Neuroanatomical considerations of isolated hearing loss in thalamic hemorrhage

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ABSTRACT

Background and importance: Thalamic lesions are associated with a wide variety of clinical syndromes. Due to the close anatomic proximity of the nuclei, many of these syndromes have considerable overlap in clinical sequelae and, as such, a lesion affecting only one modality is exceedingly rare.

Clinical presentation: In this case, a 55 year-old right handed man with a past medical history significant for hypertension, poly substance abuse, and a 25 year history of seizure disorder following clipping of a middle cerebral artery aneurysm presented with isolated bilateral hearing loss.

Conclusion: Presumably, this neurological deficit was caused by a hypertensive hemorrhage in the posterior right thalamus. The following case and discussion will review the potential neuroanatomical pathways that we suggest could make isolated hearing loss be part of a “thalamic syndrome.”

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1. Introduction

Thalamic nuclei are composed of 5 major functional classes: sensory nuclei that relay inputs from all sensory modalities, effector nuclei concerned with motor function and aspects of language, reticular nuclei that subserve arousal and nociception, associative nuclei that participate in high-level cognitive functions, and limbic nuclei concerned with mood and motivation. Damage to thalamic nuclei has consistently been found to induce a variety of motor, sensory and neuropsychological impairments with varying degrees of overlap between the functionally distinct modalities. To date, a consistent clinical picture of a thalamic syndrome has not emerged. Due to the close anatomic proximity and overlapping vascular supply, a lesion which affects only one modality while sparing others is rare [1–3]. In this case, a patient presented with isolated bilateral hearing loss from a presumed hypertensive hemorrhage in the posterior right thalamus.

2. Case report

The patient is a 55 year-old right handed man with a past medical history significant for hypertension, poly substance abuse and a 25 year history of seizure disorder following clipping of a middle cerebral artery aneurysm. The patient presented to the Emergency Department after suffering three generalized tonic-clonic seizures over the 24 h prior to admission. The patient reported that over the past 25 years he has had multiple seizures (approximately one per month). His antiepileptic medication had been switched to keppra recently and his seizure frequency had subsequently significantly decreased (no seizure episodes in the prior 2 years). On the date of admission the patient suffered three generalized tonic-clonic seizures. After the third seizure that reportedly lasted greater than 10 min the patient developed profound hearing loss and tinnitus. The seizures were described as generalized tonic-clonic, the last 2 seizures prior to presentation reportedly lasted 10 min each separated by a 30-minute interval. The patient reported loss of consciousness with all seizure episodes and postictal confusion. The patient’s neurologic exam on presentation to the emergency department was awake, alert, and oriented to person place and time, comprehension was intact, with normal writing and fluent speech. Cranial nerves II–XII were intact and bilaterally symmetric except bilateral hearing loss. Detailed hearing examination demonstrated that Weber test was midline, while Rinne test was significant for bilateral sensorineural hearing loss. The patient’s motor exam was normal and symmetric in all extremities and sensation to light touch, pin prick and joint position sense were intact. A head CT scan revealed an isolated 1.5 by 1.0 by 0.5 cm hemorrhage centered in the posterior aspect of the right thalamus, along with multiple clips in the region of the left middle cerebral artery (Fig. 1A). An MRI was unable to be performed for safety concerns due to the possibility of incompatible aneurysm clips. A CT angiogram was done which did not show any vascular anomalies. However, audiometric testing on admission revealed severe bilateral sensorineural hearing loss. On admission, the patient was started on a second seizure medication. He had no further seizures during his hospital stay and showed no signs of further neurologic deterioration. The Otolaryngology service was consulted and determined that
there was no evidence of cochlear involvement and that the patient’s deficits were likely isolated sensorineural hearing loss. Audiometric testing done on hospital day 3 showed no improvement over the initial test. Repeat head CT showed the clot to be stable in size without mass effect or signs of hydrocephalus, and so the patient was discharged home in stable condition on hospital day 3 (Fig. 1B). Following discharge the patient failed to follow up with any of the clinical services involved in his care.

3. Discussion

Thalamic lesions are associated with a wide variety of clinical syndromes. Due to the close anatomic proximity of the nuclei, many of these syndromes have considerable overlap in clinical sequelae and, as such, a lesion affecting only one modality is exceedingly rare. In this case, a patient presented with isolated bilateral hearing loss from a presumed hypertensive hemorrhage in the posterior right thalamus. This
region draws its vascular supply from the inferolateral arteries which are composed of 4 to 10 arteries that arise from the P2 branch of the posterior cerebral artery [2]. Patients with inferolateral artery infarction typically present with the thalamic syndrome described by Dejerine and Roussy, namely, sensory loss to a variable extent, with impaired extremity movement and sometimes post lesion pain [1,2]. The complexity of the penetrating arteries that constitute the inferolateral arteries explains why small vessel disease in this territory can have distinctly different presentations. In this patient an isolated hearing deficit can be postulated, but not entirely explained, by infarction of the right medial geniculate nucleus, one of the principle nuclei supplied by this vascular territory (Fig. 2).

Auditory impulses travel from the cochlea via the cochlear nerve along the anteroinferior quadrant of the internal auditory canal through the cerebellopontine angle to synapse at the dorsal and ventral cochlear nuclei at the junction of the medulla and the pons. The dorsal and ventral cochlear nuclei contain the second-order neurons and give rise to projections to the contralateral brainstem which ascend as the lateral lemniscus which projects to the central nucleus of the inferior colliculus. These projections include the dorsal acoustic striae, the intermediate acoustic striae and the ventral acoustic striae, which is part of the trapezoid body [1,4]. Decussating fibers of the trapezoid body and fibers from the contralateral superior olivary complex ascend in the lateral lemniscus and terminate in the inferior colliculus. The inferior colliculus contains the third order neurons which project to the medial geniculate body which then project forth order neurons primarily to the primary auditory cortex, Brodmann area 41 with a small contribution to the association auditory cortex Brodmann 42 [1]. There are multiple connections along the auditory pathway. The superior olivary complex receives bilateral input from crossed fibers, there are connections between the two cochlear nuclei, the dorsal nuclei of the lateral lemniscus and between the inferior colliculi. There are however no connections at the level of the medial geniculate body. Unilateral neural sensorineural hearing loss (SNHL) may result from the involvement of only the cochlear nerve or nuclei [1,4]. Lesions in the more proximal intraaxial auditory pathway result in bilateral SNHL, which is often more noticeable in the contralateral side.

In this case bilateral hearing loss cannot be explained entirely on the basis of the anatomical destruction of the right MGN as the significant crossover and of the system should allow for some sound perception from both cochlear nuclei via the crossed paths. One potential mechanism for the observed bilateral hearing that may not have been demonstrated on standard audiometric testing is the concept of auditory hemispatial extinction which is described as the ability to gate auditory input to the cortex [5]. It has been demonstrated that lesions in the posterior part of the right thalamus including the pulvinar and medial geniculate body produce deficit not only in the processing of complex auditory stimuli but also in the allocation of attention to input from one ear to the other. In this patient the profound bilateral sensorineural hearing loss and tinnitus could be explained by a combination of the destruction of one of the key relay nuclei in the auditory system with the inability to focus the cortical input from the intact contralateral system [1,5].

4. Conclusion

This case illustrates a rare presentation of an isolated right thalamic hemorrhage causing bilateral sensorineural hearing loss in a patient with no prior neurological deficits. We speculate this new deficit may be the result of a hemorrhagic lesion in the right thalamus which likely involved the medial geniculate nucleus and other posterior thalamic and basal ganglia nuclei. Due to significant commissural connections within the auditory system between the cochlear nerve and the final thalamic relay in the medial geniculate body bilateral hearing loss cannot be explained completely based on the lesion with which this patient presented. However, our case study lends support to the concept of isolated hearing loss being part of a “thalamic syndrome.”

Disclosure

The authors have no personal financial or institutional interest in any of the drugs, material, or devices described in this article.

References