A Left Jugulare Paraganglioma with a Hypoglossal Nerve Palsy and longstanding Hearing Loss Treated with SRS: A Case Report

Onkaram Likhitha Priya
Research scholar, Department of pharmacy practice, Santhiram college of pharmacy. Nandyal.5185112 Andhra Pradesh

***Abstract*—Glomus jugulare tumours are rare, slow growing tumours and very vascular tumours which arise from the jugular bulb. These paragangliomas present treatment challenges because of their locations and vascularity increase the risk of neurovascular injury. Surgery in this area carriers a real risk of nerve injury, so stereotactic radiosurgery (SRS) which is a non-invasive treatment option. A 47-year-old woman with hypothyroidism presented with left-sided hearing loss for about 5 years and deviation of tongue to the left for about 6 months. Examination demonstrated as MRI showed an intensely enhancing left jugulare foramen tumour about 2.3 to 2.5 cm enhancing the part of left internal carotid artery. Audiometry showed moderate to severe hearing loss on the left. following multidisciplinary review, she underwent stereotactic radiosurgery using a 6 MV flattening filter free (6FFF) photo beam 24Gy in 3 fractions (8 Gy per fraction) to the GTV plus a 2mm margin (PTV). At follow-up the symptoms were occasional tightness over the left ear, giddiness and no other significant complaints. A follow-up MRI was done about 12 months after treatment. Procedure hypo fractionated SRS is a safe and non-invasive option which is used for a tumour of this size which lies close to the brainstem, inner ear and lower cranial nerves.**

***Index Terms*—Glomus jugulare,Jugular paraganglioma; stereotactic radiosurgery, Gamma Kinfe; Hypofractionation, hearing loss, hypoglossal nerve palsy**

I. INTRODUCTION

Glomus jugulare tumors -they are also named as tympan jugular paragangliomas which are rare and highly vascular neuroendocrine neoplasms which arise from paraganglion cells near the jugular bulb. These highly vascular skull-base tumor that most often present left-sided hearing loss, pulsatile tinnitus and a retro tympanic vascular mass(1).Treatment option which is Stereotactic radiosurgery chosen to remove the tumour but it can also carry a higher chance of new lower cranial nerve palsy, swallowing problems and cerebrospinal fluid leak,long term tumour control is similar with SRS (2,3)The present report describe a woman with a left glomus jugular tumour, of hearing loss and hypoglossal nerve palsy treated with hypo fractionated LINAC based SRS.

II. CASE PRESENTATION

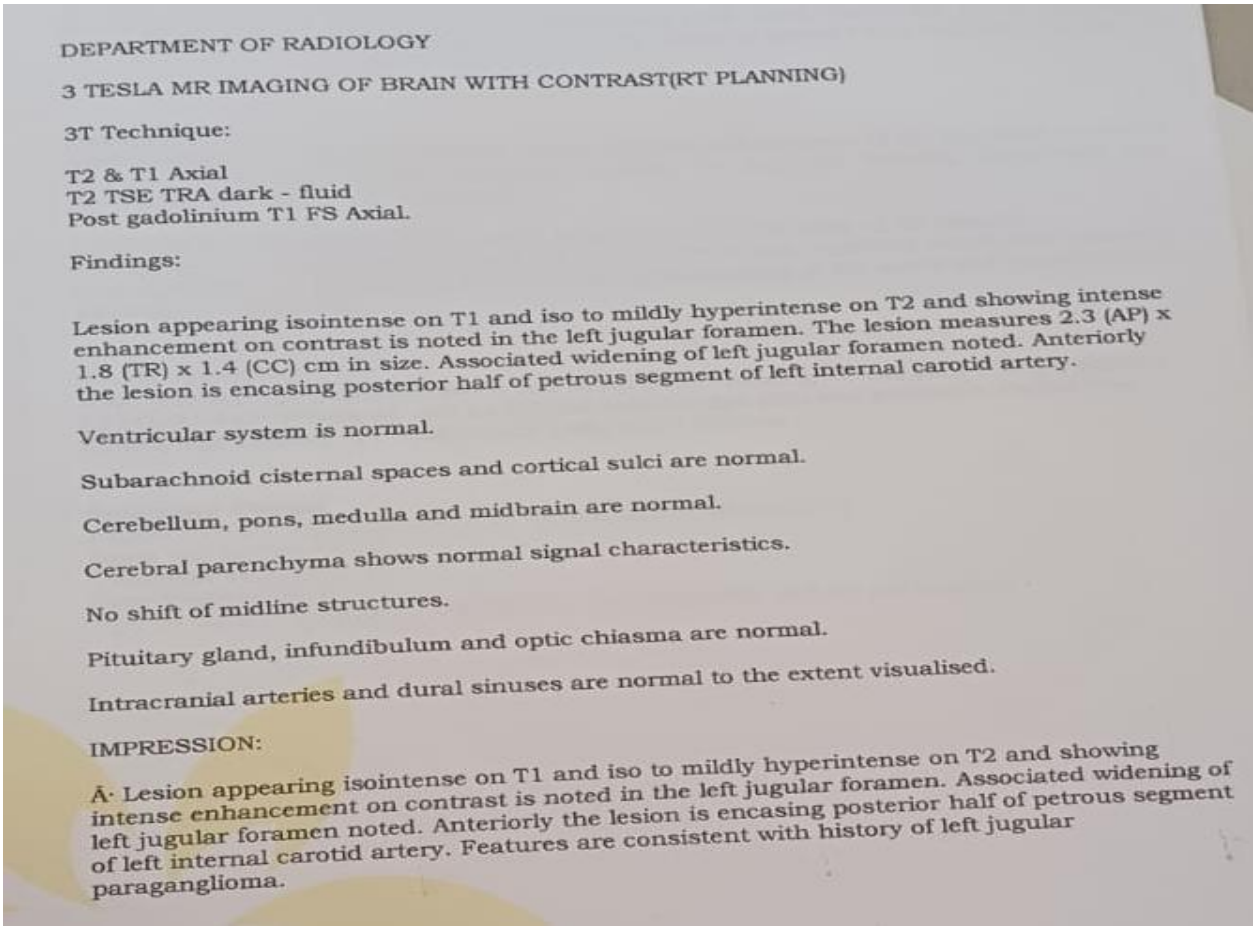
A 47-year-old woman presented to department of Neurology with 5 years of hearing impairment in the left ear and known history of hypothyroidism with a dental implant and symptoms began deviation od the tongue to the left side for about 6 months, difficulty in swallowing. The patient reported absence of dysphagia,seizures,giddiness or loss of consciousness. Patient appetite was reduced. She had no allergies and no pain. Her ECOG performance status was 1 and higher mental functions were normal. Patient also had pterygium in both eyes, which was incidental.

Investigations

MRI of the brain showed a 1.8×2.5×2 cm lesion in the left jugular foramen with vivid contrast enhancement .it also implies a separate 4mm lesion lateral to the cochlear promontory. Audiometry showed moderate to severe sensorineural hearing loss on the left screening ultrasound of the left jugular region was normal.

A3 Tesla MRI with contrast for radiotherapy planning was done. the lesion lay in the left jugular foramen. The lesion was isointense on T1 and iso to mildly

hyperintense on T2, with intense contrast enhancement and widening of the left jugular foramen. Anteriorly it encased the posterior half of the petrous segment of the interna carotid artery. It measured 2.3×1.8 (transverse)×1.4(craniocaudal) cm. The radiological impression was a left jugular paraganglioma.



DEPARTMENT OF RADIOLOGY

3 TESLA MR IMAGING OF BRAIN WITH CONTRAST(RT PLANNING)

3T Technique:

T2 & T1 Axial
T2 TSE TRA dark - fluid
Post gadolinium T1 FS Axial.

Findings:

Lesion appearing isointense on T1 and iso to mildly hyperintense on T2 and showing intense enhancement on contrast is noted in the left jugular foramen. The lesion measures 2.3 (AP) x 1.8 (TR) x 1.4 (CC) cm in size. Associated widening of left jugular foramen noted. Anteriorly the lesion is encasing posterior half of petrous segment of left internal carotid artery.

Ventricular system is normal.

Subarachnoid cisternal spaces and cortical sulci are normal.

Cerebellum, pons, medulla and midbrain are normal.

Cerebral parenchyma shows normal signal characteristics.

No shift of midline structures.

Pituitary gland, infundibulum and optic chiasma are normal.

Intracranial arteries and dural sinuses are normal to the extent visualised.

IMPRESSION:

Â· Lesion appearing isointense on T1 and iso to mildly hyperintense on T2 and showing intense enhancement on contrast is noted in the left jugular foramen. Associated widening of left jugular foramen noted. Anteriorly the lesion is encasing posterior half of petrous segment of left internal carotid artery. Features are consistent with history of left jugular paraganglioma.

Treatment and follow up

Patient was immobilised with a stereotactic (SRS) mask. A planning CT with 2mm slices and a planning MRI with and without contrast with 1mm slices were done. The gross tumour volume (GTV)was outlined on the T1 and T2 MRI sequences, and a 2mm margin was added to make the planning target volume (PTV). Treatment used as 6 MV flattening filter free (6FFF) photo beam. The bean has a high dose rate, so the prescription of dose for each session is as follows 24 Gy in 3 fractions of 8 Gy each, and treatment was given for 3 days.

Along with supportive medications were prescribed which include Dexamethasone 4 mg tapered over 15 days (three times a day for 5 days, then twice a day for 5 days, then once a day for 5 days), pantoprazole-domperidone once daily for 15 days, vitamin B complex neurobian forte once daily for 1 month and gabapentin 100mg and ondansetron 8mg only when needed for pain and nausea or vomiting.

Follow-up

At the follow up visit after 12 months, the patient complained only of occasional tightness over the ear and temporal region, headache and no other significant complaints. Her appetite, sleep, bowel and bladder habits were normal. she was in fair general condition. Her ECOG performance status was 1 and her Glasgow Coma Scale was E4V5M6(15/15). Higher mental functions and gait were normal, and power was 5/5 in all limbs. Pterygium in both eyes was noted again.

in short, before treatment she had left hearing loss and tongue deviation to the left. After treatment she had only occasional tightness over the left ear and temporal region, progressive decrease of tongue deviation, with no other significant complaints recorded.

III. DISCUSSION

The section Below involves the discussion of how the patient tumour affecting the hearing pathway and the hypoglossal nerve. Sections include imaging, results and importance of radiosurgery and dose used to treat tumour.

Imaging

MRI is the main investigation for these tumours and as well as suits pattern in our patient fits a jugular paraganglioma The lesion lay in the left jugular foramen ,which was isointense on T1 and iso to mildly hyperintense on T2 and enhanced intensely with contrast paragangliomas are very vascular ,which typically enhance brightly and often shows a salt and pepper look on T1 and T2 made of small bright spots and dark flow voids.(4)

Up to 10% of patients have more than one paraganglioma and it was good that screening ultrasound of the neck was done and was normal.

After radiosurgery, the usual MRI response is a stable size or slight shrinkage. In the 27 patient LINAC series MRI imaging observed partial remission in 12 patients and stable disease in 15 with no tumour progression (5) the follow-up MRI pf patient was done about 12 months after treatment.

Comparison of radiosurgery to other surgeries

A meta analysis study found long term recurrence of about 15% after surgery and 7% after SRS,SRS had lower rates of CSF leak, swallowing difficulty and palsies of the cranial nerves VII,IX,X,XI,XII (3). An earlier systematic review found that tumour control failure , major complications and cranial nerve palsies were more frequent in surgical therapies than in radiotherapies(2). In this case the patient tumour encased the petrous carotid artery and the hypoglossal nerve was already weak so surgery would have risked further and vascular injury. So to prevent the risk SRS was chosen in this case.

Single dose radiosurgery results

Single dose SRS,usually around 15 Gy to the tumour edge .in a series of 18 patients treated with gamma Kinfe, the tumour was controlled in 17 of 18 and more than half of the patients improved clinically (6). A LINAC based series of 21 patients reported 100% local control at a median follow up of 49 months.(7) .A Mayo clinic series of 85 patients treated with gamma knife in which 60 had MRI volume measures reported excellent long term control with a median of follow up of 66 months .three tumours grew more than 10 years after treatment so long term follow up matters (8)

Splitting the dose into 2 to 5 fractions is used when the tumour is larger or lies close to important structures (9) A study used 24 or 27 Gy in 3 fractions for vagal paragangliomas and came to a final conclusion that hypo fractionated SRS was safe and effective while protecting nerve function(10).In this report patient received 24 Gy in 3 fractions of 8 Gy.Splitting the dose to normalize tissue between fractions, which may protect the brain stem, cochlea and cranial nerves (11).

IV. CONCLUSION

Hypo fractionated SRS is a safe and non-invasive option for a tumour of the size that lies close to brain stem, inner ear and lower cranial nerves. This case of 47-year-old women with a 2.3 to 2.5 cm left glomus jugulare tumour causing hearing loss and hypoglossal nerve palsy and encasing carotid artery and was treated with 24 Gy in 3 fractions. At follow up she had only occasional tightness over the ear and temporal region and improved tongue deviation. These tumours grow slowly and can regrow late, long term clinical and MRI follow up should be done. SRS gives tumour control of about 95% or more with lower nerve injury than surgery, hypofractionation is an option for larger tumours. The central shift is toward functional preservation alongside growth control; the principal unresolved question is whether control and neurological outcomes remain durable over decades.

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