STEREOTACTIC RADIOTHERAPY FOR BASE OF SKULL PARAGANGLIOMA: CASE REPORT

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Summary: We present a case of atypical skull base paraganglioma involving intrasellar, suprasellar, sphenoid sinus and nasopharyngeal area. The patient presented with right-sided headache and tinnitus. Stereotactic Radiotherapy was selected to be the method of treatment for this patient because of the site of the tumor and the critical normal surrounding structures. The result showed satisfactory symptomatic improvement even if complete tumor involution did not occur.

INTRODUCTION

Paragangliomas are tumors that arise from widely distributed paraganglionic tissue, cell of neural crest tissue. The common locations of paragangliomas in head and neck are the carotid bifurcation, along the vagus nerve and the jugulotympanic area. The main treatments are surgery in resectable area and radiation for the area that is difficult to approach surgically. Stereotactic radiosurgery has been reported in a few series recently as effective treatment with minimal complication for these glomus tumors.[1-4]

Paraganglioma at intrasellar and suprasellar areas are rare locations with less than 10 cases reported.[5-7] They are classified as paraganglioma of uncertain origin because normally there is no paraganglionic cell in this area. There are some hypothesis that try to explain this; Khamlichi A.[8] proposed that it originated from the neural crest following the trigeminal nerve particularly the opthalmic division, Bibao JM.[9] proposed that it originated from abnormal migration of the neural crest in the development of the pituitary gland.

Treatment for the tumor in this area has to be considered about many important factors:
1) This tumor is mostly benign and slow growing so it takes many years before the patient can detect the symptoms. So the tumor is usually huge in size when it is detected.
2) Paraganglioma is highly vascularized tumor. Excessive bleeding from the tumor during surgery was founded in case report.[2] Preoperative embolization may give benefit.
3) Optic chiasm, optic nerve, cranial nerve 3,4,5,6 and the major vessel in sellar area are frequently involved or close to the tumor. Surgery and conventional radiation therapy are limited.

From these reasons; Stereotactic Radiosurgery (SRS) and Stereotactic Radiotherapy (SRT) which was reported to be the treatment of other diseases in this area such as pituitary adenoma, meningioma and AVM seems to be useful for the treatment of paraganglioma in this area. For that it helps to avoid complication from surgery and preserves the structure nearby.

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CASE REPORT

A 47-year-old woman presented with off and on right-sided headache for 3-4 years. She had been diagnosed migraine and sinusitis and had got treatment for these diseases for years but the symptoms seemed to get worse. When she first came to our department, she had severe right-sided headache for twice to three times a day. From the retrospective review, we found that she also had right ear tinnitus for about 2 years. No eyes or other cranial nerve symptoms or endocrinologic disturbance were detected. On physical examination once, she was found to have a vascular mass with smooth surface at right middle turbinate of the nasal cavity. Then the patient was referred to our institute for additional evaluation and treatment.

A computed tomographic (CT) scan was obtained, it showed extensive enhancing mass, 5x5x5.5 cm. in maximal diameter, occupying in the midline from base of skull to suprasellar region superiorly and nasopharynx inferiorly. There were destruction of base of skull, sphenoid bone, sellar turcica and planum sphenoidale (figure 1). The tumor mass involved both parasellar area, more prominent on right side. A biopsy of the mass from right nasal cavity was done and the pathological report was paraganglioma. Agyrophilia, a feature of this site of tumor, was demonstrated in Grimelius stain.

She was treated by Stereotactic Radiotherapy with doses of 685 cGy x 5, daily fractions in 5 days in October 1997. The treatment was planned with 7 photon beams. After the treatment, her symptoms still persisted and the CT scan 1 month after treatment showed slightly decrease in size especially the intracranial part (figure 2). Then she got the second course of Stereotactic radiotherapy with dose 440 cGy. x 5 daily fractions equivalent to those in December 1997 (figure 3). The treatment interval was 80 days between the first and the second course. After the second course of SRT right-sided headache was almost completely disappeared. The CT scan 2 months after the treatment showed significant decrease in size of the suprasella lesion but there was still large residual tumor in the nasopharynx, sphenoid and ethmoid sinuses (figure 4). The patient was close follow up with good quality of life, no visual defect was detected. One year after the treatment the patient began to have repeated epistaxis, the CT scan showed persistent large lesion in the nasopharynx and sphenoid sinus (figure 5). The repeated biopsy from anterior rhinoscope showed residual paraganglioma. This time the patient was treated by 3 beams nasal irradiation, doses of 404 cGy x 10 fractions in 2 weeks in January 1999 (figure 6). The epistaxis was stopped. The CT and MRI studies 7 months after the treatment showed inhomogeneous enhancing mass occupied right parasella, pituitary fossa and right sphenoid sinus about 3x2 cm. in diameter (figure 7). The perfusion study of MRI showed only marginal perfusion of the tumor. The patient has best quality of life and no visual or any other complication is detected. Her last followed up was in September 2000.

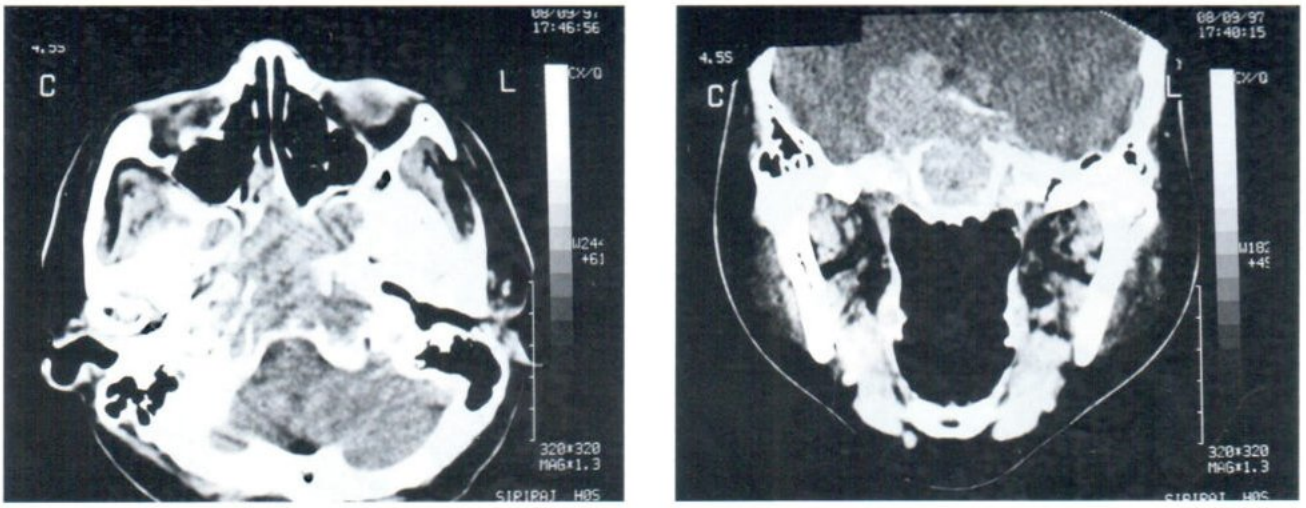


Fig. 1 Axial and sagittal CT images showed extensive enhancing mass, 5x5x5.5 cm. in maximal diameter, occupying in the midline from base of skull to suprasellar region superiorly and nasopharynx inferiorly with destruction of base of skull, sphenoid bone, sellar turcica and planum sphenoidale

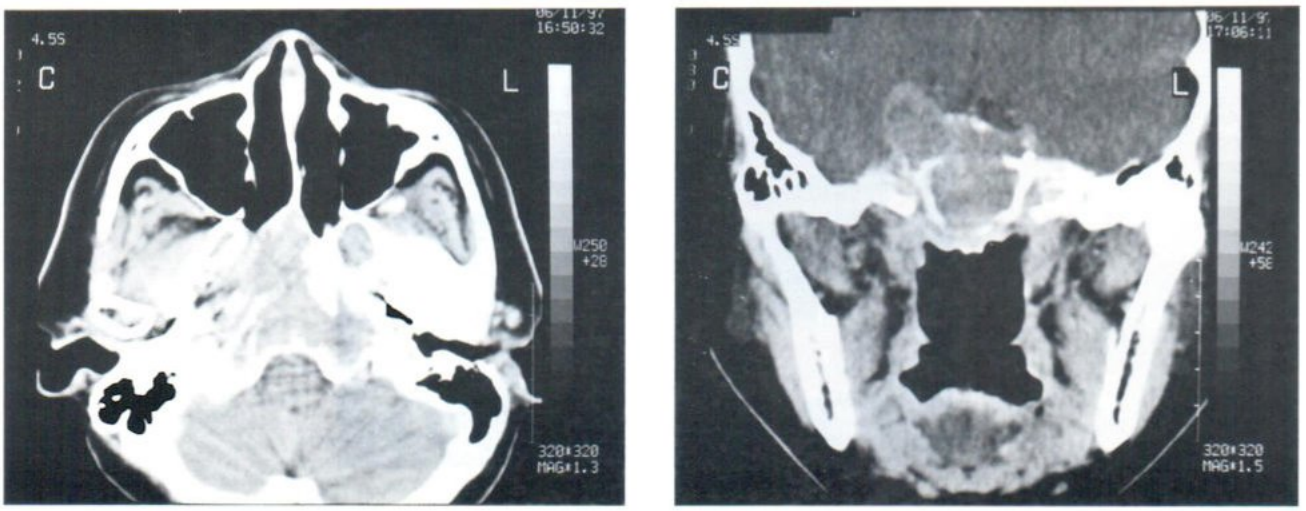
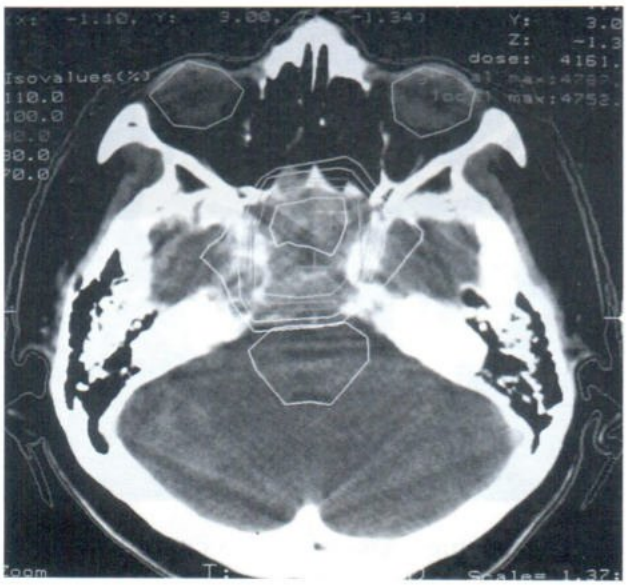
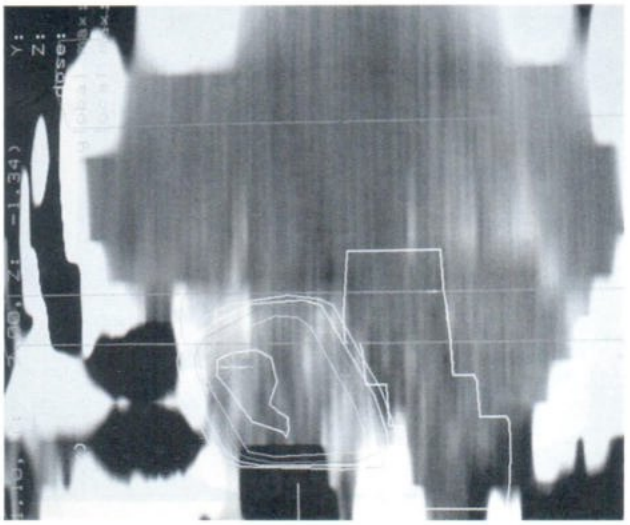


Fig. 2 Axial and sagittal CT images 1 month after the treatment showed slightly decrease in size especially the intracranial part



3A



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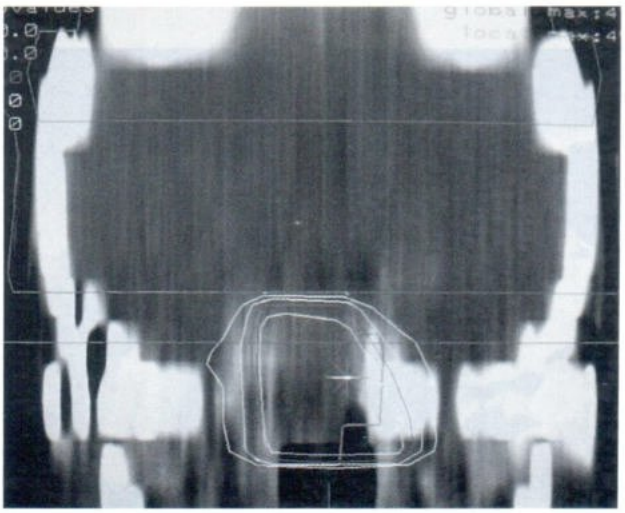

3C

Fig. 3 Isodose distribution of second SRT treatment planning White line represents tumor border
INNERMOST line shows 100% isodose
INNER line shows 90% isodose
OUTER line shows 80% isodose
OUTERMOST line shows 70% isodose
A. Axial view
B. Sagittal view
C. Coronal view

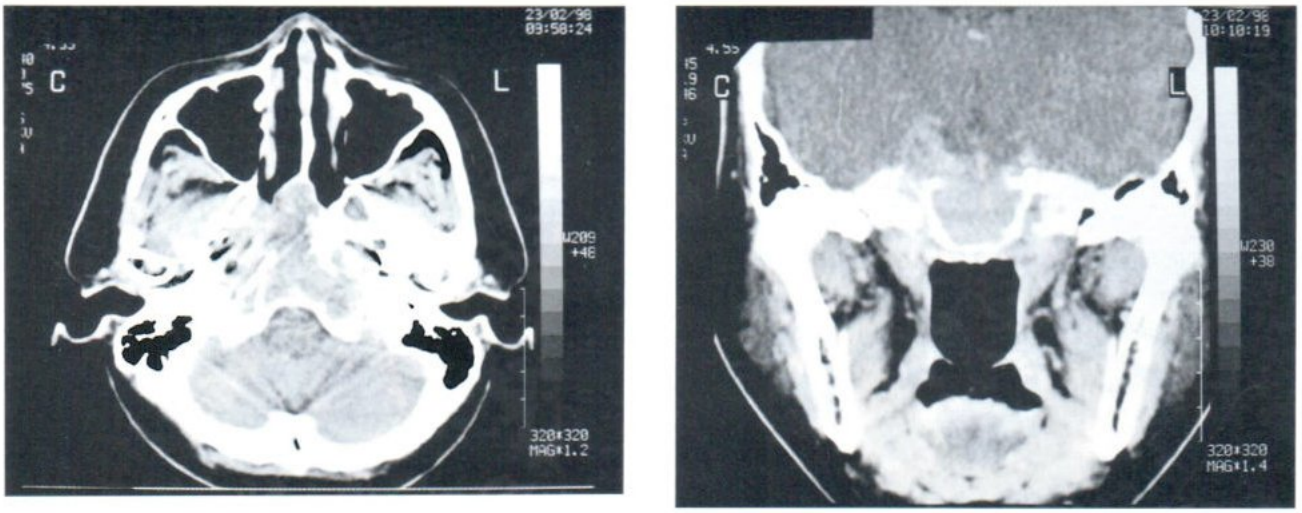

Fig. 4 Axial and sagittal CT images 2 months after the second SRT treatment showed significant decrease in size of the suprasella lesion but there was still large residual tumor in the nasopharynx, sphenoid and ethmoid sinuses

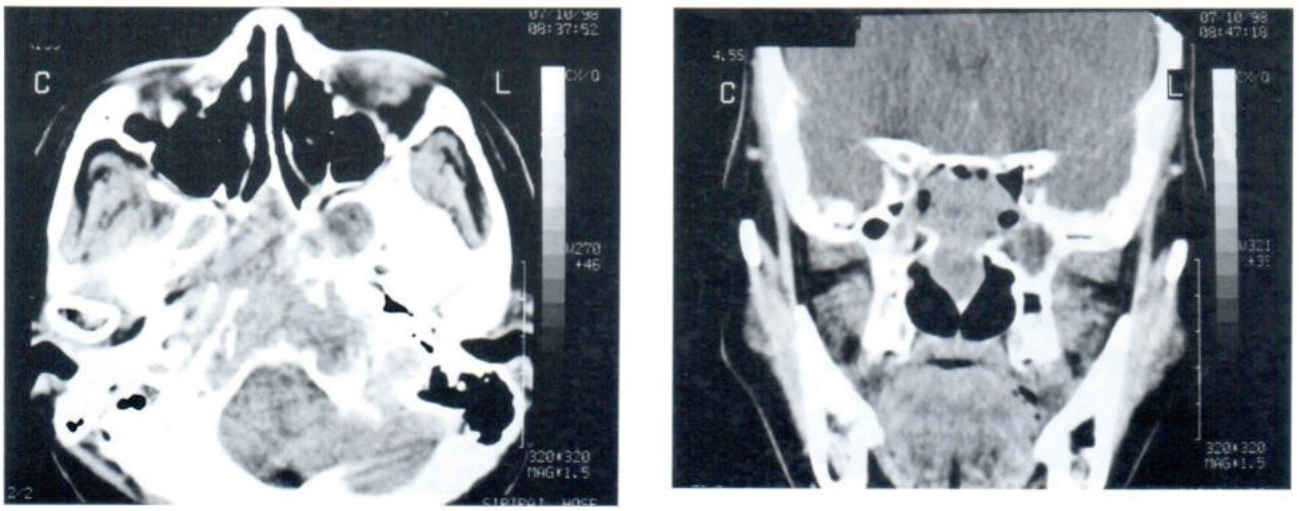

Fig. 5 Axial and sagittal CT images 1 year after the second SRT treatment showed persistent large lesion in the nasopharynx and sphenoid sinus which caused epistaxis

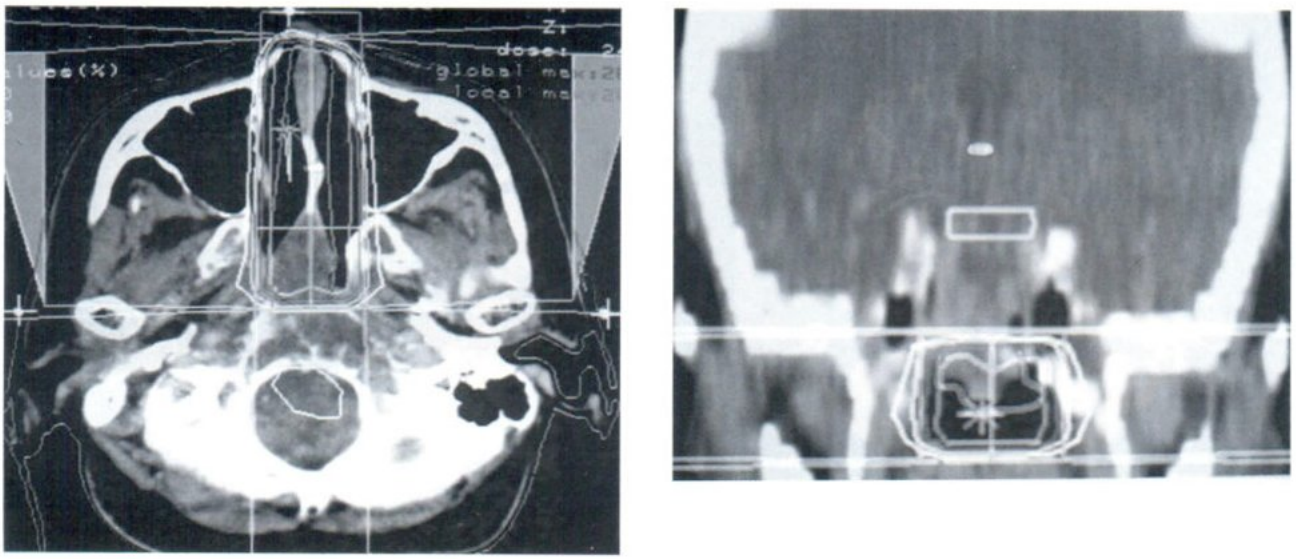

Fig. 6 Isodose distribution of the third SRT treatment with 3 photon beams aiming to stop epistaxis

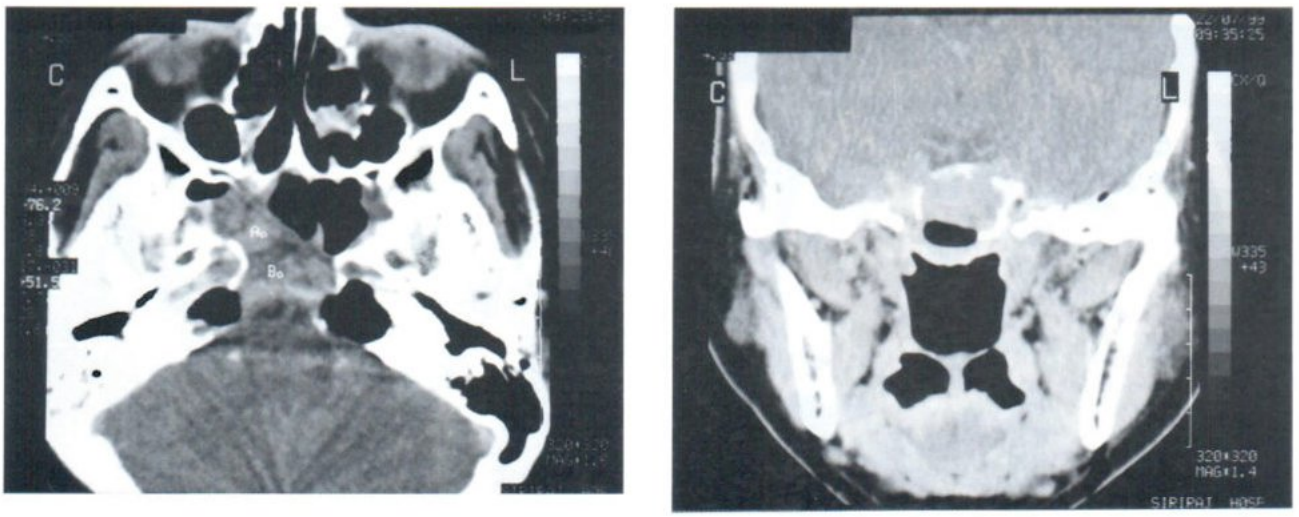

Fig.7 Axial and sagittal CT images 7 months after the third SRT treatment showed inhomogeneous enhancing mass occupied right parasella, pituitary fossa and right sphenoid sinus about 3x2 cm. in diameter

DISCUSSION

There had been reports [10-13] showing good results of the patients with paraganglioma in other areas treated by megavoltage radiotherapy alone (3500-5200 cGy conventional dose). The 10-year control rate is more than 90%. Series from 6 European centers[3-4] using SRS as the treatment for glomus jugulare tumors in 52 patients with 24 months median follow up time showed good control rate with 40% decrease in tumor size and 60% no change. The other series from Austria[1-2] using SRS for skull-base glomus tumor showed similar result with 36% decreased in tumor size and 64% had symptomatic improvement after mean follow-up time of 42 months.

Stereotactic radiotherapy gives the same benefit as radiosurgery with higher doses to the tumor and less side effect to the surrounding structures. Even there was no literature so far using hypofractionation SRT for the treatment of this type of tumor, we selected SRT to be the treatment for this patient because of the location which is close to cranial nerve and hypofractionation for the reason of cost-effectiveness. The result showed satisfactory symptomatic relief even if the image still showed residual tumor, it was smaller in size and less vascularized. This is considered to be successful treatment. However paraganglioma is a slow growing tumor and we need up to 10 year follow-up time to evaluate the result of the treatment.

Radiation of paraganglioma and other slow growing tumors mainly affects in proliferative and perivascular fibrosis with minimal alterations in the chief epithelial cells.[14] In this case angiography or perfusion MRI scan may be useful to evaluate the result of the treatment than using the imaging to see the tumor size alone. Liscak R, et al[4] compared pre and post treatment angiography in 6 patients, pathological vascularisation was completely disappeared in one patient, reduction in size in 2 and no change in 3 patients. The risk for cranial nerve complication in this patient is also high and needs to be closely followed.

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