

Intratumoral Hemorrhage in Jugular Foramen Schwannoma after Stereotactic Radiosurgery: A Case Report

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Case report

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Abstract

Background:

Clinically significant intratumoral hemorrhage is an extremely rare complication of stereotactic radiosurgery (SRS) for benign tumors.

Case Presentation:

Here, we present the case of a 64-year-old man who underwent SRS for a relatively large dumbbell-shaped left jugular foramen schwannoma and thereafter developed intratumoral hemorrhage. On post-SRS day 3, he developed lower cranial nerve palsies with radiographically evident tumor expansion. His neurological conditions had gradually improved thereafter; however, he suddenly developed headache, dizziness, and mild hearing deterioration at seven months due to intratumoral hemorrhage. We managed the patient conservatively, and eventually his symptoms improved except for slight ataxia and hearing deterioration. Follow-up images at three years from SRS demonstrated significant tumor shrinkage. This is the first report describing intratumoral hemorrhage after SRS for jugular foramen schwannoma.

Conclusions:

Transient expansion of the tumor and subsequent venous stasis around the tumor may have played a role in the hemorrhage. Intratumoral hemorrhage should be considered as a rare but potential complication of SRS for jugular foramen schwannomas.

Background

Jugular foramen schwannoma (JFS) is a rare brain tumor arising from cranial nerves IX, X, or XI, accounting for 2.9–4% of all intracranial schwannomas that in turn accounts for 8% of all primary intracranial tumors.^{1,2} While JFS typically causes dysphasia, hoarseness, hearing loss, and tinnitus, asymptomatic, incidentally found JFS cases have also been increasing because of improved access to non-invasive high-resolution imaging studies, such as magnetic resonance imaging (MRI). As for the treatment, surgical resection is a standard modality of treatment, but is associated with considerable invasiveness, as well as a 10–48% risk of permanent lower cranial nerve deficits.^{3–7} In contrast, stereotactic radiosurgery (SRS) is generally accepted as a reasonable alternative, providing favorable tumor control with minimum invasiveness.^{8–11} SRS has been accepted as a minimally-invasive treatment option for intracranial schwannomas, with the 5- and 10-year cumulative tumor control rates being 78–97% and 79–94%, respectively, and the rates of persistent adverse radiation being 3–15%.^{8–18} To the best of our knowledge, there are no reports on hemorrhagic complications subsequent to SRS for JFS. Here, we present a detailed case report of JFS complicated with subacute tumor expansion and an intratumoral hemorrhage after SRS.

Case Presentation

A 64-year-old man without known significant medical history was referred to our hospital for the treatment of an incidentally found left JFS. He was found to be completely intact on examination. MRI revealed a

dumbbell-shaped solid mass at the left cerebellopontine angle with marked enlargement of the affected jugular foramen and no sign of dural tail, which was 25 × 27 × 18 mm and 22 × 25 × 19 mm for the intradural and extradural portions, respectively, and 8.4 ml in total volume. The mass mildly compressed brainstem and had no evident sign of hemorrhage (Figure A). MR venography demonstrated that the left jugular bulb was obliterated due to tumor compression (Figure B). The tumor showed no signs of calcification or hypervascularity, and positron emission tomography with fluorodeoxyglucose did not exhibit increased uptake suggesting malignancy or any evidence of the other tumor (Figure C). Based on observations from the above examinations, a diagnosis of JFS was made by two independent radiologists. After thorough discussion, stereotactic radiosurgery using Gamma Knife (Elekta AB, Stockholm, Sweden) was performed without any complications with a marginal and maximal dose of 13 and 26 Gy, respectively (Figure D, E).

Post-radiosurgical course

Three days after SRS, the patient developed dysphagia, hoarseness, and leftward deviation of the tongue that were confirmed by otorhinolaryngologic examinations, suggesting injuries of the left vagal and hypoglossal nerves. MRI on post-SRS day 5 showed tumor expansion without peritumoral edema (Figure F). Administration of dexamethasone gradually relieved the symptoms except for vocal cord palsy that later required additional laryngological interventions.

Although his symptoms improved, follow-up MRI at five months from SRS showed further tumor expansion with central necrosis and peritumoral edema, albeit symptomatic (Figure G). This was considered as a slightly more aggressive change than transient tumor expansion typically seen in vestibular schwannomas, and thus prompted further close follow-ups.

At seven months, he suddenly developed headache, dizziness, mild hearing deterioration (20 dB decrease in pure tone audiometry), nausea, and vomiting. Imaging studies revealed intratumoral hemorrhage with exacerbation of peritumoral edema (Figure H). At that time, he was not on any blood thinners, and coagulation tests demonstrated normal function. Since he showed no further progression after re-starting administration of dexamethasone and osmotic diuretics, we continued the conservative management without surgical intervention. He finally recovered almost completely within two months after the hemorrhage, and follow-up images at three years from SRS demonstrated significant tumor shrinkage (Figure I).

Discussion And Conclusions

We experienced a JFS case which was accompanied with acute tumor expansion after SRS and subsequent clinically significant intratumoral hemorrhage. Intratumoral hemorrhage of intracranial schwannoma, mostly vestibular schwannoma, has been recently considered as more common than previously believed owing to advances in imaging studies and larger analyses. The etiology of intratumoral hemorrhage in intracranial schwannoma has not completely defined and is likely to be multifactorial. Anticoagulation therapy, high tumor vascularization, hypertension and large tumor have been recognized as a risk factor.¹⁹⁻²⁴ Especially, rapid tumor growth causes relative shortage of blood supply and tumor necrosis, resulting in increased intratumoral pressure and hemorrhage. These repeated microhemorrhage also lead to cystic changes and

further expansion.^{23, 25-27} Although microhemorrhage alone is asymptomatic in most cases, some patients infrequently present with the abrupt symptom progression.²⁷ Regarding vestibular schwannoma, about 50 cases have been reported in literature.^{27, 28} Carlson et al. estimated that the rate of intratumoral hemorrhage in untreated vestibular schwannoma was 0.4%, which decreased to only 0.2% after excluding patients on anticoagulation.¹⁹ The symptoms are headache, nausea and vomiting, cranial neuropathy, ataxia and so on. Special care is needed for them because these symptoms can occasionally be life-threatening due to vicinity of brainstem.

Influence of radiation on the intratumoral hemorrhage of intracranial schwannoma has not been fully elucidated. In particular, limited to clinical significant hemorrhage, there are eight reported case in literature (Table 1).^{22, 28-33}

Table 1
Literature review about cases with symptomatic intratumoral hemorrhage after SRS for intracranial schwannoma.

Author, year	Age, sex	Disease	Modality	Dose	Interval from SRS to hemorrhage	Main symptom	Treatment
Iwai et al., 2003 ²⁹	70, F	VS	GK	12 Gy (NR)	60 months	ataxia	resection
	NR	VS	GK	NR	80 months	pain	conservative
Karampelas, 2007 ³¹	53, M	VS	GK	13 Gy (46%)	27 months	headache, facial spasm	conservative
Dehdashti, 2009 ²²	47, F	VS	GK	NR	18 months	headache, ataxia	resection
Mandl, 2009 ³³	59, F	VS	NR	25 Gy/5Fr (80%)	75 months	headache, ataxia, papilledema	resection
Miki, 2015 ³⁰	48, M	VS	GK	12 Gy (50%)	46 months	facial palsy	resection
Thombre, 2019 ²⁸	63, M	VS	GK	12 Gy (50%)	10 days	vertigo, facial palsy	resection
Noureldine, 2020 ³²	71, F	FS	CK	21 Gy/3Fr*	3 days	headache, facial palsy	resection
Present case	64, M	JFS	GK	13 Gy (50%)	7 months	headache	conservative
CK = CyberKnife, FS = facial schwannoma, GK = Gamma Knife, JFS = jugular foramen schwannoma, NR = not reported, SRS = stereotactic radiosurgery, VS = vestibular schwannoma							
* Hemorrhage occurred 3 days after the first fraction, and 7 Gy had been irradiated to tumor.							

Some mechanism could be assumed from previous studies. First, radiation induces microhemorrhage as well as other estimated factors, leading to necrotic expansion and rarely massive hemorrhage.³⁴ Second, radiation would also trigger thrombosis of irradiated endothelial cells, with increase in intravascular outflow resistance and progress in venous congestion, contributing spontaneous intratumoral hemorrhage.³⁵ It is noteworthy that our case provided sequential MRIs before hemorrhage, which showed relatively rapid tumor expansion with peritumoral edema, finally resulted in symptomatic hemorrhage. Considering venous compromise which had already existed prior to SRS, it is possible that exacerbated venous congestion promoted subsequent hemorrhage in this case.

The optimal treatment for the intratumoral hemorrhage with brainstem compression is basically resection. The previously reported post-SRS cases were managed surgically in six cases and conservatively in two other cases, largely ending up in a good recovery (Table 1).^{22, 28–33} Although our patient experienced headache, nausea and mild cranial neuropathy due to intratumoral hemorrhage, he quickly improved and the tumor demonstrated remarkable shrinkage without further intervention, suggesting that necrotic changes mainly caused intratumoral hemorrhage and did not necessarily mean failed tumor control. Based on our experience, conservative treatment with osmotic diuretics and corticosteroids would be also reasonable unless progressive neurological deterioration is evident. Nevertheless, this is just a case report; thus, accumulation of additional cases and further research is needed to validate these findings. This is the first case reported to demonstrate intratumoral hemorrhage after SRS for JFS. With conservative treatment, subsequent tumor shrinkage was observed in this case. While the etiology is not completely understood, radiation-induced tumor expansion and possible venous compromise are likely to cause the intratumoral hemorrhage. Although surgical resection is needed to be considered at first, conservative management could control the condition unless the hemorrhage is devastating or results in remarkable brainstem compression. Intratumoral hemorrhage should therefore be recognized as one of potential sequelae after SRS for JFSs.

Abbreviations

SRS
stereotactic radiosurgery; JFS:jugular foramen schwannoma; MR:magnetic resonance; MRI:magnetic resonance image

Declarations

Ethics approval and consent to participate: This study was approved by the institutional review Board (#2231), and informed consent was obtained from the patient.

Consent for publication: Written informed consent for the publication of clinical details and images was obtained from the patient.

Availability of data and materials: The patient's data and medical images can be found on the database of our hospital.

Competing interest: The authors declare that they have no competing interests.

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Authors' contributions: MK carried out the literature search and image and data collection. MK and HH drafted the article. SY performed the patient's observation and revised the manuscript. MS and NS made substantial contributions to the manuscript by revising it critically. All authors read and approved the final manuscript.

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Figures

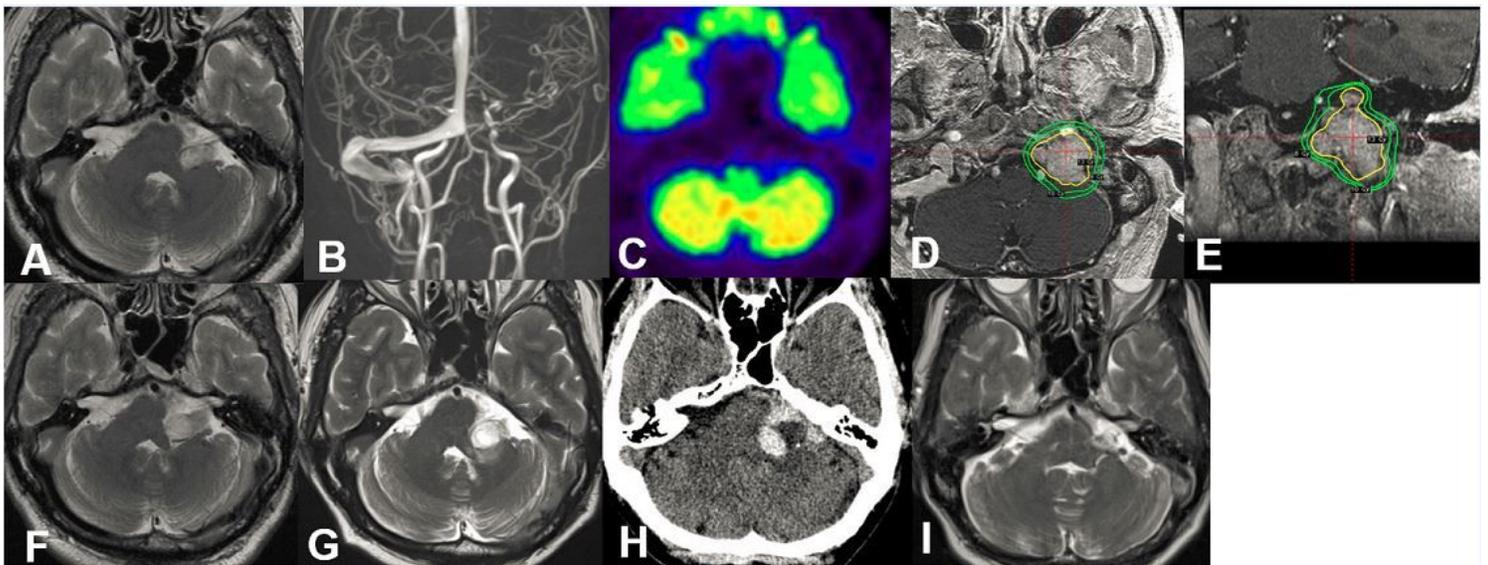


Figure 1

Pre-SRS images of the left jugular foramen schwannoma: (A) pre-SRS MRI T2-weighted axial image, (B) MR venography showing the obstructed left jugular bulb, (C) positron emission tomography with

fluorodeoxyglucose. Stereotactic MRI used in radiosurgery, (D) T1-weighted axial image with gadolinium contrast, and (E) T1-weighted coronal image with contrast, respectively. These images revealed a well-enhanced, dumbbell-shaped, solid mass extending the intra- and extracranial space, causing expansion of the jugular foramen and invading hypoglossal canal. Post-SRS chronological change in T2-weighted axial MRI: (F) tumor expansion was observed at five days after SRS. (G) Further expansion with peritumoral edema at five months after SRS. (H) Intratumoral hemorrhage occurred at seven months after SRS. (I) Evident tumor shrinkage at the last follow-up three years after SRS. SRS = stereotactic radiosurgery; MR = magnetic resonance; MRI = magnetic resonance image