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Case Report

Essential Palatal Myoclonus: A Case Report

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ABSTRACT:

Introduction: Palatal myoclonus is a rare condition presenting with clicking sound in ear or muscle tremor in pharynx. There are two varieties: essential and symptomatic. Various treatment options exists ranging from watchful observation to botulinum toxin injection. We have not found any reported case of palatal myoclonus from our country. Here we present a case of essential palatal myoclonus managed with clonazepam. **Case report**: A young female presented in Ear Nose and Throat clinic with complain of auditory click and spontaneous rhythmic movement of throat muscles for eight months. On examination, there was involuntary, rhythmic contraction of bilateral soft-palate, uvula, and base of tongue. Neurological, eye, and peripheral examination were normal. A diagnosis of essential palatal myoclonus was made. It was managed successfully with clonazepam; patient was still on low dose clonazepam at the time of making this report. **Conclusion**: Essential palatal myoclonus can be clinically diagnosed and managed even in settings where MRI is not available or affordable.

Keywords: botulinum • case report • clonazepam • palatal myoclonus

INTRODUCTION:

Palatal myoclonus, also known as palatal tremor or palatal nystagmus, is an uncommon condition.[1] It is characterized by involuntary brief rapid rhythmic jerky movements of soft palate and peritubal musculature causing clicking sound in the ear.[2]

Palatal tremor has two forms: essential and symptomatic, salient differentiating features between them are outlined in Table 1. Essential palatal tremor has no pathologic basis and is associated with rhythmic activation of the tensor veli palatini, often recognized by the patient noting an audible click that ceases with sleep.[3] It can be subjective which is perceived only by the patients as clicking tinnitus with strong muscle spasms felt at the back of throat or objective if also heard by the examiner. Several treatment options like anxiolytics, anticonvulsants, botulinum toxin, and surgery has been tried with variable success.[2,4] We were unable to find any previous case report from Nepal.

Table 1: Salient features of two types of palatal myoclonus

Features	Essential palatal myoclonus	Symptomatic palatal myoclonus
Muscle involved	Tensor veli palatine	Levator veli palatini
Audible click	Present	absent
Myoclonus during sleep	Absent	Present
Laryngeal involvement	Much less common	Sometimes
Ocular involvement	Never	Frequently
Extremity involvement	Never	Rarely
Etiology	Unknown	Vascular lesions, tumors, trauma, encephalitis
Medullary lesions	Usually no lesion	Visible in MRI

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CASE REPORT:

A 20-year-old female presented to the Ear, Nose and Throat (ENT) department of Lumbini Medical College, Nepal on 12th of February, 2017 with 8-month history of spontaneous movement of soft palate and uvula along with auditory click. She was distressed by self-movement of her oral cavity. She denied any hearing loss, vertigo, ear discharge, and otalgia. She had no history of nasal regurgitation, nasal intonation, headache, head trauma, or weakness of any part of body and difficulty in swallowing. She was house wife, non-alcoholic, and non-smoker.

Her physical examination of oral cavity revealed rhythmic contraction of both side of soft palate, uvula, and base of tongue (Fig: 1) (Video: 1, available online). Contractions were asynchronous with her pulse. No other abnormality was found on ophthalmological, otological, rhinological, neurological, and neck examination. As patient was neurologically normal and had no ophthalmic findings, she was diagnosed with essential palatal myoclonus. As we do not use botulinum toxin, she was prescribed clonazepam 0.5 mg at bed time daily for 10 days and explained about possible adverse effects of the drug and called for follow up after 10 days. On first follow up, her symptom and frequency of palatal contraction were reduced and she was feeling better. We continued the same treatment and called monthly. On third visit, she was symptoms free but had contractions when she attempted to show her throat. After third visit, she was continued on clonazepam 0.25 mg daily. She has made four follow up visits, is asymptomatic, and still on 0.25 mg of clonazepam daily.

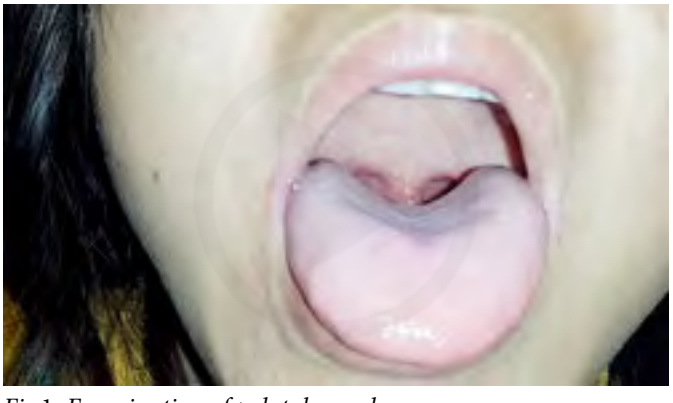

Fig 1*: Examination of palatal myoclonus.*
Click over the figure to see the video (available online and opens in a web browser)

DISCUSSION:

Essential palatal myoclonus is a rare condition, without demonstrable lesion, causing pulsatile tinnitus and soft palate tremor.[1,2,3] The condition usually affects adults and it has been reported in children as young as seven years of age. [5] The condition is benign but distressing when not treated.

There are two types of patatal myoclonus, essential and symptomatic. In the former type, there are no demonstrable lesion by MRI of brain and is mostly associated with audible click and objective tinnitus due to involvement of tensor veli palatine muscle. Myoclonus is absent during sleep and never involve muscles of eyes and extremities. In the later type, lesions are demonstrable in brainstem or cerebellum by MRI, myoclonus is present even during sleep, and ocular muscles are often involved. [1,2,3] Palatal myoclonus have been reported even after dental surgery.[6]

MRI of the brain is an important investigation for work-up of these cases. In our case, we did not do an MRI because there were no involvement of muscles of eyes and extremities, no other focal neurological features, and above all it is a costly investigation in a developing world.

Essential myoclonus may resolve spontaneously.[7] Many drugs has been tried in treatment of essential palatal myoclonus. These include anticonvulsants like clonazepam, carbamazepine, or sodium valproate. Cannabis has also been tried but botulinum toxin injection has found to be most successful in controlling the symptoms for up to three months.[5] In our case, we prescribed clonazepam and during these four months, it has found to be successful in controlling the condition. We have reduced the dose but we are not sure about stopping it completely and relapse is a possibility. We never use botulinum toxin in our center.

We believe this case report along with the accompanying online video will contribute to the fellow clinicians in the country and the region to diagnose the case, differentiate it between essential and symptomatic on clinical grounds, and start treatment for essential variety before referring them to tertiary centers in case the treatment is ineffective. Vast area of our country in north and hilly area lack trained professionals and well equipped treatment centers.

CONCLUSION:

Essential myoclonus is an uncommon condition and can be diagnosed clinically in areas where MRI is not available or feasible. Clonazepam can be successfully tried in such cases before referring them to tertiary centers.

Conflict of Interests:

Anup Acharya did not take part in any editorial decisions.

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