Musical Ear Syndrome in a Patient with Unilateral Hearing Loss: A Case Report

Faten Mane Aldhafeeri

Corresponding Author: Faten Mane Aldhafeeri, e-mail: fmaldhafeeri@uhb.edu.sa

Financial support: None declared

Conflict of interest: None declared

Patient: Male, 62-year-old

Final Diagnosis: Hearing impairment

Symptoms: Hallucinations • hearing impairment

Medication: —

Clinical Procedure: —

Specialty: Audiology

Objective: Unknown etiology

Background: Hearing music that has no source is known as musical ear syndrome, also known as musical hallucinations (MH), and is often associated with hearing impairment. This report is of a 62-year-old man with a 20-year history of unilateral hearing loss and continuous MH. We investigated the neural basis of MH in a subject without any known neurological or psychiatric disorders.

Case Report: A 62-year-old man had a history of 20 years of symptomatic mild hearing loss accompanied with continuous MH in the form of multiple tones. The MH were unilateral in the left side and much more likely to be experienced as externally located and uncontrollable. He underwent structural and functional magnetic resonance imaging (fMRI). Results indicated increased activity and reduced cortical thickness in multiple cortical areas, such as the prefrontal cortex (PFC) and temporal and limbic regions, indicating complex processing and involvement.

Conclusions: The neurological findings indicate differentiated or multiple-area involvement in MH. These morpho-functional changes may represent a neural reorganization causing MH to arise. The altered or activated regions are all related to the processing of emotions and the processing of episodic memory, which has been seen in the MH of schizophrenic patients. This report also highlights that patients with hearing loss may present with continuous MH, and that these patients should not be assumed to have delusions or psychosis, but should be investigated for underlying auditory abnormalities and treated appropriately.

Keywords: Auditory Perception • Hallucinations • Magnetic Resonance Imaging • Hearing

Full-text PDF: https://www.amjcaserep.com/abstract/index/idArt/936537
Musical ear syndrome, also known as musical hallucinations (MH), is a relatively rare phenomenon in which a person perceives auditory stimulus, such as musical sounds, harmonics, timbres, melodies, and rhythms, in the absence of any external stimuli [1,2]. In some cases, musical ear syndrome is not associated with any pathological condition, in which case it is called idiopathic. However, in the vast majority of cases, musical ear syndrome is associated with hearing impairments and other neurological and psychiatric disorders, and so can be said to have an underlying cause [2,3]. Although the condition is sometimes termed musical hallucinations, it is not a symptom of psychosis and is not associated with schizophrenia [4,5]. There is an existing body of literature indicating a connection between MH and several mental health factors such as psychiatric disorders [6], aging [7], and hearing impairment [8]. Some people with MH experience this phenomenon as a mild and tolerable condition, while others experience it as a distressing condition that affects their quality of life [2]. The pathophysiological mechanism underlying MH is still poorly understood because it is a rare phenomenon and there are thus no large studies on MH [6]. The published studies involved only case studies or a few patients with MH. A multi-model neuroimaging study has revealed increased activation in auditory regions, anterior or cingulate, and frontal regions in addition to microstructural alteration of the left uncinate fascicle [9]. A cortical activation network including inferior frontal cortices and posterior temporal regions in deaf subjects with MH has been reported previously [9]. Similarly, Placido et al [10] reported increased activation in the right temporal cortex in MH patients who had a stroke. The aim of this study was to investigate the morpho-functional mechanism of musical hallucinations.

To fully understand this phenomenon, it is important to describe musical hallucinations, and to define the difference between a MH and a typical hallucination. A hallucination is a false perception or the perception of an image that is not real, when the subject is awake [2,11]. These perceptions are not the consequence of any specific stimulus in the physical environment and can be perceived by any of the senses [11]. This can be contrasted with MH, which are always auditory in nature, and which can be defined as the perception of a musical experience, or more simply put, hearing music, in the absence of an environmental source [1]. MH are a relatively rare perceptive experience and can include a part of a song or a complete melody [9]. The content of the MH is often familiar, or a song or tune that the individual having the hallucination already knows, and can be vocal, instrumental, or a combination thereof [12]. This should be separated from the more common phenomenon of having involuntary musical imagery and is instead a full and intrusive experience of the song, and perception that it is independent of subjective control [13]. The phenomenon, in general, is more common in females [2,14], but has been documented in both genders [3].

Early research indicated, and more current research has supported the theory that, MH often come from psychiatric disorders and/or focal brain lesions [15-17]. However, other studies described a pathological response, like hearing loss [2,18]. This raises the question of how hearing loss leads to development of auditory hallucinations, or what brain activity or neural causation is related to the psychiatric and pathological causes. It is this connection that the current study seeks to describe. Razdan et al [18] reported the case of a patient with MH and associated hearing loss, with no neurological or psychiatric disorders, confirming the association between hearing loss and MH. Nevertheless, the nature of this association is still poorly understood.

Many of the cases in the existing literature have been case studies, involving patients experiencing MH, but most focused on the details of the experience [1,3]. There is very little published research on brain activity during MH. There have also been related studies on the differences between brains creating standard hallucinations and brains of those that do not have hallucinations [5,19]. These studies have also considered the differences between the brain during standard hallucinations and the brain on days when hallucination do not occur [5,20,21]. These have not focused on, or found to be significant, a single cortical area, indicating that a single area of the brain cannot be credited with creating all hallucinations or MH. Further, other parts of the brain have also been found to be involved, including the orbital frontal cortex, basal ganglia, and precuneus [22].

More recently, Cavaliere et al [9] studied the fronto-temporal circuits involved in MH to determine if there is a morpho-functional circuit consistently involved in the development or experience of MH. That study, like previous research [23], showed functional connectivity in multiple cortical regions, or that there is inconsistent involvement of subcortical sections.

There is currently no known treatment for the symptoms of MH. The most effective method for treating MH is to focus on the etiological process responsible for mediation of these experiences [24]. In most patients, effective treatment depended on resolving the underlying cause, such as suspending certain medications that might have such adverse effects. In addition, neurologic evaluation, mental evaluation, and ear, nose, and throat (ENT) evaluation are advised, as well as MRI head imaging, as part of the treatment plan [25]. MH may be self-limiting, particularly when it occurs after brain injury; therefore, not all patients require treatment. At the onset of MH, nonpharmacological treatments such as cognitive behavioral therapy are recommended [25].
In the current study, we aimed to investigate the neural basis of MH in a subject who had no known neurological or psychiatric disorders. We used functional and structural MRI to investigate the neural mechanism of MH. In the fMRI, we tried to stimulate MH by introducing music while the subject was in the MR scanner. To investigate the structural alterations that might be associated with MH, we performed high-resolution MRI on our MH patient. This report is of a 62-year-old-man with a 20-year history of unilateral hearing loss and continuous MH.

**Case Report**

A 62-year-old man presented for the first time in the clinic, reporting that he had been hearing musical tones since the last 6 months in the absence of any stimulus around him. The musical sounds were unilateral in the left side and much more likely to be experienced as externally located and uncontrollable. He described the hallucinations as familiar songs like nursery rhymes and occurring several times a day, particularly in a quite environment. The sounds were intrusive and disruptive to his daily life. The onset was abrupt and was not associated with any other neurological symptoms. The patient did not report any history of social isolation and he was not on any medications. Pure tone audiometry was conducted to assess hearing loss level at 4 different frequencies: 500, 1000, 2000, and 4000 Hz. The tones were presented at different sound intensities that ranged from -10 to 120 dBHL (Audiology 2004). **Figure 1** shows the pure tone audiometry of the patient. The patient had a history of 20 years of symptomatic mild hearing loss. To assess the patient’s cognitive and clinical profile, we used the Hamilton Rating Scale to assess depression, the Mini Mental State Examination was used to assess cognitive impairment, and the Neuropsychiatric Inventory was used to explore the presence of neuropsychiatric symptoms. The patient scored 8 on the Hamilton Rating Scale for Depression (normal range=10–24), and 10 on the Neuropsychiatric Inventory (normal range=1–30). MRI of the brain with and without contrast was normal. The present study was carried out after being approved by the Local Research Ethics Committee. Written informed consent was obtained from the patient.

The functional MRI experiment consisted of a block design of 10 epochs of 10 s, during which an auditory stimulus was binaurally delivered using MRI-compatible noise attenuation headphones. After each stimulus, a blank screen was presented for 10 s. The auditory stimuli were instrumental musical clips obtained from the International Affective Digitised Sound System (IADS) (Bradley and Lang 2007). The patient was familiar with the musical clips. Blood-oxygen-dependent echoplanar imaging was performed (TR=3000 ms, TE=30 ms, FOV=192 mm, and matrix size=64×64 mm).

For CTA, high-resolution T1 MR images were acquired using the following parameters: (TR=1600 mm, TE=3 ms, slice thickness=1 mm, FOV=256 mm, matrix size=256×256 mm).

**Image Pre-Processing**

fMRI data were processed as head motion correction, a slice-time correction, and spatial smoothing. The functional images were co-registered with the 3-D isovoxeled anatomical data, and a volume time course (VTC) file was created. The extent of activation was measured by the number of voxels, which was thresholded at ≥10 contiguous voxels at P_{FDR}<0.05. Figure 2 shows the activation map of the MH patient. Significant increased activation was detected in the right superior temporal gyrus (coordinates: 57, -21, 8), cingulate gyrus (coordinates: 3, -6, 41), inferior frontal gyrus (coordinates: -49, 20, 18), and left parahippocampal gyrus (coordinates: -36, -25, -15). Figure 2 shows the activation map of the MH patient.

Cortical thickness analysis showed a significant decrease in the left medial orbitofrontal cortex (MH: 2.35, CTRs: 2.51±0.05; P=0.004), right superior temporal gyrus (MH: 2.6, CTRs: 2.96±0.01, P=0.05), and the right inferior temporal gyrus (MH: 3, CTRs: 3.6±0.12, P=0.03).
Musical hallucinations remain a condition that has no known cause, although the literature demonstrated some associations between MH and age, psychiatric disorders, and sensorineural hearing loss [27]. No conclusive evidence points to a single root cause of musical hallucinations. Hearing impairment is a common finding, but it is not necessarily an underlying cause of musical hallucinations [28]. MH is considered a rare phenomenon, with diverse neuroanatomical factors, often directly related to an ontological, psychiatric, or neurological underlying condition or disease. During MH, music is heard in the absence of any external stimuli [15].

Here, we describe the case of an otherwise healthy 62-year-old man who had MH with unilateral hearing loss and in the absence of any neuropsychiatric disorder. The current case is comparable to a case reported by Razdan et al [18] in which a patient had MH and hearing loss without any associated neuropsychiatric disorder. Such findings indicate that hearing loss is a common occurrence in individuals with MH [19,23,27]. To assess our patient’s experience of MH, a combination of clinical and cognitive assessment was used, in addition to functional and structural MRI, to investigate the neural basis of MH. The patient had a normal cognitive profile, with some symptomatic hearing loss and a known history of MH.

Our patient had significantly increased activation in the right temporal lobe, cingulate and frontal cortices, and the left parahippocampal region. Several studies have reported similar findings in MH [9,20]. Kasia et al [20] reported increased cerebral blood flow in the right temporal and inferior PFC cortices. The increased activation in the temporal lobe where the auditory cortex is located suggests the continuous perception of MH as reported by the subject during the fMRI experiment. The increased fMRI activation is commonly believed to represent a functional reorganization in neural activity [29]. The physiological alterations characterized by an increase in the fMRI BOLD signal in the superior temporal gyrus may point toward the underlying structural lesion that appeared as a reduction in the cortical thickness. Another explanation for the involvement of the superior temporal gyrus is that increased activation in this region may reflect a premature neuropathogenesis of MH that later developed into structural neurodegeneration [30]. Decreased levels of gamma-aminobutyric acid-ergic (GABAergic) inhibitory neurotransmitters was found to cause increased cortical excitability [31]. This neural hyperexcitability may be responsible for the increased BOLD signal as an antecedent for the structural deficits. It is also possible that reduced GABAergic function has a role in MH pathogenesis as a causative factor. Reduction of GABAergic function can cause neuronal hyperactivity that is experienced as phantom perception (eg, MH) and also has other neurodegenerative effects [31].

Our findings suggest the involvement of the medial PFC in MH. Griffiths [21] reported increased regional cerebral blood flow in the medial PFC, which is in line with the current
findings. A magnetoencephalography (MEG) study conducted by Kumar et al [12] found an increase in theta and alpha oscillation in the orbitofrontal region in MH subjects which, in turn, confirms the involvement of the PFC in MH. The medial PFC has reciprocal connections with the amygdala and the limbic and the medial temporal cortices, and such connections are primarily involved in emotion processing [32]. Increased activation of the medial PFC region may suggest the negative emotions associated with MH perception. Evidence suggests the people with MH have neural plasticity in brain regions involved in sound perception, attention, emotions, and memory.

Considering possible explanations of activation in alternative portions of the brain may be possible when comparing the outcomes of the current study to other studies that show similar activation of the brain correlated with more specific medical conditions or sound-related stimuli. Osnes et al [33] found that increased activation in the superior temporal gyri is a function of memory as it relates to sound morphing or phonetic sound. The completion of an fMRI shows that while any sound, including both phonetic and non-phonetic auditory information, can increase activity in the superior temporal gyri, the interaction with the second auditory cortex and related processes is more significantly increased with sounds involving the building blocks of language [33]. Thus, as it relates to the current study, it may be of greater interest to consider whether MH involves instrumental or vocal musical stimuli, as it relates to the level of increased activity detected in the superior temporal gyri.

Another recent study [5] indicated that both simple and complex auditory phantoms demonstrate an increase in theta-gamma activity. Specifically, this was demonstrated in increased beta activity in the limbic areas, and differential alpha band activity in the auditory context, with differentiated beta activity in the cingulate cortex. Thus, it is possible that the increased activity in the cingulate gyrus is related to tinnitus, or related changes in the structure of the hearing mechanisms, and hearing loss, and is not directly related to the MH. The authors point out that auditory phantoms like MH may be associated with activation of areas of the brain that are related to music and language processing. This is consistent with both the findings of Osnes et al [33] and the current study, which find that multiple areas of the brain may be associated, as it relates to the type of music being processed and whether or not language is involved in that processing. It also creates a link between MH, brain activation, and memory. Given that MH is a frequent experience of music, which makes the subject familiar with it, certain brain regions may show increased activity because of being familiar with the music. According to Freitas et al [34], the brain areas involved in processing familiar music are distinct from those involved in processing unfamiliar music.

Other studies have also indicated that brain activation during MH may be related to the processing of memory. Bosso et al [35] studied the neurophysiology and neurobiology of the processing of musical experience and found that there is a close association between brain pathways used for emotional memory processing, and musical processing, including increased activity in the cingulate cortex, and the hippocampus, all of which were also found to be increased in activity in the current study. Thus, increased activation in these specific areas may be explained by the brain’s processing of a musical experience, which causes changes in activity in multiple areas of the brain, as memory, emotion, and auditory function are all involved in the neurobiological processing of music.

It is especially interesting to compare the results of the current study to the results of Cavaliere et al [9], who investigated the morpho-functional circuits of MH, in a similar-age healthy female, and considered brain activity using both PET and MR scans. The study showed increased activity in the superior temporal gyri, anterior cingulate, left orbital frontal, and medial temporal cortices. This is similar to the findings of the current study, placing special emphasis on the activity of the superior temporal gyri, which may be activated in all MH subjects.

**Conclusions**

This report highlights that patients with hearing loss may present with continuous musical hallucinations, and that these patients should not be assumed to have delusions or psychosis, but should be investigated for underlying brain abnormalities and treated appropriately. Considering this, it can be said that in the present study, the neurological findings indicate a differentiated, multiple-area involvement in MH. This is, at least in part, consistent with perception of external audio stimulus, and may simply be indicative of the underlying brain mechanism creating the perception of an external auditory input. However, the activity in the orbitofrontal cortex and the related gyrus is key to the experience of MH. Overall, these findings suggest an alternate processing of reality that occurs when MH is perceived as an external stimulus.

Further, the altered or activated regions are all related to both the processing of emotions and the processing of episodic memory. This is also seen in the MH of schizophrenic patients. These results support that cortical activation is specifically associated with the continuation or activation of MH.

**Declaration of Figures’ Authenticity**

All figures submitted have been created by the authors who confirm that the images are original with no duplication and have not been previously published in whole or in part.
References:


Indexed in: [PMC] [PubMed] [Emerging Sources Citation Index (ESCI)] [Web of Science by Clarivate]

This work is licensed under Creative Common Attribution-NonCommercial-NoDerivatives 4.0 International (CC BY-NC-ND 4.0)