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Clinical Records

Middle-ear myoclonus

Terrence E. Zipfel, M.D., Srinivas R. Kaza, M.D., J. Scott Greene, M.D.

Abstract
Tinnitus produced by repetitive contraction of the middle-ear muscles is a rare condition. We present an interesting case of bilateral middle-ear myoclonus causing incapacitating tinnitus in a patient with multiple sclerosis. Otological examination demonstrated rhythmic involuntary movement of the tympanic membrane. These movements correlated with a rhythmic 'rushing wind' noise perceived by the patient. Oropharyngeal examination showed no evidence of palatal myoclonus. Impedance audiometry confirmed rhythmic change in the middle-ear volume. Medical management was unsuccessful. The patient's tinnitus was subsequently cured with bilateral sectioning of the tensor tympani and stapedial tendons.

Key words: Tinnitus; Myoclonus; Middle ear; Multiple sclerosis

Introduction

Tinnitus is a common auditory phenomenon that has been associated with many otological diseases. There are essentially two types of tinnitus: subjective and objective. Subjective tinnitus is more common and reflects an abnormality of the auditory system. It is characterized by an individual's perception of sound in the absence of any physical source. Hearing loss is reported to be the primary risk factor for subjective tinnitus.[1] The exact pathophysiological mechanism of subjective tinnitus, however, is unknown.

Objective tinnitus is much less common that subjective tinnitus. Objective tinnitus refers to a sound that is produced mechanically within the head and is often heard by both patient and examiner. Objective tinnitus is uncommon and may be caused by a variety of conditions including vascular abnormalities, temporomandibular joint crepitus, a patulous eustachian tube, palatal myoclonus, and middle-ear myoclonus.[2] We report a case of bilateral middle-ear myoclonus that resulted in incapacitating tinnitus.

Case report

A 44-year-old female with a history of multiple sclerosis presented with complaints of an intermittent 'blowing, gushing wind' noise in both ears. She stated that the noise had been occurring hundreds of times a day for several months. She also complained of pressure in the left ear. She denied any change in hearing. The tinnitus became extremely annoying to her. She denied any precipitating factors. On physical examination the patient demonstrated rhythmic movement of both tympanic membranes. These movements coincided with the tinnitus. Immittance audiometry confirmed spontaneous rhythmic changes in the middle-ear volume (Figure 1). Tympanometry was normal. Pure tone audiometry demonstrated a bilateral high-tone sensorineural hearing loss (Figure 2). Direct examination and fibre-optic nasopharyngoscopy showed no signs of palatal myoclonus. Magnetic resonance imaging (MRI) of the brain revealed only multiple plaques consistent with the patient's underlying diagnosis of multiple sclerosis. A diagnosis of middle-ear myoclonus was established. Medical management with benzodiazepines was unsuccessful. The patient was ultimately treated with simultaneous bilateral tympanotomy and sectioning of the tensor tympani and stapedial tendons. She had been counselled pre-operatively that if difficulties were encountered during the surgery, only a unilateral procedure would be performed. The patient gained immediate relief from the tinnitus post-operatively.

Discussion

Tinnitus secondary to middle-ear myoclonus is rare. Few reports exist in the literature. Badia *et al.*[3] reported six cases of middle-ear myoclonus. In each case, the tinnitus was unilateral. Others have reported cases of tinnitus produced by stapedial muscle spasm.[4,5] We believe our case is the first reported case of bilateral middle-ear myoclonus.

Middle-ear myoclonus is defined as a rhythmic movement of the tympanic membrane secondary to repetitive contraction of the tensor tympani and stapedial muscles. These myoclonic movements are a form of segmental myoclonus and involve muscles innervated from a limited segment of the brainstem. Segmental myoclonus from brainstem pathology can result in ocular, palatal, jaw, facial, tongue, and middle-ear myoclonus. The aetiology of segmental myoclonus may be vascular, infectious, demyelinating, tumour, trauma or idiopathic. Myoclonus may be caused by semiautonomous discharge from medullary

From the Department of Otolaryngology – Head and Neck Surgery, Penn State Geisinger Medical Center, USA.
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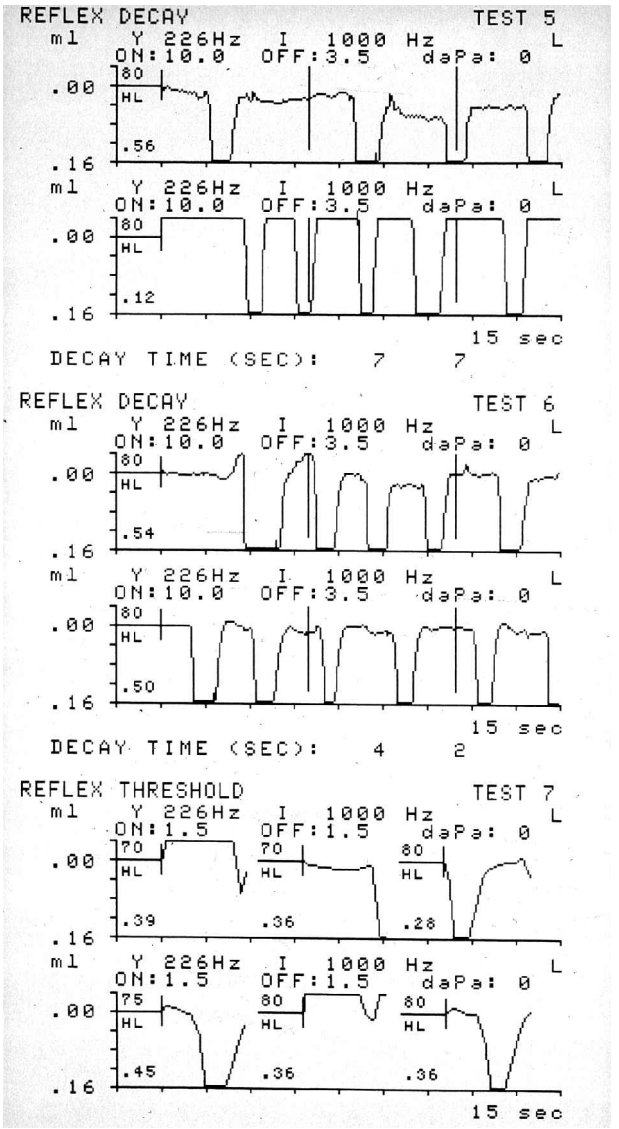

FIG. 1

Immittance audiometry using an 80 dB (10 dB suprathreshold) stimulus shows spontaneous rhythmic changes in middle ear compliance rather than the expected acoustic reflex decay.

centres that have been released from higher inhibitory control.[6] In this case, the middle-ear myoclonus was most likely due to demyelination in the brainstem secondary to multiple sclerosis.

The mechanism of tinnitus production in middle-ear myoclonus is unknown. Tensor tympani contraction is said to produce a clicking sound,[7] whereas stapedial contraction produces a buzzing noise.[5] Tinnitus could be due to propagation of the muscle contraction noise. It may also be due to potential vibration of the tympanic membrane during contraction of the intratympanic muscles, stimulation of the tympanic plexus, or alteration of the cochlear microphone potential.[3]

The diagnosis of middle-ear myoclonus is not always evident on otoscopy. In our case, however, rhythmic movements of the tympanic membranes were easily visualized on otoscopic examination. The frequency of clicking has reportedly been as high as one to two second. Clicks are most easily recorded with the use of impedance audiometry. Clicks appear as slight peaks over the baseline, indicating tympanic membrane movement.

The main differential diagnosis of objective tinnitus is palatal myoclonus. Tinnitus associated with palatal myoclonus is usually bilateral.[8] Several theories exist to explain the origin of the noise. Pulec and Simonton[8] postulated that the sound could result from the eustachian tube snapping open or the breaking of surface tension as the walls of the eustachian tube open under the action of peritubal muscles. The diagnosis of palatal myoclonus can usually be made on direct oral cavity examination. Fibreoptic examination of the nasopharynx may also assist in the detection of palatal movement. Electromyography (EMG) has also been used to record palatal muscular activity. In our case there was no evidence to support a diagnosis of palatal myoclonus on either oral cavity examination or flexible nasopharyngoscopy.

A variety of options exist for the treatment of middle-ear myoclonus. Medical management with sedatives such as benzodiazepines or carbamazepine has been used with varying success. In Badia *et al.*[3] tinnitus secondary to middle-ear myoclonus was controlled with sedatives in two to six patients. Hypnosis, psychotherapy, acupuncture, and blockage of the otic ganglion have all been attempted as treatments without much success. Several surgical therapies have been used in the treatment of middle-ear myoclonus. The placement of pressure equalization tubes

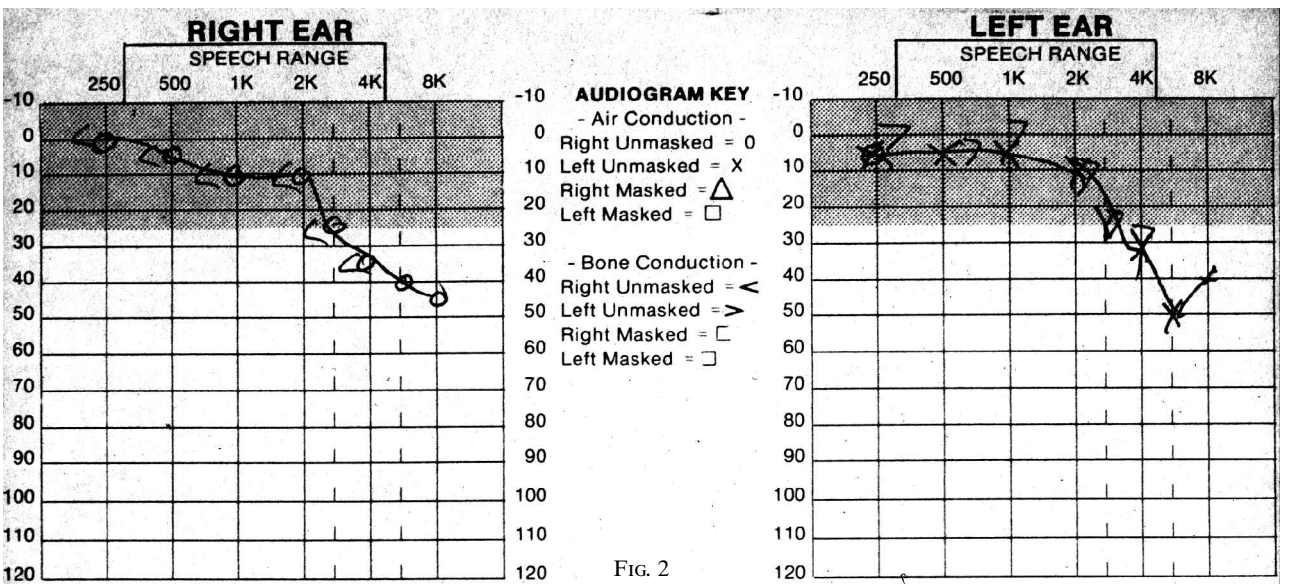

FIG. 2

Pure tone audiogram showing bilateral high-frequency sensorineural hearing loss.

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had been performed by Badia *et al.*[3] and was shown to be of no benefit. The main surgical intervention has been tympanotomy with sectioning of the tensor tympani and stapedial muscle tendons. Three of the six patients in the Badia *et al.*[3] series failed conservative treatment measures and gained significant relief after sectioning of the tensor tympani and stapedial tendons. Two cases of tinnitus produced by stapedial muscle spasm were successfully treated by Marchiando *et al.*[4] with stapedial tendon lysis after conservative measures failed. In our case, the patient was successfully treated with bilateral tympanotomy and sectioning of the tensor tympani and stapedial tendons.

Recently, *Clostridium botulinum* toxin has been injected into palatal muscles in cases of palatal myoclonus. Botulinum toxin has been used to relieve tinnitus and control palatal myoclonus.[9] This intervention was also used by Badia *et al.*[3] to control unilateral middle ear myoclonus associated with blepharospasm in a patient with underlying facial nerve pathology. Objective clicking tinnitus has also been successfully treated with tinnitus masking devices.[10]

Conclusion

The most common mechanical causes of objective tinnitus are palatal myoclonus and middle-ear myoclonus. We have presented a case of bilateral middle-ear myoclonus that was effectively treated by tympanotomy with division of the tensor tympani and stapedial muscles. Primary treatment of this problem is medical. However, patients may be so disrupted by this problem that surgery becomes necessary if medical therapy fails. Surgical sectioning of the tensor tympani and stapedial tendons via tympanotomy is a straightforward procedure. This surgical therapy has been effective and no adverse effects have yet been encountered or reported.

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Address for correspondence:
Srinivas R. Kaza, M.D.,
Department of Otolaryngology – Head and Neck Surgery,
100 North Academy Avenue,
Danville, PA 17822-1333,
USA.

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