The Journal of Laryngology & Otology (2013), ***127***, *605–606*.

doi:10.1017/S0022215113000297

CLINICAL RECORD

Transitory stapedial myoclonus in a patient with benign fasciculation syndrome

F BRIGO[1,2], M STORTI[3], P LOCHNER[2], R NARDONE[2,4]

[1]Department of Neurological, Neuropsychological, Morphological and Movement Sciences, Section of Clinical Neurology, and [3]Department of Medicine, Franz Tappeiner Hospital, Merano, Italy, [2]Department of Neurology, Franz Tappeiner Hospital, Merano, Italy, and [4]Department of Neurology, Christian Doppler Clinic, Paracelsus Medical University, Salzburg, Austria

Abstract

Objective: We report a previously undescribed association between transitory stapedial myoclonus, objective tinnitus and benign fasciculation syndrome.

Method: Case report and review of the world literature regarding stapedial myoclonus.

Results: A 30-year-old man with a diagnosis of benign fasciculation syndrome abruptly developed severe, low-pitched tinnitus on the right side. Otoscopic examination revealed rhythmic movement of the tympanic membrane, which was synchronous with the tinnitus. No palatal spasm was noted on nasopharyngeal examination. Brain magnetic resonance imaging and pure tone audiometry were unremarkable. Based on these findings, a diagnosis of objective tinnitus due to stapedial myoclonus was made. The objective tinnitus spontaneously disappeared within 48 hours of its appearance, but in the following days the patient suffered frequent, brief episodes of objective tinnitus lasting only a few seconds.

Conclusion: The occurrence of stapedial myoclonus in this patient indicated the presence of an underlying motor unit hyper-excitability. This case suggests that, in some patients, stapedial myoclonus may represent the clinical expression of diffuse motor unit hyper-excitability.

Key words: Stapedius; Ear, Middle; Myoclonus; Tinnitus; Fasciculation

Introduction

Tinnitus is defined as the perception of sound in the absence of corresponding external stimulus, and may be classified as subjective or objective. Subjective tinnitus is characterised by an individual's perception of sound in the absence of any physical source, whereas objective tinnitus refers to a sound that is produced mechanically within the head.

Stapedial myoclonus is a rhythmic movement of the tympanic membrane secondary to repetitive contraction of the tensor tympani and stapedial muscles, and represents a segmental myoclonus. It is a rare cause of objective tinnitus,[1] with only a few cases reported in the literature.

We describe a previously unreported case of objective tinnitus caused by transitory stapedial myoclonus in a patient diagnosed with benign fasciculation syndrome.

Case report

A 30-year-old man presented with severe, low-pitched tinnitus on the right side which had developed suddenly while he was sitting down watching television. His previous medical history was notable for benign fasciculation syndrome. He did not take any drugs. The tinnitus was described as an unpleasant, buzzing noise resembling that experienced during aircraft landing, but occurring in a rhythmic fashion, dozens of times a minute. There was no pressure in the left ear nor any change in hearing status. The patient denied any precipitating factors, although performing the Valsalva manoeuvre and placing pressure on the tragus were reported to slightly increase the frequency and intensity of tinnitus.

On physical examination, the tympanic cavity was clear. Otoscopic examination revealed rhythmic movement of the tympanic membrane, which was synchronous with the tinnitus. No palatal spasm was noted on nasopharyngeal examination.

Brain magnetic resonance imaging and pure tone audiometry were normal.

Based on these findings, a diagnosis of objective tinnitus due to stapedial myoclonus was made.

Electromyography showed diffuse fasciculation potentials with normal motor unit recruitment, without features indicative of chronic denervation (i.e. neurogenic enlarged motor unit potentials) and/or active denervation (i.e. fibrillation and positive, sharp waves).

The patient's objective tinnitus spontaneously disappeared within 48 hours of its appearance. In the following days, he suffered frequent, brief episodes of objective tinnitus lasting only a few seconds.

Discussion

Stapedial myoclonus is a rhythmic movement of the tympanic membrane caused by repetitive contraction of the tensor

Accepted for publication 28 June 2012 First published online 12 March 2013

tympani and stapedial muscles. It represents a segmental myoclonus, and is a rare cause of objective tinnitus, with only a few cases reported.

A convincing proposal for a pathogenetic mechanism of this condition is still lacking. Furthermore, there are no previously published reports of stapedial myoclonus associated with benign fasciculation syndrome.

Fasciculations are random muscle twitches which can be observed clinically, and are associated with electrical events recorded by a needle electrode as fasciculation potentials. A fasciculation potential therefore represents the spontaneous discharge of a motor unit or part thereof. Fasciculation potentials are seen in many pathological conditions (e.g. peripheral neuropathy, radiculopathy, peripheral nerve hyper-excitability syndromes and, especially, motor neuron diseases).[2]

Conversely, fasciculation potentials may occur in otherwise healthy subjects.[3] This phenomenon, termed benign fasciculation syndrome, is a non-progressive, chronic condition characterised by fasciculation, usually in the lower limbs (predominantly the distal leg muscles), which is not associated with any other clinical abnormality and which, unlike motor neuron diseases, has an excellent long-term prognosis.[3]

The origin of benign fasciculation potentials is unknown. It has been suggested that they may arise by ephaptic transmission from irregularly fibrillating fibres,[4] although a more proximal origin has also been suggested.[5]

To the best of our knowledge, the presented patient represents the first report of co-occurrence of stapedial myoclonus and benign fasciculation syndrome. The coexistence of a rare otological disorder such as stapedial myoclonus together with benign fasciculation syndrome could simply be a coincidence. However, the possibility of a non-coincidental association should also be considered, and we present below some considerations regarding a possible pathogenetic relationship.

- **Stapedial myoclonus is rhythmic tympanic membrane movement due to repetitive tensor tympani and stapedial contraction**
- **It represents a segmental myoclonus, and is a rare cause of objective tinnitus**
- **The presented patient had stapedial myoclonus plus benign fasciculation syndrome**
- **This suggests the presence of underlying motor unit hyper-excitability**
- **Some cases of stapedial myoclonus may be due to diffuse motor unit hyper-excitability**

It is noteworthy to consider that stapedial myoclonus has been quite frequently described during the course of recovery from peripheral facial nerve paralysis,[6] a condition in which muscle synkinesis is thought to occur (due to abnormal reinnervation of facial muscles following denervation, or to ephaptic transmission leading to formation of an 'artificial synapse', an ephapse, at the site of nerve injury). Furthermore, antiepileptic drugs and botulinum toxin administration have been proposed as treatment for this disorder.[7] Interestingly, both drugs reduce neuromuscular transmission, either by stabilising the muscle membrane and reducing neuronal firing (in the case of anticonvulsants) or by blocking acetylcholine release at the neuromuscular junction (in the case of botulinum toxin). The occurrence of stapedial myoclonus in our patient suggests the presence of an underlying motor unit hyper-excitability. It is worth considering that, in our patient, both the Valsalva manoeuvre and pressure on the tragus were reported to slightly worsen his tinnitus. Since fasciculations may be elicited by muscle percussion, it can be hypothesised that increased tension of the tympanic membrane, leading to stapedius muscle stretching, may mechanically further increase the local hyper-excitability.

Conclusion

The presented case suggests that, at least in some patients, stapedial myoclonus may represent a clinical expression of a functional hyper-excitability of a motor unit or part thereof.

References

1 Zipfel TE, Kaza SR, Greene JS. Middle-ear myoclonus. *J Laryngol Otol* 2000;**114**:207–9
2 Mills KR. Characteristics of fasciculations in amyotrophic lateral sclerosis and the benign fasciculation syndrome. *Brain* 2010;**133**:3458–69
3 Mitsikostas DD, Karandreas N, Coutsopetras P, Piperos P, Lygidakis C, Papageorgiou C. Fasciculation potentials in healthy people. *Muscle Nerve* 1998;**21**:533–5
4 Janko M, Trontelj JV, Gersak K. Fasciculations in motor neuron disease: discharge rate reflects extent and recency of collateral sprouting. *J Neurol Neurosurg Psychiatry* 1989;**52**:1375–81
5 Sindermann F, Conrad B, Jacobi HM, Prochazka VJ. Unusual properties of repetitive fasciculations. *Electroencephalogr Clin Neurophysiol* 1973;**35**:173–9
6 Watanabe I, Kumagami H, Tsuda Y. Tinnitus due to abnormal contraction of stapedial muscle. An abnormal phenomenon in the course of facial nerve paralysis and its audiological significance. *ORL J Otorhinolaryngol Relat Spec* 1974;**36**:217–26
7 Badia L, Parikh A, Brookes GB. Management of middle ear myoclonus. *J Laryngol Otol* 1994;**108**:380–2

Address for correspondence:
Dr Francesco Brigo,
Department of Neurological,
Neuropsychological,
Morphological and Movement Sciences,
Section of Clinical Neurology,
University of Verona,
Piazzale LA Scuro,
10 - 37134 Verona, Italy

Fax: +39 0458124873
E-mail: dr.francescobrigo@gmail.com

Dr F Brigo takes responsibility for the integrity of the content of the paper
Competing interests: None declared

https://doi.org/10.1017/S0022215113000297 Published online by Cambridge University Press