



Clinical Reasoning: A 45-year-old woman with reversible bilateral hearing loss

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SECTION 1

A 45-year-old woman complained of a progressive 2-month history of bilateral hearing impairment and diplopia on upward gaze. She had a history of a recurrent pineal region ganglioglioma with repeated tumor excision, adjuvant radiotherapy, and a ventriculo-peritoneal shunt performed 12 years prior. Subsequent imaging studies 6 years ago showed a pineal region cyst with progressive increase in size and a Rickham reservoir (Codman; Johnson & Johnson, Raynham, MA) was placed for percutaneous cyst fluid aspiration. The size of the cystic lesion remained static upon follow-up CT scans for several years.

On the day of admission, the woman did not complain of headache and there were no blurring of vision. On physical examination, the patient was fully conscious with significant bilateral hearing impairment and upward conjugate gaze palsy. Her visual acuity was 20/40 in each eye and her visual fields were intact. Fundoscopy showed no evidence of papilledema. The ventriculo-peritoneal shunt valve was compressible and refilled quickly.

Questions for consideration:

1. What other physical signs could be elicited in view of her past medical history?
2. What are the main differential diagnoses to consider?

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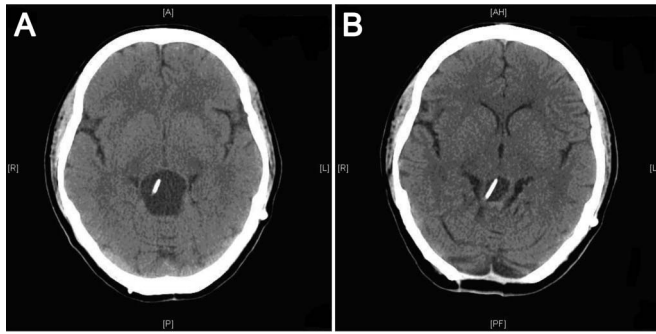
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Figure 1 CT of the head



(A) CT scan before pineal region cyst aspiration. (B) CT scan after cyst aspiration. The tip of the Rickham catheter can be seen within the pineal region cyst.

SECTION 2

On further physical examination, there was bilateral loss of the direct pupillary light reflex, but preservation of the accommodation reflex. No convergence nystagmus could be observed.

The main differential diagnoses were tumor progression, postradiation therapy cystic change, and recurrent hemorrhage with cystic transformation. Tumor progression could be diagnosed by an increase in solid component of the tumor.

The first investigation (after a complete bedside clinical examination) was a noncontrast CT scan

of the head to assess the known pineal region cyst, reservoir and shunt placement, and any other intracranial pathology. MRI would yield higher resolution images, but may not be urgently available. Her CT scan showed a large pineal region cyst with a significant interval increase in size compared to her last imaging study a year ago. The intracranial Rickham reservoir catheter was in situ within the cyst (figure 1A). The ventriculo-peritoneal shunt catheter was also in position and there was no hydrocephalus. Percutaneous aspiration of the cyst via the Rickham reservoir was attempted and a total of 20 mL of straw-colored fluid was yielded. Her bilateral hearing improved dramatically and her upward conjugate gaze palsy was completely resolved after cyst aspiration. A repeat CT scan showed a marked reduction in size of the pineal region cyst (figure 1B). MRI brain scan revealed a pineal region cyst with thick enhancing wall compressing against the midbrain and posterior third ventricle. The aspirated fluid was sent for cytologic examination, returning negative for malignant cells.

Questions for consideration:

1. Why would fluid reaccumulate in the pineal region cyst?
2. What further investigations should be performed to evaluate her hearing loss?

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SECTION 3

Fluid reaccumulation in the pineal cyst was postulated to be related to recurrent microhemorrhage or active secretion from the cyst wall.

Pure tone audiogram (figure 2A), speech discrimination assessment (figure 2B), and brainstem auditory evoked potential investigations (figure 2C)

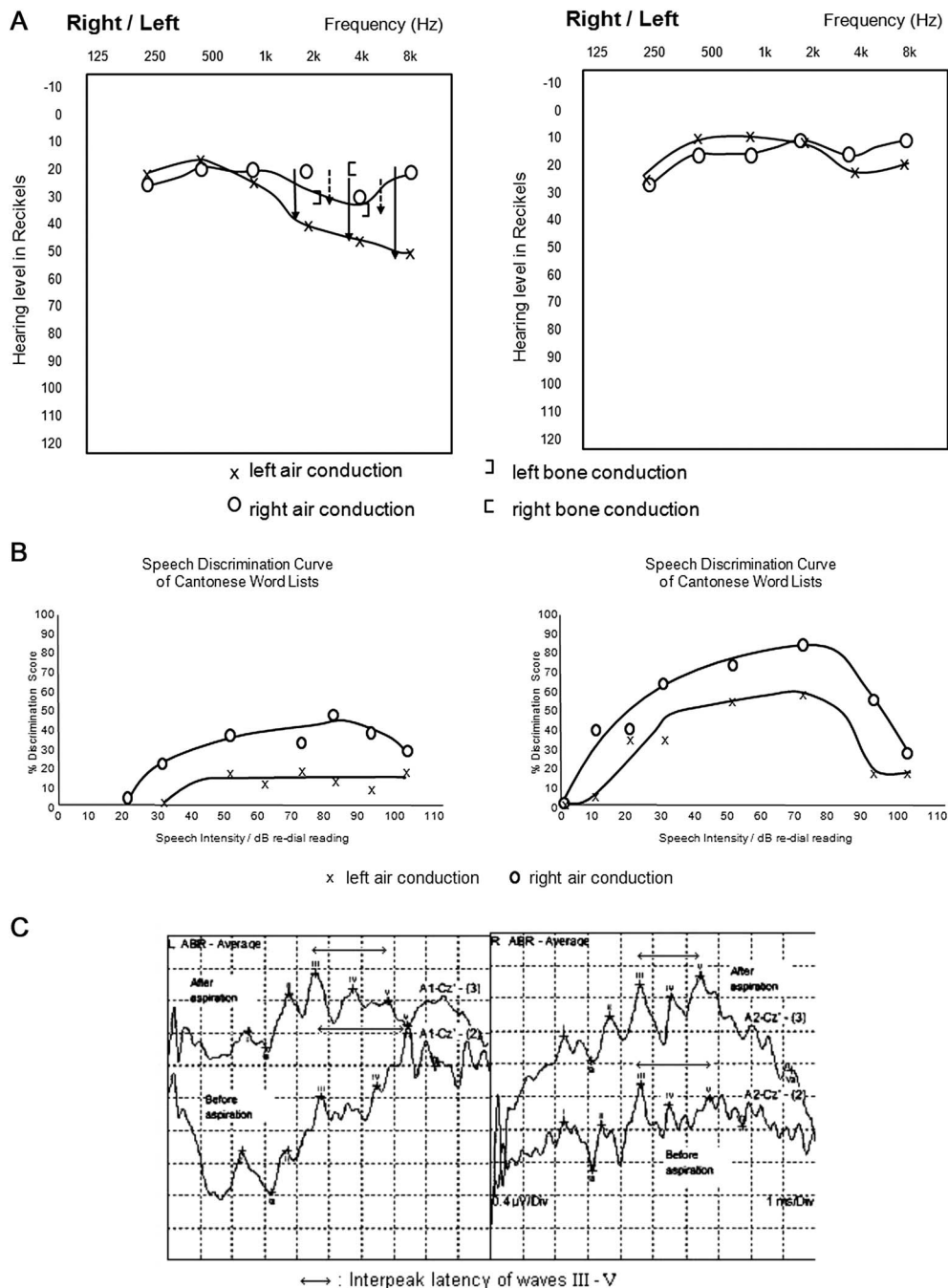
were performed before and after the aspiration of the cyst.

Question for consideration:

1. How would you interpret the pure tone audiogram (PTA), speech discrimination assessment, and brainstem auditory evoked potential (BAEP) results?

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Figure 2 Pure tone audiogram, speech discrimination assessment, and brainstem auditory evoked potential



Pure tone audiograms (A), speech discrimination assessment (B), brainstem auditory evoked potential (C) before (left) and after (right) pineal region cyst aspiration.

SECTION 4

PTA showed a significant left high-frequency hearing impairment on both air and bone conduction (suggested neurosensory hearing impairment), which recovered after aspiration. Speech discrimination assessments showed bilateral impairment (left more than right), which improved significantly after aspiration. The characteristic BAEP patterns for this patient suggest a direct compressive effect of the lesion against the tectal plate. Preaspiration BAEP recordings revealed bilateral increased interpeak latency of waves III–V (left more than right), which reduced after aspiration.

The patient developed recurrent bilateral hearing loss and diplopia 2 weeks later. A repeat CT scan showed re-emergence of the pineal region cyst. A cysto-peritoneal shunt was performed and there was a transient improvement in her symptoms that corresponded with the radiologic findings of a decompressed cyst. Her symptoms recurred 4 weeks later and eventually an infratentorial-supracerebellar approach craniotomy with total excision of the lesion was performed. Final pathology confirmed a pineal region ganglioglioma (WHO grade I glioma). Her symptoms improved after the operation and follow-up imaging did not show recurrence of the pineal region tumor.

DISCUSSION

Patients with pineal region lesions classically present with Parinaud syndrome, a tetrad of signs comprising upward conjugate palsy, Argyll-Robertson pupils, i.e., loss of the direct pupillary light reflex with preservation of the accommodation reflex, convergence-retraction nystagmus, and eyelid retraction, as a result of direct compression of the dorsal midbrain, otherwise known as the tectal plate or the corpora quadrigemina.

A clear understanding of auditory pathway neuroanatomy is crucial in explaining this patient's hearing impairment as a consequence of a large pineal region tumor. Fibers from the cochlear nerve are received by their ipsilateral pontine cochlear nuclei, which relay intricate ascending brainstem projections to the bilateral superior olivary nuclei, lateral lemnisci, and the midbrain's inferior colliculi. Thus both medial geniculate bodies of the forebrain individually receive binaural information that is relayed to the auditory cortex.¹ Due

to these extensive auditory interconnections that result in binaural representation, pineal region lesions must compress the pontine lateral lemnisci and the midbrain inferior colliculi (anatomical generators for waves IV and V, respectively) bilaterally in order to produce bilateral neurosensory hearing loss.^{2–4}

One reason for the indeterminate incidence of such symptoms is that they can easily be overlooked. In a case series of 12 pineal tumor cases, 5 patients had hearing loss with evidence of tectal plate compression upon autopsy.⁵ In another case series of 72 patients with pineal region tumors, 13 (18%) were noted to have measurable hearing impairment.⁴

Bilateral hearing loss can occur in patients with pineal region tumors and should not be disregarded. They may even be reversible after appropriate treatment as in our patient. The main mechanism was direct bilateral compression of the inferior colliculi and was confirmed with immediate hearing improvement after tumor excision.

AUTHOR CONTRIBUTIONS

G.K. Wong and X.L. Zhu conceived the paper; P.Y. Woo and J.Y. Teoh drafted the paper; all authors contributed and agreed to the final version of the paper.

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DISCLOSURE

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