Telephonic Conversational Difficulty as the Sole Symptom in a Patient with Cerebellopontine Angle Epidermoid: A Case Report

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Abstract The objective of this case report is to highlight the importance of audiological investigations in a patient whose sole complaint was difficulty hearing over telephone. The patient is a 45 year old man, approached for an amplification device for his reported difficulty of hearing over the telephone in the left ear of five years duration. A telephonic conversation test revealed functionally adequate hearing bilaterally. However, detailed audiological investigations suggested left retrocochlear lesion with possible involvement of the brainstem. This audiological formulation led to neuroimaging studies, which identified an epidermoid in the cerebellopontine angle. Thus, this case report emphasizes the importance of detailed audiological work-up in patients who are neurologically asymptomatic to unravel the neuroauditory lesions.

Keywords CPA Tumors, Dichotic Digits Test, Auditory Brainstem Response, Telephone Conversation

1. Introduction

The cerebellopontine angle (CPA) is one of the most common sites of intracranial tumors and approximately 10% of them originate in the CPA. The primary tumors of the CPA include vestibular schwannoma (VS), meningioma, epidermoid, arachnoid cyst, schwannoma of the Vth, VIIth, IXth, Xth and XIth cranial nerves, primary melanoma, hemangioma, lipoma, dermoid and teratoma[1]. Epidermoid tumors account for approximately 1% of all primary intracranial tumors and amongst them 40% appear in the CPA site[2]. Hearing impairment has been reported to be one of the common manifestations in them accounting for 37.6% followed by trigeminal neuralgia, 29.7%; dizziness or vertigo, 19.4%; facial palsy, 19.4%; headache, 17.9%; and diplopia, 16.7%[2]. However, difficulty listening over the telephone as the sole manifestation of hearing impairment is rare. The present case study reports this unusual feature in a patient who incidentally sought hearing aid to his left ear for his reported difficulty to engage in telephonic conversations for the last five years. The patient had a history of one episode of brief lasting pain over the left face a year ago. He was clinically diagnosed to have left trigeminal neuralgia and treated with carbamazepine successfully. He had not reported hearing impairment at the time of neurological consultation. Following the diagnosis of left trigeminal neuralgia, few months later, reported to have experienced a brief lasting dizziness that remitted spontaneously. He denied any other neurological or systemic symptoms at the time of audiology consultation. He was able to carry on otherwise, with his professional as well as personal duties efficiently.

2. Case Report

Mr. BK aged 45 years, male, sought for a suitable hearing aid to his left ear for his reported difficulty to engage in telephonic conversations for the last five years. The patient had a history of one episode of brief lasting pain over the left face a year ago. He was clinically diagnosed to have left trigeminal neuralgia and treated with carbamazepine successfully. He had not reported hearing impairment at the time of neurological consultation. Following the diagnosis of left trigeminal neuralgia, few months later, reported to have experienced a brief lasting dizziness that remitted spontaneously. He denied any other neurological or systemic symptoms at the time of audiology consultation. He was able to carry on otherwise, with his professional as well as personal duties efficiently.

3. Methods

The audiological approach was in the following order; 1. Telephonic conversation test. 2. Puretone audiometry and Speech audiometry tests (Speech Reception Threshold-SRT, Speech Discrimination Score-SDS & Performance Intensity for Phonetically Balanced test-PIPB). 3. Dichotic Digits Test (DDT). 4. Immittance audiometry. 5. Auditory brainstem response (ABR) and 6. Distortion Product Otoacoustic Emission (DPOAE) measurements. All audiological tests were carried out with standard procedures and test situations.

4. Results
A telephonic conversation test that was carried out demonstrated no difficulty in speech understanding in both ears. Appreciating his ability to hear in both ears with equal efficiency, the patient shunned the plan of hearing aid fitting in the alleged hearing impaired left ear. However, on inquiry, he reported that ‘speech signals’ in the left ear were not ‘as sharp’ as that of the right ear. This was probably the salient description of his reported hearing difficulty. Thus, he was further subjected to puretone and speech discrimination tests.

The pure tone audiometry showed hearing within 20 decibel (dB) in both ears at all octave frequencies from 250 Hz-8 kHz on air conduction testing. The SRT was 20 dB in both ears. The speech discrimination (SD) in quiet was 100% in both ears. No roll-over effect was observed for PIPB. Despite tests showing normal speech discrimination, the patient was subjected to DDT because of his reported feeling of ‘reduced sharpness in speech’. Prior to the administration of the DDT, monaural digits test (MDT) score was ascertained which yielded 100% score in both ears. The DDT demonstrated left ear extinction (0%) while the right ear score was 70% (Table 1). This finding implicated a central auditory disorder making further audiological investigations inevitable.

Impittance audiometry revealed normal “A” type tympanograms in both ears that suggested normal middle ear function. The acoustic stapedius reflex was absent left ipsilaterally and right contralaterally. It was present at normal sensation level right ipsilaterally but elevated left contralaterally (Table 1).

<table>
<thead>
<tr>
<th>Right Ear</th>
<th>Audiological Tests</th>
<th>Left Ear</th>
<th></th>
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<tbody>
<tr>
<td>20 dB</td>
<td>Speech Reception Threshold</td>
<td>20 dB</td>
<td></td>
</tr>
<tr>
<td>100%</td>
<td>Speech Discrimination Score</td>
<td>100%</td>
<td></td>
</tr>
<tr>
<td>No roll over</td>
<td>Performance Intensity for Phonetically Balanced Test</td>
<td>No roll over</td>
<td></td>
</tr>
<tr>
<td>100%</td>
<td>Monaural digit test</td>
<td>100%</td>
<td></td>
</tr>
<tr>
<td>70%</td>
<td>Dichotic digit test</td>
<td>0%</td>
<td></td>
</tr>
<tr>
<td>A Type</td>
<td>Tympanogram A Type</td>
<td></td>
<td></td>
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<tr>
<td>110 dB, 110 dB, 105 dB</td>
<td>Ipsilateral acoustic reflex at 500 Hz, 1 kHz and 2 kHz respectively</td>
<td>No response (NR) at all frequencies</td>
<td></td>
</tr>
<tr>
<td>NR at all frequencies</td>
<td>Contralateral acoustic reflex at 500 Hz, 1 kHz and 2 kHz respectively</td>
<td>120 dB, 120 dB, NR</td>
<td></td>
</tr>
</tbody>
</table>

The ABR for rarefaction clicks at 90 dB normal hearing level (nHL) & 100 dB nHL showed no identifiable peaks in both ears. Repeated recordings for both rarefaction and condensation click demonstrated poor replicability and morphology. For this reason though there is Vth peak like appearance (Figs 1&2), the same was not considered when other traces were examined. The DPOAE testing revealed normal cochlear function in both ears. The otoacoustic emissions (OAEs) were robust at all distortion product (DP) frequencies (Figs 3 & 4).

Note: A Vth peak like presence is observed around 5.5 ms for one rarefaction and condensation click recording. However, if other traces are examined, there is no replication of the pattern nor the morphology. Hence the pattern was considered as absent.

Figure 1. Left ear ABR for rarefaction clicks at 90 dB nHL. LSLR refers to left stimulation with left recording.
Figure 2. Right ear ABR for rarefaction clicks at 90 dB nHL. RSRR refers to right stimulation with right recording.

Figure 3. Left Ear DPOAE.
The audiological findings in this patient raised the suspicion of a retrocochlear lesion. Thus, the patient was further subjected to neuroimaging. Magnetic resonance imaging (MRI) revealed a well defined extra axial lesion in the left CPA extending into the internal auditory meatus on the left and crossing across the midline to the right CPA compressing the anterior and right lateral aspect of the mid pons. Superiorly the lesion was reaching up the tentorial hiatus and inferiorly the lesion was seen up to the lower pons. The lesion had signal intensity similar to cerebrospinal fluid (CSF) on T2 and was heterogeneous and hyper intense to CSF on T1 weighted images. The basilar artery was encased and displaced posteriorly (Figs 5 & 6).

5. Discussion

Congenital intradural epidermoid tumors are the third most common tumors of the CPA site[3]. Epidermoids are slowly progressive tumors[4]. Therefore, symptoms may surface late in the course of the illness. However, patients often present relatively early when the predominant symptom is trigeminal neuralgia[5]. The symptoms and signs are caused by displacement of the adjacent neurovascular structures. Approximately 64% of epidermoid tumors have been reported to have secondary trigeminal neuralgia[6]. Our patient had a history of one episode of brief lasting pain and was clinically diagnosed to have left trigeminal neuralgia. He was treated with carbamazepine successfully.

Greater proportion of normal hearing in nonacoustic tumors of CPA site has been reported by Hirsch & Anderson [7]. In their study 50% of patients in the nonacoustic group had demonstrated normal hearing as against 23% in the VS group and the difference between these two groups was statistically significant. They also have reported greater prevalence of poor speech discrimination in acoustic tumor ears on comparison with nonacoustic tumor ears (47% versus 35%). Our patient had also demonstrated normal hearing bilaterally with normal speech discrimination score. Higher proportion of normal hearing in the nonacoustic type tumors may be due to relatively less involvement of auditory nerve. However, Dawes and Jeannon[8] reported no difference in the pattern of hearing loss with respect to tumor type. Their patient population consisted of VS and facial neuroma and the findings noted were attributed to the close proximity of
both VS and facial neuroma with the auditory nerve. Lakhdar et al[9] have reported that the audiometric explorations provided only orienting contribution in their surgical series of 10 patients.

The left ear extinction on DDT implicated suppression of left sided caudal brainstem auditory pathways in this patient in the presence of good performance on MDT as well as 100% speech discrimination bilaterally. This was rather a defining point for a suspicion of retrocochlear pathology. The clinical utility of the DDT in identifying brainstem lesion has been well documented[10, 11].

In our patient the pattern of acoustic stapedius reflex abnormality strongly suggested left ear retrocochlear pathology with possible involvement of the brainstem. There was no clinical evidence of facial nerve disorder in the patient. Hirsch and Anderson[5] have reported acoustic reflexometry findings in CPA tumor patients classified based on tumor type. They observed comparatively higher efficacy for stapedius reflex tests in identifying nonacoustic CPA tumors. Efficacy of contralateral stapedius reflex estimation in CPA tumors identification has also been reported by Mangham et al[12] and Ferguson et al[13].

The ABR which reveals auditory nerve synchrony, showed absent response in both ears in our patient and suggested dysfunction at the brainstem level. It is interesting to note the discrepancy between the results of ABR and speech discrimination score in our patient. This may be due to the epidermoid tumor type itself, being creaky in nature exerts less effect on stimuli rich in external redundancy like the test used in the current study such as, speech discrimination, PIPB or MDT. The tumor presence was unraveled only when we employed less redundant stimuli such as dichotic test or short duration stimuli like click used in the ABR. Bilateral absence of ABRs in our patient may be due to extensive tumor invasion. The presence of DPOA E in both ears also suggested intact vascular supply to the cochlea in the presence of the tumor. This finding goes in correspondence with Mobley et al[14] study which has shown fewer propensities of nonacoustic tumors of the CPA to cause sensory hearing loss via blood supply disruption to outer hair cells.

Our patient’s audiological findings suggested two possibilities; left ear retrocochlear pathology with brainstem involvement or bilateral retrocochlear lesions. The former was considered most likely diagnosis as the performance on DDT in the right ear is 20% below the normal score that lessened the possibility of bilateral lesion. These findings were consistent with his reported difficulty of telephonic conversation in the left ear. The audiological diagnosis led to an MRI scan, which identified a space occupying lesion in the CPA site. Generally epidermoid tumors are located anterolateral or posterolateral to the brainstem and tend to expand where physical resistance is low and may extend around the brainstem into the contralateral cistern[15]. In congruence with the nature of the epidermoid tumor, our patient also had extensive invasion extending into the right CPA region with no demonstrable hearing loss. Audiollogically the only complaint was of conversational difficulty over the telephone. No surgical intervention was planned as the patient continued to be asymptomatic neurologically and is so till date.

A similar presenting symptom of difficulty in conversing over the telephone in an eight year old child and ultimately the audiological evaluation led to the diagnosis of a space occupying lesion which was later confirmed by the imaging study revealing a cerebellar pilocytic astrocytoma was reported by Berg et al[16]. Their patient had mild sensorineural hearing loss in the right ear and slight low frequency sensorineural hearing loss in the left ear with poor speech recognition in the right ear. The acoustic stapedius reflex was absent right ipsilaterally and left contralaterally. The ABR was normal in the left ear and only wave I was present in the right ear. In contrast, our patient had audiometrically bilateral normal hearing with good speech recognition score, absence of acoustic stapedius reflex left ipsilaterally and right contralaterally as well as elevated left contralaterally and extinction of the left ear on dichotic test. The only common symptom between these two patients was their inability to follow conversation over telephone. The audiological diagnosis in the present case could have been missed if only puretone audiomistry was done as adopted in the traditional way of audiological testing.

6. Conclusions

It was the hearing difficulty over the telephone in the left ear that prompted this patient to seek the help of an audiologist for hearing aid fitting. The audiological detection of a space occupying lesion which was confirmed by the imaging studies emphasizes the importance of audiological investigations in identifying the nature of the disorder. The highlight of the present report is the importance of ascertaining auditory behavior and administration of auditory tests in identifying retrocochlear lesions. Thus, audiological investigations could well be a preliminary diagnostic tool in identifying the silent lesions affecting the auditory system.

REFERENCES


