CASE REPORT

Treatment of orbital solitary fibrous tumour with gamma knife radiosurgery and systematic review of literature

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SUMMARY

Solitary fibrous tumours (SFTs) are relatively rare tumours that were originally thought to arise from the pleura but have thereafter been demonstrated as occurring anywhere in the body. These tumours are generally considered benign but have frequently been noted for recurrence and local invasion. Furthermore, their indolence is controversial due to increasing evidence implicating the existence of a spectrum that includes hemangiopericytoma (HPC). Stereotactic radiosurgery (SRS) has been well characterised in the treatment of benign, malignant and vascular conditions, and it appears to be a reasonable option as adjuvant or recurrent treatment for intracranial SFTs. We present in this case the first complete description of an SFT of the orbit treated by SRS as well as a systematic review of available English literature for intracranial SFTs treated by SRS. We report effective local tumour control in our case and conclude that SRS is a reasonable treatment option for recurrent SFT.

BACKGROUND

Solitary fibrous tumours (SFTs) were classically thought to be tumours arising from the pleura. However, after abundant case reports and reviews of extrapleural SFTs, they are now recognised as tumours that can occur anywhere in the body.¹ Intracranial SFTs are relatively rare and characterised as indolent tumours with generally benign behaviour with recurrence more commonly associated with subtotal resection and rare malignant transformation.² Stereotactic radiosurgery (SRS) has been increasingly utilised in the past decade for the treatment of benign and malignant conditions as well as vascular malformations. Rapidly growing, early-responding lesions (ie, malignant primary tumours and brain metastases); slowly growing, late-responding lesions (benign tumours of the bone, pituitary, meninges or Schwann cells) and functional disorders (pain, essential tremor, etc) have shown to benefit from SRS in terms of local tumour control rates and symptom management, respectively.³ SRS utilises precise and calculated doses of radiation to a tightly conformed treatment plan to destroy target lesions without damage to adjacent cells.⁴ A role for SRS in the treatment of SFTs has already been posited: it appears to be a reasonable option as adjuvant treatment following incomplete resection or for the treatment of recurrence, although there remains a dearth of characterisation and evidence in this area.^{2 5 6} Further, since

the description of the first SFT of the orbit in 1994, only one treated by radiosurgery has been described in English literature to the extent of our knowledge, and the details were provided in aggregate with a case series.^{7 8}

CASE PRESENTATION

The patient presented as a right-handed male aged 33 years in the summer of 2002 with symptoms of decreased vision in the right eye accompanied with protrusion and pressure for the previous few months. An orbital tumour was found and histologically diagnosed as SFT after subtotal resection in August 2002. At the time of initial surgery, the patient's visual acuity was severely impaired (20/200 OD and 0/10 on colour palettes), and he showed dramatic improvement in vision and symptoms (up to 20/20 OD with correction and 9/10 on colour). He was followed without further resection due to concern regarding tumour adherence to the optic nerve. In 2005, he developed severely worsening vision and proptosis of the affected eye and had a second subtotal resection. Pathology confirmed a spindle cell neoplasm of mesenchymal origin (positive for CD34 and negative for S100, a neuroectodermal marker) and diagnosed it as recurrent SFT. Postoperatively, he had no light perception in the right eye and developed ophthalmoplegia and ptosis. In 2006, follow-up imaging showed recurrent tumour with two distinct foci, which demonstrated interval slow growth over the years (figure 1A, B). He had a reconstructive cranioplasty along with exploration for the midorbital tumour in 2013.

TREATMENT

The patient was referred to gamma knife radiosurgery (GKRS) and was treated in June 2014 for two discrete areas of recurrent SFT in the orbit. Target volume was 1.69 and 0.834 cm³; both were treated with a margin dose of 16.0 Gy to the 50% isodose line (figure 2).

OUTCOME AND FOLLOW-UP

On follow-up, the patient remained stable clinically with unchanged ophthalmological and neurological examination and without any additional symptoms or new symptoms. Both treated lesions showed effective tumour control on last follow-up, 18 months post-GKRS. The lesions regressed in volume to measure 1.17 and 0.27 cm³, respectively (figure 1 C, D vs E, F).

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Figure 1 Sample patient follow-up. MRI T1WI fat saturation sequences. (A and B) May 2013, status post-two surgical resections (2002, 2005). The observed tumour shows growth and local recurrence. (C and D) June 2014 at the time of gamma knife radiosurgery. (E and F) November 2015, 18 months follow-up showing tumour volume regression.

Figure 2 Sample patient gamma knife treatment plan. MRI T1WI with gadolinium in axial (A), coronal (B), sagittal (C) and 3D representation of the treatment volume (D). (E) Skull geometry. (F) Histogram for each target with treatment parameters. Two targets, 5.95 and 9.45 cm³, were treated with a margin dose of 16 Gy to the 50% isodose line.

Table 1 Solitary fibrous tumours treated by radiosurgery: systematic review of the literature

Author	Age, sex	Tumour location	Treatment summary	Tumour volume	Margin, max dose (Gy)	Histology	Post-SRS recurrence	Outcome and F/u*
Caroli <i>et al</i> (2004) ¹⁰	29, F	Cerebellar tentorium	STR; LINAC	–	20, –	Typical fibrous SFT	No	Alive; 36 months
Nakahara <i>et al</i> (2006) ⁵	50 F	Occipital	STR; SRS	1.25 cm ³	21, 24	Typical fibrous SFT	No	Alive; 48 months
Metellus <i>et al</i> (2007) ¹³	67 F	Cerebellar tentorium	STR; GKRS	–	–	SFT with partial HPC characteristics	No	Alive, 10 months
Yin <i>et al</i> (2010) ⁶	32 M	Sellar	STR+GKRS	3.2×2.6×2 cm	–	SFT with partial HPC characteristics	No	Alive, 44 months
Bisceglia <i>et al</i> (2011) ⁹	76 M	Posterior fossa	GTR; GKRS; GTR; RT	–	–	Typical fibrous SFT	Yes	Deceased, 8 years
Reames <i>et al</i> (2011) ¹⁵	62 F	Sagittal sinus	NTR; STR; GKRS	–	13.5, 27	Typical fibrous SFT	No	Alive, 20 months
	37 F	Posterior fossa	Crani (6); GKRS (2)	0.135 cm ³ ; 1.70 cm ³	22, 44; 20, 40	Typical fibrous SFT	Yes	Alive, 15 months
Vassal <i>et al</i> (2011) ¹⁷	66 F	Sellar	STR; STR; STR+GKRS; STR	–	14	SFT with atypical (increased) proliferation	Yes	Deceased; 52 months
	39 F	CPA	STR; STR+GKRS; GKRS	2.5×2.5×1.5 cm	14	SFT with atypical (increased) proliferation	Yes	Alive, 45 months
Chen <i>et al</i> (2012) ¹¹	55 M	Cavernous sinus	STR; STR+RS	–	–	Typical fibrous SFT	–	–
	51 F	Cerebellar tentorium	GTR; RS	–	–	SFT with partial HPC characteristics	Yes	–
	11 M	Parafalcine	STR+RS	–	–	SFT with partial HPC characteristics	No	Alive, 23 months
	39 F	Pineal gland	STR; RS	–	–	Fibrous SFT with atypical S100+	–	Alive, 6 months
Jo <i>et al</i> (2012) ⁷	–	Orbit	Multisession GKRS	–	–	–	–	Alive, 5 months
Kasper <i>et al</i> (2012) ¹²	70 F	Parafalcine	CKRS; STR	–	30, 36	Postradiation; could not be graded	Yes	Deceased; ~32 months
Mindermann and Reisch (2014) ¹⁴	57 M	Parasagittal	GKRS (5), Crani (2)	21.1 cc	14, 28	Typical fibrous SFT	Yes	Alive, 17 years
	70 F	Tentorium	Crani (2), RS(1)	1.2	14, 28	Typical fibrous SFT	Yes	Alive, 6 years
Santa Maria <i>et al</i> (2014) ¹⁶	41 M	Jugular foramen	STR; STR+GKRS; STR	–	–, 32	Typical fibrous SFT	Yes	Alive, 11 years
Wanibuchi <i>et al</i> (2015) ¹⁸	44 M	Cavernous sinus	Crani; GKRS; GTR	–	–	SFT with high proliferation	Yes	Alive, >21 months†
Present study	33 M	Orbit	STR; STR; GKRS	1.69; 0.834	16, 32; 16, 32	Typical fibrous SFT	No	Alive, 17 months

*Follow-up presented as time after stereotactic radiosurgical treatment.

†Twenty-onemonths F/u after postresection surgery, timeline of GKRS not stated.

CKRS, cyber knife radiosurgery; CPA, cerebellopontine angle; Crani, craniotomy; GKRS, gamma knife radiosurgery; GTR, gross total resection; HPC, hemangiopericytoma; NTR, near-total resection; RT, radiotherapy; SFT, solitary fibrous tumour; SRS, stereotactic radiosurgery; STR, subtotal resection.

DISCUSSION

Intracranial SFTs are generally thought to be a benign tumour, and in the realm of fibrous tumours, they are nearly considered benign variants of hemangiopericytomas (HPCs).² Classically, they were also considered as tumours arising from the pleura, while more recently, they have been acknowledged as ubiquitous in the body.¹ Intracranial SFTs have been increasingly characterised in the past decade, culminating in a review of 220 cases published by Bisceglia *et al*² in 2011. Surgical resection remains a primary modality for initial treatment. However, there remains a role for SRS in the treatment of these tumours particularly following subtotal resection or recurrence.^{2 5 6}

A systematic review of intracranial SFTs treated with SRS was performed through a PubMed search with the following fields: '(solitary fibrous tumor) AND ("radiosurgery" OR "stereotactic radiotherapy")' and '(central nervous system solitary fibrous tumor) AND ("radiosurgery" OR "radiotherapy")'. The search yielded 31 results, from which 5 were removed as duplicate. Thirteen studies were included as they described treatment of an intracranial SFT with a stereotactic radiosurgical modality. Of the remaining 13 studies, 12 were excluded, because they did not either involve primary intracranial tumours (ie, were tumours of the bone or pleura or were metastatic) or involve treatment with SRS. One study was excluded as it mentioned the treatment of two cases with 'adjuvant radiosurgery' but did not specify modality or provide any further detail. The results of the systematic review outlined in table 1 demonstrate that in 7 of 17 cases (41.2%) with appropriate follow-up (>6 months), including the case reported here, the patient did not experience a recurrence after treatment.^{6 7 9–18} The intracranial SFTs treated by radiosurgery tended to be recurrent tumours after surgical resection (15/20 (75%) were recurrent). In the case presented, the patient showed recurrence within a year after a second subtotal resection along with a development of an additional, adjacent focus. GKRS utilised for both lesions (12 years after initial resection) provided effective tumour control and demonstrated regression of the tumours on follow-up. GKRS therefore appears to be a suitable option for the treatment of recurrence.^{5 13 15}

The tumour in our patient was diagnosed as an SFT and from the available pathology appears to be a typical SFT. In the proposed spectrum that attempts to recharacterise SFTs and HPCs, this typical SFT would be termed a 'fibrous SFT', while the entity referred to as an HPC would be termed a 'cellular SFT'.¹ Indeed, this spectrum is consistent with the recent WHO classification that has recharacterised SFTs and HPCs as ends of a histological and immunological spectrum.¹⁹ Therefore, we may be underestimating the potential for aggression in SFTs if we relegate them as benign variants of HPCs. Admittedly, intracranial SFTs in particular are still widely considered a distinct entity because of stark differences in biological behaviour compared with intracranial HPCs: intracranial SFTs may have a recurrence and metastatic rate of perhaps <15% (interpreted with caution due to the scarcity of long-term follow-up); while intracranial HPCs may have a local recurrence rate of 80–85% and an extracranial metastatic rate of 20%.²

A recent multicentre study by Cohen-Inbar *et al* provided the largest series of HPCs treated by SRS as well as the most comprehensive review of series and reports in the literature, combining for a total of 253 patients with 446 lesions treated by SRS. The reviewed set of these series reported rates of tumour control ranging from 46% to 100%, and they included a range of 8–58 lesions studied. The recent study involved 90 patients

with 133 tumours and reported a tumour control rate of 55%.²⁰ However, while the evidence may roughly appear to distinguish the two as separate entities, we encounter the issue that previous studies and diagnoses were not attuned to the pathological spectrum. Furusato *et al*²¹ exemplify this issue of changing definitions in a series in which they reexamined 41 fibroblastic orbital tumours originally diagnosed between 1970 and 2009 as HPCs, fibrous histiocytomas, mixed HPC/fibrous histiocytoma or giant cell angiofibromas. They found that on histological review, all cases were reclassified as SFT. Bernardini *et al*²² further comment that "before 1994, the diagnosis [of] orbital SFT was confused with other benign orbital tumors, such as fibrous histiocytoma and hemangiopericytoma because of a lack of use of immune-histochemical techniques."

Sharper discrimination of this currently muddled group of tumours intimates a clearer picture of biological behaviour for each subpopulation. Understanding the relationship between the two and delineating the spectrum and the gradient of characteristics is necessary to develop more reliable prognostics, especially as even intracranial SFTs have demonstrated local invasion, recurrence and rare malignant transformation with extracranial metastases.^{2 11 12 14} Similarly, the pathology and immunohistochemistry available for SFT in the case presented here were limited; thus, while they support a typical fibrous SFT as the diagnosis, additional studies and microscopy may retrospectively place it at a more moderate position in the spectrum rather than the 'benign' extreme. In addition, because SFTs treated by radiosurgery tend to be recurrent tumours, they may conceivably demonstrate greater frequency of HPC-like or atypical characteristics.

As with other publications, this study is limited by the follow-up of the case described, and long-term follow-up of these tumours is advised. Further research in the form of a multicentre study of SFTs is necessary to provide valuable data on clinical outcomes that would be comparable to the available data for HPCs, for both outcomes after SRS as well as pathological features of those tumours treated by SRS as part of a further characterisation of the spectrum of SFT pathology and its correlation with biological behaviour.

Learning points

- ▶ Stereotactic radiosurgery (SRS) appears to be a reasonable treatment option in for recurrent solitary fibrous tumour (SFT).
- ▶ A systematic review of SFTs treated by SRS demonstrates the possibility of recurrence after treatment, necessitating long-term follow-up.
- ▶ SFTs may not be as benign and indolent as previously thought, and appear to be on a pathological spectrum with hemangiopericytomas (HPCs).
- ▶ Further research is needed to characterise SFTs treated by SRS and delineate their relationship on the spectrum with HPCs.

Contributors The paper is completely original, written by the first author, and with critical editing and revision by the coauthors. All papers are cited as requested. All tables are original. The first author, AT, collected and analysed portions of the data, prepared tables and wrote the manuscript. The second author, OC-I, played a role in the conception and design of the report, collected and analysed portions of the

data, prepared figures and critically reviewed and edited the manuscript. The corresponding author, JPS, had the primary role in the conception and design of the report as well as critical review and editing of the manuscript.

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