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A Palatal Myoclonus Case with Facial Involvement Responding To Botulinum Toxin Therapy

Ayhan Köksal *, Devrimsel Harika Ertem **, Yavuz Altunkaynak *, Ayten Ceyhan Dirican *, Nesliha Sütpideler Köksal ***

**Bakırköy Ruh ve Sinir Hastalıkları Hastanesi, Nöroloji Kliniği, **Cerrahpaşa Tıp Fakültesi, Nöroloji Anabilim Dalı, ***İstanbul Eğitim ve Araştırma Hastanesi, Nöroşirurji Kliniği*

ABSTRACT

Palatal myoclonus is a syndrome characterized by involuntary, continuous or nearly continuous, rhytmical movements in soft plate, pharynx, larynx, diaphragma and rarely facial muscles. Mostly a variety of causes like vascular diseases, multiple sclerosis, encephalities, neoplasms and infections are blamed in the etiology.

A 35 year-old female patient who had ischemic infarct due to left vertebral artery dissection suffered from obvious involuntary movements on the left side of her face. Palatal Myoclonus was considered after neurological and electrophysiological examinations. Botulinum toxin was applied as there was no response to medical therapies and good response was obtained.

Palatal myoclonus rarely expands to facial muscles and is more responsive to Botulinum toxin than medical therapies. For this reason this case is reported to review the etiology, symptoms and therapy.

Keywords: *palatal myoclonus, botulinum toxin, cerebrovascular disease*

ÖZ

Botulinum Toksin Tedavisine Yanıt Veren Fasyal Tutulumlu Bir Palatal Miyoklonus Olgusu

Palatal miyoklonus, yumuşak damak, farinks, larinks, diafram ve nadiren yüz kaslarını tutan istemsiz, sürekli veya sürekliye yakın ritmik hareketlerle karakterize bir sendromdur. Vasküler nedenler, multiplskleroz, ensefalit, neoplazma ve infeksiyon gibi çeşitli nedenler etiyolojide suçlanmaktadır.

Sol vertebral arter diseksiyonuna bağlı iskemik enfarktı olan 35 yaşında kadın hastanın yüzünün sol tarafında belirgin istemsiz hareketleri mevcuttu. Nörolojik ve elektrofizyolojik değerlendirme sonrası palatal miyoklonus tanısı düşünüldü. İlaç tedavilerine yanıt olmaması nedeniyle botulinum toksin uygulandı ve tedaviye iyi yanıt alındı.

Palatal miyoklonus ender olarak yüz kaslarını etkiler ve botulinum toksinine ilaç tedavisinden daha iyi yanıt verir. Bu nendenle bu olgu etiyoloji, semptomlar ve tedavinin gözden geçirilmesi amacıyla sunulmuştur.

Anahtar kelimeler: *palatal miyoklonus, botulinum toksin, serebrovasküler hastalık*

INTRODUCTION

Palatal myoclonus is a rare type of segmental myoclonus characterized by involuntary, unilaterally or bilaterally, synchronous or non synchronous, rhytmical ossilations affecting pharynx, larynx, neck, eye, respiratory and rarely facial muscles which are originated from the same embriological bronchial arcus. The structures affected from palatal myoclonus come from first five brancial arcus [(1)].

The exact pathophysiology of palatal myoclonus is unclear and for this reason it is difficult to prescribe specific pharmacologic therapy. Pharmacologic therapy consists of cholinergics, benzodiazepam, antiparkinsonism drugs, anticonvulsants, muscle relaxants, and lithium [(2)]. Another therapeutic option for palatal myoclonus is aimed to control parts of the brain using botulinum toxin, which blocks somatic input and output through peripheral blockage of involuntary muscle movement [(3,4)]. This neurotoxin promotes the inhibition acetylcholine release, which results in muscle relaxation in the targeted organ.

CASE

A 35 year-old female patient admitted to our neurology department with nausea, vomitting, vertigo and

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Yazışma adresi: Uzm. Dr. Devrimsel Harika Ertem, Cerrahpaşa Tıp Fakültesi, Nöroloji Anabilim Dalı, İstanbul
e-posta: hkaozhan@gmail.com

left sided hemihypoesthesia. She told that her symptoms occured after she worked hard in a field and carried a heavy load on her back 4 days ago. She had no history of diabetes mellitus, hypertension, oral-genital wound, abortus or intrauterine exitus. Smoking or oral contraceptive usage was not defined.

Physical examination of the patient was normal. Neurological examination revealed left sided hemihypoesthesia-hypoalgesia and left sided ataxia and she could walk by help with wide-based. Other neurological findings were normal.

Cranial computed tomography and magnetic resonance imaginations (MRI) were consistent with acute ischemic infarct. Lesions were at the areas supplied by left posterior inferior cerebellar artery, posterior cerebral artery and bilateral superior cerebellar arteries. Significant decrease in left vertebral artery flow and meaningful irregularity in wall of artery were determined in Cervical MRI Angiography which was consistent with vertebral artery dissection. Patient was treated by heparin and oral anticoagulant therapy. Cranial MRI angiography showed tightness and irregularity of V2 segment of left vertebral artery which looked like a rat tail. Behcet's disease excluded after dermatological and ophtalmic examinations. Antinuclear antibodies were negative, anticardiolipin antibodies Ig M and Ig G were normal. Transthoracical echocardiography was normal. When she was discharged from hospital with oral anticoagulant therapy, there was no neurological pathology other than minimal ataxia.

Involuntary palatal movement developed dominantly on left side of soft palate and face about two months after stroke. As synchronised, bilateral, left side dominant, continuous bursts of abnormal electrical activity with a frequency of 160 per minute was determined in orbicularis oris, mentalis and soft palate muscles in electromyography (EMG), she was diagnosed as palatal myoclonus.

Following an ineffective 6-months course of medical therapy with a combination of muscle relaxant (baclofene), anticonvulsants (carbamazepine,valproate) and anxiolytic agent (clonazepam), the patient was treated with an injection of botulinum toxin A under the guidance of EMG. Botulinum toxin A was injected into orbicularis oris and mentalis muscles (10 U each). It was not injected into soft palate muscle as it could cause difficulty in swallowing and there was no disturbing click sound. On the re-examination at week after injection of botulinum toxin A, symptomatic recovery occured and EMG showed decreased frequency of bursts of abnormal electrical activity.

DISCUSSION

Palatal myoclonus is a rare neurological disease of soft palate and the other oropharyngeal muscles, most oftenly seen in early adulthood and middle age periods.

Palatal myoclonus may be both unilateral or bilatarel. In bilateral type soft palate deviates to superior and posterior side with uvula while posterior pharyngeal pililer are deviating to middle and anterior side synchronously. In unilateral type, soft palate and uvula deviates to the affected side. Clonus in facial muscles, pharynx, larynx may accompany. Movements may spread out to diaphragma, neck and upper extremity muscles like branchial muscles [5]. Mean frequency of contraction is 100-150/minute, between 20-600/minute [1]. In oculopalatal myoclonus, eyes are affected by two ways, laterally or on middle line, eye movements are synchronised with palatal movements, patients often complain visual failure like ossilopsy [6].

Palatal myoclonus is a continuous or nearly continuous movement. It can not be stopped by sleep, IV barbiturate infusion, carotid sinüs stimulation, coma or oncoming death. It may be difficult to differentiate faciculations due to motor neuron diseases from palatal myoclonus as both of them are not stopped by IV barbiturate infusion. Contraction frequency, amplitude and its spread to other muscles are not affected much from position of chin or repressive movements.

According to etiologic factors, platal myoclonus can be classified as symptomatic palatal myoclonus, which is a condition that is secondary to brainstem or cerebellar diseases (70 % of them are vascular infarcts), and essential palatal myoclonus, with the absence of a brain lesion [2,7]. Palatal myoclonus occurs some times later from the primary etiology, for example 2-49 months after vascular lesions, 16 hours-5

months after traumas [8,9]. Our patient had palatal myoclonus after two months of cerebrovascular disease.

An objective tinnitus may accompany. Studies show that click sound occurs due to opening movement of eustachi tube by tensor veli palatini muscle. Tinnitus can be easily heard as click sound by the examiners also. It may be heard in one or both ears [10].

In EMG of palatal myoclonus cases, bursts of abnormal electrical activities are so regular. Contraction frequency is 100-150/minute, every contraction persists for 60-150 milisecond. In diffuse cases, facial-cervical muscles are the ones that are activated just after the pharyngeal muscles [11].

Specific pathological finding for palatal myoclonus is hypertrophic degeneration of inferior olive nucleus (ION). Symptoms are caused by the lesion of the connections between the dentate nucleus, red nucleus and inferior olivary nuclei (Guillain-Mollaret triangle). Positron emission tomography shows ION hyperactivity in palatal myoclonus cases [12].

Pharmacologic treatment consists of several medications, including cholinergics, benzodiazepam, anticonvulsants, muscle relaxants, 5-hydroxytryptopha, levedopa and lithium [1,2]. Surgical therapy is another option, which dissection of the palatal muscles [13]. Another strategy for palatal myoclonus treatment is using botulinum toxin, which blocks somatic input and output through peripheral blockage of involuntary muscle movement [4,14,15]. In our case, systemic pharmacologic therapy were not effective and the symptoms are mostly controlled with Botulinum toxin A injection.

Palatal myoclonus is an uncommon issue, rarely expanding to facial muscles and more responsive to Botulinum toxin than medical therapies. The aim of this case report is to review the etiology, symptoms and therapy of it. Botulinum toxin has been stated to be effective for many neuromuscular disorders, and this was also useful in our case, we believe that it can be considered to be primary theraphy for palatal myoclonus.

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