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Essential palatal myoclonus in monozygotic male twins

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Sirs: Palatal myoclonus (PM) is an uncommon movement disorder characterized by vertical oscillation of the soft palate at 1 to 3 Hz and usually bilateral. Occasionally some other brainstem-innervated muscles may be involved. Some authors call it a palatal tremor [10, 12].

We report monozygotic male twins with essential palatal myoclonus. To our knowledge this is the first report in which essential PM can be genetically linked.

39 year old twin brothers were admitted to hospital for treatment of involuntary movements of the soft palate and clicking in both ears. In their medical history it was learned that symptoms developed at the age of 14–15 years in one of them and at the age of 37 years in the other. The patients' parents were healthy and unrelated.

The patients' general and neurological examination was unremarkable except easily audible clicking in time with the visible rhythmical movements of the soft palate. It was not associated with synchronized movements of eyes, tongue, larynx, diagram, intercostal muscles, extremity or trunk. The patient's whose soft palate frequency had developed at the age of 14–15 years was 2Hz. symptoms, and was higher than his brother's whose frequency was 1,5Hz. Chronic treatment with clonazepam, piracetam, valproic acid, amitriptiline and carbamazepine failed to improve PM in the first patient and clonazepam, carbamazepine, valproic acid were administered with no effect in the second. Because the different drugs did not improve the PM, five units of botulinum toxin (Botox®/Allergan) in 0.2 ml. saline solution were injected into each side of tensor veli palatini through a special needle under EMG guidance. The first patient reported some initial difficulty with swallowing, in the second patient there was no severe side effect. Three days after the injections PM and clicking began to diminish and had completely disappeared after 6 days in both of them. They remained free of the PM and clicking at the every three months follow-up visit for one year in the first patient and six months in the second. The investigations including routine blood chemistry, computed tomography (CT), magnetic resonance imaging (MRI), electroencephalography (EEG), somatosensory evoked potentials (SEP) and brainstem auditory evoked potentials (BAEP) were all normal.

There are two forms of palatal myoclonus. Essential palatal myoclonus is characterized by a vertical oscillation of the soft palate at 1 to 3 Hz. unaltered by voluntary actions, and an annoying ear click. There are no other neurological signs or symptoms, and the pathophysiology of essential palatal myoclonus is unknown. Symptomatic palatal myoclonus is nearly always associated with syncronous movements of the eyes, larynx, pharynx, face, diaphragm, cervical and axial muscles. Some authors, however, point to difficulties in classifying their patients with palatal myoclonus as either the essential or the symptomatic type [1, 8, 9, 13]. Symptomatic palatal myoclonus produced by a lesion involving the pathway connecting dendate, red and inferior oliver nuclei. Olivary enlargement can be visible on MRI in some symptomatic cases [4, 5, 9, 13].

The patients may complain only of annoying ear click, the origin of which is uncertain, although many believe it arises from muscles contracting around the eustachian tube.

The potential role of heredity in the etiology of essential palatal myoclonus is unknown. These monozygotic twin brothers with essential palatal myoclonus in which symptoms developed at the age of 14–15 years in one of them and at the age of 37 years in the other are of interest for suggesting the hereditary etiology of essential palatal myoclonus. In our monozygotic male twins the first symptoms of palatal myoclonus were noted 22 years later than in the other one. These findings indicate that there is a role of inheritance in the pathogenesis of essential palatal myoclonus but also that some other factors contribute to the clinical presentation of palatal myoclonus. Similar findings have been described in monozygotic twins with idiopathic dystonia and Huntington's disease [3, 6, 11]. As the relationship between the disease and its genetic linkage could be confirmed in our patients, it might also be thought that the pathology may not be inherited.

Successful treatment of essential palatal myoclonus by botulinum toxin has been reported several times [2, 7]. In our monozygotic male twins, five units botulinum toxin in 0.2 ml. saline solution were injected into each side of tensor veli palatini and both cases were rendered free of their myoclonus a few days after injection until 11 months in the first patient and 6 months in the second one.

In conclusion, the essential

palatal myoclonus may be based on a genetic defect. But possible influences of prenatal and postnatal environmental factors are presently unknown. Tensor veli palatini botulinum toxin injection is an effective treatment for essential palatal myoclonus and long-term benefits are possible.

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