

Pre-op Deaf Post-op Dumb: A case report of a Tectal Plate Glioma and a Brief Literature Review

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Abstract

Any disorder of the brain stem that involves the auditory pathways and/or centers may produce hearing abnormalities. Auditory pathways are tonotopically organized, and they cross the midline at several points both below and at the level of the inferior colliculus but the central nucleus of the inferior colliculus and higher centers have a clear contralateral dominance. Ipsilateral auditory abnormalities are caused by lesions below or within the cochlear nuclei; bilateral auditory abnormalities are caused by lesions rostral to cochlear nuclei or they may be silent. A patient with a tectal plate glioma can present with bilateral hearing impairment due to the involvement of both inferior colliculi.

Our case is a 28-year-old male who presented with progressive bilateral hearing impairment and a generalized headache despite normal pure-tone audiometry. Computed tomography (CT) of the brain revealed a tectal plate glioma (TPG) extending into the right thalamus, and an obstructive hydrocephalus. Following a ventriculoperitoneal shunt for cerebrospinal fluid (CSF) diversion, navigation-guided tumour excision was performed via a supracerebellar infratentorial approach. Postoperatively he developed unexpected aphasia probably due to ischemic changes in the cerebellar vermis and a focus of the adjacent right cerebellar hemisphere. This case highlights the rarity of bilateral auditory impairment in a TPG, surgical challenges posed by the tumour's location, and potentially rare post-op complication of cerebellar mutism syndrome.

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Introduction

Any disorder of the brainstem that involves the auditory pathways and/or centers may produce hearing abnormalities. Ipsilateral auditory abnormalities are caused by lesions below or within the cochlear nuclei; bilateral auditory abnormalities are caused by lesions rostral to cochlear nuclei or they may be silent (1).

According to literature, auditory testing of unilateral lesions in the inferior colliculus are, normal bilateral pure tone audiogram (PTA), tinnitus in the contralateral side, normal or impaired bilateral or contralateral speech discrimination, defective threshold tests, defect in localization of sound contralateral to the lesion, difficulty in understanding speech in a noisy environment and bilateral abnormal auditory brainstem response (2).

In theory, bilateral hearing loss could occur from lesions in the central hearing pathways but in practice, this is unlikely because the involvement of vital structures in the neighboring brainstem and hemispheres will produce vast neurologic disability masking the hearing loss (3). Here we present a case of tectal plate glioma involving bilateral inferior colliculi, initially presented as progressive bilateral hearing impairment, its anatomical consideration, surgical management, and postoperative complication of aphasia with possible cerebellar mutism syndrome.

Case report

A twenty-eight-year-old male presented to the general medical unit with progressive bilateral

hearing impairment, right-sided tinnitus, faintishness, and generalized headache for three weeks duration. He had no other motor or sensory symptoms attributable to other cranial nerves. His neurological examination was normal other than bilateral hearing impairment. There were no papilledema, cerebellar signs or signs of Parinaud syndrome. Other system examinations were unremarkable. Bilateral tympanic membranes were normal on examination.

Pure tone audiometry (PTA) was performed, and it revealed normal hearing except mild sensorineural hearing loss (SNHL) at 250 Hz on the right side.

He was managed initially as sinusitis following which he had further progression of hearing impairment, worsening of headache, and blurring of vision. Non-contrast CT of the brain was performed, and it revealed a lesion in relation to the tectal plate extending to the right thalamus with obstructive hydrocephalus (Figure 1).

The patient was referred to the Neurosurgical unit and an emergency ventriculoperitoneal (VP) shunt was performed following which headache and blurred vision improved but hearing impairment persisted.

Contrast enhanced magnetic resonance imaging (MRI) of the brain was performed which revealed further details of the lesion which arose from the tectal plate and filled the quadrigeminal cistern. Ventricle size has been reduced to the normal (Figures 2, 3).

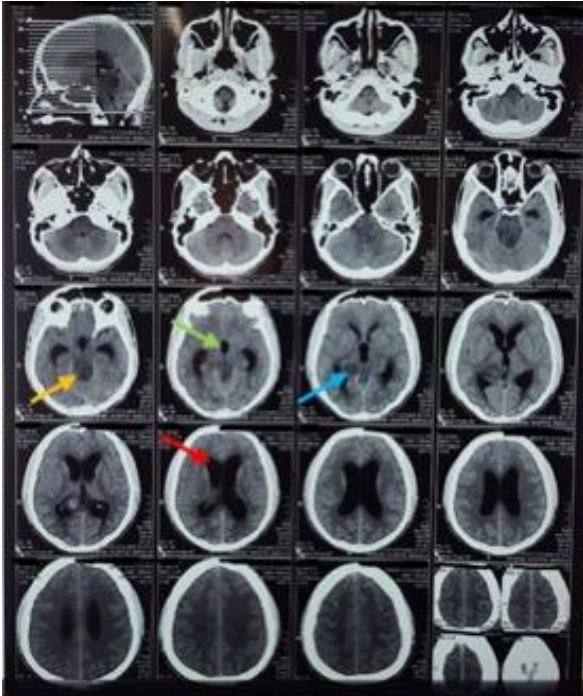


Figure 1: CT showing the lesion (Yellow arrow) in tectal plate infiltrating the thalamus (Blue Arrow) and Obstructive Hydrocephalus with enlarged 3rd (Green arrow) and lateral ventricles. (Red arrow)



Figure 2: MRI in sagittal plane showing the tumor in relation to the tectal plate. (Yellow arrows)

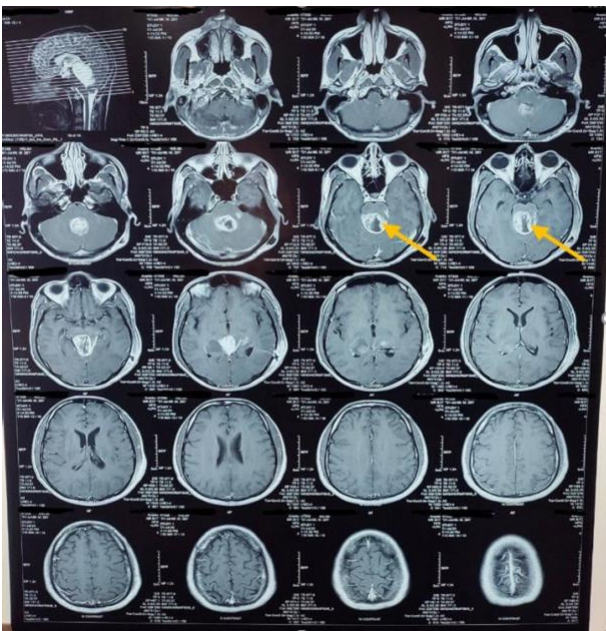


Figure 3: MRI in the transverse plane showing the tumor in relation to the tectal plate. (Yellow arrows)

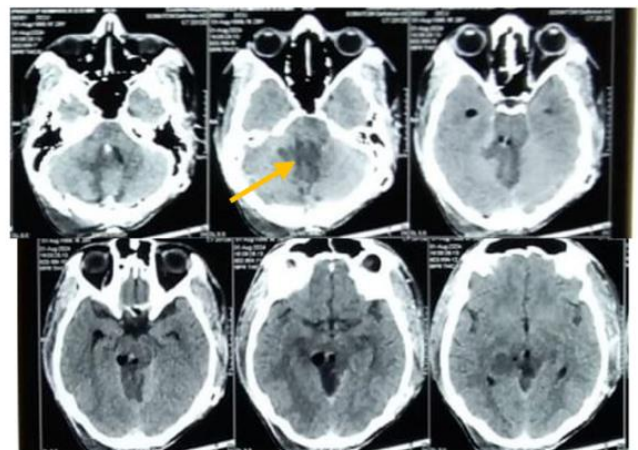


Figure 4: Post-op CT showing Ischemic changes in the Cerebellar vermis and the focus of adjacent right cerebellar hemisphere. (Yellow arrow)

While awaiting surgical excision of the tumour, level of consciousness of the patient deteriorated without prior seizure or aspiration.

The patient underwent navigation-guided tumour excision under general anesthesia. The patient was placed on prone position in head fixator device, a midline straight incision was made in the suboccipital region, muscles were dissected, and the incision was deepened into the bone. A suboccipital craniotomy was done and the bone flap was elevated. Dura was opened and an infratentorial supracerebellar approach was used to reach the tumour.

The tumour was identified filling the quadrigeminal cistern and navigation-guided tumour excision was done. Samples were sent for histopathology and wound was closed in layers after replacing and fixing the bone flap.

The patient was monitored in the critical care unit following the surgery. Intravenous Dexamethasone 8 mg tds was given to reduce surgically induced tissue edema, and Levetiracetam 500 mg bd was continued.

The patient regained consciousness, but the postoperative period was complicated by aphasia and aspiration pneumonia. Post-operative CT scan of the brain showed ischemic changes in the cerebellar vermis and focus of the adjacent right cerebellar hemisphere (Figure 4).

Aspiration pneumonia was improved with intravenous antibiotics and chest physiotherapy, but aphasia persisted even after 4 weeks following surgery.

Post-op histopathological assessment revealed a glial tumour with focal necrosis with a predominant microcystic pattern and focal gemistocytic areas with mild nuclear pleomorphism, Mitotic figures were not detected but focal microvascular proliferation was noted.

Discussion

Brainstem gliomas most commonly occur in the pediatric population and their incidence is much lower in the adult population. According to Reyes-Botero et al. brainstem gliomas account for 1% - 2% of all gliomas in adults, and tectal plate gliomas represent a small fraction of these (4).

Focal deficits are rarely observed and are mostly reversible after cerebrospinal fluid (CSF) diversion.

Conservative management, surgical excision, radiotherapy, or stereotactic radiosurgery are management options for TPGs. Because of the quiescent nature of these tumours management has followed a conservative approach. But sometimes these tumours behave aggressively despite their radiological appearances (5).

Anatomical consideration

Mechanical energy of sound waves are transformed into bioelectrical action potentials at the cochlear hair cells and are transmitted to the spiral ganglion comprised of the auditory nerve. At the cochlea high-frequency sounds stimulate the basal components and low-frequency sounds stimulate the apical component demonstrating the tonotopic

organization of the cochlea. Similarly in the auditory nerve, fibers from the low-frequency apical portion of the cochlea are located centrally and fibers from high-frequency basal portions are located peripherally. These signals traverse an ascending path through a series of nuclei, the cochlear nuclei, superior olivary complex, lateral lemniscus, inferior colliculi, and medial geniculate body (6,7).

The first relay point of the auditory pathway in the brainstem is the cochlear nucleus which is situated in the posterolateral portion of the brainstem. It contains low-frequency fibers in the ventral region and high-frequency fibers in the dorsal region. Neural fibers decussate across the midline into the contralateral superior olivary complex (6,7). Fibers then ascend through the lateral lemniscus to the ipsilateral inferior colliculus (8). The medial geniculate body receives afferent input from the inferior colliculus and transmits to the tonotopically organized auditory cortex (6).

Even though auditory pathways cross the midline at several points both below and at the level of the inferior colliculus, the central nucleus of the inferior colliculus and higher centers has a clear contralateral dominance. The primary auditory cortex is in the posterior half of the superior temporal gyrus and transverse temporal gyri (Gyri of Heschl) (9) (Figure 5).

Both inferior colliculi are contained within the tectal plate which also accommodates the superior colliculi, posteriorly to the mesencephalic aqueduct hence the name quadrigeminal plate.

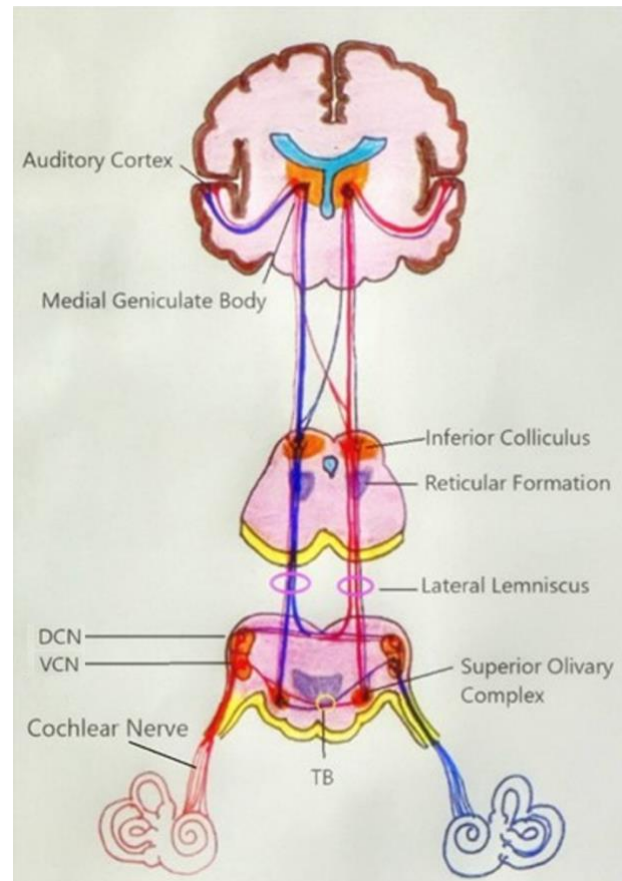


Figure 5: Auditory Pathway, VCN: Ventral Cochlear Nucleus, DCN: Dorsal Cochlear Nucleus, TB: Trapezoid body

Clinical presentation

Theoretically, a patient with a tectal plate glioma can present with bilateral hearing impairment due to the involvement of both inferior colliculi (10). Features of Parinaud syndrome may be present due to the involvement of the dorsal midbrain region. Headache and blurred vision/double vision with or without reduced level of consciousness can be present due to obstructive hydrocephalus because of compression and obstruction of the aqueduct by the lesion (11). Nevertheless, in a retrospective cohort study including 39 adult patients with tectal plate gliomas, none of the patients had bilateral

hearing impairment as their presenting symptom (12). Our patient had progressive bilateral hearing impairment, headache and blurred vision. The latter 2 symptoms were relieved following CSF diversion.

Hoistad et al. documented a case of central nervous system lymphoma partially involving bilateral inferior colliculi which presented with central hearing loss. This patient had relatively normal pure tone averages with severely reduced bilateral word recognition scores (13).

Vitte et al. reported 2 cases of word deafness which occurred in one patient after a head injury and in the other patient following a therapeutic embolization of an arteriovenous malformation that had induced a venous infarction of the two inferior colliculi. In these patients, MRI demonstrated bilateral lesions of the inferior colliculi, but brainstem auditory-evoked potentials (BAEP) were within normal limits, thus demonstrating bilateral inferior colliculi lesions induced pure word deafness without affecting BAEP (14). Another case report revealed hearing impairment due to compression of the inferior colliculus by a pineal gland cyst, in which hearing improved following excision of the cyst (15).

Our patient had normal PTA except mild SNHL at 250Hz on right side. BAEP and word recognition tests were not performed as the patient underwent emergency surgery. Post-operatively hearing was not improved. The reason may be the infiltrative nature of the tumour which has damaged the bilateral inferior colliculi.

Conservative Management

Due to the slow-growing nature of TPGs conservative management is considered first before moving on to surgical management, especially in asymptomatic patients or patients with minimal symptoms. This is supported by the fact that many TPGs remain stable without considerable clinical progression (16).

Management of hydrocephalus is prioritized while monitoring the tumour with regular imaging and clinical evaluation (17). In cases where hydrocephalus is involved, conservative management may contain CSF diversion techniques such as Endoscopic third ventriculostomy (18,19) And ventriculoperitoneal (VP) shunt.

Our patient had features of increased intracranial pressure at the presentation and therefore underwent VP shunting. Following CSF diversion hydrocephalus was relieved with clinical and radiological improvement, but the patient again deteriorated which led to the definitive surgery.

Surgical approach

Surgical access to the tectal plate tumours has been described as suboccipital supra and transtentorial approach, infratentorial supracerebellar approach, and endoscopic approach.

Lapras et al. reported a case series of 12 patients with a mean age of 19 years who were treated with direct surgery. Their MRIs showed 4 intrinsic, 6 exophytic, and 2 ventrally infiltrating tectal tumours. In every case, suboccipital supra and transtentorial

approaches were used. Resection of tumours at the level of the superior colliculi was generous but at the level of the inferior colliculi resection was limited due to the auditory risk (20). Fumiaki Oka et al. reported a case of hemorrhagic tectal pilocytic astrocytoma in which a 21-year male underwent total resection via occipital transtentorial approach using a neuronavigation system. The right dorsolateral midbrain was explored through this approach and acute hemorrhage was observed through the pia matter (21).

In a retrospective analysis of eight patients (mean age 16.6+/- 11.5) with low-grade astrocytoma total resection was achieved only in seven patients using an infratentorial/supracerebellar approach but in one case due to the infiltrative pattern of the lesion radical removal was not possible (22). In a study by Sima Sayyahmelli et al. they detailed the microsurgical techniques and specific considerations in using a transcollicular approach in resection of a tectal glioma. The subject was a 37-year-old male presenting with a well-defined, non-enhancing cystic mass located in the dorsal midbrain, exerting pressure on the aqueduct and causing hydrocephalus. They used an infratentorial, supracerebellar, transcollicular approach for surgical excision of the tumour. Motor and somatosensory evoked potentials were monitored during the procedure and the operation proceeded without complication. Post-operative recovery was uneventful. The follow-up MRI showed gross total resection of the tumour (23).

Perin et al. performed an endoscopic third ventriculostomy and tumour biopsy in a 24-

year male patient with TPG, with a flexible endoscope through a single burr hole, thereby achieving CSF diversion and histopathological diagnosis at the same time (24).

Our patient underwent suboccipital craniotomy and the tumour was accessed through infratentorial/supracerebellar approach with neuro navigation. Intra-operative period was uneventful, and the tumour was grossly excised. Post-operative CT scan of the brain showed gross total resection except the part infiltrating into the right thalamus.

Postoperative aphasia

De Smet et al. summarized the various hypotheses regarding the functional role of the cerebellum in language procession. The first hypothesis, "diaschisis" describes changes in cerebral blood circulation secondary to decreased excitatory impulses from the cerebellum potentially leading to impairment of higher cortical functions. The second hypothesis "timing hypothesis" states that language disorders arise from the failure of time-dependent modulation of linguistic functions. The third hypothesis encompasses the opinions that maintain the cerebellum's direct involvement of language processing (25).

Mariën et al. showed hypoperfusion in the left medial frontal area in a patient with right superior cerebellar artery infarction using Single photon emission computed tomography (SPECT) analysis (26). Similarly, Zettin et al. presented a case of expressive agrammatism following a hemorrhage in the right cerebellum associated with left fronto-temporal

hypoperfusion identified with SPECT (27). Additionally, Silveri et al. reported a case involving dysarthria and agrammatic speech, associated with word errors that emerged after a right cerebellar infarction (28).

It is generally accepted that right cerebellar lesions lead to linguistic disorders and left cerebellar lesions lead to deficits in visuospatial function because the cerebellum is linked with the contralateral cortex (29).

Furthermore, damage to tissue structures related to the cerebellar vermis and its close vicinity to the surgical approaches to offending lesions have been recognized as critically important in development of cerebellar mutism syndromes (30).

Our patient had postoperative aphasia attributable to ischemic changes of the cerebellar vermis and adjacent right cerebellar hemisphere seen on postoperative CT scan.

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Conflicts of interest

The authors report no conflict of interest.

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