Movement Disorders
CLINICAL PRACTICE

Forehead Tremor: A Clinical Presentation of Myasthenia Gravis?

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Myasthenia gravis (MG) is an autoantibody-mediated disease of neuromuscular junction, usually occurring with diplopia and ptosis. Clinical presentation of MG with involuntary movements has been observed in an exiguous number of cases,[1–3] but the association between MG and forehead tremor has not previously described. We report the case of a patient with seropositive MG presenting with forehead tremor.

A 59-year-old man was admitted to our clinic complaining of involuntary eyebrow movements improving after rest for seven months. Neurological examination showed bilateral ptosis and forehead tremor with right predominance, observable at rest and increasing in up-gaze (Video, Segment 1). EMG of head district and four limbs excluded a myopathic pattern. The clinical finding of bilateral ptosis supported the diagnostic hypothesis of MG. Therefore, single-fiber EMG (SFEMG) was performed. SFEMG of orbicularis oculi (OO) and extensor digitorum communis (EDC) muscles documented a mean jitter of 92 μs in OO (n.v. $<$ 30 μs) and a mean jitter of 78 μs in EDC (n.v. $<$ 34 μs). Low-rate repetitive nerve stimulation, brain-MR, and chest-CT were normal, while a positive titer of anti-acetylcholine receptor antibodies was detected (22 nmol/L, n.v. $<$ 0.45 nmol/L). Patient also underwent an electrophysiological study to investigate forehead tremor. With the patient sitting on a chair while watching a fixed point in front of him and during up-gaze, tremor activity was recorded by two pairs of needle electrodes from the frontalis and OO muscles. An irregular bilateral activity of frontalis muscles was observed increasing in frequency and amplitude during up-gaze. EMG recording of OO muscles activity showed electrical silence (Video, Segment 2).

He was treated with pyridostigmine 90 mg q.i.d. and prednisone 50 mg per day with a significant improvement of ptosis and forehead tremor.

The association between involuntary movements and MG was first pointed out in 1967 with the description of palpebral minor tremor in a MG patient in a non-English language article.[1] Subsequently, Wilfong and colleagues reported the co-occurrence of MG and opsoclonus-myoclonus syndrome.[2] Hemichorea had also been reported such as unusual MG presentation.[3]

Forehead tremor has been very rarely described in some patients with Parkinson's disease or essential tremor.[4–6] Erro and colleagues described a series of eight patients affected by idiopathic focal dystonia with tremor involving the upper-middle part of the face, of whom four showed eyebrow tremor.[7]

To our knowledge, this is the first clinical description of forehead tremor as presentation sign of MG. EMG tremor pattern recording is usually featured by the presence of a rhythmic alternating contraction or a synchronous co-activation of agonist-antagonist muscles. In our case, we did not find the typical tremor pattern described by Piboolnurak,[4] as OO muscles activity was absent. Moreover, the clinical finding of ptosis and the recording of an irregular electromyographic activity in Frontalis muscles, unable to maintain a steady contraction, could explain the onset of forehead tremor in trying to keep eyes open as a compensatory act of weak Frontalis muscles. Therefore, muscular fatiguing gave rise to a tremor-like phenomenon.

Forehead tremor has been described as a rare feature of essential, dystonic or parkinsonian tremor. However, when a patient presents with forehead tremor, with or without visual disturbances, clinicians should consider also MG as diagnostic hypothesis, in order to start rapidly the correct treatment.

Author Roles

(1) Research Project: A. Conception, B. Organization, C. Execution; (2) Statistical Analysis: A. Design, B. Execution, C. Review and Critique; (3) Manuscript Preparation: A. Writing of the First Draft, B. Review and Critique.

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Supporting Information

Videos accompanying this article are available in the supporting information here.

Segment 1. Bilateral ptosis and right predominant forehead tremor in a patient affected by seropositive Myasthenia gravis.

Segment 2. EMG frontal tremor recording. EMG channels 1 and 2 show muscle activity of right Frontalis muscle and right Orbicularis Oculi muscle, whereas EMG channels 3 and 4 show muscle activity of left Frontalis muscle and left Orbicularis Oculi muscle, respectively.