

## Case Report

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## Radiosurgery with Gamma Knife in Central Neurocytoma: Case Report

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### ABSTRACT

Central neurocytoma is considered a neoplasm classified as grade II by the WHO, of intraventricular location, although they can be aggressive. Due to the rare incidence, there are no fixed treatment rules or radiation therapy recommendations. Macroscopic total resection is usually curative, however, it is only achieved in 30-50% of cases due to the central location. Adjuvant treatment for optimal neurocytoma management should be considered. Stereotactic radiosurgery is now increasingly recommended as an adjuvant treatment. A case report of a patient treated with postoperative radiosurgery with high local control is presented, and the literature review is also performed.

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### Introduction

Neurocytoma are benign, rare tumors, with neuronal differentiation, derived from the precursor cells of the subventricular matrix, with an incidence of 0.1 - 0.5% [1,5]. The first patient was described in 1982 by Hassoun et.al [6]. Most have a favorable prognosis, but they can present with a more aggressive clinical behavior, affecting young adults, 70% being between 20 and 40 years old, it is found more frequently in the ventricular system, for this reason the term central neurocytoma [1,4].

The clinical manifestations are variable, generally the patients present headache, nausea, vomiting, seizures, hydrocephalus and intracranial hypertension [1,4]. The treatment of choice is surgical, it consists of total resection of the tumor, however, as it is centrally located, this type of resection is rarely achieved; in cases of subtotal resection, adjuvant treatment with radiotherapy or chemotherapy is often necessary [1,9].

Adjuvant therapy traditionally consisted of conventional radiation therapy, but produces associated cognitive deficits and other neurotoxicities [3,5,6,8,9]. Recently, stereotactic radiosurgery is increasingly being used as an alternative modality due to fewer fractions and toxicities [1,2,6,7]. Several studies have demonstrated equivalent tumor control and fewer complications with adjuvant radiosurgery compared to conventional radiation therapy [1,2].

A quantitative systematic review conducted by Park and Steven in 2012 (62 patients) demonstrated the efficacy of radiosurgery in

the treatment of central neurocytoma [1,2,6]. We report the case of a patient who received radiosurgery as adjunctive treatment for a voluminous tumor treated with subtotal surgery and gamma Knife radiosurgery with excellent results.

### Clinical Case

A 20-year-old woman presented stiffness and inability to mobilize her neck. The patient reported a previous history of vision alterations, including moments of temporary blindness (Table 1). A brain CT scan revealed a large intraventricular brain mass causing obstructive hydrocephalus. Further examination with a 1.5 Tesla Magnetic Resonance Imaging (MRI) imager with gadolinium administration revealed a 6.2 cm cerebellar lesion compressing the third ventricle and involving the intraventricular septum and resolving venous infarction of the veins. left basal ganglia. The lesion demonstrated T1 and T2 isointensity to the brain, multiple cystic areas, diffusely restricted diffusion, and moderate contrast enhancement.

### Management and Pathological Findings

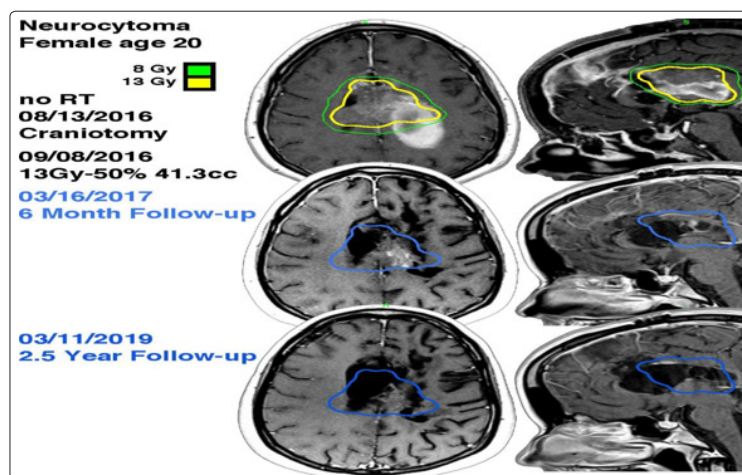
The patient underwent a right frontal craniotomy and interhemispheric transcallosal approach for resection of the intraventricular tumor. A ventriculoperitoneal shunt was placed in the left occipital horn with a medium-high pressure valve. The histological study showed small round cells compatible with a central neurocytoma. Immunohistochemical stains for synaptophysin were positive and for GFAP they were negative. The diagnosis after surgery was confirmed to be a cytologically benign cell neoplasm compatible with a central neurocytoma (Table 1).

After surgery with partial resection, the patient received adjunctive treatment with Gamma Knife radiosurgery using a cobalt-60 source. The treatment plan consisted of 1 site with 15 isocentres that used all 15 collimators. The patient received a 13 Gy dose administered to the 50% isodose line with a volume of 41.3 cc (Figure 1). MRI after radiosurgery observed resolution at intervals of increased intracranial pressure and normalization of ventricular size. Reduction in the size of complex intraventricular hemorrhage (Figure 1).

**Table 1: Summary of the clinical, radiological and histopathological characteristics of the patient**

Clinical features		Characteristics of the tumor		Immunohistochemical staining	
Sex	Female	Location	Intraventricular	GFAP	Negative
Age	20 years	Volume (cm <sup>2</sup> )	6.2		
Symptoms	Neck stiffness, blindness.	Metastases	none	Synaptophysin	Positive
Treatment	CFD, DVP, RC	Lymph nodes	negative		

CFD = right frontal craniotomy, DVP = ventriculoperitoneal shunt, CR = radiosurgery



**Figure 1:** Follow-up of the patient after treatment with radiosurgery

### Discussion

The treatment of choice for these tumors is surgical, consisting of total gross resection, with an excellent prognosis and minimizing the chances of recurrence [10,11]. Total tumor resection has been reported in one third to one half of patients [10,12]. Which significantly increases the possibility of local tumor control, compared to subtotal resection, for example, the three and five year local control for total resection was 95% and 85% versus 55% and 46% for subtotal resection, respectively [10,11]. When the resection is subtotal, adjuvant treatment with radiotherapy or chemotherapy is necessary to improve local control. Adjuvant therapy traditionally consisted of conventional radiation therapy, but produces associated cognitive deficits and other neurotoxicities [1,3,5,6,8,9].

Our patient underwent a right frontal craniotomy and interhemispheric transcalsal approach for resection of the intraventricular tumor, in addition, a ventriculoperitoneal shunt was placed in the left occipital horn with a high-medium pressure valve. The diagnosis after surgery was confirmed to be a cytologically benign cellular neoplasm compatible with a central neurocytoma. The first publication on the use of Radiosurgery in Neurocytoma was in 1997 and subsequently multiple case series and systematic reviews have been reported [13].

Stereotaxic radiosurgery has emerged as an important tool in the treatment of neurocytomas, due to its favorable characteristics such as rapid dose reduction and short-term treatment, compared to conventional radiotherapy [2,10,14,15]. Radiosurgery appears to offer superior results in terms of reducing tumor recurrences and radiation-associated complications [1,2,7,10]. Local tumor control

over 5 years was 100% with stereotaxic radiosurgery compared to 87% with conventional radiation therapy [8,10].

Adjuvant radiosurgery, demonstrated the reduction of the mean tumor volume in 14 patients after the mean follow-up of 70 months, was 69% (range 47 to 87%) without tumor progression [10,16]. Patel et al reported that adjuvant radiosurgery for patients with subtotal resection demonstrated a 100% tumor control rate compared to an 87% tumor control rate for patients with conventional adjuvant radiotherapy [4,17]. Garcia et al, had a tumor control rate greater than 93% with radiosurgery versus 88% with conventional radiotherapy [4,18]. In comparison with the present case, excellent local control with gamma knife radiosurgery could be observed after surgical treatment.

A systematic review of 150 cases with radiosurgery indicates that in order to have a local control rate > 90%, it suggests a standard dose of 13-18 Gy regardless of the tumor [1,19], compared to the case presented, he received radiosurgery with Gamma Knife, the dose given being 13 Gy administered to the 50% isodose line with a volume of 41.3 cc. Magnetic resonance imaging of the brain after radiosurgery showed resolution of increased intracranial pressure, normalization of ventricular size, and reduction in the size of complex intraventricular hemorrhage.

Kim treated 20 patients (10 patients received Gamma Knife radiosurgery (GKRS) as primary treatment and 10 patients who received GKRS as secondary treatment) with a radiological follow-up period ≥36 months. The mean duration of radiological follow-up was 100 months (range 43-149 months). The mean

tumor volume was 10.4 cm<sup>3</sup> (range 0.4-36.4 cm<sup>3</sup>) and the mean dose was 15.4 Gy (range 9-20 Gy). Failure of local control was found in six patients at the last radiological follow-up. The general local actuarial control rates were 89.5% at 5 years and 83.1% at 10 years. The primary group GKRS included two cases with local failure, with cyst formation or local recurrence. In contrast, in the secondary GKRS group, local control failure was found in four cases (including three cases with a pattern of 'recurrence outside the treatment field') and occurred earlier compared to the primary GKRS group. The study suggests that GKRS could be a primary or secondary treatment option for central neurocytoma [20].

Hung conducted a retrospective study comprising 60 patients (median age 30 years), the median tumor volume and the marginal dose were 5.9 cm<sup>3</sup> and 13 Gy. With a median clinical follow-up of 61 months, post-SRS tumor recurrence occurred in 8 patients (13%). The local tumor control rates at 5 and 10 years were 93% and 87%, respectively. The 5-year and 10-year progression-free survival rates were 89% and 80%. Previous radiation therapy was a predictor of distant tumor recurrence ( $p = 0.044$ ). Distant tumor recurrence and spread were observed in a small proportion of patients, underscoring the importance of close post-SRS surveillance of patients with central neurocytoma [21].

Schild et al, reported a 5-year survival rate for patients who received radiation therapy or radiosurgery after surgical resection of 88%, while the 5-year survival rate for patients without adjuvant radiation was only 71% [4,22]. The patient showed an excellent improvement with respect to the reduction of the tumor size, after surgery she had a residual tumor of 41.3 cc, with the controls with images showing a significant reduction at 6 months. A second follow-up control at 2 years and 5 months no longer shows a lesion by magnetic resonance images, with a completely normal state and performance of the patient. Due to the rarity of this disease, there are few research studies regarding the use of gamma knife radiosurgery in the treatment of central neurocytoma.

### Conclusion

Central neurocytoma is a rare, benign tumor common in young adults. Due to the rarity of this disease, there are few research studies regarding the use of gammaknife radiosurgery, which show that it is an effective and safe treatment for the patient. Likewise, the high late control of the local tumor and the low incidence of late neurotoxicities have been observed with the use of this technique. Future prospective studies should be conducted to determine the long-term efficacy of radiosurgery in the treatment of central neurocytoma.

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