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Case Report

Auricular Myoclonus: A Case Report and Literature Review

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Auricular myoclonus is an extremely rare disorder that manifests as involuntary semi-rhythmic movements of the auricle. We report the case of a 15-year-old female who presented to our outpatient clinics with bilateral spontaneous, uncontrolled movements of the auricles (auricular myoclonus) that are only briefly suppressible by some facial movements and completely disappear during sleep. Needle electromyography revealed baseline tonic motor unit activity with bursts of higher motor units amplitude in the posterior and superior auricularis muscles. Her symptoms improved with pregabalin intake, however, with incomplete resolution. This paper will review previously reported cases, as well as the different treatment modalities that have been used.

KEYWORDS: Ear, electromyography, muscle contraction, myoclonus, pregabalin

INTRODUCTION

Focal or segmental dystonic syndromes involving various cranial muscles are well-recognized entities.[1] Auricular dyskinesia, in particular, is an extremely rare entity with very few cases reported in the literature. The pathogenesis of this disorder is yet to be elucidated. It has, however, been linked to psychological stressors and tics.[2] The spectrum of management has practically been narrowed to a single modality of treatment, namely injection of the extrinsic auricular muscles with botulinum toxin A.[3]

CASE PRESENTATION

A 15-year-old female patient, previously healthy, presented to our outpatient clinics with bilateral abnormal and uncontrollable auricular movements of 3-4 months duration. The movements are continuous throughout the day and resolve during sleep. They are described by the patient as a "pulling of her auricle up and to the back." The patient does not recall any specific triggering event and denies any previous psychological, psychosocial, or physical stressors. A magnetic resonance imaging (MRI) of the brain had been performed in an outside facility, was reread in our medical center, and was perfectly normal. A complete blood count with differential and chemistry was done on an outpatient basis and was found to be normal. She had also failed a trial of a selective serotonin reuptake inhibitor (SSRI) and a benzodiazepine.

The patient denies tinnitus, hearing loss, and aural fullness. Otologic examination was normal, and all cranial nerve functions were intact. Both auricles were seen to be spontaneously and synchronously moving in a continuous semi-rhythmic pattern with an upward and backward displacement at a rate of approximately 90 events per minute. They were aborted with the elevation of the eyebrows and with smiling. The contractions were not synchronous with the temporal artery pulse. (Video 1). The rest of the physical examination was normal.

Needle electromyography (EMG) of the auricularis superior and the auricularis posterior was performed in our neurophysiology center and showed a constant baseline of tonic motor unit firing activity with brief bursts of higher motor units amplitude, solitary or in clusters (Figure 1).

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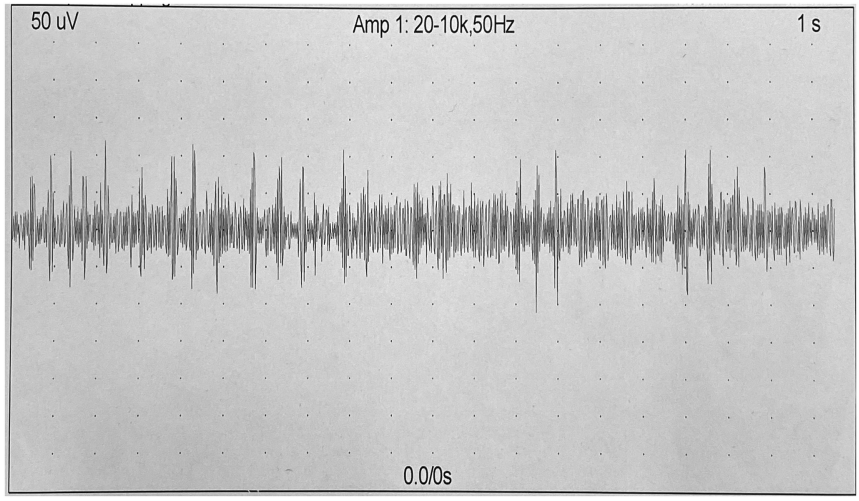

Figure 1. Needle EMG showing background firing activity interrupted by bursts of higher amplitude.

The patient was prescribed pregabalin 75 mg once daily. On follow-up, 2 weeks later, the contraction frequency had markedly decreased to around 20 minutes every hour. Increasing the dosage to 75 mg twice daily resulted in additional improvement. Treatment with botulinum toxin A injection was then offered; however, the patient's natural guardian declined and opted for continuing with medical treatment.

DISCUSSION

Myoclonus, whether generalized or focal, is defined as a brief muscular movement that is involuntary and insuppressible.[4] Myoclonus of the extrinsic auricular muscles also referred to as "moving ear," auricular dyskinesia, and "ear wiggling" is a rare disorder with yet unclear understanding of its pathophysiological mechanism.[3] It may originate from the cerebral cortex, the subcortical structures, the brainstem, the spinal cord, or from peripheral nerves.[5] No clear etiology has been described. Some reports have however suggested that drugs such as SSRIs[6] and neuroleptic preparations[7] may be possible causes, albeit unlikely.

With regards to the laterality, auricular myoclonus has been equally distributed in patients, unilateral in some[4,8,9] and bilateral in others.[1,3,6,7,10]

The strongest association of ear movements with psychological stressors and disorders was initially described in 1988 by Keshavan,[2] who had labeled them as tics and not as myoclonic phenomena, since the patients had some component of voluntary control for suppression.

Alonso-Navarro et al[8] however, reported on a 40-year-old patient with right ear myoclonus but with no previous psychological inciting events.

Auricular myoclonus may not be an isolated finding. Many associated symptoms have been described in the literature, such as peri-aural discomfort,[1,9] peri-aural pain,[1,7] headaches,[1,3] tympanophonia,[11] and tinnitus.[7,10] Patients may be asymptomatic,[7,8] with normal blood workup,[4,8] unexplained eosinophilia, and normal cerebrospinal fluid (CSF) examination.[7] Serum ceruloplasmin, antinuclear antibody (ANA), rheumatoid factor, syphilis serology, and biochemical profile have also been reported to be normal by Kirk et al.[4] Magnetic resonance imaging findings are usually normal, except in the case described by Caviness et al[1] whereby a 36-year-old lady was found to have a white matter lesion posterior to the trigone of the ipsilateral lateral ventricle.

With regards to EMG findings, the muscles involved were firing within a range of 100-500 ms, results that include the ones found in our patient. Our results, however, are different than the ones found in the literature in terms of chronicity. The patient exhibited a constant muscular baseline of high-frequency tonic muscle activity that is interrupted by bursts of higher motor units amplitude (Figure 1).

Our patient presented with a 3 months history of bilateral auricular myoclonus, a period that is within the range that is reported in the literature. As with other reported cases, our patient could not voluntarily suppress her auricular movements,[1,7,9] and the movements disappeared completely during sleep.[1,3,6,9]

We have observed the disappearance of the phenomenon with the elevation of the eyebrows, a finding that was not described before in patients with a similar presentation. Caviness et al[8] reported not only worsening of the movements with stress but also suppression with voluntary eye closure, voluntary facial movement, and phonation. These findings were also reported by Lee et al.[10] Srirompotong et al[3] and Godeiro-Junior et al.[9]

In addition, Kirk et al[4] reported a decrease in a patient's ear movements with the repetitive performance of complex hand gestures.

The pathophysiology of auricular movements by the activation of some facial muscles, even hand gestures, is unclear. In their study on nerve bundles within the internal auditory canal of cadavers, Ozdoğmuş et al[12] showed a cross neural connection between the facial, vestibular, and cochlear bundles on scanning electron microscopy. We postulate the existence of a neural connection between the various cranial nerves that may explain the phenomenon of auricular myoclonus and its modulation by facial movements.

To our knowledge, this entity has a poor long-term prognosis when left untreated. Several modalities of treatment and management

have been proposed, most of which were either unsuccessful or partially successful, with the exception of the use of botulinum toxin A.

A trial of baclofen 10 mg and clonazepam for 4 weeks was attempted by Lee et al[10] resulting in minimal improvement. Chaudhuri et al[7] however, report considerable improvement in 1 patient when oral clonazepam was used. Caviness et al[1] reported on a 25-year-old male patient with bilateral ear movement who did not show improvement with carbamazepine intake. Oral valproic acid 250 mg twice daily did not show any benefit when used on a 30-year-old patient with right ear myoclonus.[9] Premastoid facial nerve block with 4 mL of 2% lidocaine has been shown by Kirk et al[4] to be effective, resulting in the disappearance of auricular movement.[4]

Our patient was prescribed oral pregabalin 75 mg once daily for 2 weeks, achieving partial resolution in terms of frequency but not of rate. Adjusting the dosage to 75 mg twice daily showed slight additional improvement. No further increase in dosage was made taking into consideration the young age of the patient and the potential undesirable side effects.

Botulinum toxin A has shown promising results when injected into the extrinsic auricular or temporal muscles, with or without EMG monitoring, achieving partial to complete resolution, yet dosage and frequency remain controversial.[3,6-10]

CONCLUSION

Auricular myoclonus, a subset of facial myoclonus, is a rare and poorly understood entity that currently has an ill-defined etiology and pathophysiology. It may be the sole manifestation, however, other associated symptoms have been described, causing heavy bearings on the quality of life of patients. Modulation of auricular myoclonus by facial movement suggests the presence of a cross neural connection between the cranial nerves, which requires further investigation. We strongly propose a course of pregabalin for managing this disease, as it shows an improvement in symptoms.

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