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Case Report

Glioblastoma multiforme at internal auditory canal

Syeda Alisha Ali Zaidi[1], Amanullah[1], Syeda Kubra Kishwar Jafri[2], Salman Sharif[1]

[1]Department of Neurosurgery, Liaquat National Hospital, [2]Department of Neurosurgery, Memon Medical Institute Hospital, Karachi, Pakistan.

E-mail: *Syeda Alisha Ali Zaidi - alish_ali@live.com; Amanullah - draman_a@yahoo.com; Syeda Kubra Kishwar Jafri - skubra.jafri@gmail.com; Salman Sharif - ssharif99@hotmail.com

***Corresponding author:**
Syeda Alisha Ali Zaidi,
Department of Neurosurgery,
Liaquat National Hospital,
Karachi, Pakistan.

alish_ali@live.com

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ABSTRACT

Background: Glioblastomas are the most common adult primary brain tumor present supratentorially. The presence of true extra-axial GBM infratentorially, especially in the internal auditory canal, is extremely rare with only three cases reported previously in the literature. We report the fourth case of primary internal auditory canal/cerebellopontine angle (CPA) glioblastoma which initially mimicked vestibular schwannoma on the basis of its location and presentation.

Case Description: A 65-year-old male presented with headache, vertigo, and progressive right ear deafness for 5 months. His preoperative magnetic resonance imaging findings were consistent with vestibular schwannoma. Maximum safe resection (near total) was done. The final histopathology report showed glioblastoma multiforme.

Conclusion: As per our knowledge, this is the fourth reported case of an extra-axial VIII cranial nerve glioblastoma located in internal auditory canal. Hence, despite being very rare, they should be considered as a differential in tumors at CPA.

Keywords: Glioblastoma, Internal acoustic meatus, Cerebellopontine angle, Vestibular schwannoma

INTRODUCTION

Glioblastomas are the most common adult malignant primary brain tumors arising from the glial cells. They are characteristically present in the supratentorial cerebral hemispheres and account for almost 20% of all intracranial tumors.[11] Its occurrence in infratentorial locations is extremely rare and mostly attributed to metastasis or extension from the supratentorial region. Few cases of primary infratentorial glioblastoma at the cerebellopontine angle (CPA) have been reported and were thought to be arising from the cerebellum or brainstem as exophytic mass.[12,15] The presence of true extra-axial glioblastomas, especially in the internal acoustic meatus (IAC), is extremely rare with only three cases reported previously in the literature.[17,19,20] We report the fourth case of primary IAC/CPA glioblastoma which initially mimicked vestibular schwannoma but after surgical resection was proven to be a glioblastoma, WHO Grade IV on histopathological grounds.

CASE PRESENTATION

This is the case of a 65-year-old male, with no known comorbid, who presented through outpatient clinic with the complaints of right-sided deafness, peripheral vertigo associated with

vomiting, and progressive headache for the past 5 months. Symptoms had become severe over the past 2 months making him completely deaf in the right ear. There was no complaint of altered level of consciousness, fits, falls, no visual symptoms, or facial weakness. His neurologic examination revealed a horizontal left beating nystagmus, right-sided sensorineural hearing loss, and no other cranial nerve dysfunction, however, Romberg's test was positive with impaired tandem walking.

Magnetic resonance imaging (MRI) of the brain with contrast showed a lesion in the right CPA, hypointense on T1-weighted sequences [Figure 1], hyperintense on T2-weighted sequences [Figure 2], and demonstrated homogeneous enhancement on T1 postgadolinium contrast images [Figure 3], with characteristic trumpet sign. The lesion was about 4 × 5 cm in size, compressing the ipsilateral cerebellar hemisphere and 4th ventricle. Based on the abovementioned findings, a diagnosis of vestibular schwannoma was made preoperatively.

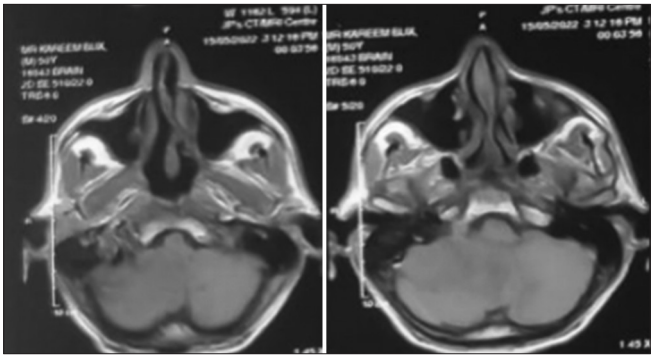

Figure 1: Magnetic resonance imaging brain T1-weighted sequences.

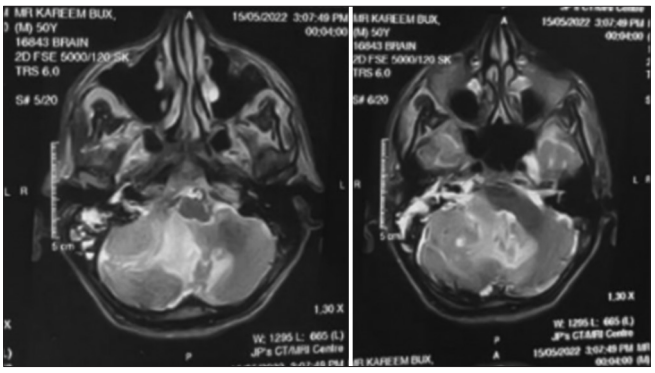

Figure 2: Magnetic resonance imaging brain T2-weighted sequences.

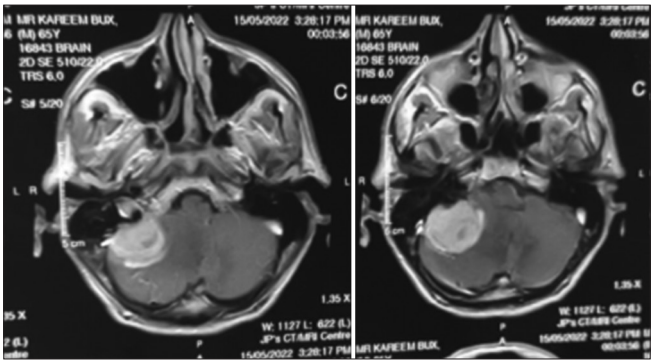

Figure 3: Magnetic resonance imaging brain T1-weighted postcontrast sequences.

Operative course

We positioned the patient in a park bench position with the right side facing up. After fixing the patient's head in the Mayfield head clamp, retrosigmoid craniotomy was done and surgery progressed in a usual fashion. The cerebellomedullary cistern was opened followed by drainage of cerebrospinal fluid. Cerebellum was retracted with the help of a Duro retractor and the tumor was identified. It was encapsulated and firm in consistency. The cerebellar invasion was evident along the medial aspect along with dense adhesions between the capsule and lower cranial nerves. After taking a biopsy, intracapsular tumor debulking was done with the help of an ultrasonic aspirator and near-total resection was achieved with the possible removal of all visible tumor mass.

Postoperative course

Postoperatively, the patient remained stable with no new neurology. His central nervous system examination was the same as the preoperative examination, and hence, he was discharged and followed up as an outpatient in the clinic. Histopathology revealed fragments of neoplastic lesions showing dense cellularity composed of markedly atypical cells with abundant eosinophilic cytoplasm and pleomorphic enlarged nuclei with vesicular chromatin and prominent nucleoli. Patchy areas of palisading necrosis were also seen. Few gemistocytic cells were seen. Mitosis of approximately 3–4/10 HPF was noted.

Immunohistochemical studies were performed by DAKO envision method using the following antibodies:

- GFAP patchy +ve
- S100 patchy +ve
- EMA –ve
- CKAE1/AE3 –ve

Ki67 approximately 8–10 % proliferative index.

All of these features favored the diagnosis of glioblastoma WHO Grade-IV.

The patient was then referred to a neuro-oncologist for further management, however, because of the poor prognosis, the patient refused further management and follow-up. On calling, after 2 months, we came to know that after remaining symptom free for about 1 month, the patient had started to have symptoms of vertigo and severe gait imbalance.

DISCUSSION AND LITERATURE REVIEW

Glioblastomas are WHO Grade IV primary intracranial tumors arising from glial cells. They commonly tend to occur in the supratentorial region, especially in frontal lobes, however, in very rare cases, gliomas (WHO Grades I–IV) may arise from other cortical areas such as brainstem cerebellum and spinal cord, in very rare instances, they arise from cranial nerves.[2,4-7,10-17,19,20] Our case is the fourth case of a true extra-axial IAC tumor mimicking vestibular schwannoma in its clinical course and radiological presentation. The three previously reported cases presented with common symptoms of sensorineural hearing loss, tinnitus, vertigo, and headache along with lower cranial nerve palsies [Table 1].[17,19,20]

Glioblastoma is classically hypointense to isointense, with a ring-enhancing pattern on postgadolinium T1 images. It is hyperintense on both T2 and FLAIR images. Magnetic resonance spectroscopy (MRS), showing larger choline, creatinine and lactate peaks, and a shallow N-acetylaspartate peak, is also used sometimes and is a more visionary imaging tool as it can aid in differentiating benign from malignant tumors.[1,20] Contrast MRI studies from the previously reported cases of IAC glioblastoma revealed similar MRI findings, however, these findings are also consistent with vestibular schwannoma which is currently the most common tumor arising from the eighth cranial nerve in the internal auditory canal, except for the vivid contrast enhancement rather than ring-enhancing pattern. Based on their MRI findings, diagnosis of vestibular schwannoma or meningioma was made in all previous cases including ours (misdiagnosed as vestibular schwannoma), which additionally demonstrated the "trumpet sign" that is characteristic of vestibular schwannoma.[3,17,19,20]

The current treatment of all newly diagnosed GBM patients is cytoreductive surgery (>98%), followed by the Stupp regimen (fractionated focal irradiation in daily fractions of 2 Gy given 5 days/week for 6 weeks (a total of 60 Gy), plus concomitant daily temozolomide (75 mg/m^2/day, 7 days/week from the first to the last day of radiotherapy), followed by six cycles of adjuvant temozolomide (150–200 mg/m^2/day for 5 days during each 28-day cycle).[18]

However, the current treatment options for vestibular schwannoma include observation, surgical resection, fractionated radiotherapy, and radiosurgery. Small, asymptomatic, or incidental tumors undergo a trial of observation versus radiosurgery whereas large tumors undergo surgical decompression, followed by fractionated radiotherapy or radiosurgery [Table 2].[8]

It has been reported that the overall survival of patients with newly diagnosed glioblastoma multiforme (GBM) is 17–30% at 1 year and only 3–5% at 2 years, despite access to state-of-the-art modalities of therapy. Young age and high KPS (Karnofsky Performance Scale) are the strongest positive predictors of survival.[1] However, 10-year progression-free survival rate for vestibular schwannoma was 87%.[9]

We additionally found a total of 14 overall reports of the tumors at a CPA out of which eight cases turned out to be low-grade gliomas, these eight cases were pilocytic and fibrillary astrocytomas mostly in children. The remaining five were tumors of unknown origin.[2,4-7,13,14,16] Only one tumor was found to be gliosarcoma. Of these, to the best of our knowledge, there are only three cases reported to be involving the internal acoustic canal and arising from the 8th cranial nerve that, on histopathological examination, was found to be glioblastoma WHO Grade IV.[17,19,20]

Following diagnostic confirmation, through histopathology, all patients were advised to undergo adjuvant therapy. Those who underwent chemo and radiotherapy had a symptom-free span of around 6 months–1 year, while patients who refused

Table 1: Previous case reports describing glioblastoma at internal auditory canal.

Study	Age (years), sex	Clinical presentation	Symptom duration	Size (mm)	Tumor origin	Chemo	RT	Ki-67 (%)	Follow-up
Wu *et al*., 2011	60, M	Facial numbness, facial palsy, hearing loss, dysarthria, dysphagia	2 months	36×35×33	CN VIII	No	No	40–50	2 months
Yang *et al*., 2018	55, M	Hearing disturbance, vertigo, gait disturbance, facial palsy, facial numbness, hoarseness, dysarthria, dysphagia, headache	3 months	40×33×28	CN VIII	No	No	ND	2.5 months
Takami *et al*., 2019	55, M	Imbalance, hearing loss, tinnitus	19 months	24×22	Possibly CN VIII/IAC	TMZ	60 Gy/30 Fr	ND	3 months
Our case	65, M	Headache, vertigo, hearing loss	5 months	40×50	Possibly CN VIII/IAC	no	no	8–10	2 months

CN VIII: Cranial nerve seven, CNVIII: Cranial nerve eight, ND: Not documented, TMZ: Temozolamide, Gy: Grey, Fr: French, IAC: Internal acoustic meatus

Table 2: Treatment outlines for vestibular schwannoma.

Clinical situation	Recommendation	Evidence class	Recommendation level
Spontaneous VS, small asymptomatic	Observation OR SRS	III	C
		II	B
Spontaneous VS, small, complete hearing loss	Observation OR	III	C
	SRS superior to	II	B
	Surgery	III	C
Spontaneous VS, large with brainstem compression	Surgery inferior to	IV	GPP
	Combination surgery+SRS	IV	GPP
NF2	Surgery and/or	IV	GPP
	SRS and/or	IV	GPP
	bevacizumab and/or	II	B
	SRT dependent on situation	IV	GPP

GPP: Good practice point, VS: Vestibular schwanoma; SRS: Stereotactic radiosurgery; NF2: Neurofibromatosis type 2. B and C are level of evidences as followed by different guidelines

any further management survived for around 1–2 months without recurrence of symptoms.[6,15,17,19,20]

CONCLUSION

There have been multiple cases reported about the presence of glioblastoma at the CPA which mimicked vestibular schwannoma or meningioma due to their location and presentation on MRI. MRS may help in differentiating between benign and malignant tumors. These tumors should be followed aggressively to detect any suspicious malignant activity to treat them early and prevent or delay the recurrence to provide maximum symptom-free survival.

Declaration of patient consent

Patient's consent not required as patient's identity is not disclosed or compromised.

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Conflicts of interest

There are no conflicts of interest.

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