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Case Report

Oliver Sacks syndrome treated with adaptation to hearing aid

Ebru Sahan, Ismet Kirpinar

Bezmialem Vakif University, Department of Psychiatry, Istanbul/Turkey

Corresponding Author:

Ebru Sahan, M.D.

Department of Psychiatry,

Bezmialem Vakif University,

Adnan Menderes Boulevard (Vatan Street) P.K. 34093

Fatih, Istanbul

Turkey

Tel: +905054028722

Fax: +902124531869

E-mail: ebrushaan@hotmail.com

ABSTRACT (137 words)

An eighty-year old woman was diagnosed with bilateral severe sensorineural hearing loss in audiometric examination and recommended to use hearing aids. The patient could not adapt to the device and was reluctant to wear it. In the psychiatric examination it was difficult to communicate with the patient due to hearing loss. No delusions were found in the thought content. When hallucinations were asked about, she noted resonance and humming in the early periods that later turned to helicopter, airplane sound and as hearing loss progressed, she heard wedding music. The patient had full insight about the sound was coming from her head. The psychiatric treatment was useful for adjustment to hearing aid and maintaining acoustic stimulation. As hearing devices were used, the ambient sounds increased, and the musical hallucinations were slowed down and communication with her relatives increased.

KEY WORDS: audiometry; hearing aid; hearing loss; musical hallucination

INTRODUCTION

Auditory hallucinations can be seen either in elementary forms such as resonance and buzzing or in complex forms such as human voices or music. Musical hallucinations have been defined as Oliver Sacks syndrome, they are a form of cortical release phenomenon in hearing loss and in this context they are also described as auditory Charles Bonnet syndrome ^[1].

CASE REPORT

An eighty-year old woman was brought to the psychiatry outpatient unit of the health board by her children for investigation about disability. On the wheelchair, she seemed a little bit surly. She was overweight and had been diagnosed with diabetes mellitus, essential hypertension and gonarthrosis during the last 25 years. She was well controlled with antidiabetic and antihypertensive drugs. For the past three years she was not hearing properly what others were saying in conversations and she had the Television (TV) on a high volume all the time. She was brought to an otorhinolaryngologist three months ago and was diagnosed with bilateral severe sensorineural hearing loss according to the audiometry tests and it was suggested to use hearing aids. The patient could not adapt to the device and did not want to wear it. Her children noticed that she had recently become more uncomfortable and stubborn, talked less with her relatives, occasionally cried, and had difficulty sleeping at night. There was no previous history of psychiatric disorder. She lost her husband 5 years ago. She lived with her children, their spouses and her grandchildren. Her appearance seemed compatible with her chronological age. It was difficult to communicate with the patient during the mental status examination due to hearing loss. When she could hear and understand the questions asked, she was responding a little bit reluctantly. She was oriented to place, person and time. No delusions were found in her thought content. When hallucinations were questioned, she confirmed that there were noises in her ears. When she was asked about the voices, it was learned that in the early periods of the hearing loss the voices were in the form of resonating and humming. Later, they converted to a helicopter, airplane noise and progressively to halay music which is composed of drum and clarion sounds played at the weddings in

the region she lived in. She said that the sounds were heard from both ears, increasing at quieter times such as night time.

A detailed audiometric examination including Bekesy and Sisi scores was done. Bekesy audiometry is based on a comparison of thresholds for constant and intermittent tones. Clinical importance has been attached to measurement of the threshold excursions. Short Increment Sensitivity Index (SISI) test measures the ability of a subject to detect a 1 dB change in intensity at a level 20 dB above threshold. This test can be used at any frequency in either ear, regardless of the asymmetry of the hearing loss [2]. There was severe sensorineural hearing loss in both ears plus air conduction deficits were noted on the left (right ear speech reception threshold [SRT] 65 dB, discrimination 40% dB 90; left ear SRT 80 dB, discrimination 40% dB 100). Tympanograms were type A and acoustic reflexes were absent bilaterally. The music was less with hearing aids, but the device was very uncomfortable. The music noise could be suppressed by television or radio noise. Different melodies and folk songs were able to change her musical hallucination temporarily. Halay sound was not as uncomfortable as the device. The patient had good insight that it was the sound coming from inside her head.

The cranial magnetic resonance imaging of the patient revealed mild cortical atrophy and widening of the sulci. Routine hemogram and biochemistry values were in the normal range. Hepatitis, AIDS, syphilis, and brucellosis were excluded by serological tests. The EEG was normal. In neuropsychological tests, simple attention can be considered immediate memory or very short term memory. Complex attention defines functions based on sustaining attention; in the event of deterioration, there is deterioration in persistence (persistence), perseverations, distractibility, reduced resistance to interference, and difficulty in suppressing the immediate, but unfavorable, tendency to react. These functions, which are based on complex consideration, are called executive or executive functions [3]. Short-term memory, simple and complex attention were preserved in our patient. There was minimal age-compatible cognitive impairment. She had a depressed mood most of the day, nearly every day, diminished interest and pleasure in almost all activities, psychomotor retardation, fatigue, and insomnia yet she did not have suicidal thoughts or tendency. Thus, she was diagnosed with major depressive disorder according to DSM-V. In order to exclude malingering, confirm diagnosis of depression and to learn antidepressant treatment response, the decision of the board was postponed for 6 months with once a month outpatient follow-up visits.

For the treatment of depression escitalopram 5 mg/day and insomnia trazodone 50 mg/day were prescribed. She was supported by her family to wear her hearing aids starting with one hour a day and to increase gradually. At the third month outpatient visit, the trazodone was discontinued because the sleeping problem had disappeared. As hearing devices were used, the ambient sounds increased thereby the sensory input suppressed myriads of nonessential information, including previously acquired memories. The musical hallucinations regressed and communication with her relatives increased. At the sixth month follow-up visit, escitalopram was discontinued.

DISCUSSION

The diagnosis of dementia was ruled out because the patient's cognitive changes were compatible with her age. Psychotic depression was ruled out as a differential diagnosis due to the fact that the musical

hallucinations reflecting past experiences were not congruent with depressive mood (Halay voice did not include a sad, pessimistic, critical or accusative theme) though she had major depressive disorder with melancholic features. It was thought that auditory hallucinations were due to hearing loss because they started at similar times with the hearing loss and they increased as the hearing loss progressed.

Sensory inputs suppress much of the information that is not necessary, such as previous memories. The 'Perceptual Release Theory' of West ^[4] influenced by Hughlings-Jackson's concept of disinhibition suggests that when a certain amount of external sensory stimulation does not go to the area of the cerebral cortex responsible for hearing (in the temporal region of the non-dominant hemisphere) due to deafness or isolation, the acoustic stimuli of recorded memories are transmitted and perceived again with familiar or new and strange forms ^[5].

Peripheral sensory organ damage by anatomical and physiologic pathways may cause central perceptions which can also be seen in Charles Bonnet and Phantom Limb syndromes. Charles Bonnet Syndrome, in which visual hallucinations occur due to loss of sight, is more similar by its origin to auditory hallucinations of patients with hearing loss. In addition, it has been reported that musical hallucinations are lateralized to the ear that can not hear in a person with unilateral deafness ^[6].

The two-factor theorists who advocate that both peripheral and central dysfunction should be present, as evidence, suggest that such hallucinations occur more frequently in older adults with a high incidence of brain pathology ^[7].

The main risk factors for musical hallucinations are impaired hearing, tinnitus, advanced age and, perhaps, also female sex; however, the latter finding may be due to an overrepresentation of females in the literature^[8].

There is no consensus for the treatment of disturbing musical hallucinations and decision is made usually according to underlying etiology. In a review, among pharmacological treatment methods investigated, antidepressants found to be possibly more helpful than antiepileptics (which are still better than antipsychotics) ^[9]. The limited use of acetylcholinesterase inhibitors was promising ^[10]. Musical hallucinations experienced in the context of brain injuries and epilepsy tend to respond well to antiepileptics. Musical hallucinations occurring as part of a psychiatric disorder tend to respond well to psychopharmacological treatments targeting the underlying disorder.

As in our case, the hypoacusis etiological group appears to respond to hearing impairment treatment. More acoustic stimulation suppresses the hallucination (e.g. our patient increased the volume of the television or music) ^[9].

We used antidepressant treatment in this patient for depressive disorder and easing adjustment to hearing aid use thereafter. In contrast to the report by Serrador-García et al. our patient had a complete response; musical hallucination did not occur when taking hearing aid out at night ^[11].

CONCLUSION

Not all adults provided with hearing aids use them, wear them regularly, or are satisfied with them ^[12]. Adjustment to hearing loss and hearing aids become much more difficult with increasing age and psychiatrists can help these people by means of dealing with this adjustment disorder.

ACKNOWLEDGMENT

None to declare.

This work represents original material, it has never been published before, it is not under consideration for publication elsewhere. It is not supported by any drug or other companies. No grants or fees were taken from any institution or firm. We have no conflicts of interest to disclose. We confirm that all identifying information has been removed, and the written consent has been obtained from the patient for submission of the report. The abstract of our manuscript was a poster presentation at 3. Psikiyatri Zirvesi, 10. Ulusal Anksiyete Kongresi, Antalya, Turkey between 15-18 March 2018.

We have no potential conflict of interest to disclose.

Contribution of each author in the manuscript:

1. Substantial contributions to the conception or design of the work; or the acquisition, analysis, or interpretation of data for the work;
2. Drafting the work or revising it critically for important intellectual content;
3. Final approval of the version to be published;
4. Agreement to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved.

1. Author's contribution: 1,2,3,4

2. Author's contribution: 1,2,3,4

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