Multimodality management of rare solitary fibrous tumor can be associated with extended survival

Thomas Mindermann, Robert Reisch

Neurosurgery Klinik Im Park, †Neurosurgery Klinik Hirslanden, Zurich, Switzerland

E-mail: *Thomas Mindermann - tmindermann@hin.ch; Robert Reisch - robert.reisch@hirslanden.ch
*Corresponding author

Received: 23 September 14  Accepted: 30 October 14  Published: 30 December 14

Abstract

Background: Little is known on the long-term course of patients treated for intracranial solitary fibrous tumors (iSFT). We therefore retrospectively reviewed the charts of our patients who underwent Gamma Knife radiosurgery (GKRS) for iSFT at Klinik Im Park in Zurich and who were treated by one of the authors. Between 1994 and 2009, two patients underwent GKRS for iSFT at Klinik Im Park.

Case Description: One patient underwent altogether five radiosurgical treatments and two craniotomies for iSFT and its local recurrences. The other patient underwent two craniotomies and one radiosurgical treatment for iSFT. Both patients maintained a Karnofsky performance score 100 during follow-up and both were long-term survivors with a follow-up of 9 and 17 years, respectively.

Conclusion: A close follow-up of patients with iSFT and repeat radiosurgery or surgery when indicated seems to lead to a favorable long-term outcome.

Key Words: Intracranial, metastasis, radiosurgery, surgery, solitary fibrous tumor

INTRODUCTION

Intracranial solitary fibrous tumors (iSFT) are rare tumors. Series with 5, 18, and 24 patients have been published[1,4,8] as well as several case reports with 1–2 patients.[2,3,5‑7,9] Two of the case reports present the outcome following radiosurgery[5,6] in a total of three patients. The longest follow-up (FU) in those three patients was 4 years. In order to add to our understanding of iSFT, we retrospectively reviewed our patients with iSFT who underwent single-session radiosurgery.

METHODS

We retrospectively reviewed the patient charts of the patients who underwent Gamma Knife radiosurgery (GKRS) for iSFT at Klinik Im Park, Zurich between 1994 and 2009 and whose surgical- or radiosurgical-treatments were performed by one or both of the authors (TM, RR). The diagnosis of iSFT was based on histology.

RESULTS

From 1994 to 2009, two patients underwent GKRS for iSFT at Klinik Im Park.

Case 1
A 57-year-old male patient initially underwent GKRS in 1997 for what was believed to be a parasagittal meningioma located over the right central region. Throughout the 17-year FU, the patient underwent altogether five radiosurgical treatments and two craniotomies for iSFT and its local recurrences. The choice of open surgery or radiosurgery depended on the
patient’s preferences. In 2005, 8 years after the initial GKRS, the patient underwent the first craniotomy for local tumor recurrence. The histological diagnosis was then meningioma WHO I. In 2010, 13 years after the initial GKRS, the patient underwent another craniotomy for tumor recurrence. Histology then revealed an SFT. Immunohistochemistry showed positive expression for Vimentin, CD34, and CD99 and negative expression for Epithelial membrane antigen, Carcinoembryonic antigen (EMA) Pancreatin, and S-100. The proliferation marker MIB 1 was expressed in 2–5% of the nuclei. The previous histology was reevaluated and the diagnosis of SFT was retrospectively confirmed for the initial surgery. The patient developed liver metastases 13 years after the initial GKRS. The liver metastases were first biopsied and then removed in liver segments II, VI, VII, and VIII. Histology was compatible with SFT: Positive expression for CD34, CD99, and BCL2; negative expression for CD20, CD3, smooth muscle Actine, S-100, HMB-45, WT1, Lu-5, CD10, CD138, Inhibine, DOG-1, Desmine, Synaptophysine, CD117, and EMA. A total of 13.5 years following GKRS, pulmonary metastases were discovered on computed tomography (CT) scan. The patient did not undergo biopsy or surgery for the pulmonary metastases. The patient did not undergo any additional systemic oncological therapy or any radiotherapy for the distant metastases since it was felt that there were no promising local or systemic therapies to be offered. Therefore, the patient underwent a wait and scan FU regime for the distant metastases. The radiosurgical procedures were done with two different technologies since the hospital changed the radiosurgical equipment from Gamma Knife to CyberKnife in 2009. Progression-free survival (PFS) following the various radiosurgical procedures was 2–5 years and 0.75–2 years following the various open surgical procedures. The patient is now 74 years old and alive. He maintained a Karnofsky performance score (KPS) 100 during the 17-year FU. At present, he has a slight spasticity of his left leg and is still skiing. The details of the clinical course are listed in Table 1, the tumor characteristics in Table 2, the radiosurgical treatment characteristics in Table 3, and the planning images for the last radiosurgical treatment with the CyberKnife are illustrated in Figure 1.

Case 2

A 70-year-old female patient had a history of resection of a malignant melanoma of the right thigh and postoperative local radiotherapy when she was 55 years old. At age 70, she underwent resection for a tumor of the right tentorium. Histology revealed an SFT with positive expression for CD34 and negative staining for EMA and Glial fibrillary acidic protein (GFAP). Throughout the 9-year FU, she underwent two craniotomies and one radiosurgical treatment for iSFT. PFS was 3 years following the initial surgery. PFS was at least 2 years following GKRS; the patient did not show up for FU until 6 years after GKRS. Nine years after the initial surgery and 6 years after GKRS, a large local tumor recurrence was seen on MRI and the patient opted for another craniotomy. Histology revealed again

| Case | Age, gender | Treatment | Year of treatment | PFS after each treatment (y) | Total FU (y) | Status, KPS |
|------|-------------|-----------|------------------|----------------------------|-------------|-------------|
| 1    | 57, male    | GKRS      | 1997             | 5                          | 17          | Alive, 100  |
|      |             | GKRS      | 2002             | 3                          |             |             |
|      |            | Surgery   | 2005             | 0.75                       |             |             |
|      |             | GKRS      | 2005             | 4                          |             |             |
|      |             | Surgery   | 2010             | 2                          |             |             |
|      |             | CKRS      | 2012             | 2                          |             |             |
|      |             | CKRS      | 2014             | NA **                      |             |             |
| 2    | 70, female  | GKRS      | 2005             | 3                          | 9           | Alive, 100  |
|      |             | Surgery   | 2014             | NA **                      |             |             |

GKRS: Gamma Knife radiosurgery, CKRS: CyberKnife radiosurgery, NA: Not applicable, PFS: Progression-free survival, FU: Follow-up *The patient did not show up for FU until 6 years following GKRS, **The last treatment was too recent to determine PFS, iSFT: Intracranial solitary fibrous tumors, KPS: Karnofsky performance score

| Case | Initial tumor location | Location at recurrence | Volume (cc) |
|------|------------------------|------------------------|-------------|
| 1    | Parasagittal right central | Local                  | 21.1        |
|      |                        | Local                  | 1.2         |
|      |                        | Local                  | NA          |
|      |                        | Local                  | 0.9         |
|      |                        | Local                  | NA          |
|      |                        | Local                  | 1.4         |
|      |                        | Local                  | 0.2         |
| 2    | Right tentorium        | Local                  | NA          |
|      |                        | Local                  | 1.2         |
|      |                        | Local                  | NA          |

NA: Not applicable because the tumor volume was not measured at surgery. iSFT: Intracranial solitary fibrous tumors

| Case | Treatment | Tumor volume (cc) | PD (Gy) | Time interval from initial tumor diagnosis (y) |
|------|-----------|-------------------|--------|---------------------------------------------|
| 1    | GKRS      | 21.1              | 14@50% isodose | NA *                                       |
|      | GKRS      | 1.2               | 15@50% isodose | 5                                           |
|      | GKRS      | 0.9               | 13@50% isodose | 8                                           |
|      | CKRS      | 1.4               | 12@70% isodose | 15                                          |
|      | CKRS      | 0.2               | 14@73% isodose | 17                                          |
|      | GKRS      | 1.2               | 14@50% isodose | 3                                           |

GKRS: Gamma Knife radiosurgery, CKRS: CyberKnife radiosurgery, PD: Prescription dose, NA: Not applicable, *Not applicable because GKRS was the initial treatment, iSFT: Intracranial solitary fibrous tumors
an SFT with positive expression for CD34 and negative staining for EMA and GFAP. The patient is alive and is now aged 79 years. She maintained a KPS 100 during the 9-year FU with no focal neurological deficits. The details of the clinical course are listed in Table 1, the tumor characteristics in Table 2, the radiosurgical treatment characteristics in Table 3, and the planning images for the radiosurgical treatment with the Gamma Knife are illustrated in Figure 2.

DISCUSSION

To our knowledge, this is the longest FU of patients who underwent surgery and radiosurgery for iSFT. It seems that a close FU and a combination of surgery and radiosurgery lead to promising results in such patients. Small tumor recurrences may be best treated by radiosurgery and larger tumor recurrences by open microsurgery even though a rather large tumor volume of 21.1 cc was treated successfully by GKRS in Case 1, which was followed by a 5-year PFS. Tumor volumes treated with radiosurgery ranged from 0.2 to 21.1 cc. In our patients, a prescription dose of 12-15 Gy to the tumor margin lead to a PFS for intervals ranging from 2 to 5 years. Repeat radiosurgery was tolerated without any problems. One of our two patients developed distant metastases 13 years after the occurrence of iSFT. SFT seem to have a high tendency for local tumor recurrence. SFT seem to respond well to radiosurgery and they seem to have a rather good long-term prognosis. We therefore advocate for a close FU of patients with iSFT. Whenever tumor recurrence is evident on MRI, patients should be offered radiosurgery or open microsurgery.

CONCLUSION

Both patients with iSFT are long-term survivors with a FU of 9 and 17 years, respectively. Patients with iSFT seem to have a rather good prognosis if they are followed closely with MRIs and if tumor recurrences are treated when they become evident. A combination of surgery and radiosurgery seems to lead to a favorable long-term outcome in patients with iSFT.

REFERENCES

1. Chen H, Zeng XW, Wu JS, Dou YF, Wang Y, Zhong P, et al. Solitary fibrous tumor of the central nervous system: A clinicopathologic study of 24 cases. Acta Neurochir 2012;154:237-48.
2. Kasper E, Boruchow S, Lam FC, Zinn PO, Anderson M, Mahadevan A. Hitting all the right markers to save a life: Solitary fibrous tumors of the central nervous system: Case series and review of the literature. Surg Neurol Int 2012;3:83.
3. Macfarlane RG, Galloway M, Plowman PN, Thomas DG. A highly vascular intracranial solitary fibrous tumor treated with radiotherapy and toremifene: Case report. Neurosurgery 2005;56:E1378.
4. Metellus P, Bouvier C, Guyotat J, Fuentes S, Jouvet A, Vasiljevic A, et al. Solitary fibrous tumors of the central nervous system: Clinicopathological and therapeutic considerations in 18 cases. Neurosurgery 2006;60:715-22.
5. Nakahara K, Yamada M, Shimizu S, Fuji K. Stereotactic radiosurgery as adjuvant treatment for residual solitary fibrous tumor. Case report. J Neurosurg 2006;105:775-6.
6. Reames DL, Mohila CA, Sheehan JP. Treatment of intracranial solitary fibrous tumors with gamma knife radiosurgery: Report of two cases and review of literature. Neurosurgery 2011;69:E1023-8.
7. Son S, Lee SG, Jeong DH, Yoo CJ. Malignant solitary fibrous tumor of tandem lesions in the skull and spine. J Korean Neurosurg Soc 2013;54:246-9.
8. Vassal F, Manet R, Forest F, Camdessanche JP, Péoc’h M, Nuni C. Solitary fibrous tumors of the central nervous system: Report of five cases with unusual clinicopathological and outcome patterns. Acta Neurochir 2011;153:377-84.
9. Yin W, Ma C, Wu J, Cai B, You C. A primary atypical solitary fibrous tumor of the sella mimicking nonfunctional pituitary adenoma: A case report. Acta Neurochir (Wien) 2010;152:519-22.