



Motor-Phonic Tic Mimicking Essential Palatal Myoclonus (EPM)

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INTRODUCTION

Essential palatal myoclonus (EPM), also referred to as essential palatal tremor, is an uncommon, idiopathic disorder characterized by rhythmic, continuous, bilateral and symmetric movement of the soft palate. Instead of an oscillatory movement typically encountered in tremor, EPM is caused by contractions of the tensor veli palatini muscle, which opens the Eustachian tube and produces a clicking noise. Tics may have a clonic phenotype, but are distinguished from myoclonus by their suppressibility, presence of a premonitory sensation which is relieved by the execution of the tic, and co-morbid neurobehavioral manifestations, such as obsessive compulsive disorder (OCD) and attention deficit hyperactivity disorder (ADHD).

METHODS

We reviewed the medical records and video recordings of two patients with Tourette syndrome (TS), who presented with ear clicking and palatal movements with features overlapping with EPM.

RESULTS

Both patients exhibited repetitive elevations of the soft palate accompanied by a synchronous clicking sound. However, in contrast to typical EPM, the movements were irregular, intermittent, and temporarily suppressible. Both patients had other features supporting the diagnosis of TS (Table).

DISCUSSION

TS rarely presents with a palatal tic associated with ear clicking, characteristic of EPM. EPM is caused by rhythmic and continuous contractions (125 ± 70 cycles/min) of the tensor veli palatini muscle, which opens the Eustachian tube and causes an abrupt decrease in the surface tension within the tube and a clicking noise. EPM is rarely suppressible. Oropharyngeal tics may have features overlapping with EPM, including bilateral ear clicking, but are usually associated with more widespread contractions of the oropharyngeal muscles. In contrast to typical EPM, palatal tics are usually more irregular, intermittent, of variable frequency, temporarily suppressible, and distractible. Furthermore, tics are usually preceded by a premonitory sensory component and usually disappear during sleep. The presence of other motor or phonic tics, and the coexistence of associated psychiatric co-morbidities (OCD, ADHD, impulse control disorder) are other clues that help differentiate the two conditions. Psychogenic palatal tremor is also part of the differential diagnosis of isolated palatal tremor / myoclonus. It results from the contraction of the tensor veli palatini and other extrapalatal muscles. Psychogenic palatal tremor is distractible, entrainable and variable in frequency. The onset is usually acute, following an emotional trigger or trivial trauma. It is responsive to verbal suggestion, non-physiologic treatment (tuning fork) or placebo.

CONCLUSIONS

Distinguishing between EPM and palatal tics is important not only because the etiologies are different, but also because the treatment differs. Antidopaminergic drugs are often effective in the treatment of tics, but botulinum toxin may also be considered as a therapeutic option. Patients with TS often have other tics and neuropsychiatric co-morbidities that may need to be addressed and treated.

Table: Patient Characteristics

Patient	1 (Video segment 1)	2 (Video segment 2)
Age (years)	21	12
Palatal movements	Onset 2 years prior Fluctuating course, partially suppressible, without a premonitory sensory component. Present intermittently. Associated with bilateral ear clicking noise ("leaky faucet") Absent during sleep	Onset 6 months prior Partially suppressible, without a premonitory sensory component. Associated with bilateral ear clicking noise Presence during sleep: not known
Initial diagnosis	Palatal myoclonus	Tic
Initial treatment	Escitalopram Clonazepam Valproic acid Topiramate	Pimozide
Age of onset of other tics	Childhood	10 years
Other tics	Lower lip sucking and licking; knuckle popping	Hand clapping, head hitting, neck extension, blinking, grimacing
Past medical history	Bipolar disorder, anxiety	Depression, asthma
Birth history and development	Normal	Normal
Family history	Tics (father and brother); compulsive behavior (trichotillomania in his father and a paternal uncle); ADHD (two maternal cousins)	ADD (father), OCB (mother)
Neuropsychiatric comorbidities	Obsessive compulsive behavior (OCB), impulse control behavior	OCB, ADD
Treatment	Fluphenazine (positive response)	Tetrabenazine (no follow up)
Other medications	Amphetamine/Dextroamphetamine 10 mg daily Diazepam 10 mg daily Escitalopram 10 mg daily	Paroxetine 60 mg daily Escitalopram 20 mg daily Valproic acid 500 mg BID Guanfacine 1 mg daily
Diagnostic tests	Brain MRI: normal Audiometry evaluation: normal	Brain MRI: normal EEG: normal
Prior exposure to neuroleptics	None	Brief trial of pimozide resulted in acute dystonic reaction

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