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Tremor and Other Hyperkinetic Movements

Video Abstracts

Facio-Oculo-Palatal Myoclonus Complicated by a Recurrent Brainstem Stroke

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Abstract

Background: A 54-year-old Thai male who has suffered from multiple episodes of ischemic and hemorrhagic strokes developed facio-oculo-palatal myoclonus (FOPM) 1 month after the latest episode of the brainstem stroke.

Phenomenology Shown: The patient presented with semirhythmic, involuntary, horizontal jerky, and rotatory ocular oscillation concomitant with asymmetrical palatal and perioral myoclonus consistent with FOPM.

Educational value: FOPM is a useful clinical clue for diagnosing brainstem lesions, specifically in the Guillain–Mollaret triangle. The commonest etiology is cerebrovascular diseases.

Keywords: Cerebrovascular diseases/stroke, palatal tremor, facio-oculo-palatal myoclonus, brainstem infarction, Guillain–Mollaret triangle.

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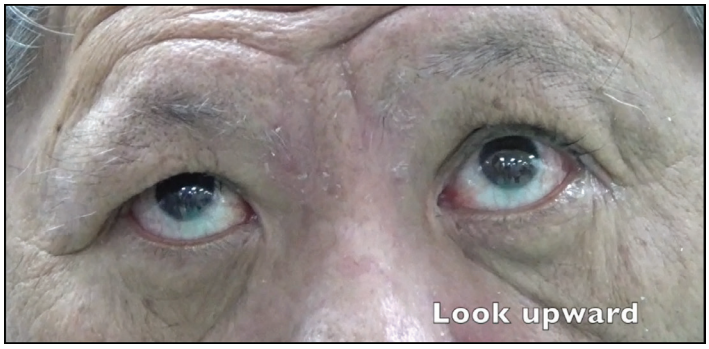
Ethics statement: All patients that appear on video have provided written consent; authorization for the videotaping and for publication of the videotape was provided.

Case summary

A 54-year-old Thai male has suffered from multiple episodes of stroke, including right pontine hemorrhage for 5 years, right basal ganglion infarction for 2 years, and multiple lacunar infarction of the brainstem and cerebellum, which was the most recent episode of stroke, for 1 year. The patient could not ambulate independently. One month after the latest episode of stroke, the patient experienced severe oscillopsia. The abnormal movements were semirhythmic, involuntary, horizontal, and jerky, with rotatory ocular oscillation concomitant with asymmetrical palatal and perioral myoclonus consistent with facio-oculo-palatal myoclonus (FOPM) (Video 1). The patient denied ear clicking sound. Brain MRI showed multiple old infarctions and old hemorrhages within the Guillain–Mollaret triangle (GMT) (Figure 1A), along with hypertrophy of the right inferior olivary nucleus (ION) (Figure 1B). Facial myoclonus improved with clonazepam 1.5 mg/day, while oscillopsia and palatal myoclonus persisted and disturbed the patient's quality of life.



Video **1. Phenomenology of the Patient.** (**Segment 1**) Multifocal facial myoclonus, torsional nystagmus while looking horizontally, multidirectional ocular myoclonus while looking vertically, and palatal tremor. (**Segment 2**) Nonentrainable palatal tremor.

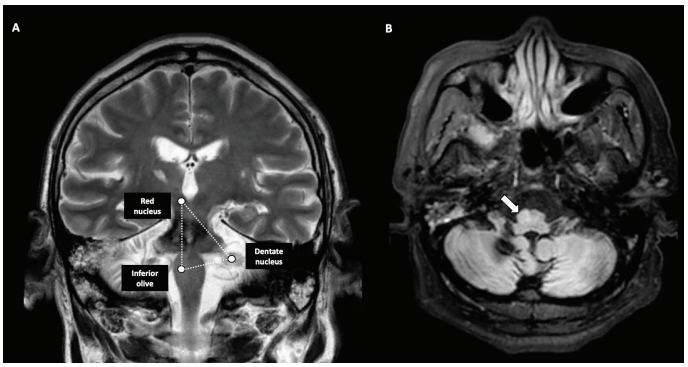


Figure 1. **Brain MRI of the Patient.** (A) Brain MRI T2 FLAIR sequence shows old ischemic and hemorrhagic lesions in the Guillain–Mollaret triangle. (B) Hypertrophy of the right inferior olivary nucleus (arrow).

Discussion

FOPM is a rare, delayed complication of brainstem or cerebellar lesions. Patients with FOPM usually present with ocular jerky movement or nystagmus, including pendular vertical nystagmus, torsional nystagmus, and, rarely, horizontal nystagmus,[1] as well as abnormal movements of the larynx, pharynx, diaphragm, and facial muscles. Interestingly, our patient showed both usual (i.e., rotatory ocular oscillation) and rare (i.e., horizontal nystagmus) ocular manifestations along with typical facial myoclonus and palatal tremor. The major differential diagnosis of these abnormal eye movements is an opsoclonus; however, the physicians can distinguish an opsoclonus from an ocular myoclonus by carefully looking at the patient's eyes. The eyes movement in an opsoclonus is involuntary, chaotic, conjugated, and multidirectional movement, including a combination of horizontal, vertical, and torsional directions with large amplitude. The frequency of an opsoclonus is approximately 10–15 Hz, and the key feature of an opsoclonus is an absence of intersaccadic interval.[2] An opsoclonus can persist during sleep or eyelid closure. It usually occurs with myoclonus which not only restricts to the cranial region but also involves limbs and trunk that may prevent the patients from sitting and standing. In addition, the myoclonus in an opsoclonus-myoclonus syndrome generally does not involve soft palate.

Circuitry disruption in the GMT (Figure 1A) associated with ION hypertrophy (Figure 1B) has been described as the possible pathogenesis of FOPM. Loss of GABAergic control at the level of the gap junction between neurons in the ION and loss of inhibitory signal from the cerebellar deep nuclei to contralateral ION are the possible pathophysiologic mechanism of FOPM.[3,4]

While the most common etiology of FOPM is vascular causes, of which hemorrhagic stroke is more frequent than infarction, the common etiology of an opsoclonus-myoclonus syndrome is immune-mediated diseases such as neuroblastoma or paraneoplastic syndromes.[2] Therefore, differentiate between two patterns of eye movement abnormality can help the physicians to narrow down the differential diagnosis and point out the possible etiology more precisely. Other etiologies of FOPM have been reported, including trauma, vascular malformation, and inflammatory diseases such as multiple sclerosis, Behcet's disease, and neurosarcoidosis.[1] The time between initial brainstem injury and manifestation of FOPM varies from 1 month to 8 years, but commonly occurs within 2–49 months.[3]

Usually, palatal tremor does not disturb patients, while oscillopsia due to ocular myoclonus is a very disabling symptom. FOPM is difficult to treat. One report showed a significant effect of gabapentin and memantine on decreasing the amplitude and frequency of nystagmus.[1]

In conclusion, FOPM is a useful clinical clue for diagnosing brainstem lesions, specifically in the GMT. The most common etiology of FOPM is cerebrovascular diseases.

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