Auditory agnosia caused by a tectal germinoma

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Abstract—The authors describe a patient with auditory agnosia caused by a tectal germinoma. Despite having normal audiometric tests, the patient failed to recognize words and musical characters. On head MRI, the inferior colliculi were infiltrated by tumor. Neuropsychological tests revealed severe impairment in recognition of environmental sounds and words, defective musical perception, and stop consonant-vowel discrimination. Inferior colliculus may play a role in the analysis of sound properties.

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Auditory agnosia is an impairment of sound recognition in the presence of adequate hearing and is usually associated with lesions in the auditory cortex. Lesions in the midbrain tectum may result in various hearing disorders, such as severe deafness, impaired auditory recognition, or pure word deafness.[1-3] However, the characteristics of these hearing disorders have not been fully examined. We performed detailed neuropsychological assessments of auditory functions to evaluate a patient with auditory agnosia caused by a tectal germinoma.

Case report. A 14-year-old boy was admitted because of progressive hearing impairment and occasional severe headache. The patient had normal development and school performance before the onset of hearing disturbances. Detailed history revealed no evidence of congenital or developmental hearing disorders. For the past 3 months, his mother had to face him and talk slowly for him to understand. During conversations, he usually interpreted others' speech by reading the speakers' lips. He had difficulty understanding when listening to telephone calls. On neurologic examination, the patient was alert and cooperative with profound hearing disturbance. There were bilateral papilledema, ptosis, and Parinaud syndrome (upward gaze palsy, retraction nystagmus, and impaired convergence). Motor and sensory examination was normal, and the plantar response was flexor bilaterally. There was dysmetria and dysdiadochokinesia, more severe on the left side. Head MRI (figure 1) revealed a tectal tumor infiltrating the inferior colliculi with obstructive hydrocephalus and extension into right posterior thalamus. Another mass was noted on the right frontal ventricular wall with heterogenous enhancement by gadolinium. Excision of the frontal tumor revealed germinoma with lymphocytic infiltration and reactive astrocytosis. Endoscopic third ventriculostomy relieved the hydrocephalus. Immunostaining for neuron-specific enolase and glial fibrillary acid protein was negative. Serum levels of α-fetoprotein and β-human chorionic gonadotropin were within normal limits. He was treated with fractionated radiotherapy at 5,000 cGy. The headache and papilledema subsided, and there was radiologic evidence of complete remission. Follow-up MRI up to 3 years after treatment did not disclose recurrence.

Audiologic and neuropsychological tests. On examination, the auditory comprehension was severely impaired and improved a little only when the examiner faced the patient and spoke slowly. Writing, naming, and reading comprehension were normal. He performed poorly on dictation, with a 20% correction rate. There were a few phonemic errors in his spontaneous speech; otherwise the speech was fluent and intelligible. On testing nonverbal environmental sounds (including a ringing bell, a telephone, and a clicking watch) recorded on a tape, he could only recognize 7 of the 16 test sounds. He had difficulty differentiating between voices of different genders (50% correct). On testing of musical ability, he could not tell whether it was piano, flute, or violin. In two-tone discrimination test, he could not differentiate between two tones whether the intervals were half, two, four, or eight steps, either in ascending or descending sequences. He had difficulty recognizing different melodies, and he could not sing as he used to.

Linguistic testing was performed in a quiet room, with the examiner speaking behind the patient so that the patient could not read the examiner's lips. He had a 90% correct rate in identifying the common vowels in Mandarin (a, e, i, o, u). The stop consonant-vowel discrimination was performed in two steps, and each test set was repeated 100 times. Four different test sets were given in both stages. In the first stage, the patient was asked to discriminate between two stop consonant-vowel sets that differed in the articulation place of the stop consonants (e.g., /pa/ vs /ta/ or /ga/ vs /da/). There was only one discriminating feature between these two test sets. He achieved a correct rate of 53% on average. In the second stage, test sets with two discriminating features were given, different in the articulation place of the stop consonants and the nature of whether they were voiced or voiceless. For example, he was asked to discriminate between /ga/ and /ta/ or /da/ and /ka/. He achieved an average correct rate of 85%.

The result of pure tone audiometry on admission was normal, with detection threshold of 10 dB over a frequency range from 125 to 8,000 Hz binaurally (figure 2). The tympanogram and stapedial acoustic reflex were normal. Brainstem auditory evoked potentials (BAEPs) were performed, and all components of BAEPs were evoked with normal latencies. The amplitudes of wave V were attenuated bilaterally.

During the follow-up period, he showed a steady but limited improvement of hearing ability. The auditory and neuropsychological tests were performed 6 months and 2.5 years later, with similar results. The dictation improved with a correct rate of 60%. Follow-up audiometry showed a mild hearing loss with a detection threshold of 20 to 30 dB (see figure 2). Wave V of BAEP was absent bilaterally.

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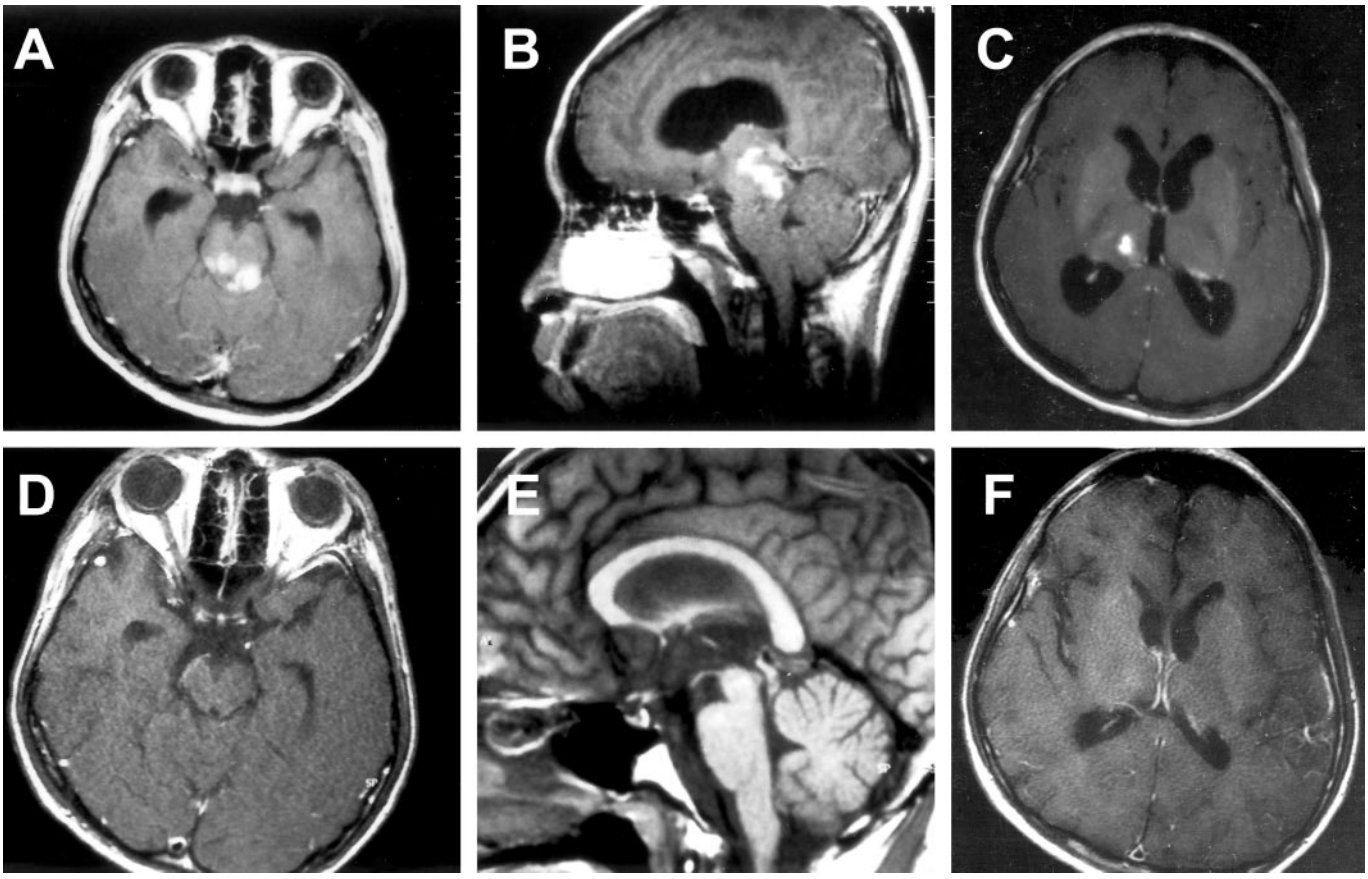

Figure 1. Contrast-enhanced, axial (A, C, D, F) and sagittal (B, E) T1-weighted MRI. (A, B) The inferior colliculi are infiltrated by the tumor. (C) The perisylvian cortices are normal bilaterally, with an enhanced lesion in right posterior thalamus and mild hydrocephalus caused by the midbrain tumor. (D, E, F) Follow-up MRI 1 year after treatment. The tumor is not recognized in the inferior colliculi, and the perisylvian cortices remain normal.

Discussion. This report demonstrated the association of auditory agnosia with a tectal germinoma. Auditory agnosia is usually associated with lesions in the auditory cortex. However, the responsible lesions in our patient resided in the inferior colliculus, as evidenced by the MRI and BAEP findings. Systemic reviews of midbrain tectal lesions and their relationship to central hearing disorders are few. In one study, abnormalities of BAEPs and middle-latency auditory evoked potentials were found in five and eight of nine patients with tectal tumors.[4] The clinical manifestations were not described. In a more recent review of 48 adult patients with brainstem gliomas, 10% had deafness of various degrees, but the anatomic locations were not specified.[5]

Although inferior colliculi are considered to be the generator of wave V in BAEPs, wave V can be preserved with bilateral inferior collicular lesions,[2,3,6,7] and additional lesions in the midbrain-pontine junction and upper pons may be necessary to abolish wave V.[6] Clinically, there seems to be two patterns of defective auditory recognition associated with brainstem lesions. The first type is generalized auditory agnosia with diminished wave V of BAEPs, and the pure-tone audiometry may be normal or abnormal.[1,8] This is seen in the current case and other reports associated with etiologies including post-traumatic midbrain hemorrhage and pineal meningioma.[1,8] The second type is pure word deafness with preserved BAEPs seen in traumatic midbrain contusion,[2,6] midbrain astrocytoma,[3,7] and arteriovenous malformation followed by embolization.[6] However, the neuropsychological profiles of these patients were rarely reported. Generalized auditory agnosia, impaired musical perception, and defective sound localization had been reported in one patient with post-traumatic hemorrhage in the inferior colliculi.[1] The vowel identification and phonemic discrimination were severely impaired, although the standard audiometry was normal. These findings were similar to those in the current case. Moreover, our patient's performance in stop consonant-vowel discrimination was improved when discriminating features in the test sets were increased. Collectively, these findings suggest that lesions in the inferior colliculi can cause impairment of auditory recognition, whereas the hearing thresholds are well preserved.

Auditory pathways in the brainstem consist of various hierarchical and parallel neuronal connections. Inferior colliculus is the first major site of neu-

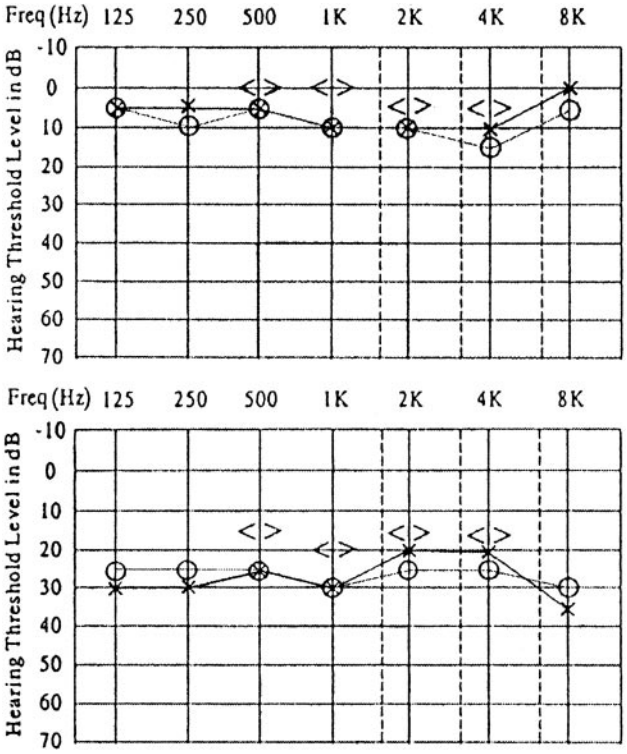

Figure 2. (Top) Audiogram before treatment, showing normal pure-tone detection thresholds. (Bottom) Follow-up audiogram 2.5 years after treatment with mild elevation of pure-tone detection thresholds. Freq = frequency; cross = left ear; open circle = right ear; bracket = bony conduction.

ral integration, receiving most of the ascending auditory inputs.[9] Animal studies suggest that inferior colliculi play an important role in the analysis of the spatial and temporal properties of sounds.[10] Consistent with these observations, the current report provides neuropsychological evidence that high-order auditory processing also occurs in human inferior colliculi. Whereas widespread destruction of brainstem auditory pathways can lead to severe deafness, restricted damage to inferior colliculi may result in impaired sound recognition with a normal detection threshold, the audiologic and neuropsychological findings of which may be indistinguishable from that caused by lesions of the auditory cortex.

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