Central neurocytoma is a rare, benign brain tumor that is usually found in an intraventricular location; it often presents with signs and symptoms of obstructive hydrocephalus, typically in the second and third decades of life. With no clear preponderance in either sex, this tumor classification is a relatively recent designation, the clinical course in these lesions has been mainly defined anecdotally. Standard treatment so far has been gross-total resection, which has been achieved in 60 to 100% of patients treated. However, local recurrence rates of 0 to 33% have been reported following subtotal resection, which may indicate that this level of resection is an adequate outcome of surgery when gross-total resection could not be safely achieved. When recurrences were found, the patients’ clinical course and prognosis were varied, ranging from continued lack of neurological deficit to death. Follow-up management in these cases has included routine MR imaging and clinical monitoring, reoperation for gross-total resection, RT, or chemotherapy. We present a series of four patients who underwent primary resection of central neurocytoma and were subsequently treated with GKS for residual or recurrent tumor. The imaging and clinical features of these cases were retrospectively reviewed to determine the efficacy of this type of radiosurgery.

**Clinical Material and Methods**

**Patient Population**

In reviewing the 935 patients treated at the New England Gamma Knife Center between 1992 and 1999, we identified three with central neurocytomas who were followed at our center after subtotal resection or local recurrence of central neurocytoma. In addition, the first patient in the series underwent GKS in November 1990 at the University of Virginia Health Sciences Center, Charlottesville, after surgical resection at our hospital.

**Diagnosis, Treatment, and Follow Up**

A biopsy-proven diagnosis reviewed by a neuropathologist was made for each tumor. All four tumors received minimum doses of 9 to 13 Gy prescribed to the 30 to 50% isodose line. Follow-up consisted of annual MR imaging and clinical evaluation by a neurosurgeon. Follow-up periods after radiosurgery for the four patients were 12, 23,
Case Reports

Case 1

This 22-year-old woman initially presented after having suffered severe headaches for some months. Neuroradiological imaging, including computerized tomography scanning and MR imaging, demonstrated an obstructing tumor of the left lateral ventricle that extended from the occipital horn to the foramen of Munro. A ventriculoperitoneal shunt was placed, which resolved the headaches; however, due to the patient’s intervening pregnancy, this procedure was delayed until 18 months postoperatively. Pretreatment imaging demonstrated that the tumor had not grown in size since her postoperative MR images 18 months before. After the residual tumor was treated using 23 isocenters with a prescribed dose of 13 Gy to the 50% isodose line, the patient remained asymptomatic. At her last follow-up review, which was performed 23 months after radiosurgery, the residual tumor volume had been reduced by 72%.

Case 2

This 32-year-old woman initially presented after a 1-year history of bifrontal headaches that were worse in the morning, and occasional nausea and vomiting when bending over. Admission MR imaging revealed a 5 × 5.5 × 4.5–cm left-sided lateral intraventricular tumor. After resection via a transcalsallos approach, a significant reduction in tumor volume was achieved; residual tumor remained in the medial and posterior aspects of the left lateral ventricle. The patient’s symptoms improved postoperatively; she experienced only occasional headaches. The residual tumor was treated with GKS by using 30 isocenters to deliver 10 Gy to the 40% isodose line. The patient tolerated the procedure well and remained asymptomatic postoperatively. Follow-up imaging revealed that the tumor volume was reduced by 81% at 42 months after GKS (Fig. 1).

Case 3

This 32-year-old woman presented initially with a 9-year history of severe headaches, which became worse 1 month before diagnosis. Physical examination revealed a neurologically intact patient with no focal findings. Admission MR images revealed a 4-cm heterogeneous mass within the body of the left ventricle that had resulted in marked ipsilateral and mild contralateral hydrocephalus and subfalcine herniation. A gross-total resection was achieved, as demonstrated on postoperative imaging. A postoperative dense hemiparesis resolved completely. However, recurrent tumor was noted in the frontal horn of the left lateral ventricle on follow-up images obtained 53 months postoperatively. This lesion extended into the third ventricle, the infundibular recess, and the supraoptic recess without hydrocephalus. Images obtained 1 year previously had demonstrated no recurrent or residual tumor. The patient was referred to our center for GKS. She was asymptomatic on presentation and was neurological-

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...ly intact on physical examination. After treatment of the recurrent tumor by using 29 isocenters with a prescribed dose of 10 Gy to the 50% isodose line, the patient remained asymptomatic, with complete preservation of vision. At her last follow-up review, conducted 12 months after radiosurgery, the tumor volume had been reduced by 77%.

Discussion

Central neurocytoma is a histological entity that was relatively recently defined by Hassoun, et al.5 These are somewhat rare tumors, with fewer than 300 cases reported in the literature since their definition in 1982. Often confused with oligodendrogliomas because of their appearance under light microscopy, and ependymomas because of their location, either special staining for synaptophysin or neuron-specific enolase16 or electron microscopy for neuronal ultrastructural features9 is required to make the diagnosis. Differentiation between central neurocytoma and oligodendroglioma is especially critical, because the clinical course, treatment, and prognosis of these tumor types are substantially different.

Central neurocytomas typically arise in intraventricular or periventricular sites, although locations such as the occipital lobe,12 cauda equina,15 cerebellum, and cervical spinal cord13 have been reported. These tumors are histologically benign, and their clinical course usually progresses very slowly until signs of elevated intracranial pressure develop due to occlusion or intermittent blockage of the foramen of Monro. Intraventricular hemorrhage9 and sudden death are extremely rare complications. Malignant variants, although very rare, have also been reported.15

Treatment of these tumors depends on both their presenting symptoms and location. Acute obstructive hydrocephalus is treated on an emergency basis through extraventricular drainage. Open resection in which microsurgical procedures are used is the most frequently cited method of initial treatment. Because these tumors are often located anteriorly in the lateral ventricles and sometimes extend into the third ventricle or across to the other lateral ventricle via the foramen of Monro, a transcannal approach frequently provides the best opportunity to achieve a gross-total resection. In many reports7,10,12 gross-total resection is said to be extremely effective in achieving local control of tumors and long-term survival, although some patients experience recurrences.

After subtotal resection and local recurrence, the strategies are more varied. In two studies9,12 no recurrence or neurological deficit following reoperation resulting in gross-total resection were reported. The use of RT postoperatively has been controversial. In several studies7,10,12,15,16 the use of conventional RT to treat subtotally resected and recurrent neurocytomas has been reported. Zentner, et al.,10 argued against the use of postoperative RT, because of the benign nature of central neurocytoma and the lack of evidence of response to this treatment. Yasargil, et al.,15 recommended that RT be reserved for tumors that exhibit malignant characteristics histologically. Sgouros, et al.,12 offered RT as a sensible approach in cases of subtotal resection; in their study they also cited the need for close prospective follow up to determine the efficacy of this adjuvant treatment. Schild, et al.,10 reported a 50% absolute increase in local control rates in three patients who received RT as adjuvant treatment after subtotal resection. Two of these patients received fractionated doses of between 50.4 and 60 Gy; the third received a single dose of 15 Gy delivered by the gamma knife method. Only in the patients receiving conventional RT was a reduction in tumor volume detected. Kim, et al.,7 also reported that fractionated RT after subtotal resection resulted in tumor volume reduction, but urged caution in the use of RT because of posttreatment complications.

Because all of these studies have limited statistical power with respect to the use of RT, no definitive recommendation concerning its use in subtotally resected or recurrent tumors can be made. Additionally, because these tumors are benign on histological studies, given their often clinically benign nature, the use of conventional RT without strong clinical data is problematic to propose. However, because there is no way to predict which subtotally resected tumors will recur or progress, the addition of an adjunctive therapy that confers documented high local control rates and increased survival rates, along with minimal complication rates or neurological sequelae, would be an attractive option in the postoperative management of central neurocytomas. In our limited experience, GKS provides this option.

The application of GKS in the treatment of central neurocytomas has only been anecdotally reported in the literature.10 In that case, as we have mentioned, the gamma knife was used to deliver a 15-Gy dose following tumor progression after an initial subtotal resection, resulting in arrest of tumor growth. Based on our series, in which follow up ranged from 12 to 99 months, the tumor volume in all four patients was markedly reduced, from 48 to 81% (Fig. 2), which was surprising in view of the benign and slow-growing nature of these central neurocytomas. It is even more remarkable that this shrinkage was achieved with a comparatively low radiosurgical dose and with no...
complications or side effects. Nevertheless, we believe that meticulous radiosurgical planning is critical for both a reduction in tumor size and avoidance of neurological complications due to collateral injury. In our cases, peripheral doses in the 9 to 13–Gy range with maximum doses to the tumor volume ranging between 20 and 30 Gy were sufficient to achieve the reduction reported. In two of our cases particularly, functionally important structures such as the internal capsule, the thalamus, and the optic chiasm were close to the tumor. Very conformal coverage was ensured to limit the exposure of these structures to radiation, preventing neurological complications.

Conclusions

Even with few observations and limited follow-up time, because of the consistency of the response without observed side effects, we have concluded that GKS may be the treatment of choice for subtotally resected and recurrent central neurocytomas.

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References


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