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The etiology, pathogeneses, and treatment of objective tinnitus: Unique case series and literature review

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ABSTRACT

We present two unique cases of myoclonus-induced objective tinnitus (OT), along with a comprehensive literature review on the topic. Primary goals include: explore the relationship between palatal myoclonus (PM) and middle ear myoclonus (MEM), highlight the embryologic, neurologic, and anatomical relationship between the involved peri-tubular muscles, exemplify the first case of OT which documents video evidence demonstrating the link between objective tinnitus and eustachian tube movement. Also, we discuss available treatment interventions and why they often do not fully resolve patients' symptoms. Finally we introduce a novel way to objectively quantify the severity of OT. Ultimately, our series hopes to inform future diagnostic and treatment guidelines.

1. Introduction

Objective tinnitus (OT), sounds produced by the ear and/or its surrounding structures, is audible to both the patient and examiner and can be both physically impairing and mentally distressing for patients [1]. OT caused by nearby muscle contraction, referred to as myoclonus-induced OT, is a well-known phenomenon and has historically been separated into two separate entities: middle ear myoclonus (MEM) and palatal myoclonus (PM) [2].

Although the topic of OT related to myoclonus has been explored extensively within the otolaryngologic literature, the relationship of MEM and PM to OT still remains obscure [2]. We present two unique cases of myoclonus-induced OT. The first case highlights a never-before-reported presentation of PM induced OT, in which the myoclonus and resulting tinnitus is elicited by palpation of various superficial skeletal muscles. The second case provides evidence that OT in PM can be attributed to sound emanating from myoclonic peritubular muscles causing the eustachian tube (ET) to open and close. Our findings support the hypothesis that PM and MEM are, in many cases, related diseases; this should be considered when contemplating optimal diagnosis and treatment. Moreover, our second case introduces a new way to quantify the degree of OT using audiological testing. Finally, this report explores the common questions regarding myoclonus-induced OT and offers insight into their answers.

2. Case report

2.1. Case #1

A 38 year-old male with a past medical history of ET dysfunction presented with right-sided tinnitus described as a clicking noise. Under direct visualization using a flexible fiberoptic scope palatal myoclonus was visualized with palpation of his skeletal muscles, including the sternocleidomastoid, trapezius, pectoralis major, brachioradialis, and rectus femoris. Moreover, the patient's palatal myoclonus coincided directly with complaints of objective tinnitus. When sitting upright and still, patient was completely asymptomatic, and no movement of his palate was observed. There was not a noticeable change in the severity of the patient's symptoms with varying degrees of palpation or with specific muscles.

2.2. Case #2

A 59 year old female with unremarkable past medical history presented to clinic with five years of “crunching sound” in her right ear. She had previously undergone placement of a right ear tube for presumed “ET dysfunction,” which did not alleviate her symptoms. Rigid nasal endoscopy revealed rhythmic contraction of the right ET in synchrony with the patient reporting sound in her ear (Video 1). Direct visualization of the palate revealed intermittent myoclonus. The patient's symptoms of soft palate movement, eustachian tube contractions, and OT coincided temporally. The remainder of her physical exam was

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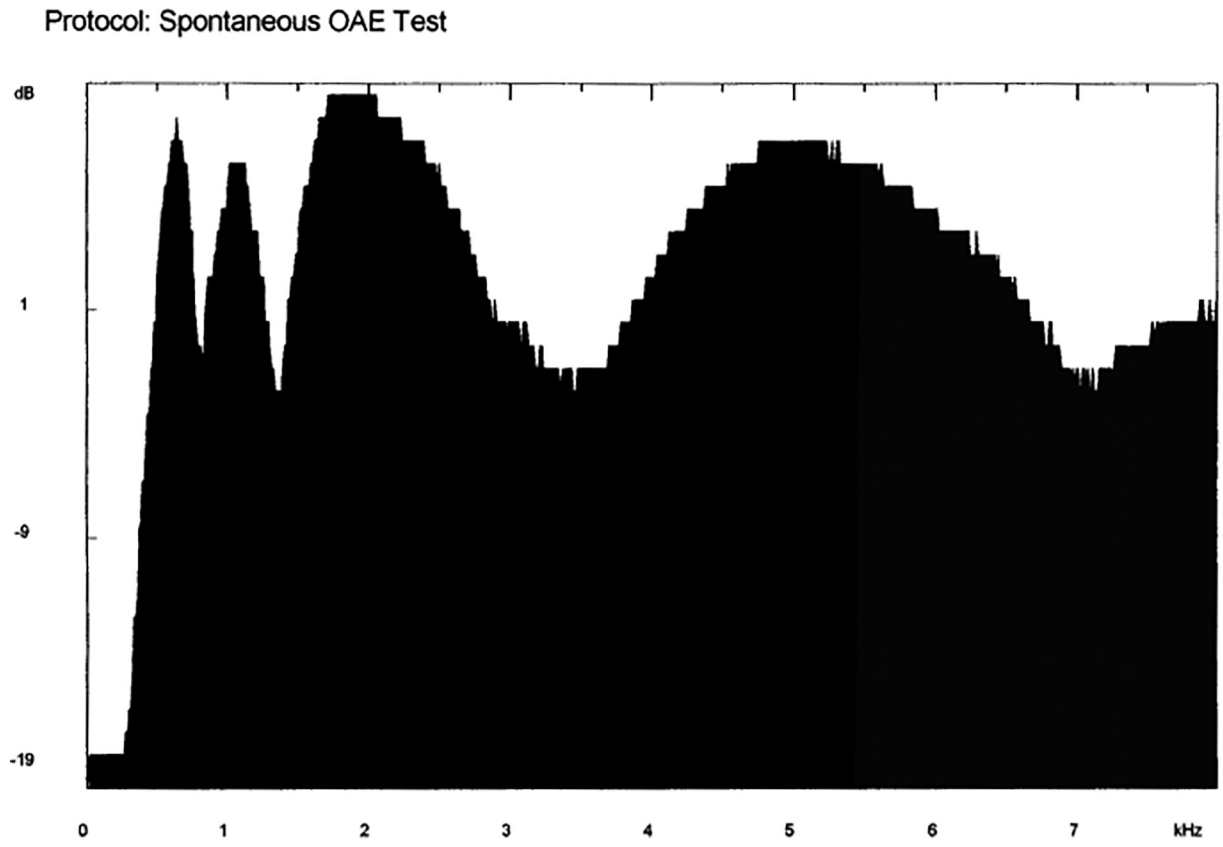

Fig. 1. Spontaneous OAE test, right ear.

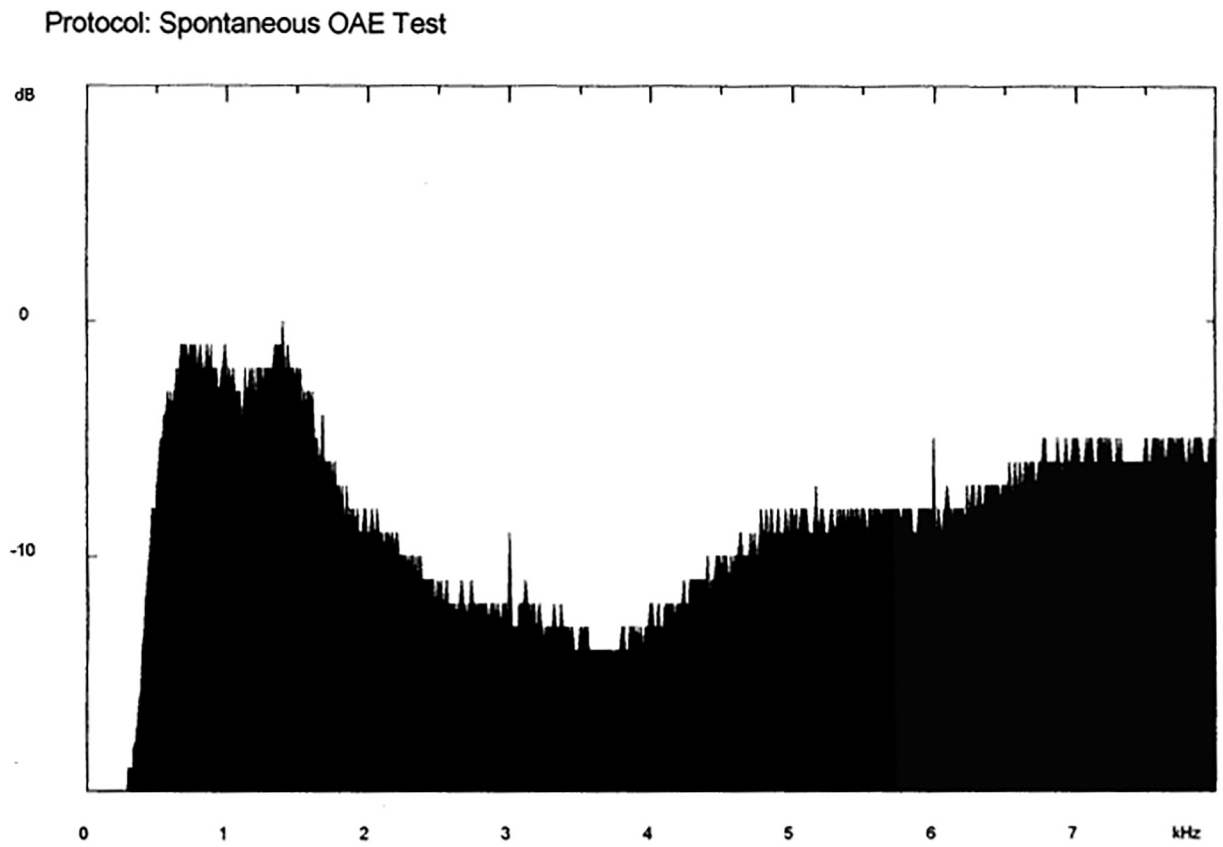

Fig. 2. Spontaneous OAE test, left ear.

unremarkable. An MRI of the internal auditory canals with/without contrast was unremarkable. A diagnosis of middle ear myoclonus was made. An audiology evaluation was remarkable for large emission responses noted during spontaneous otoacoustic emission (SOAE) testing of the right ear (Fig. 1)–the SOAE testing in the left ear is shown for comparison (Fig. 2). These asymmetric emission responses were noted in the affected ear and coincided temporally with patient's objective tinnitus. Steady state tympanometry was normal bilaterally. Several months later patient underwent section of the tensor tympani tendon. At her four-month post-operative visit, the frequency and severity of the patient's symptoms had decreased, however patient continued to experience symptoms of OT.

3. Discussion

While not the sole causes of objective tinnitus, both palatal myoclonus and middle ear myoclonus have both been implicated in objective tinnitus patients. The exact etiology and optimal treatment of these diseases remains unclear.

In both of our cases, we were able to directly visualize palatal myoclonus and verify that the myoclonus temporally coincided with the OT, further agreeing with the literature that these two entities are likely linked [2]. Anatomically, the soft palate is thought to be linked to the ET via the tensor veli palatine (TVP) muscle, which partially arises from the cartilaginous lateral wall of the ET and inserts onto the palate [2]. As a result, a clonus in the TVP may cause the lateral wall of the ET to collide with the medial wall, which we were directly able to observe with an endoscope, causing an audible noise. Indeed, Pulec et al. found that patients reported tinnitus immediately after onset of electrical activity in the region of ET walls [3]. It is important to note that the literature has historically considered MEM to be a separate cause of OT, mediated by a clonus of the tensor tympani muscle (TTM). In this mechanism, the TTM muscle (originating within the cartilaginous ET) causes OT due to its insertion onto the handle of the malleus. However, a clonus in the tensor tympani may also cause the ET to collide upon itself, through its attachment to the ET. With this in mind, and having directly visualized ET movement, we thought to treat our second case via a myotomy of the TTM. This improved symptoms, but did not result in full resolution—likely because the TTM remained connected to the ET even after sectioning of its attachment to the malleus handle. Moreover, this furthers our contention that PM and MEM are in fact part of the same disease process, as treatment of the TTM alone did not completely alleviate symptoms. This finding may be consequential in both diagnostic and treatment settings.

The anatomical and neurologic relationship between TTM and TVP seems to support our hypothesis. As discussed, both muscles not only directly attach to the ET, but also contribute to the opening/closing of the ET. In fact, anatomical studies have demonstrated that a tendon physically links the TTM and TVP; Ramirez Aristeguieta et al. declared that these muscles are in fact “nearly the same muscle by continuity” [4,5]. This finding has been confirmed by several authors [1,6,7]. Furthermore, they are both derived from the first branchial arch and innervated by a branch of the same nerve — the medial pterygoid nerve, a motor branch of V3 [5]. Taken together, the TTM and TVP appear to be integrally connected, meaning that contraction of one also means movement in the other; this would cause a collision between ET walls, leading to an objective sound — i.e. objective tinnitus.

Due to the proposed link between PM and MEM, treatment choices should be tailored to the individual patient based on their clinical exam. Treatment has classically been divided into three categories: reassurance, medical treatment, and surgery. Though a benign condition, patients often seek treatment to address their OT as it can be emotionally distressing. Medical treatments include sedatives, muscle relaxants, anti-convulsants, Botox injections, ET occlusion, tinnitus masking devices, psychotherapy, and zygomatic pressure maneuvers [8–10]. Surgical treatments include TTM, TVP, and/or SM tenotomy [1,2]. We feel the efficacy of the above interventions has varied widely, in part, because clinicians are selecting treatments that do not address the underlying etiology of the OT. In cases of PM and MEM that involve the TVP and TTM, our proposed etiology of OT indicates that treatment should be directed at both muscles, as ET contraction can contribute to OT in these patients. This is why in cases of PM, Botox injection into the TVP alone will show improvement of OT, but not complete resolution of symptoms, as the TTM may remain myoclonic [9]. Similarly, in cases of MEM involving the TTM, selective TTM tenotomy may improve patients' symptoms, but not result in complete resolution of symptoms as the attachments of the TTM and TVP to the eustachian tube remain active. Both our Case #2 and several other cases have supported this notion [2].

Hence, when designing a treatment plan to address OT, clinicians should always thoroughly inspect for both PM and MEM. If both are present, then both the TVP and TTM (which represent a functional unit) are likely involved. Thus, treatment should be directed towards both muscles for optimal results, with the primary aim of treatment being preventing the opening/closing of the ET. On the other hand, in cases of isolated MEM caused by the stapedius muscle (SM), tenotomy of the SM should be curative. Nevertheless, in refractory cases of MEM, treatment of the TVP muscle ought to be considered, as Ellenstein et al. found that it is possible to have non-visualized, pathogenic TVP movement creating OT [5]. As the TVP is the only palatal muscle to attach to the ET, myoclonus of other palatal muscles should not cause OT.

Of note, as an ancillary finding, we found that SOAE may be useful in assessing the degree of tinnitus in patients with OT; to date, objective measurement of OT has remained elusive. Various techniques have been used to quantify the degree of OT, such as listening to the outer ear via stethoscope, volume variation registered by impedance bridge, and recording sound in the external ear canal via microphone [2,11]. While these methods may attest to the presence or absence of OT, they fail to quantify the degree of OT. In Case #2, results of otoacoustic emissions testing were abnormal and asymmetric in the patient's affected ear, revealing large emission responses (Fig. 1). While OAE testing is typically used to assess sounds generated within the inner ear (e.g. the cochlea), the same test may be adapted to test for sounds emanating from the middle ear. Although more data will be required to create severity parameters, SOAE could eventually be used to create treatment guidelines in cases of OT. To our knowledge, this is the first case to use SOAE to characterize the severity of tinnitus in patients with PM/MEM.

4. Conclusion

In conclusion, the above cases illustrate that providers should recognize MEM and PM as either related, or possibly even the same, diseases. This notion is supported by the embryologic, neurologic, and anatomical relationship between the TTM and TVP. Our case series provides further evidence that the source of OT in MEM/PM is indeed derived from the ET walls slapping together, and is the first case to document video evidence demonstrating the link between OT and ET movement.

To appropriately treat these conditions, clinicians must first identify the muscles involved in creating symptoms. Both the TTM (causing MEM) and the TVP (causing PM) can cause OT by opening/closing the ET. Hence, in cases involving the TTM or the TVP, treatment of both muscles may result in improved outcomes. Moreover, in refractory cases of MEM, clinicians should look for evidence of PM and vice versa; as it is possible to have non-visualized, pathogenic TVP movement creating OT, treatment of the TVP muscle should be considered in refractory cases of MEM. Conversely, in cases of isolated SM myoclonus causing MEM and OT, tenotomy of the SM should be curative. As the TVP is the only palatal muscle to attach to the ET, myoclonus of other palatal muscles should not cause OT.

Finally, our case series is the first to introduce the utility of SOAE as

a possible method for assessing the severity of tinnitus in patients with OT; this may ultimately inform future treatment guidelines.

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Disclosure

The authors have not published any related papers from this manuscript. This paper is not under review at any other journal or publication venue.

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Conflicts of interest

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References

[1] Virtanen H. Objective tubal tinnitus: a report of two cases. J Laryngol Otol 1983;97(9):857–62.
[2] Oliveira CA, Negreiros Júnior J, Cavalcante IC, Bahmad F, Venosa AR. Palatal and middle-ear myoclonus: a cause for objective tinnitus. Int Tinnitus J 2003;9(1):37–41.
[3] Pulec JL, Simonton KM. Palatal myoclonus: a report of two cases. Laryngoscope. 1961;71:668–71.
[4] Ramirez Aristeguieta LM, Ballesteros Acuña LE, Sandoval Ortiz GP. Tensor veli palatini and tensor tympani muscles: anatomical, functional and symptomatic links. Acta Otorrinolaringol Esp 2010;61(1):26–33.
[5] Ellenstein A, Yusuf N, Hallett M. Middle ear myoclonus: two informative cases and a systematic discussion of myogenic tinnitus. Tremor Other Hyperkinet Mov (N Y) 2013:3.
[6] Kierner AC, Mayer R, v Kirschhofer K. Do the tensor tympani and tensor veli palatini muscles of man form a functional unit? A histochemical investigation of their putative connections. Hear Res 2002;165(1–2):48–52.
[7] Barsoumian R, Kuehn DP, Moon JB, Canady JW. An anatomic study of the tensor veli palatini and dilatator tubae muscles in relation to eustachian tube and velar function. Cleft Palate Craniofac J 1998;35(2):101–10.
[8] Park SN, Bae SC, Lee GH, et al. Clinical characteristics and therapeutic response of objective tinnitus due to middle ear myoclonus: a large case series. Laryngoscope. 2013;123(10):2516–2520.
[9] Scozzafava J, Yager J. Images in clinical medicine. Essential palatal myoclonus. N Engl J Med 2010;362(21):e64.
[10] Pollak N, Azadarmaki R, Ahmad S. Feasibility of endoscopic treatment of middle ear myoclonus: a cadaveric study. ISRN Otolaryngol 2014;2014:175268.
[11] Alicandri-Ciufelli M, Soloperto D, Palma S, Marrara A, Genovese E. A clockwork ear. Neuropediatrics 2012;43(3):146–8.