



(CASE REPORT)



GAMMA KNIFE RADIOSURGERY FOR INTRACRANIAL HEMANGIOBLASTOMAS: A CASE SERIES

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ABSTRACT

Hemangioblastomas are benign tumors, which develop mainly in the brainstem and cerebellum, sporadic in approximately 75% of cases or associated with von Hippel-Lindau disease (VHL). While surgical resection is considered the best treatment for hemangioblastomas, stereotactic radiosurgery has recently emerged as an attractive alternative to surgery. We present a series of 11 cases of hemangioblastomas treated by radiosurgery at the National Center for Rehabilitation and Neurosciences in Rabat over a period of 6 years from 2019 to 2024.

Keywords: Gamma knife, Hemangioblastomas, Stereotactic radiosurgery, Von-Hippel-Lindau disease.

1 INTRODUCTION

Hemangioblastomas are benign tumors, which develop mainly in the brainstem and cerebellum, sporadic in approximately 75% of cases or associated with von Hippel-Lindau disease (VHL). While surgical resection is considered the best treatment for hemangioblastomas, stereotactic radiosurgery has recently emerged as an attractive alternative to surgery. We present a series of 11 cases of hemangioblastomas treated by radiosurgery at the National Center for Rehabilitation and Neurosciences in Rabat over a period of 6 years from 2019 to 2024.

Objective

The objective of this study was to analyze our experience with Gamma Knife radiosurgery (GKRS) for treatment of intracranial HB and to explore the efficacy, safety, and long-term outcomes of this technique, reviewing clinical studies, dosimetric parameters, and prognostic factors to optimize treatment strategies for this challenging pathology.

2 METHODS

This retrospective study was conducted on 11 patients of hemangioblastoma who underwent SRS at the National Center for Rehabilitation and Neurosciences in Rabat, Medical records and radiological studies of all the patients were reviewed to study the radiosurgical details and the clinic-radiological outcomes.

The patient population included three women and eight men with a mean age of 42 years, ranging from 26 to 59 years.

3 RESULTS

Eleven patients (8 men and 3 women). The mean age was 42 years. The tumor was located in the cerebellar hemispheres, vermal regions, and brainstem in 6 patients (54%), 2 patients (18%), and 2 patients (18%), respectively. The tumors in 7 patients (64%) were predominantly cystic, while in 4 patients (36%), they were solid or had a small cystic component. Two patients (18%) underwent CSF shunting procedures prior to tumor surgery. Seven patients (64%) underwent surgery for tumor resection, while five experienced recurrences (45%), and four patients (36%) were treated with gamma knife alone. Tumor volume at the time of GKS ranged from 0.1 cm³ to 83 cm³ (mean: 17.53).

The tumor received a median dose of 17.33 Gy. Two patients (18%) died postoperatively.

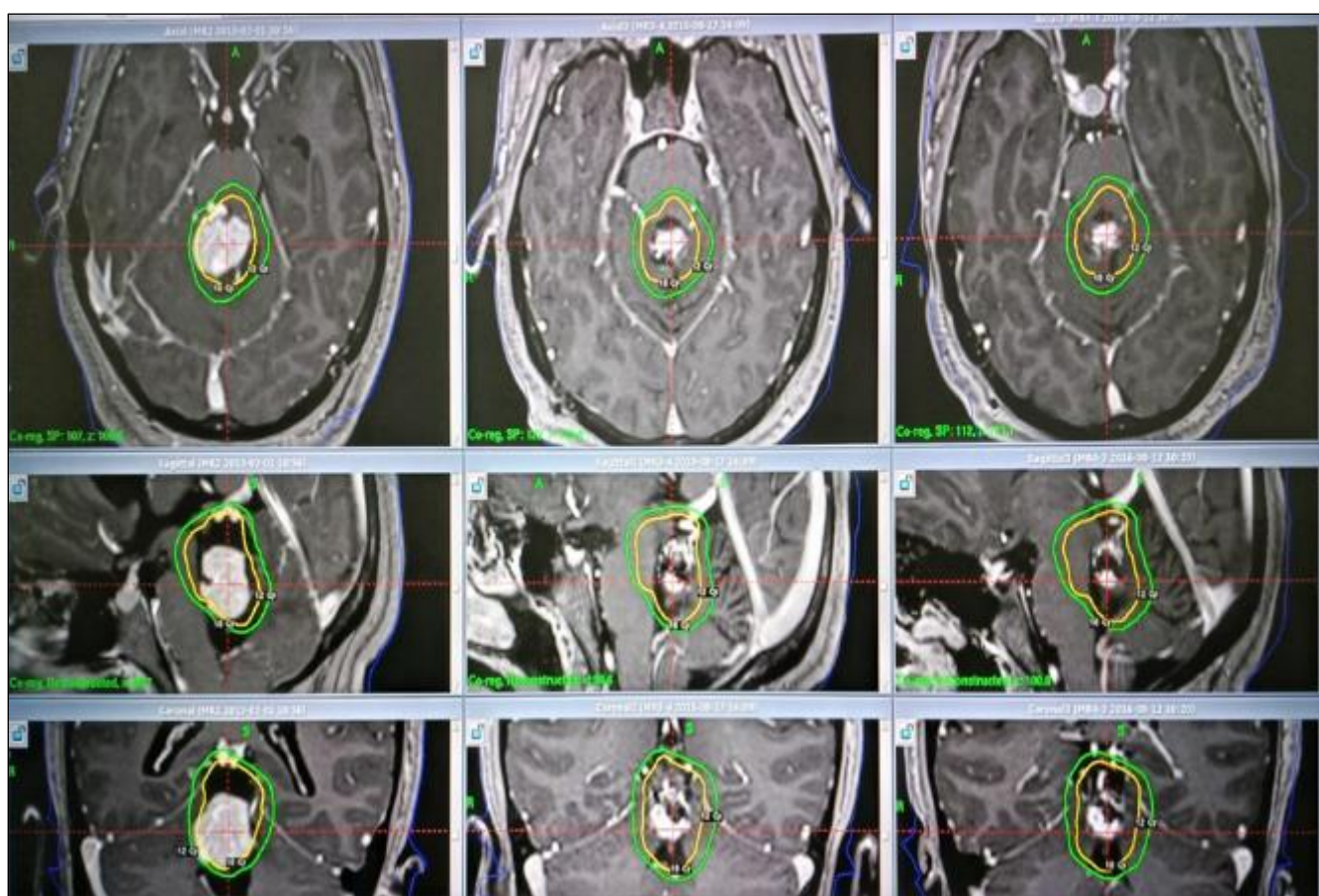


Figure 1: 26-year-old patient followed for VHL disease, treated by GK for solid Hemangioblastoma of the floor of V4 receiving a dose of 18 Gy, with last MRIs show a clear tumor regression.

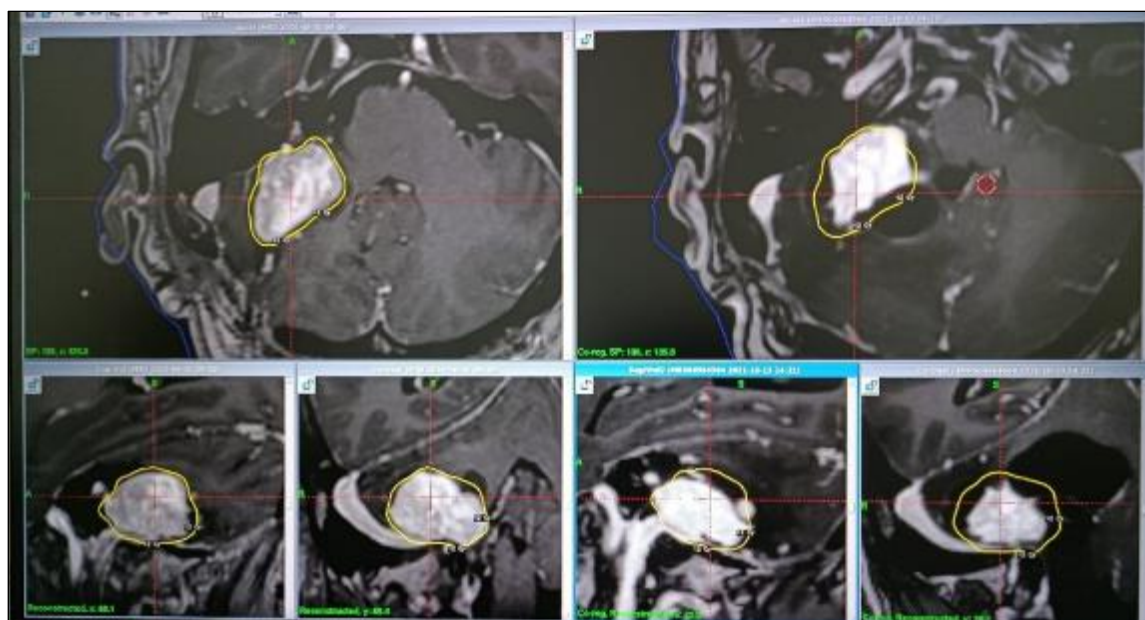


Figure 2: Case of 59-year-old patient followed for multi-operated hemangioblastoma, treated in 2015 by GK, control MRI in 2021 shows the appearance of a cyst opposite, then in 2022 we note an increase in the volume of this cyst which was punctured, followed by a GK session at a dose of 15 Gy on a volume of 24 cc.

4 DISCUSSION

Hemangioblastomas are benign tumors of the central nervous system that are well-circumscribed and extremely vascular. They only make up 7–12% of posterior fossa tumors and 1–2.5% of total cranial tumors. They are classified as WHO grade I tumors of unclear origin and are made up of mast cells, pericytes, stromal cells, and many capillaries [1].

According to radiological study, cerebellar hemangioblastomas are frequently divided into four types. Five percent of posterior fossa hemangioblastomas are type 1 cysts, which are simple cysts without a macroscopic bulge. Type 2, a cyst with a mural nodule, accounts for 60% of posterior fossa hemangioblastomas. According to reference [2], type 3 comprises solid tumors without a cystic component (26%) while type 4 comprises solid tumors with small internal cysts (9%).

Young adults are most commonly affected by hemangioblastomas. VHL syndrome is linked to over 20% [3]. Similar to findings in the literature, where the age ranged from 38 to 49.5 years, the age range in our study was 26 to 59 years (mean 42 years) [4]. Four of the patients (36,37%) in our study had VHL disease, which is concomitant with the 23–40% recorded in the literature [5].

Similar to previous research, the tumor was found in the brainstem (18%), vermillion (18%), and cerebellar hemispheres (54%), as well as the cavernous sinus (10%).

The presenting symptoms may include nystagmus, headache, and cerebellar indications, depending on where the lesion is. It is thought that this is because the neoplastic tissue secretes a chemical that resembles erythropoietin [6].

In most cases, symptoms develop gradually, allowing the treating physician to customize the treatment plan.

For hemangioblastomas, surgical excision has long been regarded as the preferred course of treatment [7]. The aim of surgery is total excision. It is not necessary to remove the cyst wall. By separating the gliotic borders from the little coagulated vascular pedicles, the solid nodular tumor mass is excised under a microscope. Large draining veins must be kept intact until the arterial feeders of the mural nodule have been identified and removed in order to prevent intraoperative edema. In order to minimize catastrophic hemorrhage, piecemeal removal must be avoided. Preoperative embolization has been recommended by certain authors [8].

For hemangioblastoma, recurrence is always the result of sub-total resection. After surgical resection, 20% of hemangioblastomas return, according to Conway et al. Hemangioblastomas can be radiosurgery because they typically have a tiny, clearly defined vascular mural nodule. A noninvasive, outpatient treatment called radiosurgery allows for the satisfactory tumor management of numerous lesions in a single session [4].

According to a study of the literature, radiosurgery for hemangioblastomas has shown promising results. In 64 lesions with at least two years of follow-up, Moss et al. found actuarial control rates of 84% and 81% after three and five years, respectively. Matsunaga et al. reported an 83.6% tumor control rate over an average of 63 months in their analysis of 67 cases. 13 patients with 46 hemangioblastomas had a 96% control rate at 10 years, according to Tago et al. [9].

However, SRS may not be an appropriate therapeutic choice for a patient who has a mass effect due to a cystic component because it only targets the solid portion of the tumor. Furthermore, research has shown that radiation might, at least temporarily, enhance vascular permeability, which can result in cyst development and peritumoral edema [10]. Three of five cystic hemangioblastomas underwent surgical evacuation following radiosurgery, according to Niemelä et al.'s analysis of 11 hemangioblastomas [11]. Five lesions had an increase in cyst volume following radiosurgery, and one of them required surgery to remove the cystic mass, according to Tago et al. [12]. While SRS was successful for hypervascular tumors, Pan et al. claimed that treatment was ineffective for cystic tumors [119]. Two of their 13 SRS patients experienced cyst growth, necessitating surgery. Six of these lesions were surgically drained and removed less than three months after SRS due to symptomatic cyst development, according to a more recent study by Moss et al. [13]. They predict that fluid collection will stop almost a year after SRS. Even after radiosurgery, two patients in our series who had type 2 hemangioblastomas with massive cystic lesions causing the cerebellar symptoms continued to experience symptoms.

Two patients in our series had type 2 hemangioblastomas with extensive cystic lesions that caused cerebellar symptoms, and their symptoms continued even after radiosurgery. These patients had surgery. The authors believe that, looking back, SRS and cyst aspiration could have been a preferable course of treatment for these two patients. For big cystic hemangioblastomas, Kano et al. suggest a similar treatment approach [14]. The authors believe that selective cases of type 2 hemangioblastomas and type 3, 4 hemangioblastomas are better candidates for SRS.

SRS is often a novel therapeutic approach for recurring instances, numerous tumors, unresectable lesions, and situations involving subtotal resection. Recurrent instances comprised five of our patients. There were several intracranial lesions in two. There are unclear indications for using SRS in hemangioblastomas. SRS is recommended by Kano et al. for unresectable tumors with mass effect (which may be followed by stereotactic cyst aspiration) and for resectable tumors following incomplete resection [14]. SRS is recommended by Moss et al. for asymptomatic hemangioblastomas smaller than 3 cm. They state that if an asymptomatic lesion in VHL is larger than 5 mm and exhibits radiological progression or a cystic component, SRS is recommended. Tago et al. found in their investigation that SRS has a greater effect on tiny lesions with high dose [9]. According to Kano et al., patients with solid tumors, tumor volumes less than 3.2 ml, and marginal doses greater than 15 Gy had improved prognoses. According to the authors, lesions with a maximum diameter of less than 3 cm and no discernible mass effect should be treated with gamma knives as the first line of treatment [13].

Hemangioblastomas are tumors that grow slowly and have a saltatory growth pattern. A period of quiescence could be the reason for the lack of growth observed at follow-up. As a result, it might be challenging to say with certainty whether SRS is truly responsible for the local tumor control rate when there is no reduction in size. According to Yamamoto et al., unless there was noticeable tumor growth prior to radiosurgery, the lack of growth would not imply treatment [15]. Therefore, before any survival benefit can be ascribed to SRS, long-term follow-up is necessary.

Five of the 13 patients treated by Tago et al. experienced recurrence outside the SRS-treated area. Of their 32 patients, 10 had new lesions outside the irradiation area, according to Kano et al. One patient in our investigation experienced a new lesion outside of the irradiation area, at the contralateral cerebellar hemisphere. Follow-up MRIs are performed at our institute six months following radiosurgery and subsequently once a year for the first five years [9].

In our study, two patients had undergone VP shunt for raised intracranial pressure prior to SRS. One of the patients treated by Page et al. had symptomatic radiation necrosis that required corticosteroid therapy and a VP shunt [16].

Two patients (18%) in the current series experienced postoperative problems, which is consistent with the literature that is currently available [17]. Meningitis and lower cranial nerve paresis were the most common postoperative morbidities.

Radiation poisoning is one of SRS's main concerns. When there are several lesions with overlapping radiation fields, it could be a serious issue. In their study, Moss et al. documented radiation necrosis in 11 malignancies [13]. Not every patient in our study experienced radiation harm as a result of radiosurgery.

The mortality rate in available literature has varied from 4-36% [17]. The mortality in this series was 18% (2 patients).

5 CONCLUSION

SRS is a viable treatment option for hemangioblastomas, particularly in cases that are unresectable, recurrent, numerous, or subtotally resected. This method effectively localizes treatment. However, tumors with cystic components may require careful monitoring due to potential cyst growth. Recent studies indicate that gamma knife surgery (GKS) is a low-complication procedure effective for stabilizing or reducing solid hemangioblastomas.

STATEMENTS ON COMPLIANCE WITH ETHICAL STANDARDS

Disclosure of conflict of interest

No conflict of interest to be disclosed.

Statement of informed consent

Informed consent and verbal permission were obtained from the patient before the submission of this article. In addition, this article follows both the Consensus-based Clinical Case Reporting Guideline and the Recommendations for the Conducting, Reporting, Editing, and Publication of Scholarly Work in Medical Journals.

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