Bilateral Tinnitus due to Middle-Ear Myoclonus

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Abstract: Tinnitus is a common otological symptom. Usually it is subjective (perceived only by the patient); very rarely is it objective (heard by both the patient and the examiner). Objective tinnitus due to middle-ear myoclonus is extremely rare, with only a few case reports published in the literature. We present three cases of objective tinnitus caused by middle-ear myoclonus. All patients were cured by tympanotomy with stapedial and tensor tympani tendon section.

Key Words: myoclonus; objective tinnitus; stapedius muscle; tensor tympani muscle

innitus, a common otological symptom secondary to a large number of etiologies, results in variable degrees of disability in the affected patient population. It is classified as subjective (perceived only by the patient) or objective (perceived by the patient and the examiner). Objective tinnitus is much less common than is subjective tinnitus: It usually refers to sounds that are produced mechanically within the head and are transmitted from nearby structures (somatosounds), whereas subjective tinnitus reflects an abnormality of the auditory system and is characterized by an individual's perception of sound in the absence of any physical source. A wide variety of anomalies have been reported to cause objective tinnitus, including various vascular abnormalities (arteriovenous malformations and shunts, vascular neoplasms, arterial bruits, and venous hums); a patulous eustachian tube; temporomandibular joint clicks or crepitus; palatal myoclonus; and middleear myoclonus [1,2].

BACKGROUND

Tinnitus caused by middle-ear myoclonus is an extremely rare but often very distressing condition, with only few case reports published in the literature [1,3-6]. It is produced by abnormal, repetitive, rhythmical contractions of the middle-ear muscles: the tensor tympani and the stapedius [1]. The tinnitus is usually unilateral and rarely bilateral [3].

Etiology

The etiology of tinnitus produced by middle-ear myoclonus is unknown. Several theories cover a gamut of possibilities—vascular disorder, infection, demyelinating diseases, excessive anxiety, tumor, or trauma [1] but none of these has any substantial scientific support [5]. In most cases, no precipitating factors can be found, and the etiology remains idiopathic [4].

Tensor tympani contraction is said to produce a clicking sound [7], whereas stapedial contraction produces a buzzing [6], a rumbling, or a crackling sound [2]; however, the description of the tinnitus may vary widely among patients. The diagnosis of middle-ear myoclonus is not always evident on otoscopy although, occasionally, rhythmical movements of the tympanic membrane are easily visualized on otoscopic examination. Pure-tone audiometry results usually are normal, and impedance tests may disclose changes in the tympanic membrane's complacency or may capture repeated movements of the drum [3]. Acoustic reflex testing also may demonstrate sustained middle-ear muscle contractions synchronous with the patient's complaint of tinnitus [5]. Marchiando et al. [5] concluded in

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their study that impedance audiometry is the only means by which clinicians should evaluate patients with potential stapedial muscle abnormalities.

Treatment

A variety of options exist for the treatment of middleear myoclonus. Several medical treatments, including the use of muscular relaxants, sedatives, and anticonvulsants, have been described, but study results have been contradictory [3]. Hypnosis, psychotherapy, acupuncture, biofeedback, masking, and blockage of the otic ganglion have all been attempted as treatments, albeit without much success [1,5]. No reports to date have cited the use of botulinum toxin (Botox) for middle-ear myoclonus [3,4].

Several surgical procedures also have been used in the treatment of middle-ear myoclonus. The placement of ventilation tubes was shown to be of no benefit [4]. Surgical sectioning of the tensor tympani and stapedial muscle tendons via tympanotomy is a straightforward and effective procedure having no adverse effects.

CASE REPORTS

We present three cases of bilateral tinnitus caused by middle-ear myoclonus. In two cases, the etiology might have been exposure to intense sudden noise or a stress reaction to this exposure. In one case, the cause could not be determined.

Patient 1

A 14-year-old boy presented with complaints of a continuous clicking tinnitus in both his ears after exposure to a firecracker. The tinnitus had been continuous for 5 months and did not respond to a number of medications, including antispasmodics, muscle relaxants, and anticonvulsive medication. The tinnitus could easily be heard at 10-15 cm distant from the patient's ears. Examination of the tympanic membrane under the operating microscope revealed a clear movement of the membrane in synchrony with the patient's tinnitus. The rest of the physical examination was unremarkable. Puretone audiometry demonstrated normal hearing in both ears. Tympanometry results were normal, but a small cogwheel effect was seen on the recordings of both ears.

As the patient's complaints were not improved by the various treatments, a simultaneous bilateral tympanotomy under general anesthesia was performed, with bilateral sectioning of the tensor tympani and stapedial tendons. Surgery was uneventful and rendered the patient asymptomatic immediately postoperatively, thus confirming the diagnosis of middle-ear myoclonus. After 1 year of follow-up, the tinnitus did not recur, and the patient is completely symptom-free.

Patient 2

A 20-year-old, healthy male soldier was exposed to intense artillery noises during a fierce battle. Immediately after the battle, the patient complained of a continuous noise in both his ears and was referred with the diagnosis of acute phonal trauma.

On physical examination, a continuous low-pitched buzzing noise of a frequency of 90-100 times per minute was easily heard from a distance of 10-20 cm. It emanated from both the patient's ears and was not synchronous with his pulse rate. Otoscopic examination demonstrated rhythmical movement of both the tympanic membranes, which coincided with the tinnitus. The remainder of the ear, nose, and throat examination was normal. Pure-tone audiometry demonstrated a normal hearing threshold in all frequencies. Tympanometry results were normal, but a fine cogwheel effect was seen on the recording, similar to the effect that was seen in patient 1. Direct examination and fiberoptic nasopharyngoscopy showed no signs of palatal myoclonus; thus, the diagnosis of middle-ear myoclonus was made. Medical management with tranquilizers and sedatives was unsuccessful. The patient was so disrupted by his problem that surgery was planned. The patient was operated on under local anesthesia. First the right ear was explored. After exposure of the tympanic cavity through the external auditory canal, the stapes and the tendon of the stapedial muscle were observed under high magnification of the operating microscope. The contraction of the stapedial muscle was found to be synchronous with the tinnitus. The tendon of the muscle was cut, and the tinnitus ceased immediately. This cessation was confirmed by both the patient and the surgeon. No attempt was made additionally to section the tendon of the tensor tympani muscle. The same procedure then was performed on the left ear, with the same findings and results. No complications were encountered during surgery or postoperatively. The patient has been followed up for a 10-month period and has not complained of recurrence of the tinnitus, hyperacusis, or hearing loss.

Patient 3

A 45-year-old woman noted in both ears a clicking tinnitus of more than 1 year's duration, with no change in her hearing. She denied any exposure to intense noise or other precipitating factors. The tinnitus was not alleviated by various medications given to her by her general practitioner, and she did not experience any relief by biofeedback and acupuncture treatments. She was finally referred to our department by her new general practitioner who, while performing a routine otoscopic examination (which had not been done by her former doctor), was able to hear a clicking noise that emanated from both the patient's ears.

The clicking noise was easily heard emanating from the patient's external auditory canals during our otoscopic examination. In addition, rhythmical movements of the tympanic membranes were seen very clearly, synchronous with the patient's complaint of clicking. Fiberoptic nasopharyngoscopy confirmed the absence of palatal myoclonic movements. Pure-tone audiometry demonstrated a mild, bilateral high-tone sensorineural hearing loss. A cogwheel effect, similar to the effect that was seen in the two previous patients, was seen on a type A tympanogram in both ears.

After a diagnosis of middle-ear myoclonus was made, the option of stapedius and tensor tympani tendon section was suggested. A bilateral tympanotomy was performed under local anesthesia, with sectioning of the tendons of the middle-ear muscles. The patient reported complete alleviation of the tinnitus even during the operation. She has been free of any tinnitus to date and has been seen in follow-up for almost 1 year.

DISCUSSION

Objective tinnitus secondary to middle-ear myoclonus is a very rare entity. As early as 1867, Schwartze saw movements of the central and posterior parts of the tympanic membrane in a patient suffering from myoclonus and attributed them to contractions of the tensor tympani muscle [8]. In 1981, Klochoff [9] described the "tensor tympani syndrome" that is caused by increased muscular tonus and various auditory or vestibular symptoms. He identified this as an anxiety-tension response and reported successful treatment with relaxation therapies. However, Klochoff conceded that tenotomy of the tensor tympani may be necessary as an alternative treatment.

Tinnitus produced by stapedial muscle spasm is even much less common than the tinnitus that is produced by tensor tympani myoclonus. Myoclonic contractions of the stapedial muscle were reported as a cause of objective tinnitus by Pulec et al. [10] in 1978. Only a few case reports of tinnitus due to middle-ear myoclonus can be found in the world literature. Watanabe et al. [6] described unilateral tinnitus produced by stapedial muscle contraction in eight patients during the course of recovery from peripheral facial nerve paralysis of various etiologies. The tinnitus occurred whenever a certain facial muscle contracted voluntarily or involuntarily. Marchiando et al. [5] described two cases of tinnitus due to prolonged, spasmodic stapedial muscle contraction, with no concomitant facial nerve disorder.

Badia et al. [4] presented six patients with clinical diagnosis of unilateral middle-ear myoclonus. In one patient, the tinnitus was associated with blepharospasm and was partially improved after botulinum toxin injection into the ipsilateral orbicularis oculi. Three patients were cured by tympanotomy with stapedial and tensor tympani tendon section. Bento et al. [3] described a case of continuous, high-frequency, objective unilateral tinnitus caused by middle-ear myoclonus. The affected patient was treated by sectioning of the stapedial and tensor tympani tendons. Recently, Zipfel et al. [1] presented a case of bilateral middle-ear myoclonus causing an incapacitating tinnitus in a patient with multiple sclerosis; it also was cured with bilateral sectioning of the middle-ear muscles' tendons.

The exact mechanism for the production of objective tinnitus in patients with middle-ear myoclonus is a controversial issue. According to some authors, this type of tinnitus is caused by muscular contraction (muscular sound) of the stapedius or tensor tympani muscles [9]. It could also be due to potential vibration of the tympanic membrane during contraction of the intratympanic muscles, to stimulation of the tympanic plexus, or to alteration of the cochlear microphonic potential [1,4-6].

We have presented three new cases of bilateral objective tinnitus due to middle-ear myoclonus: one case of stapedius muscle myoclonus, one case of tensor tympani myoclonus, and one case of myoclonus of both muscles. In the first two cases (patients 1 and 2), the etiology might have been either exposure to intense sudden noises or a stress and anxiety reaction to this exposure, or both. In patient 3, the cause could not be determined. Both loud noise and anxiety were suggested by some authors as possible causes of middle-ear myoclonus [1-5,9].

These cases are unique in that all three patients had a bilateral tinnitus and in that two patients' complaints started immediately after exposure to sudden, very intense noises. In all the other reported cases in the literature, the patients had long-lasting complaints of tinnitus [1,3–6], as seen in our third patient. In all the other cases reported, except one described by Zipfel et al. [1], tinnitus was unilateral.

CONCLUSION

We described two cases of bilateral objective tinnitus caused by middle-ear myoclonus after exposure to highly intense noises and another case of bilateral objective tinnitus due to middle-ear myoclonus without a known etiology. Surgical section of the stapedial and tensor tympani tendons via a tympanotomy is a straightforward procedure and should be performed if medial therapy fails. This procedure was found to be effective and produced no complications or adverse effects.

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