

# Postoperative Management of The Primary Single Intracranial Rosai-Dorfman Disease: Case Report and Literature Review

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## Case report

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## Abstract

The isolated intracranial Rosai-Dorfman disease (RDD) is extremely rare. It remains unclear for the best management of the residual lesions. In this report, the authors describe a case of primary single intracranial RDD treated with Gamma knife radiosurgery after partial resection, review all the related literatures and summarize the postoperative management data, in order to provide useful therapeutic information.

A 23-year-old male presented with a more than 10-month history of dizziness, blurred vision and weakness. Preoperative MR imaging revealed a giant lesion involving the left middle cranial fossa and extending to the left cavernous sinus. He underwent fronto-temporal craniotomy and eventually achieved partial resection of the lesion. Postoperative histopathological results suggested a diagnosis of intracranial RDD. The residual lesion was treated with Gamma knife radiosurgery about 1 month after surgery. The follow-up imaging showed an obvious decrease in the size of the residual lesion.

We are the second to provide the detail parameters of gamma knife therapy, which may be a reference for clinical neurosurgeons. Compared with the firstly reported case, our patient with a longer follow-up time has different therapeutic dose. Based on our own experience and an updated literature review, adjuvant stereotactic radiosurgery may be a safe and effective treatment for primary single intracranial RDD patients with incomplete excision. Other adjuvant therapies can be as an option in refractory patients.

## Introductions

Rosai-Dorfman disease (RDD) is a rare non-Langerhans cell histiocytosis (nLCH), which is first described In 1965 by a French pathologist, Pierre Paul Louis Lucien Destombes. At that time, upon histologic analysis, he reported 4 children and young adults with lymphadenopathy and sinus histiocytosis[6]. In 1969, Juan Rosai and Ronald Dorfman analyzed 34 cases of the same entity under the name sinus histiocytosis with massive lymphadenopathy[12]. The classical symptom for patients with RDD is bilateral, massive, and painless cervical lymphadenopathy, but 43% of these patients manifest extranodal disease[5].

The central nervous system (CNS) is involved in less than 5% of cases, among which 75% occurs as intracranial and 25% as spinal lesions[23, 25]. Patients with intracranial RDD always go to see a doctor when they have the onset of symptoms which include headaches, seizures, gait difficulty, motor or sensory abnormalities, and cranial nerve deficits due to the mass effect. In some severe cases, the patients can have disturbance of consciousness[23]. In addition, it is difficult to differentiate the intracranial RDD with meningioma and lymphocytoma depending on the clinical demonstrations and radiologic imaging[23]. So, surgical operation has been considered as first therapy and thought to be essential for the diagnosis[1, 14, 23, 30]. Intracranial RDD can be solitary extraaxial dural masses, as well as parenchymal lesions, which may be adjacent to the vital cranial nerves and brain tissues, so most cases are difficult to achieve total removal[23]. And incomplete resection is associated with a higher rate of recurrence (20-25%)[9]. However, there is no standard postoperative management for the residual lesion, due to the rarity of the disease, short follow-up period and poor reporting of postoperative treatment data[16, 23]. Here, we report a case of primary single intracranial RDD treated with Gamma knife radiosurgery after partial resection, review all the related literatures and summarize the postoperative management data, in order to provide useful therapeutic information.

## Case Report

A 23-year-old male presented with more than a 10-month history of dizziness, blurred vision and weakness. These symptoms had worsen significantly 1 month before he went to see a doctor. On neurological examination, the patient was cooperative. He had loss of vision in rough measurement, and the vision of his left eye was worse than that of his right eye. There was no evidence of peripheral Lymphadenopathy. Rest of the neurological examinations didn't show any abnormalities.

Endocrinological evaluations also did not reveal any abnormalities. Preoperative magnetic resonance imaging (MRI) revealed a homogeneous, Gd-enhancing giant lesion involving the left middle cranial fossa and extending to the left cavernous sinus. The lesion was well-defined but irregular in shape and mild peritumoral edema could be observed (Fig. 1: A1-A4). The preoperative diagnosis was meningioma based on the radiological imaging.

He underwent maximal safe resection via frontotemporal craniotomy approach and achieved optic nerve decompression. But he eventually had partial resection because the lesion invaded the cavernous sinus and adhered closely to the left internal carotid artery (ICA). Intraoperative findings included a hard to firm, moderately vascular tumor, compressing the optic nerve, oculomotor trochlear nerve and trigeminal nerve more on the left side. Postoperative MRI also provided a proof for partial residual lesion in the left temporal fossa and cavernous sinus after 1 month of the operation [Fig. 1: B1-B4].

Postoperative histopathological report revealed that the tumor was composed of variable numbers of histiocytes intermixed with plasma cells and lymphocytes (Fig. 2: A1-B2). And these cells were distributed in focal and scattered manner. Some of the tissue cells were large, had abundant cytoplasm and showed features of emperipolesis (Fig. 2: B1 and B2). Immunohistochemical studies of the histiocytes showed strong positivity for S-100 protein, CD68/PGM-1, CD163, CD1a, and part EMA (Fig. 2: C-G). IgG4 was also positive and could be seen at high magnification (16/HPF) (Fig. 2: H). The results also showed that immune negativity for Langerin, PR, and GFAP (Fig. 2: I-K). It was negative for acid fast stain, PAS stain, and periodic acid-silver methenamine (Fig. 2: L-N). The results of TB/PCR test were negative. These pathological results suggested a diagnosis of intracranial RDD.

Gamma knife radiosurgery was applied in this patient about 1 month after the incomplete resection (The gamma knife therapeutic plan was showed in Fig. 3). The residual lesion was irregular in shape, extending along tentorial margin of cerebellum and extending from the petrous apex across the clivus and into the left cavernous sinus (Fig. 1: B1-B4). A combination of 11 isocenters of irradiation (tree collimators with a diameter of 14 mm and seven collimators with a diameter of 8 mm) was used. The 45% isodose volume enclosed the residual lesion and conformed closely to the shape of the residual mass. A maximum dose of 31 Gy was administered in the center of the lesion. No procedure related morbidity occurred.

The patient received clinical and imaging follow up after Gamma knife radiosurgery. After 3 months of the gamma knife therapy, the imaging showed an obvious decrease in the size of the residual lesion (Fig. 1: C1-C4). The same result could also be seen from the MRI 9 months after the gamma knife therapy (Fig. 1: D1-D4). Compared with the MRI of 9 months after the gamma knife therapy, the latest follow-up imaging (24 months after the gamma knife therapy) revealed the size of residual mass suspiciously increased a little (Fig. 1: E1-E4). But the patient had almost recovered from blurred vision, dizziness and weakness, and he had no new clinical symptoms. It's very gratifying that he had returned to his own work and normal life. So, we suggested that he could choose the treatment of a second gamma knife. He also could choose regular follow-up, if he didn't want the aggressive treatment. He decided to accept a second gamma knife after deep consideration.

## Discussion

The isolated intracranial Rosai-Dorfman is an extremely rare benign disorder. Almost all of the reported intracranial lesions appear to be dural-based, so preoperative imaging is difficult to identify RDD with meningioma [1, 9, 23]. The definitive diagnosis of RDD is generally made based on histopathologic immunohistochemistry of the surgical specimen. The typical characteristics of immunohistochemistry show S100 (+) and CD68 (+), and lesional histiocytes demonstrate variable frequency of emperipolesis [5, 6, 12, 16, 18, 20]. Operation is always applied to most patients with symptomatic intracranial RDD [16, 20, 23]. The goal of surgical resection is to relieve mass effect, to improve neurologic deficits and to obtain a diagnostic specimen. RDD is a benign tumor. When achieving gross total resection, patients generally have a good outcome without risk of recurrence [16, 23]. However, if the lesion is in technically difficult regions, invading or surrounding the critical structures, complete surgical resection is impossible. In this situation, subtotal or partial resection of the lesions should be done to provide the diagnostic specimen and symptomatic relief. Subtotal resection is associated with a higher rate of recurrence (20–25%) [9]. The long-term outcome were limited because only 37% of patients had more than one-year follow-up

information [1]. So, the true recurrence rate may be higher than 25%. It remains unclear for the best postoperative management of the residual lesions. The reason is that the disease is rare and almost all are presented in case report, which hampers the progress toward identifying an optimal treatment strategy[16, 20, 23]. Here, we reported a case of primary single intracranial RDD treated with incomplete resection and postoperative Gamma knife radiosurgery. In addition, we reviewed all related literatures, and the cases who had primary single intracranial RDD and underwent postoperative adjuvant treatment after incomplete resection were included. We hope that this will provide useful information for the postoperative therapy of intracranial RDD.

We searched English literature using PubMed and other search engines, which covered a period from 1969 to 2020. For the literature search, we used the keywords *Rosai-Dorfman disease, sinus histiocytosis with massive lymphadenopathy, and central nervous system*. In addition, we reviewed references cited in all the collected manuscripts to identify additional reports of RDD presenting with CNS involvement. At last, we read all the reported cases in those articles, and just 24 patients who had primary single RDD and the therapy of incomplete resection and postoperative adjuvant treatment were included (Table 1)[2-4, 7-11, 13, 15, 17-19, 21, 23-27, 29], including the patient reported here. The primary single intracranial RDD had been described in both adults and children, with a mean age at presentation of 32.09 years (range from 2 to 78 years) and a strong male prevalence (male/female: 2.8: 1) (Table 1).

As we can see from the Table 1, 11 (47.83%) patients accepted incomplete resection alone. During the follow-up period, 6 (6/11) patients achieved stable for the residual lesion and the remaining mass of 2 (2/11) cases appeared to be enlarged, apart from 3 (3/11) cases without follow-up information (Table 2).

Gamma knife radiotherapy has a good therapeutic effect on the residual intracranial RDD after incomplete resection. In 2003, Hadjipanayis CG firstly reported that the patient with primary single intracranial RDD underwent gamma knife radiosurgery for residual lesion 2 months after the partial resection[9]. For the patient, the maximum dose of 24 Gy was administered in the center of the residual mass, and the 50% isodose volume, which measured 6.4 cm<sup>3</sup>, enclosed the tumor and conformed closely to the shape of the tumor (Table 2). 13 months after the gamma knife surgery, the patient had improved clinical symptoms and decrease in the size of the residual tumor. Here, we reported the second case of primary single intracranial RDD. The patient was treated with maximum central dose of 31 Gy and 45% isodose line. 9 months after the gamma knife therapy, the imaging showed an obvious decrease in the size of the residual lesion. Compared with the MRI of 9 months after the gamma knife therapy, however, the imaging of 24 months after the gamma knife therapy revealed suspicious recurrence of the residual mass. We consider that the size increase may be caused by the bias of the visual effect. One reason is that the slice thickness and base line of MRI scanning are different between the two times. The other reason is that this patient had almost recovered from blurred vision, dizziness and weakness, and that he had no new clinical symptoms. It's very gratifying that he had returned to his own work and normal life. So, we suggested that he could choose the therapy of a second gamma knife. He also could choose regular follow-up, if he didn't want the aggressive treatment. He decided to accept a second gamma knife after deep consideration. We will continue the follow-up and report the following result of this patient.

To our knowledge, whole brain radiotherapy (WBRT) is not a good therapeutic option for patients with residual intracranial RDD after surgery, because of the poor therapeutic effect, the related complications or side effects. In the Table 1 and Table 2, 6 (26.09%) patients with primary single intracranial RDD underwent WBRT after incomplete resection. Symss NP[19] reported two cases with primary single intracranial RDD, a male patient and a female one. For the male patient, only subtotal resection of the mass could be obtained because it was too extensive. Six months after the operation, He presented recurrence of clinical symptoms and the imaging showed progression of the residual lesion. Then the patient received WBRT (20 Gy). At his routine follow-up for 8 years, he didn't get improved clinical symptoms and repeated imaging demonstrated the increase in the size of the residual mass (extending into the cavernous sinuses up to the orbit). For the female patient, she only achieved subtotal excision of the tumor, because the mass was densely adherent to the hypothalamus and arachnoidal adhesions were existed. She underwent WBRT (20Gy) following the operation. One year later, she had the complication of hydrocephalus and required a ventriculoperitoneal shunt. Two years later, she was symptom free and the imaging showed

further resolution of the lesion. But there was no detail therapeutic dose or follow-up information for the rest 4 reported cases who underwent WBRT after operation (Table 2) [10, 18, 21, 29].

For the complicated cases, systemic chemotherapy can be selected. But we need to take obvious and severe side effects into consideration, especially for pregnancy or young patients [22, 28]. Penetration of the blood-brain barrier will affect the therapeutic effect, so we should consider this factor when we decide to choose which kind of chemotherapy agents [28]. Meanwhile, the response of chemotherapy is not uniform. There are two reports about 2 (8.69%) patients with a challenging management (incomplete resection, WBRT and Chemotherapy/ Steroid) (Table 1 and Table 2). In 2017, Das S [3] reported a male patient with primary single intracranial RDD in the suprasellar cistern. The patient had maximal safe resection and postoperative contrast MRI showed minimal dural enhancement along the right temporal lobe. Symptomatic stable for 1 year, thereafter, he presented with gradually progressive symptoms and the repeated MRI suggested the recurrence of the lesion. He took steroids for two weeks, but his symptoms became increasingly severe. Therefore, a total dose of 20 Gy in 10 fractions over 2 weeks was delivered to the mass by three-dimensional conformal radiotherapy (3D-CRT). He had improvement in hearing. 20 months after completion of radiotherapy, the patient was considered for systemic chemotherapy in terms of progressive and diffuse leptomeningeal disease. But the chemotherapy strategy and the following outcome were not reported in the literature. In 2014, Sandoval-Sus JD [23] reported a female with primary single intracranial RDD originating from the clivus. She had subtotal resection of the lesion and postoperative adjuvant radiotherapy (a total dose of 45 Gy in 1.8 Gy daily fractions), and achieved improvement of the hemiparesis. The patient suffered recurrence of the lesion three years later. She underwent locally re-irradiated (45 Gy) followed by 1 cycle of adjuvant temozolomide (75 mg/m<sup>2</sup>). Three years after the chemotherapy, she had improved symptoms and the follow-up imaging demonstrated no progression of the tumor.

Steroid has been used in a few reports with modest efficacy. Md. Taufiq [25] reported a male with a huge, irregular lesion in the parasellar region. The patient was treated with incomplete removal of the space occupying lesion and steroid following the operation. About two years later, he got improvement of some symptoms. In 1993, Shaver EG [24] reported a boy with a lesion in the left cavernous sinus and extending over the tentorial margin into the posterior fossa. The boy achieved subtotal excision of the lesion. Postoperatively, he took 4 mg dexamethasone every 6 hours for 1 week and then the dose decreased by 50% each week for 6 weeks. After 1 month, the boy got resolution of the mass and clinical symptoms.

In 2018, there were consensus recommendations for the diagnosis and management of RDD which were collaboratively put forward by a group of pathologists, pediatric and adult oncologists, hematologists, and internists [20]. It's a pity that neurosurgeons didn't take part in this consensus recommendations. They recommended that the use of surgery, corticosteroids, chemotherapy, immunomodulatory therapy, targeted therapy, and radiotherapy for patients with intracranial RDD should be discussed in further details. Sandoval-Sus JD [23] proposed that surgery should be the first therapeutic option for symptomatic patients with intracranial RDD, and a gross total resection of the lesion should be attempted if possible. For intracranial RDD patients with incomplete resection, the optimal postoperative adjuvant therapy was still unclear, due to the lack of randomized prospective data on therapeutic approaches and short follow-up period [16, 20, 23]. Based on our own experience and an updated literature review, adjuvant stereotactic radiosurgery may be a safe and effective treatment for primary single intracranial RDD patients with incomplete excision. Other adjuvant therapies can be as an option in refractory patients.

## Conclusion

It remains a common problem for recurrence after surgery of patients with primary single intracranial RDD, which is usually difficult to manage. The role of postoperative management is not well defined due to the rarity of this lesion. We report one case of our own experience, review all the related literatures, and summarize the postoperative therapeutic approaches and related therapeutic dose. We hope that this will provide valuable information in therapeutic options for these patients. However, to develop rational and normalized therapy strategies to improve the prognosis of these patients, international multicenter should collaborate to provide enough and valuable data.

## Abbreviations

RDD: Rosai-Dorfman disease; nLCH: non-Langerhans cell histiocytosis; CNS: The central nervous system; MRI: magnetic resonance imaging; ICA: internal carotid artery; WBRT: whole brain radiotherapy; 3D-CRT: three-dimensional conformal radiotherapy

## Declarations

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### Author Contributions

Wei Wang: Study concept and design

Xiaowei Liu: Review all the literatures, have access to the data and draft the manuscript

Yangyang Xu, Yang Wu, Rong Wen, Yuan Gao: help collect some of the data and revise the manuscript

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### Availability of Data and Materials

Please contact author for data requests.

### Ethics Approval and Consent to Participate

In this report, the subject has given his written informed content to publish his case (including publication of images).

### Competing Interests

The authors declare that they have no competing interests.

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## Tables

Table 1  
 presentation and management of 23 patients with primary single  
 intracranial Rosai-Dorfman disease as reported in the literature.

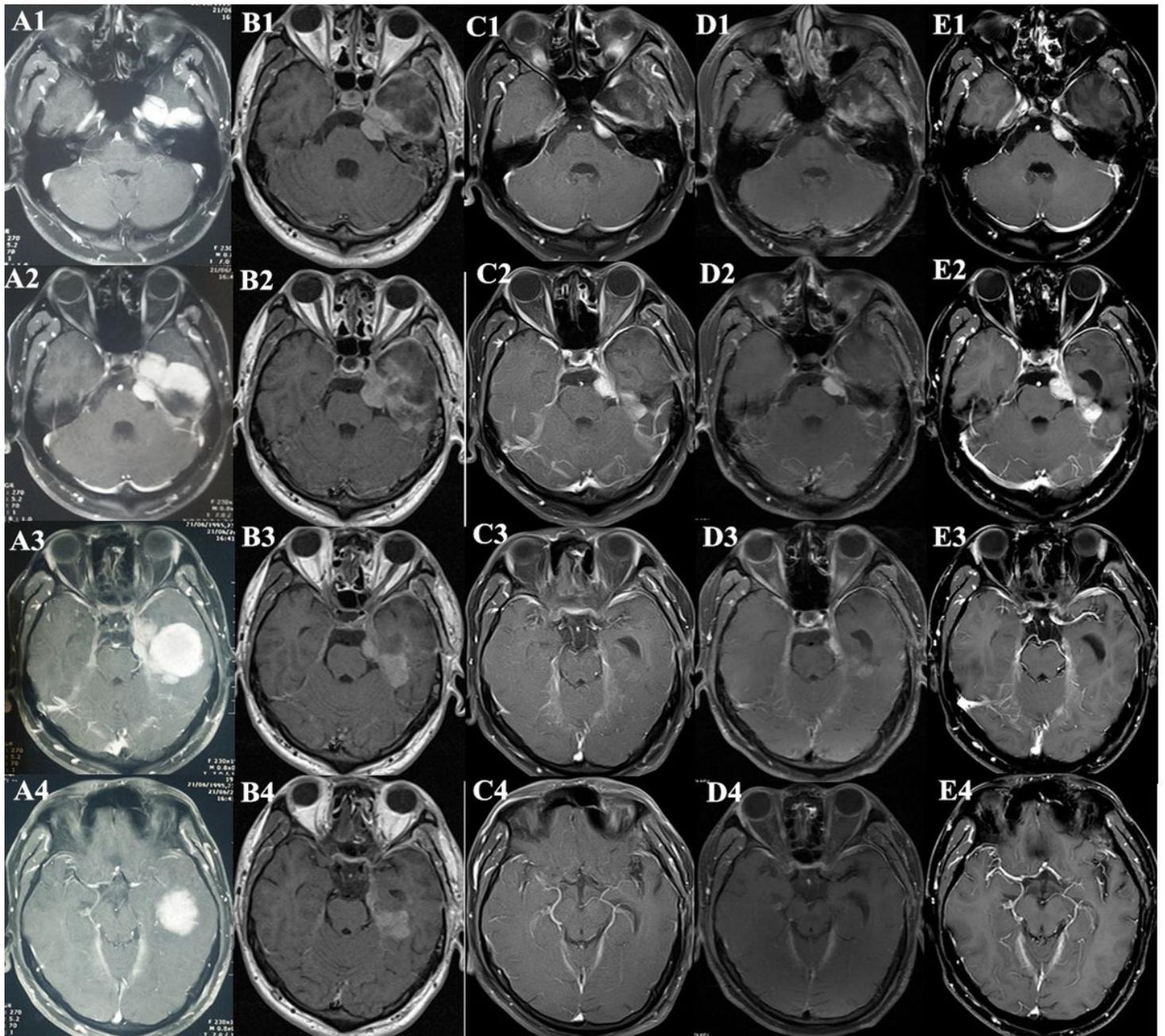
<b>Male, n (%)</b>	<b>17 (73.91%)</b>
<b>Female, n (%)</b>	<b>6 (26.09%)</b>
Mean age at presentation (range) (years)	32.09 (2–78)
Management	11 (47.83%)
Incomplete (subtotal + partial) resection alone	2 (8.69%)
Incomplete resection and λ Gamma knife radiosurgery	6 (26.10%)
λ conventional radiotherapy	2 (8.69%)
λ conventional radiotherapy and Chemotherapy/ Steroid	2 (8.69%)
λ Steroid	
Follow-up time	4 (17.39%)
≤ 1 year	11 (47.83%)
1–3 years	4 (17.39%)
> 3 years	4 (17.39%)
Not available	
Prognosis	14 (60.87%)
Stable disease/decrease	5 (21.74%)
Relapse/growth	4 (17.39%)
Not available	

Table 2  
Summarize the detail data of reported cases in the literatures

	age(Y) /sex	postoperative management				follow-up period	follow-up results
		Gamma knife	conventional radiotherapy	Chemotherapy	Steroid		
Joshi SS, 2017	58, M	no	no	no	no	60 months	stable
Huang BY, 2016	18, M	no	no	no	no	36 months	stable
Lüdemann W,2015	2, F	no	no	no	no	16 months	stable
Varan A, 2015	5, M	no	no	no	no	6 months	recurrence
Kumar YA, 2014	43, M	no	no	no	no	NA	NA
Gupta K, 2011	14, M	no	no	no	no	NA	NA
Di Rocco F, 2007	13, F	no	no	no	no	3 months	recurrence
		no	no	no	no	12 months	stable
Gupta DK, 2006	15, M	no	no	no	no	12 months	stable
Toh CH, 2005	60, F	no	no	no	no	4 months	stable
	59, M	no	no	no	no	30 months	stable
Bhattacharjee MB, 1992	35, M	no	no	no	no	NA	NA
Hadjipanayis CG, 2003	52, M	maximum dose of 24 Gy, 50% isodose volume 13 isocenters, 8-mm- diameter collimators				13 months	decrease
Our case	23, M	maximum dose of 24 Gy, 45% isodose volume, 13 isocenters 8-mm- and 14-mm- diameter collimators				24 months	decrease

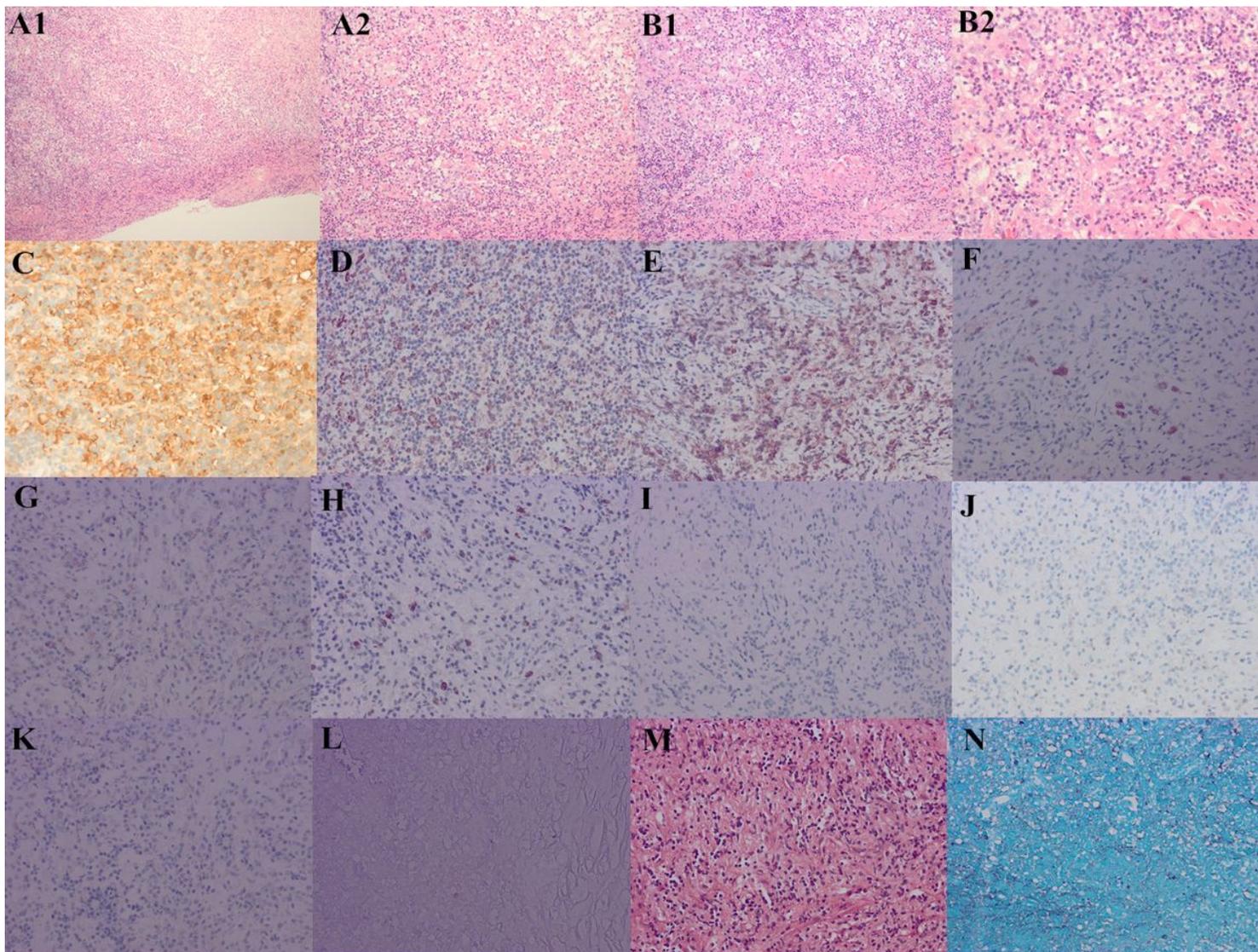
	age(Y) /sex	postoperative management		follow-up period	follow-up results
Hollon T, 2016	26, M	the dose NA		24 months	decrease
Symss NP, 2010	21, M	20 Gy		96 months	recurrence
	17, F	20 Gy		36 months	decrease
Walid MS, 2010	60, F	the dose NA		NA	NA
Petzold A, 2001	78, M	the dose NA		120 months	stable
Trudel M., 1984	28, M	the dose NA		14 months	stable
Das S, 2017	50, M	total dose of 20 Gy in 10 fractions, over 2 weeks	corticosteroids	20 months	recurrence
Sandoval-Sus JD, 2014	32, F	total dose of 4500 cGy in 180 cGy daily fractions	Temozolomide 75 mg/m2, one cycle	72 months	stable
Md. Taufiq, 2016	24, M		steroid: the dose NA	24 months	stable
Shaver EG, 1993	5, M		dexamethasone:the dose NA	1 month	stable
NA: not available; Y: years					

## Figures



**Figure 1**

Preoperative MRI revealed a homogeneous, Gd-enhancing giant lesion involving the left middle cranial fossa and extending to the left cavernous sinus (A1-A4). Postoperative MRI showed the residual lesion was irregular in shape, extending along tentorial margin of cerebellum and extending from the petrous apex across the clivus and into the left cavernous sinus (B1-B4). The imaging showed an obvious decrease in the size of the residual lesion (C1-C4 and D1-D4). Compared with the MRI of 9 months after the gamma knife therapy, the latest follow-up imaging (24 months after the gamma knife therapy) revealed the size of residual mass suspiciously increased a little (E1-E4).



**Figure 2**

HE showed the tumor was composed of variable numbers of histiocytes intermixed with plasma cells and lymphocytes (A1-HPF×100; A2-HPF×200). HE showed obvious feature of lymphocyte emperipolesis(B1-HPF×200; B2-HPF×400). Immunohistochemistry showed strong positivity for S-100 protein, CD68, CD163, CD1a and EMA (C-HPF×400, D-HPF×400, E-HPF×400, F-HPF×400 and G-HPF×400, respectively). IgG4 was also positive and could be seen at high magnification under an electronic microscope (16/HPF) (H-HPF×400). Immunohistochemistry also showed that immune negativity for Langerin, PR and GFAP (I-HPF×400, J-HPF×400 and K-HPF×400, respectively). It was negative for acid fast stain, PAS stain and periodic acid-silver methemamine (L-HPF×400, M-HPF×400 and N-HPF×100, respectively).

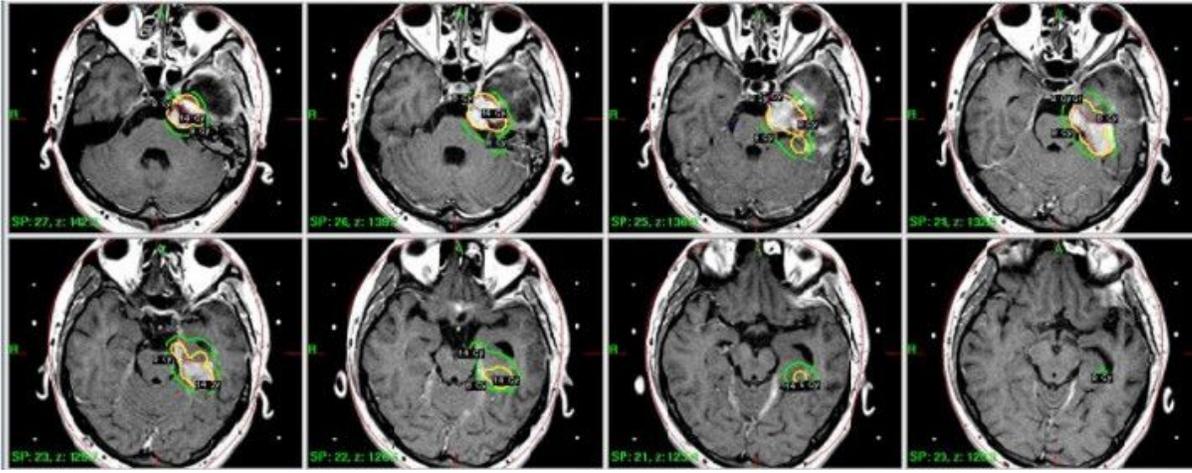


Figure 3

Leksell Gamma Plan images with a total of 11 isocenters in which 14-mm-diameter and 8-mm-diameter were used.