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A rare case report of moving ear syndrome

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Abstract
Focal or segmental dyskinesia is a rare phenomenon manifesting as an involuntary movement. We report a 15-year-old female with bilateral spontaneous, semi-rhythmic, involuntary movements of the ear briefly suppressible by distraction and completely disappeared on sleep. Her symptoms didn't improve with medications. In this case report, we have reviewed the existing literature and it seems that further studies will improve our knowledge regarding this syndrome and its treatment modalities. However, meanwhile based on the available evidence, we propose a trial of botulinum toxin for the treatment.

Keywords: Moving ear syndrome, dyskinesia, botulinum toxin

Introduction
Movement disorders involving the ear (moving ear syndrome) have rarely been described in the literature. It includes auricular myoclonus, focal motor seizure, or dystonia [1, 6]. The ear is surrounded by three vestigial muscles namely the anterior, superior, and posterior auricular muscles [7]. The intrinsic musculature is involuntary, although some people can move their ears with the help of extrinsic scalp muscles [8]. There are reports of ear movement disorders being successfully treated with botulinum toxin injections [2, 5, 9] and one case was treated with unilateral pallidothalamic tractotomy [10].

Case Description
A 17-year-old female studying in high school presented with complaints of headache and involuntary movement of both external ears. Headache started around 1 year back and preceded the onset of ear movements and it was low to moderate in intensity and would remain for most of the days every week. Headache was not associated with photophobia, phono-phobia, chewing movements of the jaw, or any dental, or eye pain. She was started on Amitriptyline 50mg/day, and subsequently, Escitalopram 10mg/day, Mirtazapine 45mg/day, Clonazepam 0.5mg/day, and Carbamazepine 600 mg/day were all tried but her headache didn't subside with these medications. Then after 6 months, she started to notice her ears were moving, upon which she was referred to our centre by her doctor.
She presented to us with both headache and bilateral ear movements. Her history suggested that the movement started bilaterally and was initially intermittent with 4-5 episodes per day and each episode lasted 2-3 minutes. Gradually, the duration of these movements went up from 2-3 minutes to 10-15 minutes. The movement in both ears was semi-rhythmic, involuntary with synchronous elevation and retraction of both external ears with equal amplitude and frequency (30-40/min) (Video 1-3). These movements were absent during sleep and decreased marginally on distraction (like asking the patient to perform mathematical calculations) and they were not associated with any palatal tremor, facial twitching, slurring of speech, or myoclonus. She didn't report any urge preceding the movements nor any major stressor during subsequent interviews.
Her past, family, and medical history were unremarkable. Her routine blood investigations, MRI brain and EEG were all normal and her NCCT head revealed bilateral ethmoidal and frontal sinusitis. Her HAM D score was 5, HAM A-10 and Dissociative Experiences Scale (DES) was 28.
Her drug regimen was simplified as she didn't have any significant improvement from her current medications. Her Carbamazepine, Mirtazapine was gradually titrated down while Amitriptyline was titrated up to 75 mg/day. Her headache improved but her ear movements persisted, so Tetrabenazine 25 mg/day (gradually titrated up to 75 mg/day over a few days)

along with clonazepam 1 mg/day were added to her drug regimen. Her movements persisted even after an adequate trial of these medicines, so Botulinum toxin injections were recommended to her but the patient and her guardian refused to undergo the procedure due to high costs and unproven effects. Therefore, we decided to give a trial of Pregabalin, her tetrabenazine was gradually titrated down and Pregabalin was titrated up from 75mg/day to 150mg/day, but even then she had no improvement in the ear movement and she complained of dizziness. Her Pregabalin was gradually titrated down and she was maintained on Tab Amitriptyline 75 mg/day, and Tab Clonazepam 1 mg/day.

Discussion

The clinical picture of our case intimately resembles segmental and focal dyskinesia affecting auricular and craniofacial muscles as the movements were non-jerky, stereotypic and lasted for a few seconds to a few minutes. Focal dyskinesia can affect various regions of the body and they arise spontaneously following either trauma, surgery, or neuroleptic drug intake but there was no history of these events in our case, so we excluded these causes.

A close differential for these movements could be disorders of the brainstem region including auricular myoclonus, palatal tremor, and reticular reflex myoclonus while auricular myoclonus consists of irregular clonic & jerky movements with a rate of 70-75/ minute affecting the antitragus and antihelix, our case had semi-rhythmic movements with less frequency (30-40/min). The palatal tremors have a frequency of 40-600/minute and that was not present in our patient as well as MRI brain didn't show any brainstem lesions thus ruling out the possibility of these disorders [4, 8, 11, 12].

Keshavan described 10 cases of ear wigglers caused by tics. Although, ear tics are unlikely in this patient as movements in our patients were involuntary and slow [8].

One rare cause of dyskinesia can be the serotonergic effect on dopamine-2 receptors. Our case also had exposure to SSRI as well as another serotonergic reuptake inhibitor (TCA) and temporally medication preceded the movements. A review of 71 cases revealed the development of motor symptoms after SSRI exposure. Dystonia was reported in 20 patients, Akathisia in 32, Parkinsonism in 10, and tardive dyskinesia in 8 cases. To date, only 1 case has been reported ear dyskinesia arising due to serotonergic medication [13].

We reviewed the relevant literature for therapeutic strategies and there is a piece of anecdotal evidence supporting Botulinum Toxin injection for moving ear syndrome. Botulinum injection has also proved to be effective for other etiological causes of movement disorders such as tics, tremors, myoclonic jerks, and stuttering [14].

Table 1: Previous cases of "Moving ear syndrome" and their characteristics

Author's name	Case characteristics	Investigation	Affected ear	Involved Musculature	Proposed aetiology	Treatment
Caviness *et al* [1]	23-year-old female	MRI: Not done EEG: Not done	Both ear			Carbamazepine
	25-year-old female	MRI: Normal EEG:150-400 ms; 2Hz	Both ear	Occipito-frontalis		Carbamazepine
	36-year-old female	MRI: Single lesion in the white matter posterior to the trigon of the left ventricle EEG:500 ms: 0.5 Hz	Left ear	Auricularis superioris and frontalis		Carbamazepine
	48-year-old female	MRI: Not done EEG:100-400 ms; 3-5 Hz	Right	Auricularis superioris		Carbamazepine
Chaudhuri *et al* [2]	23-year-old male	MRI: Normal EEG: Normal	Both	Auricularis superioris and frontalis	Tardive dyskinesia	Clonazepam
	32-year-old male	MRI: Normal EEG:200-300 ms:2 Hz	Left	Auricularis superioris and posterior		Botulinum toxin type A – 40 U
Kirk and Helman *et al* [4]	20-year-old male	MRI: Not done EEG: Irregular	Right	Anti-tragus		Pre-mastoid facial nerve block with 4 ml of 2% lidocaine
Carluer *et al* [5]	57-year-old female	MRI: Normal EEG: 280 ms; 2 Hz	Both	Auricularis superior	Drug-Induced paroxetine	Botulinum toxin type A - 40U
Kim *et al* [7]	57-year-old female	MRI-Normal	Both ear	Anterior, Posterior and superior uricular muscle	Focal auricular dystonia	Botulinbotulinum injection
Godeiro – Junior *et al* [9]	30-year-old male	MRI: Normal EEG: 1.5 – 2 Hz	Right	Temporalis		Botulinum toxin type A - 60U
Jabbour *et al* [15]	15-year-old female	MRI-Normal EEG- f tonic motor unit firing activity with brief bursts of higher motor units amplitude, solitary or in clusters	Both ear	Superior and Posterior Auricularis	Dyskinesia	Pregabalin 75mg/day

Conclusion

The presentation of complex movements become a difficult differential diagnosis. From tics to dystonia to dissociative disorders, there are multiple possibilities. A careful evaluation and assessment of individual cases is needed to offer optimal results.

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