Neuroscience | Case Report

Hearing Impairment in Pineal Tumors

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Background: **Patients with pineal tumors commonly present with symptoms due to increased intracranial pressure and tectal deformation. However, hearing loss and tinnitus are likely under-represented, underestimated symptoms caused by pineal tumors.** *Case description:* **A 32-year-old male presented with symptoms of acute lethargy, confusion, headaches, nausea, vomiting, and slurred speech in addition to subacute visual and hearing impairment. Prior to his acute presentation, both an audiogram and a brainstem auditory evoked response (BAER) test were performed and found to be normal. He was found to have a large pineal germinoma, which was debulked. His subjective hearing impairment improved postoperatively with the rest of his symptoms.** *Conclusions:* **The case presented demonstrates the importance of proceeding with intracranial imaging in patients with subjective auditory symptoms, even with normal audiologic testing, and serves as a reminder for the clinician to keep pineal tumors in the differential diagnosis for hearing loss/tinnitus.** *Journal of Nature and Science, 1(3):e48, 2015.*

Brainstem auditory evoked response test | germinoma | hearing loss | pineal tumor | speech audiometry

Introduction

Pineal region tumors comprise 1-2% of primary intracranial tumors in adults and 3-8% in children [1]. Neuro-ophthalmologic findings, classically Parinaud syndrome, are common. Hearing loss and tinnitus have also been described in patients presenting with pineal tumors. These symptoms are explained by the proximity of the pineal region to the auditory pathways in midbrain, upper brainstem, and diencephalon.

Hearing problems in patients with pineal tumors occur more commonly than expected, with some case series showing a prevalence of up to 20% [2-7]. We present a case that highlights the importance of keeping pineal tumors in the differential diagnosis of patients with subjective hearing loss, even those with normal auditory testing. A literature review follows that demonstrates the heterogeneity of audiometric and brainstem auditory evoked response (BAER) tests in these patients and the various trajectories of improvement seen after the lesions are treated.

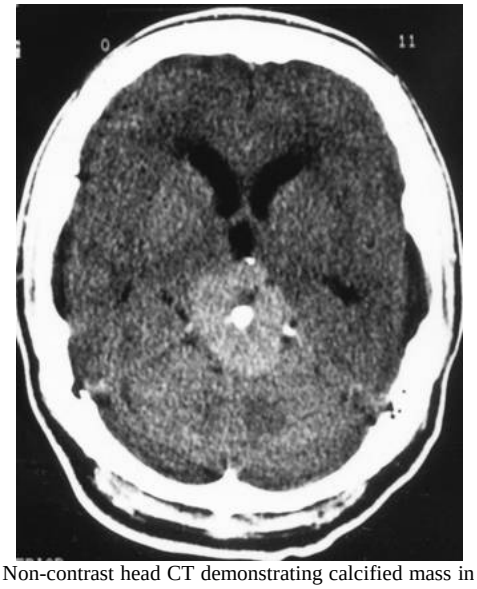

Figure 1. Non-contrast head CT demonstrating calcified mass in the pineal region with resultant obstructive hydrocephalus.

Case description

History

A 32-year-old, right-handed male presented to the Emergency Department for symptoms of lethargy, confusion, headaches, nausea, vomiting, and slurred speech. He had been complaining of decreased hearing for 3-4 months, and had been treated for recurrent ear infections during that time. Nineteen days prior he had a normal audiogram. One week before admission a BAER test was performed and was also completely normal.

Examination

On examination, the patient was lethargic but easily aroused. Cranial nerve examination was significant for Parinaud syndrome. Speech was dysarthric, and his responses were often inappropriate, with difficulty following commands. Unenhanced head CT showed a moderately dense mass in the pineal gland region, with central calcification (Figure 1). The mass obliterated the posterior third ventricle with resultant moderately enlarged lateral ventricles. MRI showed the mass to be densely enhancing (Figure 2).

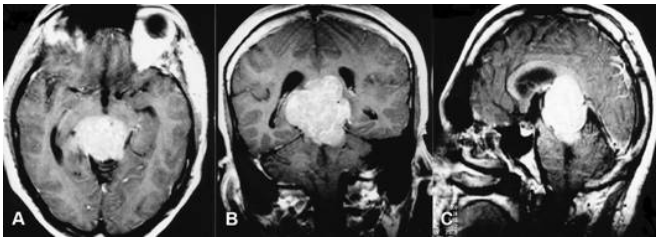

Figure 2. Axial (A), coronal (B), and sagittal (C) T1-contrast MRIs of brain showing extent of tumor.

Operation

An external ventricular drain was placed; drainage resulted in immediate improvement in lethargy, but his other symptoms did not resolve. He then underwent surgery via supracerebellar infratentorial approach to debulk the mass. Histologic diagnosis was germinoma.

Postoperative Course

Postoperatively, the patient's preoperative symptoms resolved and his hearing returned to normal. He underwent a course of radiation therapy after healing from his operation. Audiogram performed 1 year postoperatively continued to be normal.

Discussion

The true incidence of hearing problems in pineal tumors is difficult to determine, mainly due to the lack of routine preoperative and postoperative auditory testing performed in these patients. Moreover, hearing impairment is rarely the predominant presenting symptom and is more likely overshadowed by the more typical complaints caused by pineal tumors. Hearing impairment associated with pineal tumors is most likely explained by the pineal gland's location near the lateral lemniscus, inferior colliculus, and medial geniculate nucleus of the thalamus, all structures important

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Conflict of interest: No conflicts declared.

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Table 1. Results of literature review for patients with pineal tumors and hearing test results

Reference	Auditory Symptoms at Presentation	Initial Audiogram Results	Initial BAER	Operation/Intervention	Postoperative Improvement in Auditory Symptoms	Post-Intervention Hearing Test Results
Starr & Hamilton [8]	None	None	Prolonged latency in waves II and III; small IV-V wave at prolonged latency	Ventriculoatrial shunt placement	N/A	N/A (patient expired)
Missori et al.[9]	Tinnitus	Severe bilateral neurosensory hypoacusia on medium-acute waves	Normal	Craniotomy	Yes; complete resolution of tinnitus	Unchanged audiometric examination
	Not stated	Bilateral impairment of acoustic sensation	Normal	Craniotomy; previously fitted with ventriculoatrial shunt	Not stated	N/A (tests refused)
	None	Slight impairment of acoustic sensation	None performed	Craniotomy	No change; no preoperative auditory symptoms	Normal audiometric testing
Gaspar et al.[7]	None initially; complete hearing loss after shunt placement	Profound bilateral perceptive hearing loss, no speech recognition or detection	Normal	Unsuccessful ventriculocisternostomy followed by placement of ventriculoperitoneal shunt and chemoradiation	Yes; return to normal function	Normal audiometric testing 4 months from start of chemoradiation
Woo et al. [10]	Bilateral hearing impairment	Significant left high-frequency hearing impairment with bilateral speech discrimination impairment	Bilateral increased interpeak latency of waves III-V	Cyst aspiration via Rickham reservoir followed by craniotomy. Previous history of pineal region ganglioglioma with repeated excisions, adjuvant radiotherapy, and ventriculoperitoneal shunting	Transient dramatic improvement after cyst aspiration; continued improvement after craniotomy	Improvement in audiometric testing and BAER after cyst aspiration
DeMonte et al. [11]	Difficulty hearing	Near normal pure tone definition; significantly disturbed speech discrimination	Increased latency of wave V bilaterally and slight increased latency of wave III on right	Craniotomy	Improvement in hearing	-"Impressive" improvement in speech discrimination scores - No change in BAER
Toshniwal et al. [12]	Progressive hearing loss and tinnitus; complete deafness	Profound bilateral sensorineural hearing loss	No response	Ventriculoperitoneal shunt followed by craniotomy and radiation	Continued hearing impairment	BAER absent
Kashlan et al.	Decreased hearing	Normal	Normal	External ventricular drain followed by craniotomy	Improved to normal	Normal audiometric testing

BAER = brainstem auditory evoked response

in transfer of information from the cochlea to the primary auditory cortex. Another possible explanation includes the ability of increased intracranial pressure to also be a false-localizing sign of hearing loss [7]. However, the former theory is likely more valid, as the patient described by Gaspar et al. developed hearing problems after his intracranial pressure was normalized by placement of a ventriculoperitoneal shunt while awaiting chemoradiation [7].

Interestingly, there is a paucity of information regarding the effect of pineal tumors on audiometric and BAER tests. We performed a literature review for reports of pineal tumors causing hearing loss that included associated hearing test results (Table 1) [7-12]. Six of the 7 patients who underwent audiogram had significant abnormality. The other patient did not have auditory complaints and therefore only had slight impairment of acoustic sensation on audiometric testing. Three of 7 patients who underwent audiograms had disturbed speech recognition. Our patient had a completely normal audiogram preoperatively. With regard to BAER results, 3 of 7 patients who underwent BAER testing had a normal examination similar to our patient. Three others had abnormalities in waves III-V, which is expected for the location of a pineal lesion. Previous results show that audiometric

testing is likely more sensitive in characterizing hearing loss due to pineal tumors than BAER testing. Moreover, it demonstrates the importance of keeping a central cause of hearing loss in the differential diagnosis for a patient with subjective auditory symptoms, even with normal audiometric or BAER testing.

Regarding postoperative improvement in symptoms, with the exception of the patient who presented with subacute complete deafness, all patients had improvement in auditory symptoms much like our patient did. In the 5 patients with post-intervention audiometric testing, 4 had concurrent improvement in their results, while 1 had an unchanged examination, even with improved preoperative symptoms. In contrast, in the 3 patients with post-intervention BAER testing, 1 had improvement while 2 others had no change. One of these 2 patients presented with complete deafness that did not improve postoperatively. These results demonstrate the ability to use audiologic and BAER testing as a correlate to improvement of functional hearing after an intervention is performed.

Conclusions

In summary, our case serves as a reminder to clinicians to keep pineal tumors in the differential diagnosis for patients presenting with hearing loss. Moreover, it also demonstrates that hearing loss can be occult to detection on hearing tests, therefore making it important to proceed with cranial imaging in a patient with continued auditory complaints and normal audiograms and/or BAER tests.

Acknowledgments

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