Optic nerve glioma treatment with fractionated stereotactic radiotherapy

Case report

NURI USLU, M.D., EBRU KARAKAYA, M.D., AYSEN DIZMAN, M.D., DINCER YEGEN, M.S., AND YILDIZ GUNEY, M.D., PH.D.

Clinic of Radiation Oncology, Dr. Abdurrahman Yurtarslan Ankara Oncology Education and Research Hospital, Demetevler, Ankara, Turkey

In the current report, the authors present a case of optic nerve glioma treated with fractionated stereotactic radiotherapy (FSRT). An 11-year-old girl was referred to our clinic with increasing proptosis over a 1-year period. At that time orbital MRI revealed a 20 × 17–mm mass in the right retroorbital lipomatous tissue, and FSRT was delivered to the tumor using the CyberKnife. During the 1.5-year follow-up, ophthalmological examinations did not indicate any treatment-related severe toxicity, and posttreatment MRI demonstrated marked regression of the lesion to 13 × 10 mm. Given the scarcity of reports on this subject, the authors support more extended studies of the CyberKnife for the effective treatment of this relatively common childhood tumor.

(http://thejns.org/doi/abs/10.3171/2013.2.PEDS11435)

KEY WORDS • optic nerve glioma • CyberKnife • oncology • fractionated stereotactic radiotherapy

The most common tumors of the optic pathways in childhood are ONGs, which have different prognoses and are generally diagnosed early in life, at a median age of 4.5 years.7 While most ONGs are low-grade pilocytic astrocytomas and their excision reveals a benign course, these lesions can also regress spontaneously. Malignant ONGs, which are rare, can cause death resulting from mechanical pressure on vital structures.7,10,12 So, the natural history of this lesion varies from a benign self-limiting disease to a rapidly growing one.2,3,7

Although there is a well-defined relationship between these tumors and NF1, most cases of ONG are sporadic.5,18,19 Approximately 15% of patients with NF1 also have ONG.20 Moreover, hemangiopericytomas are another pathological tumor type that can be seen in the optic pathways.19

Signs and symptoms in patients with ONGs include diminished visual acuity (monocular and binocular), exophthalmos, strabismus, nystagmus, optic disc changes (papilledema or optic atrophy), and, especially in cases of intracranially located tumors, increased intracranial pressure and visual field defects.4

Close observation, surgery, chemotherapy, and radiotherapy, either alone or in combination, are the therapeutic options for optic pathway gliomas, and the treatment of choice is debated.5,8,16

Few studies have shown the effectiveness of stereotactic Gamma Knife surgery for ONG, and these have a limited number of patients.16,17,22 To our knowledge, there is only 1 case study documenting neovascular glaucoma after FSRT using the CyberKnife.14 Thus, in the present study we revealed the clinical findings and effect of FSRT via the CyberKnife on visual acuity and local control in a child with an ONG.

Case Report

History and Examination. An 11-year-old girl presented with a 1-year history of gradually increasing proptosis in her right eye. Ophthalmological examination revealed her visual acuity as 7/10 in the right eye and 9/10 in the left eye. Orbital MRI demonstrated a 20 × 17–mm contrast-enhanced mass in the right retroorbital lipomatous tissue with extension superomedial to the orbit (Fig. 1). The patient did not have biopsy confirmation, but MRI criteria presumed an ONG. For treatment planning, the

Abbreviations used in this paper: FSRT = fractionated stereotactic radiotherapy; NF1 = neurofibromatosis Type 1; ONG = optic nerve glioma.
patient was immobilized with a mask only. For the planning procedure, the slice thicknesses of the CT scan (Fig. 2) and MR image were 1.5 and 3 mm, respectively.

_Treatment_. In August 2009, over the course of 5 days, the patient received FSRT via the CyberKnife (Accuray, Inc.). A prescription dose of 21 Gy was delivered to the 83% isodose line in 5 fractions. The gross tumor volume was 5.207 cm³ (Figs. 3 and 4). For treatment planning, a sequential optimization technique with a fixed collimator was used.

_During CyberKnife radiotherapy we did not observe any severe, treatment-related side effects. Only conjunctivitis-like symptoms and dry eye were observed, which were Grade 2 symptoms according to the Radiation Therapy Oncology Group acute radiation morbidity scoring criteria._

_Posttreatment Course_. After completion of therapy, the patient was followed up with routine ophthalmological examinations and MRI studies. Posttreatment MRI revealed a gradually decreasing maximal tumor diameter of 20, 18, 13, and 13 mm at 2, 5, 7, and 17 months, respectively.

At the last MRI follow-up (Fig. 5), the maximal tumor dimension was 13 × 10 × 7 mm with minimal contrast enhancement. In addition, there were inflammatory changes on the right eyelid and preseptal region. Ophthalmological examination at 17 months posttreatment...
revealed that right-eye visual acuity was 20/20, and the patient’s endocrine status was completely normal. We did not observe any chronic serious side effects.

Discussion

The term “optic nerve glioma” usually means glial tumors involving the anterior visual pathways, optic nerves, optic chiasm, and optic tract, and these lesions make up 1% of all intracranial tumors.13,21 Because of the variable natural history of these rare tumors, treatment choice is a matter of debate.13 Some studies in the literature report spontaneous regression and improving visual acuity without any treatment.11,13 In our case, the patient did not show any spontaneous regression for a 1-year period after symptom onset. Moreover, she had no family history of NF1 or signs such as Lisch nodules or endocrinopathies.

Another controversial point is the indication for surgery in ONG. If there is high intracranial pressure, meaningful neurological deficits, or tumor progression, then surgical treatment should not be delayed.1,8,15 Note that partial excision of the tumor can result in subsequent tumor growth.8

Through high-precision conformal techniques using modern imaging modalities like MRI, radiotherapy is advancing. Debus et al.9 reported their results with FSRT for optic glioma. In their study of 10 patients, 8 were treated with FSRT for progressive recurrent tumor following partial excision and 2 were treated after the initial surgery to remove the tumor. They applied a total dose of 52.4 Gy in 1.8-Gy daily fractions via a noninvasive stereotactic fixation system using a multileaf collimator connected to a linear accelerator. All patients were free from local tumor progression, except one patient who had been treated for recurrent tumor after previous radiotherapy. Authors from the same center reported their experience during a longer median follow-up of 97 months in 15 patients with optic pathway gliomas.8 The ages of patients were between 21 months and 33 years. Among these 15 patients, 12 had a partial resection, 1 had a biopsy, and 2 had no histological confirmation. Fractionated stereotactic radiotherapy was delivered in 6- or 15-MeV photon beams via a linear accelerator using a multileaf collimator. The median dose of 52.2 Gy (biologically effective dose: 99.2 Gy, α/β 2) was applied in median fraction doses of 1.8 Gy. The 5-year survival and progression-free survival rates after FSRT were 90% and 72%, respectively. After radiotherapy, vision improved in 6 patients, remained stable in 7, and was impaired in 2.

In our patient, we applied a total of 21 Gy (biologically effective dose: 65.1 Gy, α/β 2) in 5 fractions via FSRT using the CyberKnife. Although our dose was lower than doses in other studies, we still observed a significantly decreased tumor size without any severe side effects such as neovascular glaucoma or endocrine dysfunction. In addition, our patient’s visual acuity was excellent at 17 months after treatment.

In the Xu et al. study,22 202 patients with orbital tumors underwent Gamma Knife surgery. While the tumor was smaller in 58.4% of patients, it remained stable in 35.1%. These authors also revealed that, while visual acuity was preserved in 129 patients, improvement in vision occurred in 72 patients. Significant impairment of visual acuity was detected in 18 of 147 patients who had functional vision before radiosurgery.

Liang et al.17 reported the results of Gamma Knife surgery for optic glioma in 2 pediatric patients with 60 and 55 months of follow-up. They delivered 11 Gy of radiation to the 50% isodose line for the optic chiasm glioma and 15 Gy to the 50% isodose line for the ONG. Nearly total disappearance of the tumors was achieved with no severe toxicity.

A case report by Jeon et al.14 revealed neovascular glaucoma as a possible side effect after FSRT using the CyberKnife for an ONG. As mentioned in Chang’s letter to the editor,6 however, the dose of 16 Gy administered in 4 fractions was not sufficient to cause any toxic effect on the retina or other eye structures except the lens. Thus, close follow-up is needed after stereotactic radiotherapy.

Conclusions

Although there are limited data on FSRT for ONG, when we take into account the steep dose gradient outside the target, the noninvasive and easy treatment delivery system, and the chance of preserving visual function, the CyberKnife can change the approach to ONG treatment. A larger number of patients and a longer-term follow-up are required for further evaluation of the efficacy and potential side effects of FSRT using the CyberKnife.

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...
Optic nerve glioma treatment with the CyberKnife

include the following. Conception and design: Uslu, Karakaya. Analysis and interpretation of data: Uslu. Drafting the article: Uslu. Critically revising the article: all authors. Approved the final version of the manuscript on behalf of all authors: Uslu. Administrative/technical/material support: Dizman, Yegen.

References


 Portions of this work were presented in poster form at the 2011 CyberKnife Robotic Radiosurgery Summit held in San Francisco, California.

Please include this information when citing this paper: published online March 15, 2013; DOI: 10.3171/2013.2.PEDS11435. Address correspondence to: Nuri Uslu, M.D., Clinic of Radiation Oncology, Dr. Abdurrahman Yurtarslan Ankara Oncology Education and Research Hospital, Demetevler, Ankara 06200, Turkey. email: nuriuslu_70@hotmail.com.