



Case Report

Essential Palatal Myoclonus and Clicking Tinnitus in a Nine-Year-Old Boy—A Case Report

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INTRODUCTION

Objective tinnitus (OT) is a sound emitted by the ear and/or its surrounding structures, which can be heard by both the patient and the examiner and can produce physical damage and emotional distress to patients. OT has two identifiable sources: blood vessels and muscles. Myogenic tinnitus can be caused by rhythmic contractions of any muscle near the ear and throat. It has been divided into two separate entities: middle ear myoclonus (MEM) and palatine myoclonus (PM). When we encounter patients with inexplicable tinnitus in clinical practice, we should be careful in differential diagnosis to avoid any misdiagnosis.

Tinnitus caused by PM is uncommon and rarely reported in the literature. This disorder has been described in adults but was rarely found in children. Deuschl et al.¹ made a meta-analysis of 77 patients with PT and found an even sex distribution, the age distribution had a mean of 24.8 ± 12.9 years. Various possible etiologies of PT have been described, including brainstem lesions, stroke, olive gland hypertrophy, Alexander's disease, or other congenital brain malformations. It also can be idiopathic without any etiology to be identified, but this is rare. We are going to introduce a case of PM that occurred in a nine-year-old boy, who was then treated by

botulinum toxin injections. Finally, this case report discusses common questions about myoclonic-induced OT clicking tinnitus and provides answers. Our report is intended to establish future diagnostic and treatment guidelines.

CASE REPORT

Our patient is a nine-year-old boy, who had a one-year history of a click noise in his left ear. The sound began spontaneously with no predisposing factor; it existed persistently and could be audible by others at a 0.5 m distance. The noise, even annoying during his sleep at night, made the boy so miserable that he even hit his ears trying to relieve the stress. He denied ear pain, hearing loss, or other symptoms. He had normal growth and development without head injury or acoustic trauma before. Of his family members or close relatives, no one suffered from ear problems, hearing loss, or tinnitus. He had no problem with blood count, thyroid tests, liver, and renal function. The results of tympanometry and audiometry were also normal. Otoscopy revealed a normal ear canal and eardrum in each ear. The click noise was measured in the external auditory canal with 77 dB SPL or 0.141 Pa peak value (Fig 1), using an electret microphone (KE4, Sennheiser, Germany). The Valsalva maneuver had no effect on the tinnitus.

With the hypothesis of a MEM, the patient underwent tympanoscopy, and botulinum was applied locally to the middle ear muscles. The tinnitus was observed to stop immediately during surgery but reappeared and continued after surgery.

After that, we did a magnetic resonance imaging (MRI) of the internal auditory canals to rule out the cause of central lesions. On palpation of the soft palate and lateral pharynx, a non-obvious, very discrete, locally circumscribed myoclonus of the lateral soft palate could be palpated. Rigid nasal endoscopy revealed rhythmic contraction of the left eustachian tube in synchrony with the patient reporting sound in his ear. The patient's symptoms of soft palate movement, eustachian tube

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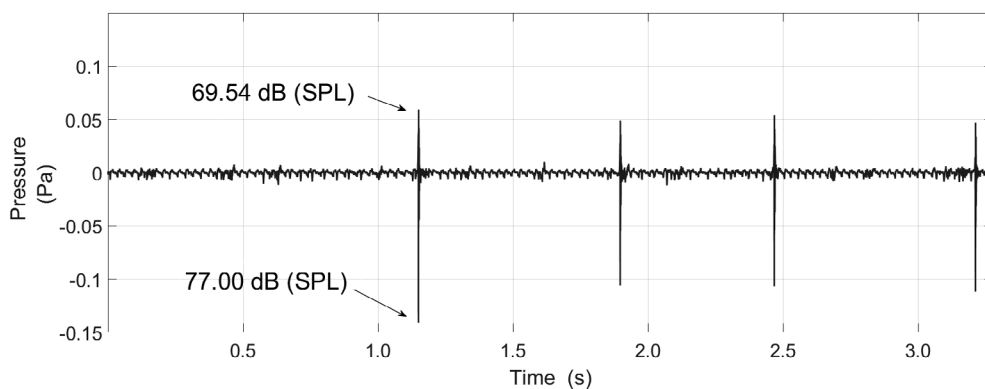


Fig. 1. Objective tinnitus measured by a microphone as sound pressure level (Pa) in the external auditory canal.

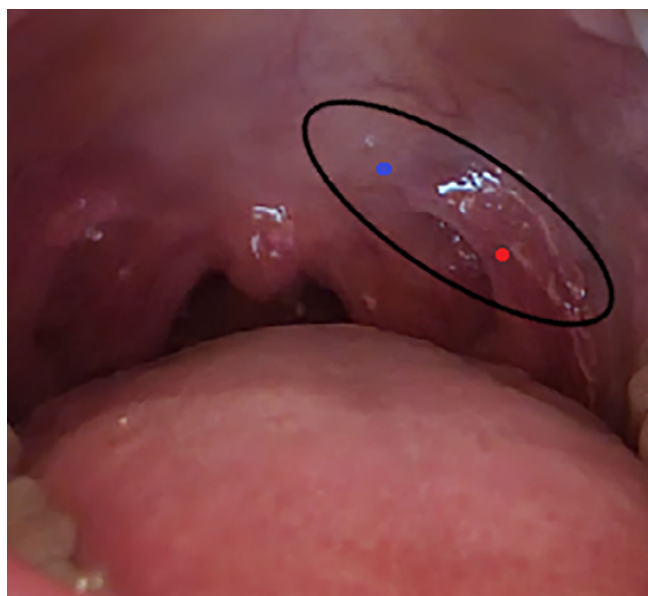


Fig. 2. Injection site of botulinum toxin (7.5 units) injected (red dot) just medial to the pterygoid hamulus (inside black ellipse) and 3 units of botulinum toxin injected just lateral to the musculus uvula (blue dot). [Color figure can be viewed in the online issue, which is available at www.laryngoscope.com.]

contractions, and OT coincided temporally. We changed the diagnosis to PM and took botulinum toxin (Botox, Allergan) injection in the area of the lateral soft palate (Fig 2). It was done in narcosis because he did not take it well in local anesthesia. There occurred slight difficulties with swallowing developed after the injection but resolved completely after one week. His symptoms diminished in the next few days, until the writing of this text, the patient has been free of symptoms for three months. However recently he mentioned the noise was back at a low volume but it would not affect his life.

DISCUSSION

The OT caused by nearby muscle contraction, referred to as myoclonus-induced OT, was separated into two separate entities: MEM and PM. The differentiation

of the MOM versus PM determines the further therapeutic procedure. Careful oral examination is essential to identify a PM and should always include palpation, if necessary also transnasal flexible endoscopy, because a wide mouth opening can lead to the PM interruption or a discrete peritubal myoclonus is easily overlooked. Acoustic impedance changes and reflex decay in audiometric diagnostics can be useful for the MOM. The therapeutic procedure for MOM includes cutting the tendons of the tensor tympani/stapedius muscle.²

Our patient's only chief complaint was tinnitus. The neurological examination and brain MRI had no abnormalities detected, consistent with essential palatal tremors (EPT). For primary palatal tremors, there is no clear treatment report. Anticonvulsants, piracetam, and psychotherapy all had no success in suppressing the tinnitus. Botulinum toxin, which is produced by the anaerobic bacterium *Clostridium botulinum*, causes temporary paralysis of muscles by blocking the presynaptic release of acetylcholine at the neuromuscular junction. In the clinic, it has been used to treat excessive muscle activity in different anatomical regions. Recently, various new indications for botulinum toxin therapy, including essential palatal tremor, have been reported.³ As far as we know, botulinum toxin has been used in a small number of patients with EPT, and Table I lists the characteristic features of previous cases. Penney et al.⁴ reported five cases of palatal tremor tinnitus treated by botulinum toxin and pointed out botulinum toxin has been proven to be a safe effective treatment for many neuromuscular disorders, and it should be considered as a first-line treatment for patients with palatal tremor.

CONCLUSION

Currently, the etiology and pathophysiology of EPT have not been sufficiently elucidated and further studies are needed.⁵ When injected in an optimally diagnosed patient suffering from objective tinnitus, because of EPT, botulinum toxin appears to be an effective treatment with few side effects and complications. In our opinion, it should be considered as a first-line treatment for patients with palatal fibrillation.

TABLE I.
Characteristic Features of Previous PM Cases.

	Penney et al. ⁴	Penney et al. ⁴	Carman et al. ⁵	Krause et al. ³	Krause et al. ³
Age	26	42	9	10	6
Sex	Female	Male	Male	Female	Male
EPT side	Bilateral	Bilateral	Left	Bilateral	Bilateral
Symptom duration	1 year	14 years	2 years	1 year	1.5 years
Rate	NR	NR	160/min	90/min	85/min
Family history of otologic diseases	NR	NR	No	NR	No
BT type	Dysport (Ipsen)	Dysport (Ipsen)	BOTOX® (Allergan)	Dysport (Ipsen)	Dysport (Ipsen)
BT total dose	5 U	15 U bilateral TVP + LVP	5 U left TVP + LVP	30 MU left TVP	1.3 MU right TVP 2.5 MU left+ right TVP
Side effects	2/52 mild VPI	No	1/52 mild nasal regurgitation	NR	NR
Symptom relief	Improved	Improved	Resolved	Resolved	Resolved

LVP = levator veli palatini; NR = not reported; TVP = tensor veli palatini; VPI = velopharyngeal insufficiency.

Though we identified the cause of the objective tinnitus as a palative myoclonus, it is not excluded that the measured acoustic signal is similarly present in the middle ear muscle-caused myoclonus. A limitation of our study is the expected ambiguity of the measured sound pressure in the ear canal. A similar objective tinnitus might be caused by activities of the musculus stapedius and/or musculus tensor tympany with similar sound pressures in the ear canal. Therefore, the measured acoustic correlation is not a unique indication of palatal myoclonus.

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