Complex auditory hallucinations (AH) are frequently a symptom of psychiatric disease. They have also been described with lesions of the peripheral auditory pathways or cerebral hemisphere. The pathophysiology of complex AH remains controversial. Their occurrence with peripheral auditory lesions suggests a role for deafferentation in the central release of auditory memories. In contrast, a direct role of the cerebrum is suggested by the association of AH with brain tumors, epilepsy and alcohol abuse. Recently, cases of AH due to brainstem stroke, haemorrhagic and ischemic, have been described. We report an association with Listeria rhombencephalitis, which to our knowledge has not been previously described. The possible mechanism of such brainstem AH will be discussed and an approach to the diagnosis of new onset AH is proposed.

CASE REPORT

A 43-year-old man presented with a prodrome of bifrontal headache, fever, nausea, vomiting and a three day history of left sided perioral paraesthesia. At the time of admission he also complained of difficulty with balance and taste. He had a past history of hypertension, hypercholesterolemia, obstructive sleep apnoea and alcohol abuse.

On admission, his temperature was 38.7°C. There was no meningeal irritation. The clinical examination was normal. Lumbar puncture revealed a clear CSF with cell counts of 7 RBC/mm³ and 143 WBC/mm³ (42% polymorphonuclear cells). The CSF to serum glucose ratio was 50%. The CSF protein level was 0.92g/L. Listeria monocytogenes was identified in blood cultures.

Despite immediate treatment with cefotaxime 1g IV Q8H and ampicillin 2g IV Q6H, his neurological condition deteriorated in the 48 hours following admission. He developed progressive deficits involving the left cranial nerves V, VII, VIII, IX and X, as well as left-sided limb ataxia. Hearing acuity was markedly reduced on the left side and the Weber test lateralized to the right. He complained of nearly constant paraesthesia. At the time of admission he also complained of difficulty with balance and taste. He had a past history of hypertension, hypercholesterolemia, obstructive sleep apnoea and alcohol abuse.

Audiometry was only done at the time of recovery. It revealed a mild impairment of speech discrimination on the left with moderate high frequency bilateral sensorineural hearing loss.

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ABSTRACT: Background: Complex auditory hallucinations have rarely been reported in cases of brainstem stroke or tumor. Method: Case study. Results: A patient with acute Listeria rhombencephalitis complained of formed musical auditory hallucinations on the side of recent sensorineural deafness. MRI revealed an abscess in the middle cerebellar peduncle with extensive surrounding edema. Conclusions: Disruption of brainstem auditory pathways may cause complex auditory hallucinations. Potential pathogenetic mechanisms are discussed and a diagnostic approach is proposed.

DISCUSSION

Listeria rhombencephalitis is a rare infection with a high mortality and frequent neurologic sequelae in survivors. As part of his acute presentation, this patient developed AH and severe unilateral sensorineural deafness. The definite musical and verbal content of his auditory experiences, as well as his preserved insight and clear sensorium establish that these are hallucinations, not auditory illusions nor simple tinnitus. Neuroimaging and clinical findings allow inferences as to the likely site of pathology. MRI showed an abscess in the middle cerebellar peduncle, with extensive edema in the neighboring medulla and pons. Clinical deficits implicating cranial nerves 5, 7, 9 and 10, with sparing of the abducens nerve, medial lemniscus and corticospinal tract would suggest involvement of the dorsolateral pontomedullary junction. Although extraaxial involvement of these cranial nerves by meningitis cannot be completely excluded, such unilateral clustering would be very unlikely and meningeal hyperintensity was not seen on the gadolinium-enhanced MRI study. In addition, there was no clinical, MRI, nor electroencephalographic evidence of supratentorial involvement. Thus the central auditory structures most likely to be implicated in this case are the left cochlear nuclei, acoustic striae, superior olive, trapezoid body and caudal lateral lemniscus. There is bilateral representation in the auditory pathways ascending from the cochlear nuclei, such that lesions of the lateral lemniscus or more rostral auditory structures cause only partial bilateral hearing loss. The profound left-sided deafness noted in our patient suggests significant involvement of the ipsilateral cochlear nuclei.

Brainstem AH have been reported previously in five cases of pontine haemorrhage, four cases of pontine tegmental infarction and one case of metastasis to the midbrain. The anatomical structures implicated ranged in caudal-rostral extent from the cochlear nuclei to the inferior colliculi. Serial neuropathological sections of the brainstem were examined only in the case of Cascino and Adams and revealed destruction of the inferior colliculus and adjacent lateral lemniscus, with sparing of more caudal auditory pathways. On the basis of clinical, MRI and brainstem evoked potential data, Lanska and Murata have postulated involvement of more caudal structures, namely the acoustic striae, trapezoid body and superior olivary nucleus, consistent with the localization observed in the present case.

In our patient, as in all previously reported cases of brainstem AH, hearing loss occurred ipsilateral to the ear in which hallucinations were perceived to arise. Elementary and musical hallucinations have been extensively described in cases of acquired chronic deafness of peripheral etiology. In this context, hallucinations tend to occur after years of sensorineural or conductive hearing loss and run a fluctuating course. They generally begin insidiously, taking the form of elementary tinnitus of variable pitch or more complex, engine-like sounds. Gordon has argued that most, if not all, AH have a peripheral generator. Indeed, individual hair cells of the bullfrog sacculus have been implicated in the production of cochlear distortions, although there is no evidence that this mechanism would lead to more complex AH. Clearly, AH which include verbal content or recognition of a specific melodic line must involve activation of cerebral cortical areas.

In the present case, complex AH appeared within a few days of an acute brainstem injury. Their disappearance was concurrent with neurologic improvement, further suggesting an etiologic link. Conceivably, in AH of peripheral or brainstem origin, the release of auditory memories is triggered by auditory input deprivation. The pathways mediating such
selective cortical disinhibition or hyperexcitability are unknown. Hypnagogic hallucinations, which may feature visual, auditory or somesthetic perceptions, may represent a useful model. They consist of a dissociation of dream-like experiences from their usual substrate, sleep. Pontine neurons responsible for ponto-geniculo-occipital waves (which also project to parietal and temporal association areas), may be the generators of hypnagogic hallucinations. These neurons appear to be inhibited by pontine raphe neurons, which have been a site of pathology in many reported cases of brainstem AH. Peduncular hallucinosis also bears similarity to brainstem AH, although here vespertal visual hallucinations are the hallmark and the pathology is mesencephalic rather than predominantly pontine.

Complex AH may be a source of great patient anxiety and often pose a diagnostic challenge. When there is a definite verbal content, a psychiatric disorder can be suspected in the context of a normal neurologic examination, prior history of psychiatric disease, impaired insight and association with other psychotic symptoms. The presence of chronic or new hearing loss should be assessed, as auditory deprivation may play an important role in AH. Although rare, focal supratentorial and brainstem pathology should also be considered. This report and others emphasize an association of complex AH with neoplastic, vascular or inflammatory disorders of the brainstem, particularly the pontine tegmentum. In all cases reported so far the brainstem localization was evident from the clinical examination. However, it is conceivable that a more limited structural brainstem lesion could cause selective auditory pathway deafferentation. Consequently, focal infratentorial pathology should be considered in the assessment of patients with complex AH.

REFERENCES