The Journal of Laryngology and Otology
May 1994, Vol. **108,** pp. 380–382

Management of middle ear myoclonus

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Abstract

Tinnitus produced by synchronous repetitive contraction of the middle ear muscles (middle ear myoclonus) is a rare condition.

We present six cases of middle ear myoclonus in whom different management regimes were successful. In two patients, the tinnitus was controlled by conservative measures. In one patient, whose tinnitus was associated with blepharospasm, significant improvement occurred following botulinum toxin injection into the ipsilateral orbicularis oculi. Three patients were cured by tympanotomy with stapedial and tensor tympani tendon section.

The aetiology of this type of myoclonus remains unclear. The diagnosis is based on the history of involuntary and rhythmic clicking or buzzing tinnitus which is invariably unilateral. The primary differential diagnosis is palatal myoclonus whilst other local aural pathologies must be excluded by careful clinical assessment. Surgical section of these muscles via tympanotomy brings guaranteed relief when conservative measures fail.

Key words: Tinnitus; Myoclonus; Middle ear

Introduction

The tinnitus associated with abnormal contraction of the intratympanic muscles is extremely disturbing to the sufferer. Myoclonus causes a repetitive, usually unilateral, sound which in some cases is audible to the examiner. Characteristically it is described as clicking or buzzing in quality, but varies widely between patients.

In patients without previous episodes of facial nerve palsy, the noise seems to be elicited by exposure to certain sounds. It can be related to hemifacial spasms or eyelid blinking. It also occurs under conditions of excessive anxiety, though in some cases no precipitating factors are found. The aetiology remains obscure, and parallels have been drawn with facial tic due to an uncontrolled contraction of specific facial nerve motor units (De Jong, 1950).

We present six patients with a clinical diagnosis of middle ear myoclonus and discuss the different treatment modalities. The detailed case histories of three patients are representative of the six.

Patients

The main features of the six cases are summarized in Table I. There were three males and three females, and their mean age was 39 years. The mean duration of their tinnitus was four years and none of them had other otovestibular symptoms.

The diagnosis was based on the features of the tinnitus, and in two cases it was audible. The rest of the physical examination excluded palatal myoclonus and a neurological or vascular cause for the tinnitus. We were unable to demonstrate any impedance changes (perhaps due to lack of sensitivity of the apparatus) and pure tone audiometry was unremarkable. Three illustrative cases are described in more detail.

Case 1

A 31-year-old financial analyst was referred for management by a neurological colleague with a diagnosis of palatal myoclonus. She gave a nine-year history of an audible clicking noise and some local discomfort in her right ear. The clicking was intermittent, with no obvious precipitating factors and she was not aware of movement in the back of her throat when her symptom was active. The noise had been continuous, and her husband could hear it even when she was asleep. The periodic clicking tinnitus became extremely annoying whenever she tried to relax. A ventilation tube in the right ear had been inserted in another unit but her tinnitus persisted. There was no history of any systemic illnesses and the patient was on no medication.

General and neurological examination was unremarkable. Otological examination confirmed an audible clicking sound emanating from her right ear. Fibreoptic nasoendoscopy confirmed that there were no palatal myoclonic movements. The right tympanic membrane was normal.

Pure tone audiometry revealed normal hearing

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Accepted for publication: 6 January 1994.

TABLE I

Case	Age	Sex	Symptoms	Precipitating factors	Duration (yrs)	Management	Results
BO	31	F	Clicking (R) ear: aural discomfort	None	9	Stapedius and tensor tympani tendon section	Tinnitus resolved
JD	30	M	Bubbling (L) ear	Low frequency sounds	5	Stapedius and tensor tympani tendon section	Tinnitus resolved
AP	56	M	Crackling (L) ear: like a grasshopper	None	4	Stapedius and tensor tympani tendon section	Tinnitus resolved
LF	61	M	Throbbing (L) ear	Associated with blepharospasm	4	Botulinum toxin injection to orbicularis	Improved 50%
EB	28	F	Tapping (L) ear: aural pressure	High frequency sounds	1½	Conservative sedatives	Partial improvement
MH	30	F	Clicking (R) ear	High frequency sounds	2	Conservative sedatives	Partial improvement

thresholds and middle ear impedance studies showed normal stapedial reflexes and normal decay. We were unable to measure any compliance changes in association with her symptoms.

We concluded that her unilateral objective tinnitus was due to the myoclonus of her stapedius and tensor tympani tendons. A right tympanotomy and section of these tendons was undertaken. At outpatient review four and eight weeks later the patient's clicking tinnitus had resolved completely.

Case 2

A 61-year-old male presented with a five-year history of left hemifacial spasms associated with ipsilateral throbbing tinnitus coinciding with the onset of his spasm. His symptoms were worse when lying down and when he got up in the morning. There were no other otovestibular symptoms. At the time of otological assessment he was waiting to undergo intramuscular botulinum toxin injections to relieve his associated blepharospasm.

On examination, the left tympanic membrane appeared healthy. Pure tone audiometry showed evidence of slight noise induced high tone sensorineural hearing loss but impedance changes with his ocular spasms were not seen. His blepharospasm was treated by repeated injections of botulinum toxin into the orbicularis muscle. Excellent control was achieved and his tinnitus became subjectively quieter by approximately 50 per cent. The option of stapedius and tensor tympani tendon section was suggested, but no further action was taken as the patient was satisfied with the degree of tinnitus improvement.

Case 3

A 28-year-old secretary complained of vibrating tapping noises in her left ear for the past 18 months which were precipitated by various similar frequency sounds. These included telephone dialing tones, fax machines, singing birds and a particular piece of music (Flor de cana). The noises lasted as long as the precipitating sounds and were associated with some preauricular dull ache radiating into the neck which seemed to be due to associated temporomandibular joint dysfunction. There were no associated vestibular symptoms nor any previous history of ear disease. She was otherwise well but admitted that she had been under a lot of stress in her personal life during the previous two years.

On examination her left tympanic membrane was normal and there was no evidence of palatal myoclonus. Impedance audiometry did not demonstrate any middle ear compliance abnormality. Pure tone audiogram and auditory brain stem evoked responses excluded any inner ear disorder with an associated recruitment phenomenon.

The patient was strongly reassured. At review one month later, the tinnitus had not disappeared but was much more tolerable.

Discussion

Tinnitus due to middle ear myoclonus is uncommon and there are few previous reports in the English literature. The differential diagnosis includes intermittent palatal myoclonus or patulous eustachian tube. Some types of eustachian tube dysfunction without middle ear effusion may also occasionally cause confusion. The source of tinnitus in these patients is from contraction of the soft palate or middle ear muscles or undue laxity of the peritubal tensor muscles.

The main differential diagnosis is palatal myoclonus, and indeed this condition was incorrectly suggested by one of the referring clinicians. The tinnitus associated with palatal myoclonus was first described in the C19th by Bigelow and Holmes (Kwee and Struben, 1972); it is usually bilateral and the sound is either due to the snapping open of the eustachian tube (Pulec, 1980) or to the breaking of surface tension as the walls of the eustachian tube open under the action of the peritubal muscles (Pulec and Simonton, 1961). Although the diagnosis is usually readily apparent on oral cavity examination, on occasions opening the mouth may suppress the myoclonus. When this is suspected flexible fibreoptic nasoendoscopy will facilitate recognition. Management with drugs, psychotherapy or surgical procedures aiming to modify palatal function, has proved difficult. Recently, white-noise masking devices have shown favourable results (East and Hazell, 1987). An alterative treatment aimed at abolishing the myoclonic activity by selective injection of the palatal muscles with botulinum toxin has proved useful in three cases (Saeed and Brookes, 1993).

Patulous eustachian tube or abnormal patency of the eustachian tube may give rise to a crackling sensation in the ear, but the most frequent symptom is autophony (O'Connor and Shea, 1981) There may be many aetiological factors, but the most constant is weight loss. Otoscopic examination reveals movements of the tympanic membrane with respiration and changes can also be measured on acoustic impedance.

How middle ear myoclonus produces tinnitus is unknown. Tensor tympani contraction is said to produce a

https://doi.org/10.1017/S0022215100126866 Published online by Cambridge University Press

clicking sound (Pulec, 1980), whereas stapedius muscle contraction produces a buzzing noise (Watanabe *et al.*, 1974). The tinnitus could be due to the propagation of the muscle contraction noise (Kirikae, 1960): this would explain how in some cases it is objective. It could also be due to potential vibration of the tympanic membrane during contraction of the intratympanic muscles; to stimulation of the tympanic plexus (Rosen, 1951); or to alteration of the cochlear microphonic potential.

The tinnitus associated with abnormal contraction of stapedial muscle in patients with facial nerve pathology has been described by several authors: Watanabe *et al.* (1974) have described eight patients with facial paralysis and subjective tinnitus with eyelid closing movements; Marchiando *et al.* (1983) described two cases of tinnitus due to stapedial muscle contraction in patients with no concomitant facial nerve disorder. Stapedial tendon section relieved their symptoms. The cause of the myoclonus in patients recovering from facial nerve paralysis seems to be VIIth nerve regeneration with synkinetic production of stapedial reflex. The cases of stapedial myoclonus correlated with hemifacial spasm are possibly due to compression of the facial nerve in the posterior fossa by aberrant or ectatic blood vessel or to some pathological process involving the facial nerve root close to the brain stem.

In those cases with no facial nerve involvement the aetiology is unclear. Hypersensitive reflex arcs have been described in myoclonus affecting other parts of the body (Swanson *et al.*, 1962). This would be corroborated by the fact that in some patients, the tinnitus was precipitated by certain factors. Klochoff (1981) describes a 'tensor tympani syndrome' due to increased tone in the muscles with impedance fluctuation and a number of variable aural and vestibular symptoms. He identified this is as an anxiety/tension response, and reported successful treatment with relaxation therapists. He admitted that an alternative method of management could be a tensor tympani tenotomy. It was of interest that stress was clearly a significant factor in one of our cases who was suffering from facio-myalgia spasms.

A number of different treatments have been tried. One patient in our series had a right grommet inserted previously which unfortunately made the clicking sounds louder. Management with anti-epileptic drugs and psychotherapy has generally proved ineffective. In our experience, those patients with stress-related tinnitus may benefit from sedatives with muscle relaxant properties, although their tinnitus often does not disappear completely, it stops being troublesome. The use of botulinum toxin for hemifacial spasm and focal blepharospasm has an established role. The toxin gives rise to pre-synaptic neuromuscular blockade by binding to the motor endplate and preventing acetylcholine release. To our knowledge it has not previously been used in the primary treatment of middle ear myoclonus, but the fact that one of our cases with associated blepharospasm gained benefit from the treatment may possibly suggest a future role.

In conclusion, surgical section of the stapedius and tensor tympani tendons via a tympanotomy is a straightforward procedure. In our experience it effectively relieves those cases of intractable tinnitus. We have not encountered any complications from the surgery, nor do there appear to be any measurable changes in functional terms as a consequence of intratympanic muscle division.

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