Schwannomas originating from the oculomotor, trochlear, and abducent nerves are extremely rare.4,7,10,19 Because of the high risk of permanent nerve injury with resection, minimally invasive alternative strategies are desirable.

We report our early experience with GKS for schwannomas originating from the nerves governing eye movement. We were interested in determining whether radiosurgery would provide a tumor response with clinical response or would prove hazardous to nerve function.

Methods

Over a 7-year period, 8 patients with schwannomas originating from the oculomotor (2 patients), trochlear (5 patients), or abducent (1) nerve underwent GKS. The mean patient age was 46.1 years (range 19–59 years). The presenting symptoms included diplopia in 5 patients, ptosis in 1 patient, ophthalmoplegia in 1 patient, and headache in 1 patient. Two patients had a history of neurofibromatosis Type 2. Gamma Knife surgery was performed as primary management in 7 patients and after prior resection in 1 patient. The median and mean tumor volumes were 0.22 and 1.32 cm³ (range 0.03–7.4 cm³). A median margin dose of 12.5 Gy (range 11.0–13.0 Gy) was prescribed to the tumor margin. Clinical and imaging follow-up data were available for all 8 patients.

Results. Magnetic resonance imaging showed tumor regression in all patients. The progression-free period varied from 4 to 42 months, with a mean of 21 months. Over a mean of 23 months, 4 of the 5 patients with a trochlear schwanna and symptoms of diplopia noted symptomatic improvement. No improvement was noted in the 2 patients with oculomotor nerve palsies. Headache was improved in the 1 patient with an abducent neuroma.

Conclusions. Gamma Knife surgery is an effective and minimally invasive approach capable of inactivating schwannomas originating from the oculomotor, trochlear, and abducent nerves. Accompanying trochlear function may improve. Longer follow-up and larger patient samples are needed to confirm the authors’ initial observations. (DOI: 10.3171/JNS/2008/109/12/S23)

Keywords: abducent nerve • Gamma Knife surgery • oculomotor nerve • radiosurgery • schwannoma • trochlear nerve

SCHWANNOMAS originating from the oculomotor, trochlear, and abducent nerves are extremely rare.4,7,10,19 Because of the high risk of permanent nerve injury with resection, minimally invasive alternative strategies are desirable.

We report our early experience with GKS for schwannomas originating from the nerves governing eye movement. We were interested in determining whether radiosurgery would provide a tumor response with clinical response or would prove hazardous to nerve function.

Methods

Over a 7-year period, 8 patients with schwannomas originating from the oculomotor (2 patients), trochlear (5 patients), or abducent nerve (1 patient) underwent GKS. The mean age of the patients was 46.1 years (range 19–59 years). Two patients had a medical history of neurofibromatosis Type 2. One patient underwent GKS for an intracanalicular vestibular schwannoma and a trochlear schwannoma simultaneously.

One patient underwent adjuvant radiosurgery for recurrence after surgery. For 7 patients, GKS was used as a primary approach. The diagnosis in these patients was based on 1) the patient’s neurological signs, which were compatible with involvement of a specific CN; 2) MR imaging findings compatible with schwannomas, including low signal intensities in T1-weighted images, high signal in T2 enhancement after gadolinium contrast, and no direct relation to dura mater; and 3) tumor location along the route of a specific CN and visualization and continuity of the ipsilateral trigeminal nerve and CN VII and VIII nerve complex on thin-slice MR imaging. The clinical signs included ipsilateral superior oblique muscle dysfunction in patients with a CN IV schwannoma and ipsilateral ophthalmoplegia and increased pupil size with impaired reactivity to light in patients with a CN III schwannoma.

Prior to GKS, 5 patients reported diplopia and 1 pa-
tient each reported ptosis, ophthalmoplegia, and headache (Table 1). For patients with a trochlear schwannoma, the interval from onset of diplopia until the tumor was identified through imaging ranged from 6 months to 4 years. One patient with an oculomotor schwannoma developed ptosis after resection was attempted 3 years prior to GKS. Three patients had been observed with regular MR imaging evaluation over 1–4 years. During the period of observation, the tumors enlarged in two of these patients, and the other patient had worsened clinical symptoms.

**Radiosurgery Technique**

The procedure began with rigid fixation of an MR imaging–compatible Leksell stereotactic frame (Model G, Elekta Instruments, Atlanta, GA) to the patient’s head using local anesthetic scalp infiltration (5% bupivacaine and 1% xylocaine) supplemented by mild intravenous sedation. High-resolution stereotactic MR imaging was performed with a fiducial system attached to the stereotactic frame. For stereotactic targeting, both 3-dimensional volume acquisition MR images using contrast-enhanced spoiled gradient–recalled acquisition and volumetric T₂-weighted images were used. Images were imported into GammaPlan dose planning software (Elekta Instruments). Radiosurgery planning was performed on narrow slice thickness axial images with coronal and sagittal reconstructions. A highly conformal dose plan using multiple collimators covering the entire lesion was created. To create conformal dose plans, we selected the isodose line (50–65%) that best conformed to the lesion. The isodose, maximum dose, and dose to the margin were jointly determined by the neurosurgeon and radiation oncologist. Tumor dose volume histograms were evaluated. The procedure was performed in a single session by positioning the head serially for each subsequent isocenter using the automated positioning system or trunnion mode until a fully conformal field encompassed the tumor volume.

The median and mean tumor volumes were 0.22 and 1.32 cm³, respectively (range 0.03–7.4 cm³). A median margin dose of 12.5 Gy (range 11.0–13.0 Gy) was prescribed to the tumor margin. In 1 patient with a trochlear schwannoma and a medical history of neurofibromatosis, an ipsilateral vestibular schwannoma was treated with GKS simultaneously.

**Postoperative Care and Evaluations**

All patients received an intravenous dose of 40 mg of methylprednisolone at the conclusion of the procedure. Patients were observed for a few hours in our unit and then discharged. After radiosurgery, all patients were followed with serial gadolinium-enhanced MR imaging scans, which were requested at 3–6 months, 1 year, and then every 2–3 years.

### Results

**Imaging Follow-Up**

Magnetic resonance imaging follow-up was obtained in all 8 patients. We measured the change of tumor volume using the X, Y, and Z diameter. Tumor regression was defined as the reduction of the length of all 3 diameters.

![Fig. 1. Case 2. Left: Gadolinium-enhanced MR image of an oculomotor schwannoma showing a small mass adjacent and anterior to the left side of the midbrain in the prepontine cistern at GKS (arrow). Right: Follow-up image 36 months after GKS showing a decrease in tumor size.](image-url)
All the tumors (100%) in these patients showed volumetric reduction (Figs. 1–3). The progression-free period ranged from 4 to 42 months, with a mean of 21 months. Tumor control was also achieved for the patient with a history of neurofibromatosis Type 2 who underwent GKS for trochlear and vestibular schwannomas simultaneously (Case 3).

Clinical Follow-Up

Clinical follow-up was obtained in all 8 patients. During the follow-up period (mean 23 months), diplopia was improved in 4 of the 5 (80%) patients with a trochlear schwannoma. Although this improvement was noted subjectively, 2 patients reported marked improvement and the other 2 patients some improvement in their diplopia. One patient who underwent GKS only recently did not report any change in diplopia.

The ptosis in the patient with an oculomotor schwannoma who had prior resection remained unchanged. In the other patient with an oculomotor schwannoma, the intensity and frequency of ophthalmoplegia with headache remained unchanged. The ipsilateral pupil size showed mild reduction (from 8 to 6 mm) but the impaired papillary reactivity to light remained unchanged. The patient with an abducens schwannoma reported improvement in headache. The tumor showed marked reduction in size on follow-up imaging. None of the patients experienced any complications or adverse radiation effects during the follow-up period.

Discussion

Schwannomas originating from CNs other than the vestibular nerve are rare, representing < 0.5% of all intracranial tumors.23 Among nonvestibular schwannomas, the most frequently affected nerves are the trigeminal and
facial nerves and the lower CNs.\textsuperscript{4,7} Schwannomas originating from the oculomotor, trochlear, or abducen nerves are extremely rare.\textsuperscript{4,7,19} Approximately 40 cases of oculomotor schwannoma,\textsuperscript{18,19} 35 cases of trochlear schwannoma,\textsuperscript{18} and 15 cases of abducen schwannoma\textsuperscript{19} have been reported in the literature. Surgical management of these tumors has been reported mainly in case reports, with meta-analysis in some recent reports.\textsuperscript{1,18,19}

Radiosurgery has been proven to be highly successful in the treatment of nonvestibular schwannomas, including schwannomas originating from the trigeminal nerve\textsuperscript{1,5,15,17,27} and nerves of the jugular foramen.\textsuperscript{12,14,23,24,28} However, in our literature search, we did not find studies describing schwannomas originating from the oculomotor, trochlear, or abducen nerve. In a report of a large series of nonvestibular schwannomas, Pollock et al.\textsuperscript{23} reported the use of GKS for a single trochlear nerve schwannoma. Chakrabarti et al.\textsuperscript{2} described a patient with an abducen nerve neurinoma who underwent linear accelerator stereotactic radiosurgery.

Currently, high-resolution volumetric MR imaging facilitates recognition of these rare tumors after neurological signs develop. In this study, all of the patients with a trochlear schwannoma had diplopia caused by the impairment of ipsilateral superior oblique muscle function. Magnetic resonance imaging showed a tumor with low signal intensity on T1-weighted images, high signal intensity in T2, and uniform enhancement after contrast. These features are typical of schwannomas.\textsuperscript{3,5,9,13,15,17,27}

In this study, GKS for schwannomas of the nerves governing ocular movement resulted in effective tumor control, although long-term follow-up is not yet available and one-half of the patients had follow-up periods of < 1 year. Radiation doses at the tumor margin ranged from 11 to 13 Gy. These relatively low marginal doses were selected for the protection of the brainstem or for nerve protection.

Pollock et al.\textsuperscript{23} reported favorable results with use of GKS for treatment of 23 nonvestibular schwannomas, which mainly consisted of trigeminal and jugular foramen schwannomas but included one trochlear schwannoma. They had used 18 Gy as the median marginal dose, which was higher than the doses currently prescribed for vestibular schwannomas. However, in the present series, the radiation doses were similar to those used for vestibular schwannomas (11–13 Gy). These lower doses were effective for tumor control and provided symptomatic improvement. During the follow-up period, we did not observe adverse radiation effect in any patients. However, careful long-term follow-up is required for some patients. It is generally theorized that sensory nerves may be more vulnerable to radiation effects than motor nerves. Because the oculomotor, trochlear, and abducen nerves are purely motor nerves, their radiation tolerance may be higher, which may be one reason why no adverse radiation effects related to the function of these CNs was noted in this patient population.

Conclusions

Gamma Knife surgery is an effective and minimally invasive approach without significant risk of CN injury in the treatment of patients with schwannomas originating from the oculomotor, trochlear, and abducen nerves. Longer follow-up and larger patient populations are needed to confirm our observations.

Disclosure

Dr. Niranjan reports that he serves as a consultant for Elekta AB, Dr. Kondziolka reports that he serves as a consultant for Elekta Instruments, Inc., and Dr. Lunsford reports that he serves as a consultant for and owns stock in Elekta AB. This work was supported by a Chonnam National University Medical School Department of Neurosurgery Academic Foundation Grant (2007–2008).

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References

Radiosurgery for cranial nerve schwannomas