and visual symptoms were improved. Kwon et al. described 2 patients with optic gliomas treated with GKS; follow-up MR imaging in these cases demonstrated a decreased tumor volume. The follow-up periods for the 2 cases were 10 and 48 months. Kurt and colleagues reported on 2 patients with optic gliomas treated with fractionated GKS. The tumors were well controlled during the follow-up period (39 and 42 months). Han et al. treated 4 children with optic gliomas using Novalis SRS. Tumor control was obtained in all patients. The follow-up period was 54 months (range 50–58 months). None of these patients displayed clinically relevant morbidity, especially endocrine dysfunction. Stereotactic radiosurgery allows exposure of adjacent critical structures to a much smaller dose of radiation and is considered to be safer than conventional RT or FSRT. In particular, it allows reduction of the dose to the pituitary gland and helps avoid therapy-induced endocrine disorders. Nevertheless, the number of cases reported in the literature is too small and the follow-up period relatively short to draw any definitive conclusions at this time.

Conclusions

Optimal management of optic gliomas is controversial because of the varied natural course of the disease. We reported 2 cases of optic gliomas that were treated with GKS and resulted in well-controlled tumors. These case reports contribute to the literature by underscoring the impressive efficacy of GKS for optic gliomas. We believe that the near-complete response in our patients without evidence of toxicity supports the use of GKS as a safe and effective treatment modality in children with symptomatic optic gliomas. However, long-term follow-up is required for further evaluation of its efficacy and potential side effects.

Disclosure

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Author contributions to the study and manuscript preparation include the following. Conception and design: all authors. Acquisition of data: all authors. Analysis and interpretation of data: all authors. Drafting the article: Liang, Liliang. Critically revising the article: all authors. Reviewed final version of the manuscript and approved it for submission: all authors. Administrative/technical/material support: Chen, Liang, Lu. Study supervision: Chen, Liang.

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C. L. Liang et al.
Gamma Knife surgery for optic glioma


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