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Bilateral objective tinnitus secondary to congenital middle-ear myoclonus

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Abstract
Subjective tinnitus (heard only by the patient) is a common otological complaint. Objective tinnitus (heard by the examiner as well as the patient) is extremely rare. There are only a few cases of objective tinnitus, secondary to middle-ear myoclonus, described in the literature.

We present the case of a child with bilateral, congenital, objective tinnitus, secondary to middle-ear myoclonus, with otherwise normal hearing thresholds (250Hz-8kHz), and with no evidence of intra-cerebral or systemic disorders. No similar case has been reported in the world literature.

Key Words: Tinnitus; Myoclonus; Middle Ear; Paediatric

Case report

The patient was originally referred aged six years by his general practitioner (GP) to the ENT department as his parents expressed some concern that he was not hearing as well as he might. He was not complaining of any pain but did report bilateral 'clicking' in his ears, for as long as he could remember. His hearing was deemed satisfactory and the diagnosis of 'very mild eustachian tube dysfunction' was made. The patient was discharged.

Four years later the patient, now 11 years old, was referred to the audiologist by a new GP, with 'bilateral audible clicking', which, though ignored most of the time, proved 'an intermittent source of irritation'.

The patient was seen in the ENT out-patients clinic by the consultant, with the above history. On examination, bilateral audible clicks could be heard by the examiner. Endoscopic examination of the soft palate by naso-endoscopy excluded palatal myoclonus (as no soft palate movements were observed synchronised with audible clicks) and no vascular or other neurological cause could be identified. The clicking noises were present continuously and no trigger or exacerbating factors could be identified. The 'clicks' were demonstrated on the tympanometry traces originally at age six and again at age 11 (Figure 1). Pure-tone audiograms showed normal hearing on both occasions (Figure 2). A 10 second recording of compliance revealed regular 'blips' in the trace (Figure 3), which were synchronised with the audible clicking. By direct observation, by otoscopy, the tympanic membrane could be seen to be moving in synchrony with the audible clicks. This confirmed involvement of the tensor tympani muscle. Myoclonus of the stapedius tendon would not cause visible movement of the tympanic membrane because of the way the incu-stapedial joint articulates.

It was decided, in view of the bilateral nature of the problem, and the fact that the patient had essentially normal ears and was little troubled by his symptoms, that intervention was inappropriate at this stage. Indeed his parents declined consent for any surgical procedure. He has now been under review for over 12 months and the problem is still of little significance to his day-to-day life. At present no further action is planned.

Discussion

Objective tinnitus due to middle-ear myoclonus is rare.[1] No previous paediatric cases are reported in the English literature. A paediatric case previously reported in the German literature, had an associated bilateral 40dB sensorineural hearing loss.[2] There was no such hearing loss in this case.

How middle-ear myoclonus produces the tinnitus is not fully understood. It is suggested that the tensor tympani and stapedius muscles undergo repetitive, abnormal contraction - thus leading to rhythmical movement of the tympanic membrane.[3] Tensor tympani contraction is said to produce a clicking sound,[4] as in the described case, whereas stapedius muscle contraction is said to produce a buzzing sound (Watanabe, 1974).[5,6]

These myoclonal movements are a form of segmental myoclonus and involve brainstem innervated muscles. The myoclonus is most often unilateral.[7]

Vascular, infectious and demyelinating disorders as well as anxiety, trauma and neoplastic disease have all been implicated in the aetiology of segmental myoclonus.[7] Idiopathic cases are also reported where no precipitating cause could be identified.[6] Other suggested mechanisms include stimulation of the tympanic plexus and alteration of the cochlear microphonic potential.[1]

The tympanograms appear to have a classical 'cogwheeling effect' (Figure 1) as previously described.[7]

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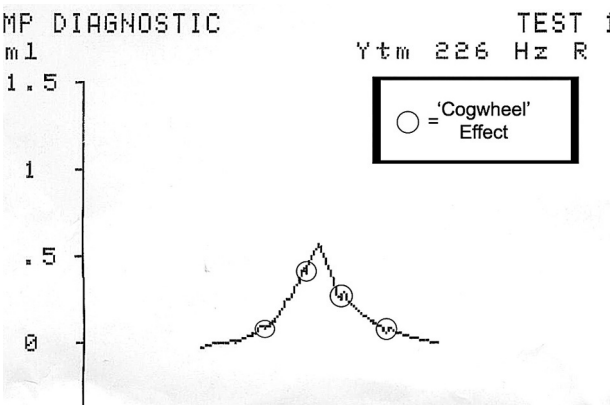

FIG.1
Tympanogram, age 11.

However, it is difficult to distinguish this from movement artefact in such a young subject. Figure 3 demonstrates a repeatable movement of the tympanic membrane, below the threshold for acoustic stimulation of the stapedial reflex. This is strongly indicative of a repetitive rhythmical middle-ear myoclonus.

The main differential diagnosis of objective tinnitus is palatal myoclonus. In this case the tinnitus is usually bilateral. The diagnosis is usually made by direct intra-oral examination or by flexible naso-endoscopy.[3] It is thought that the noise is produced by the snapping open of the eustachian tube, or secondary to peritubular muscles, causing a break in surface tension as the walls of the eustachian tube pull open.[8]

A number of treatments, both medical and surgical, have been advocated for middle-ear myoclonus. Muscle relaxants, benzodiazepines and anticonvulsants have been used with differing degrees of success.[7] Other suggested medical treatments have included hypnosis, psychotherapy, acupuncture, biofeedback, masking and otic ganglion blockade. These have been of limited success.[3]

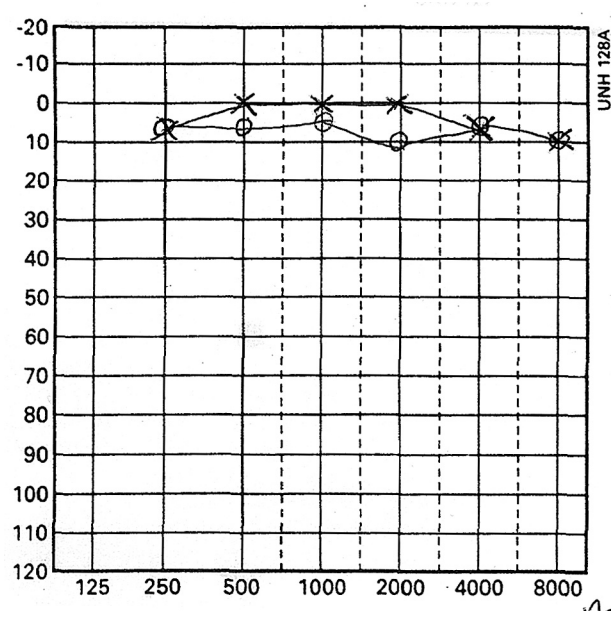

FIG. 2
Pure-tone audiogram, age 11.

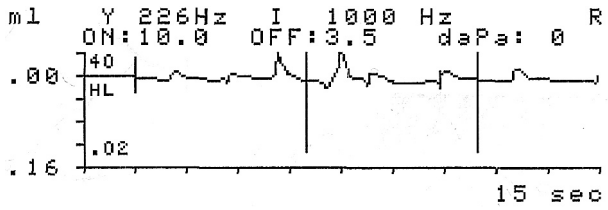

FIG. 3
Rhythmical contractions of the tympanic membrane.

The use of botulinum toxin has been described, although this was in a case with associated focal blepharospasm.[6] Its role in the primary treatment of middle-ear myoclonus is yet to be established but its effectiveness for palatal myoclonus is, however, well documented.[9]

The mainstay of surgical treatment is the surgical sectioning of the stapedius and tensor tympani tendons via a tympanotomy. This has proved effective where medical treatments have failed,[3,6] and no adverse effects have yet been encountered or reported.[3]

Conclusion

We describe what appears to be the first reported paediatric case of an objective, bilateral tinnitus, secondary to middle-ear myoclonus involving the tensor tympani muscle. The child had normal hearing and no known intracerebral or systemic pathology.

There are recognised medical and surgical options available to treat this potentially distressing condition. At present, the degree of discomfort caused by the condition does not justify intervention in this child.

- **Objective tinnitus secondary to middle-ear myoclonus is extremely rare and the mechanism is poorly understood. It is often associated with intracranial or systemic disease**
- **Congenital middle-ear myoclonus is bilateral**
- **The outcome of medical treatment is variable, but effective surgical treatment options are available**

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