

CASE REPORT

Essential rhythmic palatal myoclonus in a 51-year-old man

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Abstract

Essential palatal myoclonus is a rare movement disorder consisting of continuous rhythmic jerks of the soft palate; it usually develops secondary to brainstem or cerebellar disease called symptomatic rhythmic palatal myoclonus. Diagnosis is usually clinical, and some patients, however, fail to show evidence of a structural lesion like our patient called essential rhythmic palatal myoclonus. We report a 51-year-old man who has suffered from the condition and showed improvements on treatments.

CASE REPORT

A healthy 51-year-old man presented with a 2-year history of progressive involuntary worsening movement of the soft palate and uvula. The movement was rhythmic and repetitive (see Supplementary Video 1 showing rhythmic movement of the soft palate and also Fig. 1 showing a snap shot of the uvula). Initially, the movement was tolerable, and the patient had no disturbance during sleep. Gradually, the uvula movement intensified to an extent that he could not tolerate it. It interfered with patient's speech and swallowing and even disturbed his normal sleeping pattern. Brain MRI was normal and did not show any evidence of a structural lesion as a cause. Workup for autoimmune conditions and cerebrospinal fluid analysis were within reference range. Echocardiography was done to exclude aortic regurgitation, and it was normal as 'Müller's sign', which is a pulsation of uvula during systole and can be seen as one of the signs of aortic regurgitation, and it might mimic this condition, although the patient did not have any cardiac symptoms and signs. Essential palatal myoclonus was considered as a diagnosis. Essential



Figure 1: A snap shot of uvula.

palatal myoclonus is a rare movement disorder consisting of continuous rhythmic jerks of the soft palate; the primary one does not have any clear cause, but there is also a secondary type that usually develops secondary to brainstem or cerebellar

Received: March 29, 2016. Revised: May 26, 2016. Accepted: May 31, 2016

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disease called symptomatic rhythmic palatal myoclonus. Diagnosis is usually clinical, and some patients, however, fail to show evidence of a structural lesion like our patient called essential rhythmic palatal myoclonus [1]. Although it resembles a tremor, the movement is repetitive rather than oscillatory and involves contraction of tensor veli palatini muscle. Auditory click might happen in essential palatal myoclonus, for which the cause is unknown, and it was present in our patient indicating that the lesion is subcortical, although the brain imaging was normal, which can be differentiated from cortical in which the click is absent. In this patient, we tried carbamazepine with no response, but he showed improvement to clonazepam within 10 days and was given 1 mg twice daily, and the dose was titrated to 2 mg twice daily and maintained for the past 1 year. He only had mild sedation with no tolerance. Other drugs like L-5-hydroxytryptphan, phenytoin, barbiturates, diazepam and trihexyphe-nidyl can be used selectively in some patients [2]. An injection of botulinum toxin into the tensor veli palatini muscle has been described in the literature in some cases and results in remarkable improvement in symptoms [2–4]. The patient still have mild symptoms in the form of sensation of uvula movement that he does not consider disturbing.

CONCLUSION

Essential palatal myoclonus can be a serious problem, and secondary causes should be always excluded; furthermore, its treatment might be challenging with variable response. It should also be differentiated from Muller's sign which could be the sign of aortic incompetence.

SUPPLEMENTARY MATERIAL

Supplementary material is available at *Oxford Journal* online.

CONFLICT OF INTEREST STATEMENT

None declared.

FUNDING

Not applicable.

ETHICAL APPROVAL

Ethical approval was provided by Shar Teaching Hospital.

CONSENT

Written informed consent was obtained from the patient.

GUARANTOR

Not applicable.

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