Case Report

Jugular foramen schwannoma: A rare tumor treated by stereotactic radiosurgery

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ABSTRACT

Jugular foramen schwannomas (JFS) are rare benign tumors located in the jugular foramen. They can present with various symptoms depending on the extent of involvement of the tumor such as hearing loss, tinnitus, headache, and ataxia. Surgical resection has been considered as the primary treatment option for JFS. Stereotactic radiosurgery (SRS) offers an equally good treatment in patients with small and residual tumors after surgery with the advantage of being non-invasive with minimal complication rates. Herein, we present a rare case of JFS treated by SRS in our institution.

Key words: Gamma knife, Jugular foramen schwannomas, Linac, Stereotactic radiosurgery

Jugular foramen schwannoma (JFS) is a rare type of benign tumor located in the jugular foramen, accounting for 2–4% of all intracranial schwannomas [1]. They can arise from the cranial nerves IX, X, or XI, with IX being the most common. JFS can present with various symptoms depending on the size and the anatomic location of the tumor. Surgical resection has been considered as the primary treatment option for JFS. However, complete resection is often difficult due to the anatomical location of the tumor and its relationship to adjacent structures. Stereotactic radiosurgery (SRS) has been considered an equally good and safer alternative to surgery, especially for small volume tumors [2].

CASE REPORT

A 59-year-old normotensive and non-diabetic male presented with hoarseness of voice and difficulty in speaking for 1 month. There was no history of headache, vomiting, or seizures. Furthermore, he had no hearing loss, tinnitus, or vertigo.

On general examination, there was no abnormality detected. The patient underwent an ENT examination at his native place which was normal.

His baseline blood counts, liver, and kidney function tests were normal. The patient was, further, investigated with computed tomography (CT) scan of the brain followed by contrast Magnetic resonance imaging (MRI) of the brain which revealed a well-defined lobulated extra-axial lesion measuring 13.7 × 9.8 × 10.0 mm, seen within the left jugular foramen, expanding it and projecting into the left cerebellomedullary cistern (Fig. 1). It was causing compression on the IX, X, and XI cranial nerves. Features were suggestive of jugular foramen schwannoma.

The patient was given the option of surgery versus SRS with the risk and benefits of each modality explained in detail. The patient chose SRS in view of the non-invasive technique. He was immobilized using a stereotactic head thermoplastic cast with rigid mouth and head fixation. Then, contrast-enhanced CT (CECT) scans were acquired at 1 mm slice thickness and imported in the Monaco™ treatment planning system for planning. The CECT images were fused with MRI images for delineation of the tumor and other critical structures. Proper optimization followed by Monte Carlo dose calculation algorithms were used for SRS plan generation. Finally, he underwent SRS treatment on 6 MV Linear Accelerator using a micro multileaf collimator to the localized and marked portal. The tumor volume was 1.01cc. The prescribed dose for the tumor was 15 Gy in a single fraction. The 80% isodose line coverage to the tumor was ensured to 98.7% volume (Fig. 2). In addition, doses received by nearby critical organs such as the cochlea, brain stem, left optic nerve, right optic nerve, and optic chiasm were found well within the tolerance level. As of now, the patient is improving clinically.

DISCUSSION

JFS is an uncommon pathological condition and represents 10–30% of all tumors seen in this location. The other pathological

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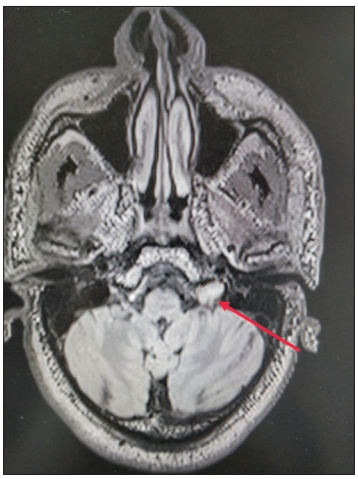

Figure 1: Contrast MRI brain showing enhancement of tumor in jugular foramen

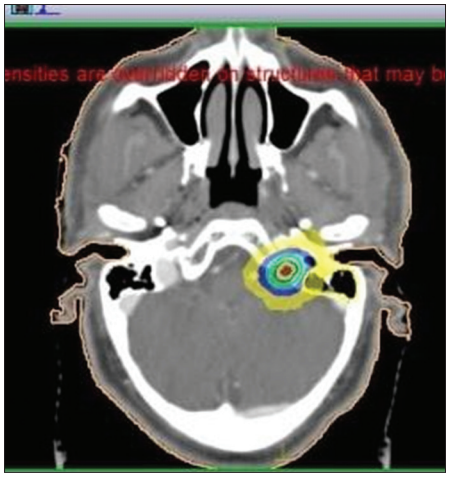

Figure 2: Isodose curve showing 80% coverage of the tumor

diagnosis may include paraganglioma, meningioma, and other nerve sheath tumors [1]. This was the first case in our institution to be treated with SRS. Pellet *et al*. classified JFS into four types [3]. Type A is primarily intracranial with minimal extension into the jugular foramen; Type B is primarily within the bone with or without an intracranial component; Type C is primarily extracranial with only a minor extension into the jugular foramen or into the posterior fossa; and Type D is dumbbell-shaped tumors with intra- and extracranial extension. According to the Pellet's classification, Type D schwannomas are the most common (40–68%), followed by Type A (25–32%), Type B (15–22%), and Type C (12–15%). Our patient belonged to Type B.

The presenting symptoms in JFS vary depending on the location. However, due to the slow growth of these tumors, symptoms do not manifest till the tumor is of large size. The most common initial symptoms are hearing loss, tinnitus, ataxia, and vertigo. Other symptoms are dysphagia, hoarseness of voice, headache, and blurred vision [4]. Our patient presented with hoarseness of voice and difficulty in speaking due to compression of the IX, X, and XI cranial nerves. Another interesting fact is that JFS mostly arises from the left side of the jugular foramen and the glossopharyngeal nerve is the most common nerve involved as in our case also. In a review of 204 patients, the tumor originated from the glossopharyngeal nerve (IX) in 47 cases (23.6%), in 26 cases (13%) from the vagal nerve (X), and in 11 cases (5.5%) from the accessory nerve (XI). In the remaining 58% of the cases, the origin of the tumor remained unknown [1].

The diagnosis is usually radiological with CT and MRI scans of the brain. On MRI, JFS is usually iso- or hypointense on T1-weighted images and iso- or hyperintense on T2-weighted images along with enhancement on gadolinium-DTPA [5]. Resection has been considered the primary treatment option for JFSs. Complete resection is often difficult due to the tumor's anatomical location and its relationship to adjacent critical cranial nerves, brain, and vascular structures. The advantages of surgery are complete removal of the tumor with immediate relief from symptoms along with confirmation of histopathological diagnosis. However, due to close proximity to the cranial nerves and vessel, surgery has a potential risk of post-operative complications such as cranial nerve palsies (15%) CSF leakage (6.5%), meningitis (2.0%), aspiration pneumonia (1.5%), and mastoiditis (1%) [6,7]. Bakar reviewed the outcomes obtained after the surgical treatment of 204 patients. He reported a gross tumor resection (GTR) in 86.9% of the patients, near GTR in 3.3% of the patients, and subtotal resection in 9.8% of the patients [1]. Park *et al*. analyzed the results of surgery in 275 cases collected in large series of jugular foramen schwannomas and found postsurgical lower cranial nerve palsies in 34.9% of the patients [8].

Near GTR followed by SRS or SRS alone offers an alternative approach in some patients. High doses of radiation induce the cessation of lesion growth or reduction in size, rather than the complete disappearance of the tumor, which is the goal of treatment. It offers the advantage of being non-invasive with minimal complication rates. However, the response may take a longer time to happen. SRS has been used as a primary treatment for medium to small JFSs and as a secondary treatment for residual or recurrent JFSs after microsurgery. Reports on SRS for JFS are still limited due to the rarity of the tumor and the limited facility of SRS in many institutions till now. Martin *et al*. reported 34 patients with JFS who underwent SRS with Gamma knife. The 10-year PFS was 94%, with a mean follow-up of 84 months [9]. Recently, Hasegawa *et al*. reported the results of an 18-institution Japanese multicenter JFS study in which 117 patients underwent SRS, with a median follow-up of 52 months. Tumor regression was found in 53% and tumor progression was detected in 11%. The PFS was 91% at 3 years and 89% at 5 years [10]. As far as the clinical improvement is concerned, hoarseness of voice and swallowing disturbances improved in 66%–63%, respectively, in their study. Factors associated with decreased tumor control include dumbbell-type tumors, brainstem edema, and tumor volume >6 cm^3 [11].

CONCLUSION

Major advances in stereotactic localization, non-invasive neuroimaging, and radiation physics made it possible to

selectively irradiate a sharply defined target and largely sparing the surrounding normal tissues. JFSs are one such group of tumors that can be effectively treated by SRS with a high tumor control rate, a high rate of cranial nerve preservation, and low morbidity.

REFERENCES

1. Bakar B. The jugular foramen schwannomas. Review of the large surgical series. J Korean Neuro Surg Soc 2008;44:285-94.
2. Kano H, Meola A, Yang HC, Guo WY, Martínez-Alvarez R, Martínez-Moreno N, *et al*. Stereotactic radiosurgery for jugular foramen schwannomas: An international multicenter study. J Neuro Surg 2018;129:928-36.
3. Pellet W, Cannoni M, Pech A. The widened transcochlear approach to jugular foramen tumors. J Neuro Surg 1988;69:887-94.
4. Vorasubin N, Sang UH, Mafee M, Nguyen QT. Glossopharyngeal schwannomas: A 100 year review. Laryngoscope 2009;119:26-35.
5. Matsushima T, Hasuo K, Yasumori K, Yoshida K, Hirakata R, Fukui M, *et al*. Magnetic resonance imaging of jugular foramen neurinomas. Acta Neurochir (Wien) 1989;96:83-7.
6. Sedney CL, Nonaka Y, Bulsara KR, Fukushima T. Microsurgical management of jugular foramen schwannomas. Neurosurgery 2013;72:42-6.
7. Nowak A, Dziedzic T, Czernicki T, Kunert P, Marchel A. Surgical treatment of jugular foramen schwannomas. Neurol Neurochir Pol 2014;48:188-95.
8. Park ES, Lee EJ, Park JB, Cho YH, Hong SH, Kim JH, *et al.* A single-institution retrospective study of jugular foramen schwannoma management: Radical resection versus subtotal intracranial resection through a retrosigmoid suboccipital approach followed by radiosurgery. World Neurosurg 2016;88:552-62.
9. Martin JJ, Kondziolka D, Flickinger JC, Mathieu D, Niranjan A, Lunsford LD. Cranial nerve preservation and outcomes after stereotactic radiosurgery for jugular foramen schwannomas. Neurosurgery 2007;61:76-81.
10. Hasegawa T, Kato T, Kida Y, Sasaki A, Iwai Y, Kondoh T, *et al*. Gamma knife surgery for patients with jugular foramen schwannomas: A multi-institutional retrospective study in Japan. J Neurosurg 2016;125:822-31.
11. Suárez C, López F, Mendenhall WM, Andreasen S, Mikkelsen LH, Langendijk JA, *et al.* Trends in the management of non-vestibular skull base and intracranial schwannomas. Cancer Manag Res 2021;13:463-78.

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