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Treatment of Stapedial Myoclonus as a Migraine-Related Phenomenon

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Objective: To describe a case series of patients with stapedial myoclonus (SM) whose conditions improved after prophylactic migraine treatment.

Patients: We present seven cases of SM reported from a tertiary care neurotology clinic. All seven patients reported SM triggers similar to those of migraine headaches and suffered from concomitant headaches and/or vertigo, and were thus treated with a standard migraine protocol used at this neurotology clinic.

Intervention: Prophylactic migraine treatment.

Main Outcome Measures: Reduction or resolution of SM.

Results: In this series, seven patients with SM were included. Six of seven subjects were male (86%), with a mean age at presenta-

tion of 44 years. Four patients noted significant improvement in their symptoms, with a reduced frequency, duration, and intensity of their symptoms with the migraine regimen. Three patients experienced complete resolution of SM with their migraine treatment.

Conclusion: We report that treatment with prophylactic migraine treatment can provide long-term relief for patients with SM, which may suggest an etiological association between migraine and SM as well as a possible treatment for SM.

Key Words: Lifestyle changes—Migraine—Migraine regimen—Migraine-related symptoms—Stapedial myoclonus.

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INTRODUCTION

Stapedial myoclonus (SM), one form of middle ear myoclonus, is defined as a rapid motion of the stapes secondary to repeating contraction of the stapedius muscle in the middle ear (1). SM is one cause of objective tinnitus (2), in which the perception of a true, mechanical somatosound occurs and can oftentimes be audible to a listener with a Toynbee tube (3). SM is a nonvascular, intermittent form of tinnitus (4) that can affect one or both ears (5) and typically presents as a rapid (>1 Hz) irregular thumping sound (6). Sounds have also been documented as tapping, throbbing, bubbling, ticking, or clicking (7–9).

Diagnosis is based on an individual's symptoms, impedance audiometry that may show changes in TM compliance

(10), a saw-toothed pattern on tympanometry (5), and absence of palatal myoclonus (PM) and patulous eustachian tube (ET) (5). Sometimes, rapid movement of the TM or the incudostapedial joint can be seen on microscopy or otoendoscopy (11). Previous reports of SM have associated the condition with benign fasciculation syndromes (12), facial nerve injury (6), noise trauma, anxiety, tumors, and vascular, infectious, and demyelinating etiologies (1). However, the pathophysiology behind SM remains unclear.

In our center, we have noticed an association between the identified triggers for SM and the established triggers for migraine headaches (MHs) (13,14). However, an association between SM and migraine has not been previously established. Furthermore, to our knowledge, the use of migraine prophylaxis for patients with SM has not yet been studied. Herein, we present our experience successfully treating seven SM patients with a prophylactic migraine treatment regimen.

METHODS

A retrospective chart review from 2017 to April 2021 was performed with institutional review board approval at a tertiary care neurotology clinic to identify patients with SM. Inclusion criteria included (1) primary diagnosis of SM, (2) no evidence of patulous ET, (3) no evidence of PM, and (4) normal magnetic resonance imaging or internal auditory canals (IACs) with gadolinium to rule out facial nerve abnormalities. We found seven patients with SM of varying severity. All patients underwent otoscopy,

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audiometric testing, and nasopharyngeal and oropharyngeal examination. All seven patients had a history of headache or vertigo, and their triggers paralleled those of MH.

Patients were advised on mitigating triggers for migraine, including stress reduction, proper sleep hygiene, proper diet and hydration, regular exercise, and avoidance of intense stimulations (e.g., bright lights, intense sound, intense motion, visual motion from computers, movies). Initial dietary control included avoidance of chocolate, red wine, caffeine, fermented foods, foods containing yeast, nuts, and nut products (foods that contain significant amounts of tyramine, glutamate, histamine, or caffeine). Patients were advised to take vitamin B₂ 200 mg twice a day and magnesium oxide 400 mg twice a day (15–18). In patients who did not experience adequate relief with this initial regimen, prophylactic migraine medications, including nortriptyline or verapamil, were prescribed.

RESULTS

Our retrospective cohort included seven patients with SM (Table 1). Six of seven were male (86%), with an average age of 44 ± 15 years. Four patients had SM in the left ear, and three had bilateral SM. The average duration of SM symptoms before diagnosis was 1.8 years (range, 2 mo–12 years; SD, 5.0 yr). No patients demonstrated evidence of ETD or PM on examination. One patient had headaches consistent with migraine without aura, and one patient had headaches consistent with probable migraine as classified by *International Classification of Headache Disorders, Third Edition* classification (Tables 2 and 3) (19). Otoscopy was normal bilaterally in all patients. All patients demonstrated hearing thresholds within normal limits. Impedance audiometry showed normal middle ear functioning in all patients. Tympanograms were type A in all patients. Three patients experienced complete resolution of their SM, and the four others reported significant improvement.

Illustrative Case Example

A 27-year-old man presented with a history of intermittent, left-sided tinnitus for 2 months that began after a flight. The patient described the tinnitus as a “clicking” sensation of 10 clicks per second of mild intensity, lasting seconds to minutes, and occurring once every 1 to 2 weeks. The only associated symptom included a headache that lasted around 10 seconds. The patient also complained of motion sickness and moderately severe left-sided headaches that occurred twice a month, lasted 10 hours, and were accompanied by nausea and/or vomiting, consistent with migraine without aura as categorized by International Headache Society (Table 2). He noted poor sleep and stress as triggers for the headaches and SM. Remaining history was negative. On examination, otoscopy was normal bilaterally. Audiometry demonstrated hearing thresholds within normal limits and type A tympanograms bilaterally. A magnetic resonance imaging IAC revealed a normal IAC and facial nerve.

TABLE 1. Demographics and clinical characteristics of patients with stapedial myoclonus

Patient	Age, yr	Sex	Tinnitus Laterality	Tinnitus Description	Triggers for SM	Other Symptoms	Treatment	Results
1	27	M	L	Fast clicking	Flying; poor sleep; stress; bright light	Headache with nausea, vomiting; vertigo; motion sickness	Migraine regimen with vitamin B ₂ and magnesium	Significant improvement
2	29	M	Bilateral (L > R)	Popping; machine gun-like	Loud noise; red wine; chocolate; nuts; weather changes	Dizziness; occasional aural fullness; headache	Migraine regimen with vitamin B ₂ and magnesium and nortriptyline; switched to verapamil (escalated to 240 mg)	Significant improvement
3	26	M	L	Vibration sensation	Loud noise; blowing nose; lying on side	Aural fullness and pressure; migraine headache; motion sickness	Migraine regimen with vitamin B ₂ and magnesium	Significant improvement
4	43	M	L	Fluttering	Coffee; screen time	Vertigo	Caffeine cessation	Resolved
5	50	M	Bilateral (L > R)	Fast clicking	None identified	Vertigo; migraine headache; motion sickness	Migraine regimen with vitamin B ₂ and magnesium	Significant improvement
6	54	M	Bilateral	Clicking and thumping	Wind; loud noise; weather changes; atmospheric pressure; flying	Otagia bilaterally; migraine headache	Migraine regimen with vitamin B ₂ and magnesium, verapamil 180 mg	Resolved
7	42	F	R	Fluttering sound	None identified	Vertigo	Diet and lifestyle modification	Resolved

TABLE 2. *Migraine without Aura: ICHD-3 diagnostic criteria (19)*

-
- A. At least 5 attacks fulfilling criteria B–D
 - B. Headache attacks lasting 4–72 h
 - C. Headache has at least two of the following four characteristics:
 - ii. Unilateral
 - ii. Pulsating
 - iii. Moderate or severe pain intensity
 - iv. Aggravation by or causing avoidance of routine physical activity
 - D. During headache at least one of the following:
 - i. Nausea and/or vomiting
 - ii. Photophobia or phonophobia
 - E. Not better accounted by another *ICHD-3* diagnosis
-

ICHD-3 indicates *International Classification of Headache Disorders, Third Edition*.

The patient was started on a migraine regimen for his suspected migraine symptoms. After initial visit, the patient decided to proceed with lifestyle modifications and magnesium (400 mg twice a day) and vitamin B₂ (200 mg twice a day) with a follow-up in 2 months. At 2-month follow-up, the patient noted improved migraine symptoms with much milder and shorter (30 min to 1 h) in duration. He also endorsed less frequent (one episode only) and improved SM symptoms. The patient was advised to continue with this migraine regimen. A telephone follow-up 6 months later revealed no further episodes of SM with rare mild headaches.

DISCUSSION

In this article, we describe our experience with treating seven SM patients with the migraine regimen. The time to achieve initial symptom improvement ranged from 2 to 3.5 months (mean [SD], 2.6 [0.75] mo). Three patients reported complete resolution, and four patients reported significant improvement in SM from baseline symptoms. In one patient, caffeine cessation alone resolved the SM. In four patients, lifestyle modifications and supplements brought substantial improvements to the patients' SM. In the other 2 patients, lifestyle modifications, supplements, and migraine prophylactic medication including verapamil significantly improved symptoms in one patient and fully resolved symptoms in the other.

Although the etiology of SM is unknown, many treatment regimens have been proposed for SM. Conservative measures, including muscle relaxants, sedatives, anticonvulsants (11), hypnosis and psychotherapy (1,10), botulinum injection into the muscle (20), ET occlusion (21), tinnitus masking devices (22), and zygomatic pressure (23), have been shown to be ineffective. The only known long-term effective treatment for SM is stapedius tendon tenotomy via tympanotomy (5). Unfortunately, tenotomy is more invasive and does not always guarantee resolution of SM (5).

Although all patients reported headaches in our case series, one had symptoms consistent with migraine without aura and one had symptoms consistent with probable migraine (19). Although migraine has not been associated with SM previously, reports are beginning to emerge regarding multiple otologic conditions related to migraine. For example, Hwang et al. (24) described an association be-

tween cochlear disorders, most notably tinnitus, in patients with migraine. Previous reports have also discussed a connection between MH and conditions such as pulsatile tinnitus (25), benign paroxysmal positional vertigo (26), persistent postural perceptual dizziness (27), Menière's diseases (28), aural fullness (29), hyperacusis (18), and mal de debarquement syndrome (30). Even more, treatment with migraine medications has been shown to provide relief of symptoms for Menière's disease and aural fullness patients. Previous studies have shown significant improvement in these migraine-related phenomena with migraine lifestyle changes, supplements, and prophylactic therapy (16,18,26,29).

Because our patients responded to migraine medication in a similar fashion to these previous studies, we believe that there is likely a connection between SM and MH. Furthermore, the identified triggers for SM in this case series are similar to migraine triggers (13,14). For example, the triggers for SM in our patient list included but were not limited to, low atmospheric pressure (flying and weather changes), poor sleep, stress, loud noises, red wine, chocolate, nuts, coffee, and screen time (Table 1). A review of the literature demonstrates these as triggers for migraine as well.

In this case series, the time for each patient to experience improved SM symptoms varied. For example, two patients' symptoms improved within 8 weeks. Other patients took 3 to 6 months to improve. As mentioned earlier, each person responds differently to the migraine regimen. In one, caffeine cessation was sufficient to resolve SM. Others, however, required dose escalation and/or medication modification. These findings demonstrate that prophylactic migraine treatment may take time for SM patients to experience improvement. However, once the correct treatment is initiated, patients with SM can experience significant improvement in their symptoms. Moving forward, prospective studies with larger cohorts are needed to both confirm and expand upon this potential relationship between migraine and SM. When working up patients with SM, physicians should ask about symptoms related to migraine as this case series supports the notion that SM can be treated with prophylactic migraine regimens.

Our study is limited by the number of cases presented, making our ability to draw inferences regarding efficacy of prophylactic migraine treatment in SM patients more difficult. The paucity of objective measurements of the change in severity of the SM is another limiting element in our study, as patients' subjective assessments of their SM symptoms were used to gauge improvement. The use of subjective measurement makes quantifying patient improvement more difficult.

TABLE 3. *Probable migraine: ICHD-3 diagnostic criteria (19)*

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- A. Attacks fulfilling all but one criteria A–D for migraine without aura or migraine with aura
 - B. Not fulfilling *ICHD-3* criteria for any other headache disorder
 - C. Not better accounted for by another *ICHD-3* diagnosis
-

ICHD-3 indicates *International Classification of Headache Disorders, Third Edition*.

CONCLUSION

Treating SM patients with a prophylactic migraine regimen can provide significant improvement or full resolution in a patient's symptoms. The successful use of a prophylactic migraine regimen also demonstrates that atypical migraine may be a possible etiology behind SM.

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