Middle Ear Myoclonus in a Pediatric Patient: A Case Report

Jennifer Dang, MD1; Katherine Dunsky, MD1,2; Robert A. Williamson, MD1,2; Yi-Chun Carol Liu, MD1,2
1Baylor College of Medicine, Department of Otolaryngology – Head and Neck Surgery, Houston, Texas
2Texas Children’s Hospital, Houston, Texas

Abstract

Objective: To describe a case of bilateral middle ear myoclonus (MEM) that was treated with tensor tympani and stapedial tendon tenotomies.

Methods: A case report and review of the literature.

Results: An 8-year old healthy girl presented with clicking sound bilaterally, more bothersome on the left than the right. She denied any triggering events or exacerbating factors. She complained of this long-standing tinnitus throughout childhood, and reports an inability to focus in school. It impaired her ability to concentrate. She believed that others could also hear this sound. She otherwise has no hearing loss, vertigo or otalgia.

During examination, a continuous, clicking sound could be heard from a distance of 5-10 cm and emanated from the patient’s bilateral external auditory canals. The remainder of the head and neck exam was normal. The soft palate did not exhibit myoclonus. Neck palpation did not reveal a mass. Tinnitus remained unchanged during compression of the internal jugular vein or head rotation, and it was not synchronous with her pulse. She had no neurologic examination findings; she did not have any facial palsy, hemifacial spasm or blepharospasm. She had no history of pain, joint tenderness, crepitus or painful clicking of the temporomandibular joint upon opening or closing of the mouth.

She underwent further workup with further testing. Her temporal bone computed tomography, and magnetic resonance imaging were unremarkable. Audiometry showed no hearing loss. Impedance testing did not disclose any significant change in membrane compliance or capture movements of the membrane. Based on all these findings, a diagnosis of MEM was established. Therapeutic options including pharmacologic treatment and surgery were discussed with the patient and her parents. Her parents did not wish to pursue conservative medical therapy with anticonvulsant or muscle relaxants. The patient was offered middle ear tendon sectioning.

Introduction

Tinnitus can be classified into subjective or objective tinnitus. Subjective tinnitus occurs more commonly, and it is characterized by abnormal, spontaneous activity within the auditory pathways, and is the phantom perception of a ringing or buzzing sound in the ears in the absence of an external sound source. It is a sensation only heard by the patient. In contrast, objective tinnitus is a sound originating from the patient’s auditory canal or adjacent structures and can be heard by the patient and the observer. Conditions that can cause objective tinnitus include vascular abnormalities, temporomandibular joint disease, a patulous eustachian tube, and tinnitus of muscle origin, specifically palatal myoclonus or middle ear myoclonus (MEM).

Myoclonus is a sudden, involuntary jerking of a muscle. MEM is a rare disorder produced by repetitive contractions of the middle ear muscles, and is defined as a rhythmic movement of the tympanic membrane secondary to repetitive contraction of the tensor tympani or stapedial muscles. The tinnitus can be described as clicking or buzzing, but can widely vary in between patients. MEM is an extremely rare disease, and only a few English-language case reports have been published. Furthermore, it is even more rare in the pediatric population. We describe a case of bilateral MEM that was treated with tensor tympani and stapedial tendon tenotomies.

Case Report

An 8 year old healthy girl complained of a continuous, clicking sound bilaterally, more bothersome on the left than the right. She denied any triggering events or exacerbating factors. She complained of this long-standing tinnitus throughout childhood, and reports an inability to focus in school. It impaired her ability to concentrate. She believed that others could also hear this sound. She otherwise has no hearing loss, vertigo or otalgia.

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Discussion

There is also associated demonstration of impedance changes on long-time based tympanometry; the tympanograms may have a classical "cogwheel effect." However, this can sometimes be difficult to distinguish from movement artifact in young patients (Howsam). Ultimately, MEM is a diagnosis of exclusion.

How MEM produces an auditory sensation is unknown. The tinnitus could be due to potential vibration of the tympanic membrane during contraction of the intra-tympanic muscles. Differentiating the diagnosis of ST myoclonus from that of TT myoclonus has been sparsely described in the literature, and at present, there are no methods of separating one from another.

MEM is very rare, and very few English literature reports of it have been published. The literature consists of mainly case reports and case series, and moreover, these are mostly adult cases. In the largest recent case series by Park et al, the mean age of the patient population was 29.8. MEM is even more rare in the pediatric population, with the literature only citing 34 cases, 12 of which were under the age of 10 years (Park). The first pediatric case report in the English literature was by Howsam in 2005.

Previous case reports have described several treatments, both medical and surgical. Presently, there is a lack of consensus regarding treatment. Medical therapy includes the use of muscle relaxants (baclofen), benzodiazepines (clonazepam), or anticonvulsants (carbamazepine). These have been used with differing degrees of success. Patients are typically advised to start with 6 months of medical therapy before considering a surgical procedure. For intractable cases after conservative treatment, both middle ear tendons are sectioned; this seems to be a very promising treatment. Bimrao et al reported several studies with effectiveness after surgery; however, the effectiveness was mostly described in adults. Several reports have indicated these procedures have no adverse effects; there is a theoretical risk that myotony of the stapedius muscle would invariably lead to hyperacusis. Therefore, there is further need for strategies for targeted tenotomy.

Conclusions

Middle ear myoclonus is a very rare and potentially disabling condition in the pediatric population, with unique characteristics upon presentation. Diagnosis of MEM is based on history, clinical exam and long-time based tympanometry.

For patients who do not receive improvement with or decline medical treatment, sectioning of the tendons of the middle ear muscles is a safe and reasonable treatment. Future studies are needed to further elicit the pathophysiology and long-term outcome of medical and surgical therapy.

References