

Hearing Symptoms in Migrainous Infarction

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Background: In case reports, migraine headaches have been associated with fluctuating low-frequency hearing loss and sudden, unilateral hearing loss. Auditory symptoms associated with migrainous infarction have not previously been emphasized.

Objective: To describe migrainous infarction presenting with acute auditory symptoms.

Design: Case reports.

Setting: Tertiary care hospitals.

Patients: A 40-year-old man with a history of migraine suddenly developed bilateral hearing loss associated with severe, throbbing, occipital headache, tinnitus, vertigo, speech disturbance, and right hemiparesis. An early audiogram showed profound, down-sloping, sensorineural-

type hearing loss bilaterally. Sixteen days later, a follow-up pure tone audiogram documented marked improvement in both sides to a pure tone average of 30 dB. Right hemiparesis and dysarthria also improved steadily for 2 months. A 25-year-old woman with a history of migraine with aura suddenly developed hyperacusis, unilateral hearing loss, and migraine headache early in migrainous infarction. Magnetic resonance imaging documented infarcts in the pons and cerebellum.

Conclusions: In these patients, acute auditory symptoms are a part of the prodrome of migrainous infarction. We speculate that these symptoms may have resulted from migraine-associated vasospasm. Migrainous infarction should be considered in the differential diagnosis of acute auditory symptoms, including sudden, bilateral hearing loss.

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MIGRAINE HAS been reported to cause sudden sensorineural hearing loss (SNHL).^{1,2} When this occurs without acute headache in a migraineur, it is questionable whether the SNHL is related to migraine. We know of only a few reports¹⁻⁴ of sudden auditory symptoms attributed to migraine. We describe 2 patients diagnosed as having migrainous infarction, each of whom presented with acute auditory symptoms (AAS)—one with bilateral sudden hearing loss, the other with tinnitus, hyperacusis, and unilateral hearing loss. Both had characteristic features of migraine at the ictus as well as a history consistent with complicated migraine.

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PATIENT REPORTS

PATIENT 1

A 40-year-old man suddenly developed vertigo, vomiting, and bilateral tinnitus.

One hour later, he abruptly developed bilateral SNHL. He was unable to hear his wife speaking. Within a few minutes, he developed severe dysarthria, right hemiparesis, and severe, throbbing, occipital headache. The headache and vertigo resolved within a few hours, but bilateral hearing loss, right hemiparesis, and severe dysarthria persisted.

The patient had a history of recurrent headaches accompanied by vertigo, hearing loss, and right hemiparesis. There was no history of visual aura. Two years before admission, the patient had had a neurologic event, described as sudden right hemiparesis, followed by throbbing occipital headache, nausea, vertigo, and right-sided hearing loss. The right hemiparesis and right-sided hearing loss persisted for 2 weeks. Similar episodes recurred 5 to 6 times during the subsequent year. One year before admission, propranolol hydrochloride and flunarizine hydrochloride had been prescribed. The frequency and severity of neurologic attacks had de-

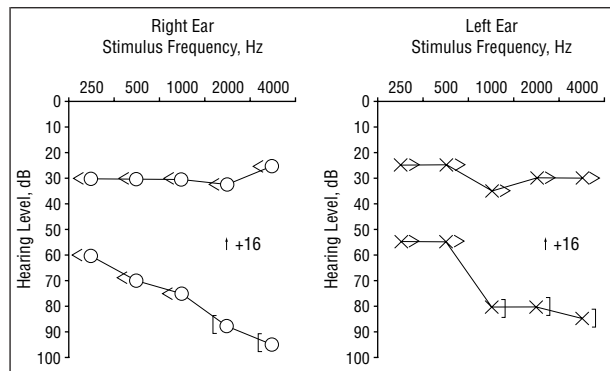


Figure 1. Pure-tone audiogram obtained on 2 different dates, 16 days apart (+16). The first audiogram was obtained May 1, 2000, and the second follow-up audiogram on May 17, 2000. The patient's hearing levels are plotted against stimulus frequency on a logarithmic scale. The decibels were measured according to the American National Standards Institute, 1989. Circles indicate air conduction on the right side; ×, air conduction on the left side; <, bone conduction on the right side; >, bone conduction on the left side; [, bone conduction (masked) on the right side; and], bone conduction (masked) on the left side.

creased remarkably with these medications. However, 6 months before admission, the patient had discontinued taking medications.

There was no history of hypertension, diabetes mellitus, coronary artery disease, or stroke. The patient's mother had migraine headaches without other neurologic symptoms. On admission of the patient, results of extensive blood tests including antinuclear antibodies, treponemal serologic testing, erythrocyte sedimentation rate, prothrombin time, activated partial thromboplastin time, activated protein C resistance, fibrinogen, antiphospholipid antibody assay, and factor V Leyden assays were negative. Brain magnetic resonance imaging and magnetic resonance angiography of the brain and neck showed no abnormalities. Transthoracic and transesophageal echocardiography with a bubble study showed no abnormalities. Audiometry showed a severe to profound down-sloping SNHL in the right ear and a moderate to severe down-sloping SNHL in the left ear. The patient was treated with calcium channel blockers and low-molecular-weight dextran. Sixteen days later, a follow-up audiogram showed marked improvement in both sides to a mild hearing loss of 30 dB pure tone average (**Figure 1**). Right hemiparesis and speech disturbance improved steadily for 2 months. At last follow-up, the patient was neurologically normal, took daily propranolol, and had had no further attacks.

PATIENT 2

A 25-year-old woman was well until her car was towed and she had difficulty regaining possession of it. She was extremely angry and noted that this may have been the most difficult day of her life. Two hours later, she suddenly developed buzzing, right-sided tinnitus, and hyperacusis. Describing the hyperacusis, she noted, "The wind sounded like an airplane." Within seconds, she developed vertigo, subjective right-sided hearing loss, diplopia, quadriparesis, and right hemibody numbness. During the following 10 hours, she had severe nausea and

vomiting; mild, persistent dysarthria; hemibody numbness; and mild, asymmetric quadriparesis. Three days later, she reported headache, blurred vision, severe hyperacusis, and dysarthria. There was a history of migraine headaches with auras described as small visual field deficits lasting approximately 5 minutes and sometimes preceding a headache. The headaches were incapacitating, with bifrontal, bitemporal, or occasionally bioccipital pressure and throbbing, associated with vomiting and occasional numbness of the right side of the face and right hand. There was a family history of migraine in both parents and a paternal grandfather.

On examination several days after the ictus, speech was mildly scanning. The patient exhibited a combined upbeat and torsional nystagmus. Lateral gaze induced predominantly torsional nystagmus, worse on rightward than leftward gaze. Downgaze was normal without nystagmus. Magnetic resonance imaging showed signal abnormalities, consistent with multiple infarcts in the bilateral cerebellum and pons (**Figure 2**). No likely stroke mechanism was disclosed by extensive testing, including complete blood cell count, electrolytes, prothrombin time, activated partial thromboplastin time, Russell viper venom time, activated protein C resistance, fibrinogen, antinuclear antibodies, anticardiolipin antibodies, homocysteine, pyruvate, blood cultures, lumbar puncture, and transthoracic and transesophageal echocardiography with a bubble study.

COMMENT

Several studies have suggested that migraine is associated with an increased incidence of stroke.⁵⁻⁹ Our 2 patients met criteria for migrainous infarction, defined according to the criteria of the International Headache Society,¹⁰ in which the pathogenesis is directly due to migraine, the incident occurs during a typical attack of migraine with aura, neurologic deficits are not completely reversible within 7 days, and the diagnosis is made when other causes have been excluded. Most striking about both these patients were the early and prominent AAS. Previous reports on migrainous infarction have not emphasized AAS as a warning of impending stroke.

ACUTE AUDITORY SYMPTOMS

Patient 1 had sudden, bilateral hearing loss at the onset of migrainous infarction. As far as we know, this is the first report of migraine associated with bilateral, reversible sudden hearing loss, documented by a pure tone audiogram. Patient 2 had acute, unilateral tinnitus, hyperacusis, and hearing loss at the onset of her migrainous infarction. Among the causes of AAS, Meniere disease, autoimmune inner ear disease, and multiple sclerosis are common.^{11,12} In many cases, however, the mechanism of AAS is indeterminate. Putative explanations of idiopathic AAS include mainly labyrinthine ischemia and inflammation.¹³ In clinical practice, AAS in a young patient without known vascular risk factors are often attributed to viral infection of the cochlea.^{13,14} However, sudden hearing loss may occur rapidly, over seconds or minutes, making infectious inflammation unlikely. The

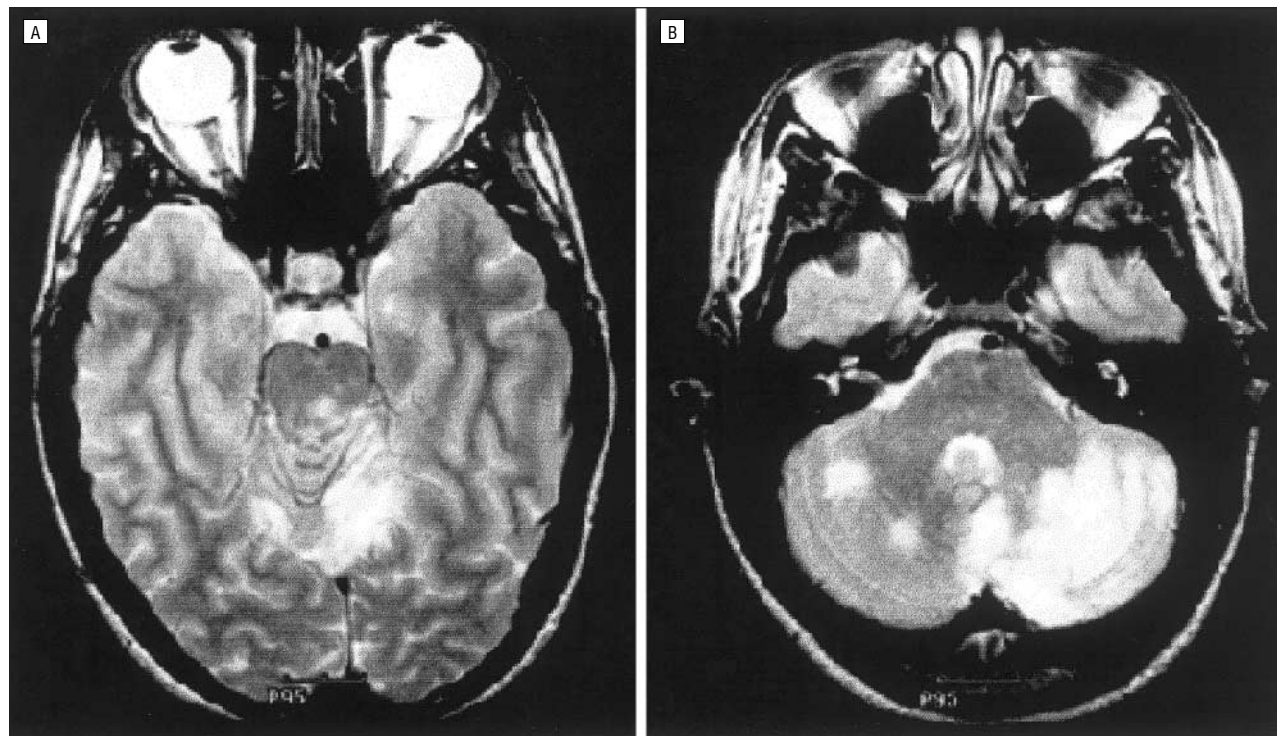


Figure 2. Axial T2-weighted magnetic resonance image showing hyperintensities involving the upper pons (A) and bilateral cerebellum (B).

AAS may be associated with basilar artery occlusion¹⁵ and with hypercoagulable states, such as polycythemia vera or macroglobulinemia.^{16,17} In our 2 patients, symptoms began abruptly, but vascular risk factors were absent, and extensive diagnostic tests did not disclose a stroke mechanism. One poorly understood mechanism for ischemia is cerebral vasospasm. We speculate that, in these 2 otherwise healthy patients with rapid onset of symptoms, vasospasm may have caused vertebrobasilar ischemia.

Vasospasm is said to be more common in patients with migraine,¹⁸ although vasospasm has an unknown role in the production of symptoms. In 1991, Gomez et al¹⁹ described a young woman with migraine with aura who had a cerebellar infarct, associated with transiently high velocities in both vertebral arteries, consistent with vasospasm. However, vasospasm is not the only explanation of migrainous infarction. In migraine, visual aura may result from a metabolic defect that slowly spreads across the occipital cortex.²⁰ In addition, patients with migraine may have increased platelet aggregation, increasing the risk of infarcts.²¹

HEARING LOSS IN MIGRAINE

We know of 4 previous studies of SNHL attributed to migraine.¹⁻⁴ In 1987, Lipkin et al¹ described a patient with sudden SNHL, associated with characteristic migraine headache. In 1991, Caplan² described sudden onset of unilateral hearing loss and aural fullness, as the prodrome of a cerebellar infarct, in a patient who had migraine with visual aura and basilar artery stenosis. In 1996, Virre and Baloh³ described 13 patients with unexplained sudden hearing loss, all of whom had long-standing migraine headaches. All of the latter patients had

abrupt onset of hearing loss and other neurologic symptoms. In 2000, Lee et al⁴ described a patient with long-standing migraine with aura who experienced sudden hearing loss at age 50 years and onset of Ménière syndrome at age 73 years. The authors suggested that migraine-associated vasospasm may have caused sudden deafness and damaged the endolymphatic sac of the initially intact ear, predisposing the patient to delayed endolymphatic hydrops. In both patients described herein, symptoms were typical of previous neurologic attacks, consistent with complicated migraine.

LOCALIZATION OF HEARING LOSS

The SNHL in patient 1 was consistent with a lesion of either the sensory end organ (cochlea) or the cochlear nerve. Despite the profound bilateral SNHL, however, vertigo in patient 1 was transient, and results of caloric tests were normal. These findings suggest that the main injury in patient 1 was to the cochlea. The high-frequency hearing loss suggests that the vasospasm may have selectively involved small arterioles within the base of the cochlea, just as it involves only a subset of retinal arterioles in ocular migraine.²² Without pathological data, it is impossible to precisely determine the disease processes responsible for hearing loss in our patients.

CLINICAL RELEVANCE

The 2 patients described herein illustrate migrainous infarction with early and prominent auditory symptoms. Each of these patients had a history consistent with complicated migraine, and past attacks were similar to the most dramatic attacks, those reported herein. Of note,

hearing loss has also been reported as a prodrome of non-migrainous infarction, in the territory of the anterior inferior cerebellar artery.¹⁵ Together, these previous observations, along with those described herein, highlight the importance of taking a good history of hearing symptoms, with particular attention to transient symptoms that could serve as a sign of impending stroke. Migrainous infarction should be considered in the differential diagnosis of acute hearing symptoms, including unilateral and bilateral hearing loss.

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