Bilateral Temporal Lobe Lesions: A Case Study

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Temporal lobes are functional centers for hearing, speech, memory, olfaction, sensation, emotion, and behavior. Each temporal lobe is separated from the frontal and anterior parietal lobes by the Sylvian fissure (*Insights Imaging.* 2016;7:265). Depending on the location and severity, temporal lobe lesions may affect a wide range of behaviors, from impaired abilities in sound awareness, speech understanding, and music appreciation to visual and auditory agnosia, amnesia, epileptic seizures, and altered behaviors. Examining this complex case lends to a clinician's better understanding of bilateral temporal lobe lesions.

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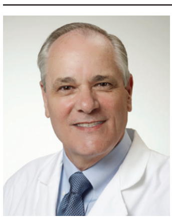
CASE DETAILS

In December 2009, a 29-year-old man was seen by his primary care physician for flu-like symptoms and given medication for bronchitis. Within nine days, the symptoms worsened and the patient was hospitalized with a tentative diagnosis of anemia or leukemia. He developed multiple system complications, respiratory and renal failure, and pulmonary infection. His bone marrow biopsy showed pre-B acute lymphoblastic leukemia. An endotracheal tube was placed to aid his breathing difficulties, but it coiled and got dislodged, causing hypoxia. Later, the patient had pneumothorax in each lung and got a tracheostomy tube. He was then put in a medically-induced coma and given intravenous chemotherapy. In February 2010, his MRI revealed bilateral temporal lobe lesions. By April 2010, the patient was released from the hospital and was asked to return on an outpatient basis. In January 2011, he received a bone marrow transplant and underwent surgery to install an intrathecal reservoir for chemotherapy. This was followed by periods of remission and reoccurrence with various complications, including seizure, stroke, unilateral vision loss, and profound central deafness.

Prior to his illness, the patient had no personal or familial history of hearing loss, tinnitus, auditory processing deficits, or cognitive impairment. He denied having any medical conditions or incidents of head, neck, or barotrauma. He had a degree in business, ran his own company, and was an accomplished pianist and guitarist.

In 2012, the patient was evaluated to determine a course for rehabilitation. Due to his auditory agnosia, receptive communication was accomplished using speech recognition software and a monitor or through writing to which the patient responded verbally. Notably throughout this process, the patient had chronic fatigue from chemotherapy. He took long breaks from voice therapy and training for sign language and lipreading, and eventually abandoned all of these except for his daily sound and music therapies.



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NEUROLOGICAL FINDINGS

The patient's radiological evaluation revealed bilateral "temporal lobe cortical infarcts with cystic encephalomalacia" (infarction with softening or loss of brain tissue) and "cortical hyperintensity" (*Br J Radiol.* 2012; Sep; 85(1017):782). His neurological examination revealed well-managed cognition, good long-term memory, and normal spatial memory. His balance, vestibulo-ocular reflex, and gait were within normal limits, and his facial strength and sensation were intact. The screening sensory examination was within normal limits. The patient's diagnosis was multiple systemic complications of acute lymphoblastic leukemia, including ischemic, cortical laminar necrosis, and central deafness with profound agnosia. The neurologist suggested using therapies designed to stimulate other areas of the brain that might compensate for some of the lost functionality. A neuropsychological evaluation was recommended to aid in the rehabilitation process.

SPEECH AND LANGUAGE PATHOLOGY

According to the patient's speech pathologist, his reading comprehension, expressive language, and written communication skills were within normal limits. However, he seemed to lack understanding of basic body language (e.g., head nod for yes), which suggested visual agnosia. He also had difficulty interpreting facial expressions, which made lipreading a challenge.

Table 1. Household Sound Perception Ratings for Loudness, Tone, Quality, and Annoyance

	A	B	C	D	E
1	**Sound**	**Loudness**	**Tone**	**Quality**	**Annoyance**
2	Toilet Flushing	Soft	Bass	Hollow	1
3	Finger Snapping	Soft	CND	CND	1
4	Clicker	Soft	CND	Tiny	1
5	Car Starting	Medium	Bass	Normal	1
6	Drill	Medium	Mid	Dull	2
7	Garbage Disposal	Medium	Bass	Tiny	3
8	Bass Drum	Loud	Bass	Holow	4
9	Shower Running	Medium	Mid	Dull	5
10	Door Knock	Loud	Mid	Reverberant	5
11	Vacuum Cleaner	Loud	Mid	Dull	5
12	Hammer	Medium	Confusing	Hollow	5
13	Hair Dryer	Loud	Mid	Hollow	5
14	Door Bell	Uncomfortable	Treble	Tiny	8
15	Hand Clapping (one person)	Loud	Mid	Reverberant	8
16	Telephone Ringing	Loud	Confusing	Harsh	8
17	Door closing	Uncomfortable	Treble	Reverberant	10
18	Blender	Loud	Confusing	Harsh	10
19	Brother Playing Drums	Loud	Confusing	Harsh	10

The patient developed difficulty controlling the loudness and pitch of his voice and presented with a staccato speech pattern. However, his dysphonia improved with voice training, which included direct and indirect approaches and vibrotactile stimulation.

SOUND PERCEPTION MATRIX

In view of the rarity of this case and the lack of usable tools to identify what the patient was perceiving, a table was created for the patient to rate common household sounds (Table 1). The patient was instructed to rate sounds for loudness, tone, quality, and annoyance using labels designed from the patient's informal comments about his sound experiences. Loudness ranged from soft, medium, or loud to uncomfortable; tone was musical in nature, using bass, mid, and treble, confusing, or CND; quality judgments were normal, harsh, reverberant, hollow, tinny, dull, or CND; and annoyance was rated on a 1/10 scale. Although it was difficult to form correlations from the responses, the patient's reported perceptions of tone and quality demonstrated how chaotic sound can be for a person with temporal lobe lesions.

AUDIOLOGICAL EVALUATION

All audiological test results were obtained in an ANSI S3.1 calibrated Tracoustics single-walled, sound-treated room using the Madsen Astera ANSI S3.6 calibrated audiometer with a circumaural headset. Otoscopic examination revealed normal-appearing ear canals free of debris and tympanic membranes with robust light reflexes and appropriate translucency, position, and color.

Pure tone air and bone conduction thresholds revealed a moderate and flat sensorineural hearing loss. Air conduction thresholds were obtained both manually and via Békésy sweep tone audiometry. The results were highly correlated, as seen in Figure 1.

Admittance testing revealed tympanograms consistent with normal middle ear pressure and mobility. Acoustic reflex thresholds were present ipsilaterally and contralaterally from either ear at 500, 1,000, 2,000, and 4,000 Hz. Distortion product otoacoustic emissions (DPOAE) were obtained for 18 data points between 516 and 10,008 Hz in either ear using the BioLogic Navigator Pro, with Scout software set to f1 65/f2 55 dB. DP-Grams showed good outer hair cell function, which was inconsistent with the audiometric findings (see Fig. 2 online).

Electrophysiology testing was conducted using the BioLogic Navigator Pro system with A1, A2, ground, and reference electrode montage. Auditory brainstem evoked potentials to clicks at 80 dB revealed waveform morphology, amplitude ratios, absolute and inter-wave latencies, and slow-fast rate (13.3 and 57.7 rps) latency changes within normal limits bilaterally (see Fig. 3 online). Latency-intensity curves were also measured down to 20 dB bilaterally. Forty-hertz responses were obtained within normal limits using click stimuli at 80 dB bilaterally. Auditory mid-latency responses to clicks were absent at 70 dB from either ear (see Fig. 4 online). It should be

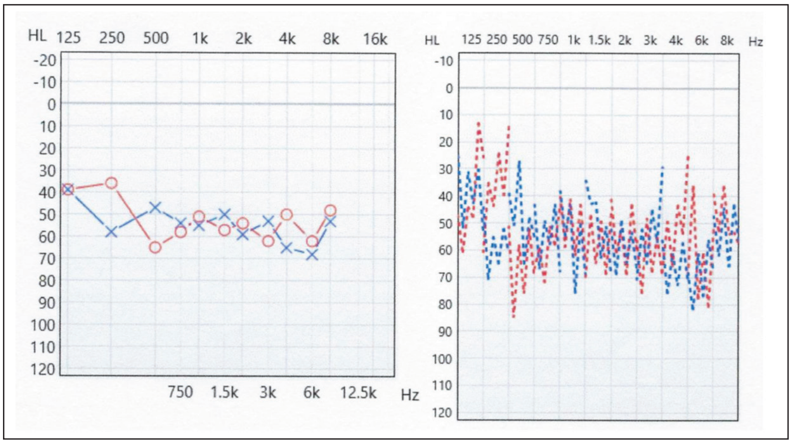

Figure 1. Pure tone air conduction thresholds obtained by manual and automatic pulse tone audiometry.

noted that testing from Northwestern University identified cABR responses within normal limits, but late latency evoked potentials were absent bilaterally.

AUDITORY PROCESSING EVALUATION

Due to the lack of receptive aural communication, verbal and non-verbal tests were employed to establish a baseline of information. The patient had abnormal gap detection for 500 Hz (90 msec), 1,000 Hz (90 msec), 2,000 Hz (90 msec), and 4,000 Hz (200 msec). He couldn't tell the difference between noise or speech stimuli from the MAC Battery Noise/Voice Test. The patient couldn't recognize any of the sounds from the Sound Effects Recognition Test (SERT). Using the Pitch Pattern Sequence pitch discrimination test with a modified response of same or different, the patient correctly identified the differences 84.2 percent of the time. On lateralization testing, the patient could accurately identify a 250-Hz pulsed tone at 70 dB HL using headphones 70 percent of the time. However, this required cuing to the onset of the signal or the patient would inconsistently identify the presence of the tone. After considerable practice, the patient was able to distinguish speech from non-speech sounds, tap to metronome beats, and perceive notes within a perfect fifth. These were important findings that could be used as the anchor for sound and music therapy.

SOUND AND MUSIC THERAPY

In an attempt to set a baseline for awareness, loudness, and tone, the therapy goals were to learn the presence of a sound (on v. off), establish an anchor for loudness perception at 60 dBA, and learn to identify low-tone pitch differences within a perfect 5th (C-G).

Due to the patient's health concerns, the therapy had to be passive at times, with some periods of active listening. To keep the therapy simple and accommodating, S-Tones (Serenade by SoundCure) centered at 1 K Hz at 60 dBA were presented using sound field speakers in the patient's living area. The patient was trained to use a sound level meter to regulate the loudness. He was prompted to randomly identify the therapy signal multiple times a day by periodically turning it on and off but to avoid quiet time. This continued for nearly two years. He learned to identify S-Tones on command within weeks of practice and became accustomed to the loudness and pitch within six months. Subsequently, the patient reported listening more to music or sound from the TV also at 60 dBA. To practice pitch and timing, the patient was encouraged to play simple songs on the piano centered around middle C (262 Hz).

DISCUSSION

The test results were consistent with the neurological findings. The auditory structure and function were within normal limits from the external, middle, and inner ears to the auditory nerve and brainstem. There were no responses from the auditory cortex. The patient's flat audiogram, impaired temporal processing and speech understanding, lipreading difficulty, and other higher auditory function issues were due to the

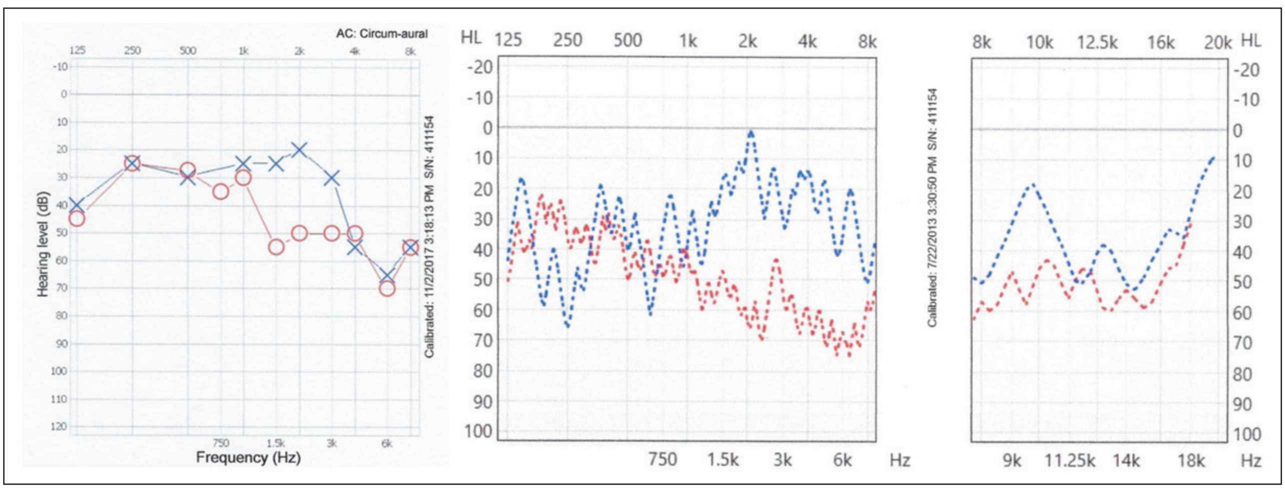

Figure 5. Pure tone air conduction thresholds obtained by manual and automatic pulse tone audiometry (2018).

bilateral temporal lobe lesions. Agnosia was the most notable symptom, and isolation was one of the most feared reactions. Therefore, the initial trials with speech reading, voice therapy, vibrotactile stimulation, and sound and music therapies were designed to immerse the patient in different activities.

There are no guidelines for therapy or training of patients with this level of temporal lobe damage. The therapies attempted were designed to avoid quiet time, establish a baseline for sound awareness, and develop some level of learning. However, after six months, the patient decided to focus on his auditory skills and abandoned lipreading and sign language.

Nearly 10 years after the onset of his illness, the patient continues to listen to the S-Tones, TV, and music. He undergoes chemotherapy treatments for leukemia, and had two recent surgeries for tumors. He reported that his hearing and sound awareness have improved, and he could recognize some sounds and types of music. His recent hearing test showed a dramatic improvement in hearing sensitivity in both ears (Fig. 5.), signaling changes within his auditory system. The patient is being re-evaluated to determine any improvement in his auditory skills, the results of which will be published in a future article. Remarkably, while undergoing treatments, the patient obtained an MBA online and became a university professor, currently teaching distance learning classes.