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Case Report

A single-session stereotactic radiosurgery for vagal paraganglioma: Effective tumor reduction and innovative treatment option

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ABSTRACT

Background: Vagal paragangliomas (VPs) are rare tumors in the upper cervical region. Although surgical resection is the standard treatment for these tumors, it carries significant risks due to the tumor's high vascularity and proximity to vital structures. Stereotactic radiosurgery (SRS) for skull base paraganglioma could be a minimally invasive alternative.

Case Description: We report the case of a 47-year-old man with a large, asymptomatic VP who was successfully treated with SRS with Gamma Knife Icon, which was performed in the parapharyngeal space (volume: 25.7 mL) using a marginal dose of 14 Gy to the 45% isodose line. This case illustrates the successful treatment of a lesion near the conventional limits (lower limit of C2 vertebral body) using noninvasive mask fixation. Excellent tumor control without neurological deficits was achieved for 25 months after SRS. The tumor volume decreased by 70% (final volume: 7.6 mL).

Conclusion: This study demonstrates the utility of Gamma Knife Icon, which facilitates optimal SRS for upper cervical lesions, including VPs.

Keywords: Gamma knife icon, Noninvasive treatment, Stereotactic radiosurgery, Vagal paraganglioma, Vagus nerve

INTRODUCTION

Paragangliomas, also known as glomus tumors, are rare neuroendocrine tumors originating from the neural crest that can grow in various regions, including the head, neck, thorax, abdomen, and adrenal glands.[2] Adrenal paragangliomas are the most common type, which are predominantly sympathetic and often produce excessive catecholamines. Conversely, paragangliomas of the head and neck are usually parasympathetic, nonsecretory, and relatively rare. These lesions are further classified based on their location in the skull base or cervical region. Skull base lesions include tympanic and jugular paragangliomas, collectively referred to as jugulotympanic paragangliomas (JTPs), whereas neck lesions include carotid body and vagal paragangliomas (VPs). Among head-and-neck paragangliomas, VPs are particularly rare, accounting for approximately 3% of such cases. Although surgical resection is the mainstay of treatment for head-and-neck paragangliomas, it carries significant risks due to tumor

vascularity and adjacent vital structures, such as the carotid arteries, jugular veins, and lower cranial nerves.[7,8] As such, alternative treatment modalities are being studied, including conventional radiation therapy, which has shown favorable tumor control in head–and-neck paragangliomas. Notably, stereotactic radiosurgery (SRS), which can deliver high-dose irradiation to precisely target tumor margins in a single session, offers the potential as a less-invasive treatment option. However, to the best of our knowledge, its application to VPs has not been previously reported. Herein, we present a case of a VP successfully treated with SRS using the Leksell Gamma Knife Icon (Elekta AB, Stockholm, Sweden) and mask fixation, resulting in favorable tumor shrinkage without neurological deficits.

CASE PRESENTATION

History, examination, and imaging

A 47-year-old male was referred to our hospital for investigation and treatment of a tumor-like lesion in the right parapharyngeal space, which was incidentally detected on computed tomography (CT) for head trauma. The patient did not present with any neurological deficits, and routine blood examinations, including catecholamine levels, yielded normal results. Gadolinium-enhanced magnetic resonance imaging (MRI) revealed a well-defined, enhancing, round mass localized between the pterygoid and longus capitis muscles (size, 35 × 29 × 55 mm; volume, 25.7 mL); [Figure 1 a]. Positron emission tomography revealed increased fluorodeoxyglucose uptake within the lesion with no evidence of metastasis to other locations [Figure 1b]. Digital subtraction angiography also showed hypervascularity of the lesion, which was primarily supplied by the ascending pharyngeal artery [Figure 1c]. Based on these findings, malignancies in the head-and-neck region, including metastases, were considered negative. Although the differential diagnoses considered were schwannoma, head-and-neck neuroendocrine tumors, and extracranial meningioma, the tumor's hypervascularity and location suggested a VP as the most likely diagnosis.

Management

Considering the patient's preference for nonsurgical treatment modalities, SRS using the Leksell Gamma Knife Icon was performed. Since the inferior margins of the tumor extended to the lower C2 vertebra, a thermoplastic mask (Icon Mask Nanor, Elekta AB) and cushion (Icon Patient Cushion Klarity, Elekta AB) were used for fixation. Leksell frame (Elekta AB) fixation was not applicable in this case due to potential shoulder collisions. Preoperatively, contrast-enhanced cone-beam CT images were obtained to confirm consistent patient positioning during irradiation, ensuring accurate tumor targeting. A single-fraction marginal dose of 14 Gy was then delivered to the 45% isodose line [Figures 1d-f].

Outcome

The patient's post-SRS course was uneventful. Serial MRIs revealed tumor shrinkage at 3 months and contrast defect of internal tumor necrosis at 6 months. At the latest follow-up (25 months after SRS), a significant tumor reduction was observed (size: 25 × 19 × 46 mm, volume: 7.6 mL, a decrease of 70%), [Figures 1g- i] without any neurological deficits.

DISCUSSION

Surgical resection of paragangliomas carries a potential risk of jeopardizing neurological function due to its high vascularity and the nature of being adjacent to critical structures. Consequently, the optimal treatment approach in such cases remains controversial. Radiation therapy offers an alternative or adjunctive therapy to surgical resection by hindering tumor progression and lowering surgical thresholds, which particularly benefits cases deemed difficult or unsuitable for surgery, cases of recurrent or unresectable tumors, or cases requiring palliative care.[9] Advancements in radiation technology have significantly improved the ability to control tumors through enhanced precision of radiation delivery. Studies have consistently reported the efficacy of radiotherapy for paragangliomas, demonstrating low complication and tumor control rates comparable to those of surgical resection.[4,16] Specifically, Suarez *et al*. documented a 78% tumor control rate for surgery compared with 92% for radiation therapy, with complication rates of 28% and 11%, respectively.[16] Studies suggest a 30–50 Gy dose range for optimal local control with minimal morbidities, achieving control rates of 82–100%.[14-19] These findings suggest the potential advantages of radiation therapy regarding efficacy and safety.

Recently, SRS has gained traction for its use against head-and-neck paragangliomas due to its ability to deliver high-dose, meticulously targeted radiation to tumor margins.[11] Several studies on SRS for JTPs have indicated favorable tumor control rates (93–97%) and low neurological complication rates (0–8%) following SRS.[1-15] A meta-analysis on skull base paragangliomas also revealed no significant differences in tumor control rates between surgery and SRS (72–84.2% vs. 83.4–100%), further supporting the effectiveness of SRS for JTPs.[4] Similarly, comparative analyses between surgical methods and radiation therapy for VP have demonstrated comparable tumor control rates and fewer complications. Notably, substantial tumor control was observed in VP cases managed with subtotal resection, implying comparable effectiveness between radiation therapy and surgery for skull base paragangliomas.[4] Given these findings regarding the radiosensitivity of paragangliomas, SRS can be presumed to be an effective treatment for VPs. Maxwell *et al*. and Künzel *et al*. reported 13 patients with VP treated with hypofractionated stereotactic radiotherapy and demonstrated

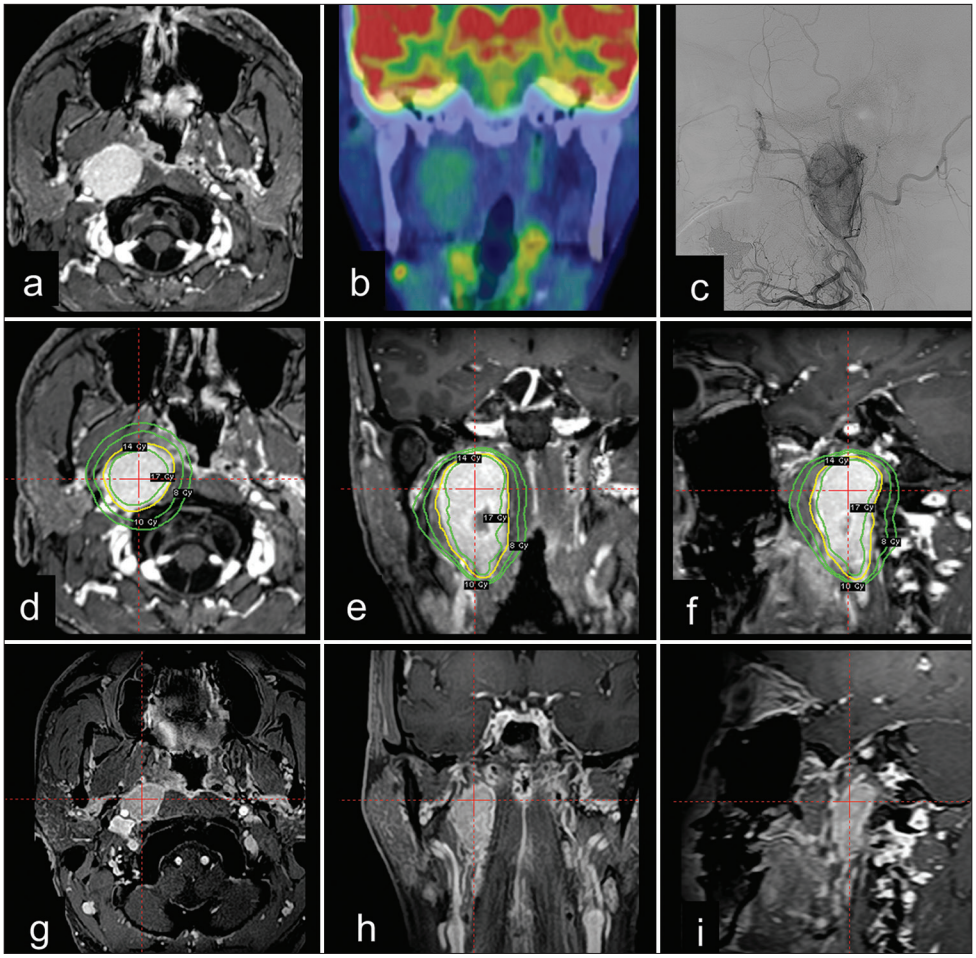

Figure 1: Vagal paraganglioma incidentally detected in a 47-year-old man. (a) Gadolinium-enhanced, T1-weighted magnetic resonance imaging (MRI) showing a well-enhanced tumor in the parapharyngeal space. (b) Positron emission tomography revealed increased fluorodeoxyglucose uptake within the lesion, with no evidence of metastasis to the other location. (c) Digital subtraction angiography showing the hypervascularity of the tumor, primarily supplied by the ascending pharyngeal artery. (d-f) Stereotactic radiosurgery (SRS) with Gamma Knife using mask fixation was performed, and radiosurgical planning images are shown in the axial, coronal, and sagittal planes. The yellow line indicates the 45% isodose line for the prescribed treatment dose of 14 Gy, and the green lines indicate the doses of 8, 10, and 17 Gy. (g-i) The patient's clinical course was uneventful, and MRI revealed significant tumor shrinkage with a 70% decrease in volume 25 months after SRS.

good tumor control without complications.[10,13] In the present case, significant tumor shrinkage was achieved with a single-fraction dose of 14 Gy, supporting the potential of SRS as an effective noninvasive treatment for VPs.

Previously, head fixation for Gamma Knife-based SRS was limited to frame-based methods, restricting treatment for C1/C2 vertebrae.[12] Earlier attempts at targeting the C3 vertebra using a maxillary frame with pin-fixation could be invasive.[17] The present case illustrates the successful treatment of a lesion near the conventional limits using noninvasive mask fixation. Furthermore, the case shows that precise co-registration in the cervical spine using pre-SRS reference CT and stereotactic cone-beam CT images is also essential, enabling the expansion of treatment feasibility for high cervical lesions.

Despite the valuable insights offered in this study, certain limitations should be acknowledged. The absence of pathological confirmation and the relatively short follow-up period may have affected our findings. A further evaluation of larger patient cohorts is necessary to evaluate the efficacy of SRS for VPs and to explore the lower limits of SRS applicability for upper cervical lesions.

CONCLUSION

This report describes the first successful application of SRS in VP. SRS with mask fixation of the head and neck might

be an alternative noninvasive treatment option for surgical resection, achieving favorable control of VP.

Ethical approval

Institutional Review Board approval is not required.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Conflicts of interest

There are no conflicts of interest.

Use of artificial intelligence (AI)-assisted technology for manuscript preparation

The authors confirm that there was no use of artificial intelligence (AI)-assisted technology for assisting in the writing or editing of the manuscript and no images were manipulated using AI.

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