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Marin-Amat syndrome

Marin-Amat sendromu

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Abstract

Involuntary contraction of one or more muscle groups during voluntary contraction of the muscle group innervated by different nerves or different peripheral branches of the same nerve is defined as synkinesia. Synkinesia is one of the most common long-term complications of facial paralysis. Marin Amat syndrome is a rare type of synkinesis that occurs when the jaw is fully opened or the involuntary closure of the eyelids during lateral movement of the jaw. It has been suggested that aberrant regeneration of nerve 7 and incorrect innervation between nerves 5 and 7 may be responsible for this syndrome. In this case report, a 31 years old male patient presented to our clinic with involuntary closure of the left eyelid during laughing and mouth opening-closing movements 1 year after a peripheral facial paralysis. Marin-Amat syndrome, a rare synkinesis, is important to remember in patients with involuntary closure of the eyelid with jaw opening or laughter after peripheral facial paralysis.

Key words: Marinamat syndrome, facial paralysis, trigeminal nerve, synkinesia, botox treatment.

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Özet

Farklı sinirler veya aynı sinirin farklı periferal dalları tarafından inerve edilen kas grubunun istemli kasılması sırasında eş zamanlı, bir veya daha fazla kas grubunun istemsiz kasılması sinkinezi olarak tanımlanır. Sinkinezi, fasiyal paralizinin uzun vadeli ve en sık komplikasyonlardan biridir. Marin Amat sendromu çenenin tamamen açılması veya çenenin laterale doğru hareketi sırasında göz kapaklarının istemsiz kapanması şeklinde ortaya çıkan nadir görülen bir sinkinezi çeşididir. Yedinci sinirin aberan rejenerasyonu ve 5 ve 7. sinirler arasındaki hatalı inervasyonun bu sendromdan sorumlu olabileceği ileri sürülmüştür. Bu olgu sunumunda 31 yaşında erkek hasta geçirilmiş periferik fasiyal paraliziden 1 yıl sonra gülme ve ağız açma-kapama hareketleri sırasında sol göz kapağında istemsiz kapanma yakınması ile kliniğimize başvuran, Marin Amat Sendromu düşünülerek botilismus toksin A uygulanan bir vaka sunulmaktadır. Nadir görülen bir sinkinezi olan Marin-Amat sendromunun periferik fasiyal paralizi sonrası gelişen çenenin açılması veya gülme ile birlikte göz kapağında istemsiz kapanma semptomu olan hastalarda akla gelmesi önemlidir.

Anahtar kelimeler: Marinamat sendromu, fasiyalparalizi, sinkinezi, trigeminalsinir, botoks tedavisi.

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Introduction

Involuntary contraction of one or more muscle groups during the voluntary contraction of the muscle group that is innervated by different nerves or different peripheral branches of the same nerve is defined as synkinesis. Synkinesia is one of the most frequent and long term complications of facial paralysis. There are many patterns of synkinesia. However, the most common is the upward movement of the lateral oral commissure and upper lip during blinking or closing. It can also be in the form of eyelid closure or autonomic synkinesia during talking or laughing. Marin Amat syndrome is a rare type of synkinesis that occurs when the jaw is fully opened or the involuntary closure of the eyelids during lateral movement of the jaw. It has been suggested that aberrant regeneration of nerve 7 and incorrect innervation between nerves 5 and 7 may be responsible for this syndrome. In this case report, a 31-year-old male patient presented

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to our clinic with the complaint of involuntary closure of the left eyelid during laughing and mouth opening-closing movements 1 year after peripheral facial paralysis, and was treated with Botilismus toxin A with Marin Amat syndrome.

Case

A 31-year-old male patient was diagnosed with peripheral facial paralysis due to left ear pain, numbness in the left half of his face and withdrawal to the right mouth, and inability to close the left eyelid. The patient complained about involuntary closure of the left eyelid when he laughed or eaten after about a year. The patient also complained of sputtering on the left eyelid and left half of the face. It was observed that the eyelid was unintentionally closed and blepharospasm-like contraction developed. This movement in the eyelid showed improvement with the termination of laughter (Figure1).



Figure 1. Significant facial synkinesia before botox treatment

Similar findings were also observed during chewing. No pathology was detected in the cranial magnetic resonance imaging of the patient. Botulinum toxin was applied to the patient. After treatment, it was found that the closure of the left eyelid was reduced concurrently with laughter (Figure 2).



Figure 2. Improvement in ptosis after botox treatment

Discussion

Facial synkinesia is defined as involuntary movement in the other part of the face accompanied by voluntary contractions in one part of the face that occurs following atypical reinnervation of the facial nerve. Facial paralysis or facial nerve grafting may cause facial nerve synkinesis [1]. In this case, faecal synkinesia occurred approximately 1 year after facial paralysis. However, the development of synkinesia in patients with severe facial paralysis may be earlier. MarcusGunn, Inverse MarcusGunn phenomena and Marin-Amat syndrome are forms of facial synkinesis that can be confused with each other and whose pathophysiological mechanisms are not fully understood [2]. Marcus Gunn phenomenon; is a common synkinesis in patients with congenital ptosis. With the opening of the jaw, the upper evelid opens and ptosis improves.

It has been shown that there are abnormal connections between the motor branches of the external pterigoid muscle innervating the trigeminal nerve and the superior branch of the oculomotor nerve that innervates the superior of the levator palpebra (false innervation between the 3rd and 5th cranial nerves) [3-5].

In reverse Marcus Gunn syndrome, the upper eyelid closes by opening the jaw. Improper firing between the third and fifth cranial nerves is thought to be responsible for this syndrome. Although it is not clear, it has been suggested that there are abnormal neural connections in the innervation of the superior muscle of the levator palpebra and that the levator palpebral superior is not related to the 3rd nucleus and that the 5th nucleus is innervated by the external pterygoid branches [1, 5]. Valve closure mechanism is caused by inhibition of levator palpebra superior rather than orbicularis oculi contraction as seen in Marin-Amat syndrome. The LPS may be linked not only to the third core, but also to the outer pterygoid portion of the fifth core. The exact level of this abnormal connection is controversial.

In Marin-Amat syndrome, blepharospasm occurs in the affected eyelid by opening the jaw or lateral movement. It is usually acquired and develops after peripheral facial paralysis. Marin-Amat syndrome has been suggested to result from acquired anastomosis of the trigeminal (CN V) and facial (CN VII) nerve innervating multiple muscle groups [2]. Whether the reverse Marcus Gunn phenomenon and Marin-Amat syndrome is different is controversial. While some authors think that both express the same disorder [3, 4], there are views suggesting that the two syndromes are different [5].

The most common treatment modalities for patients with Marin-Amat syndrome today are: Botulinum toxin type A injection, facial neuromuscular training, biofeedback method, selective neurolysis or myectomy [6]. Botulismus toxin treatment may be beneficial on blepharospasm [6, 7]. In our case, it was found that the closure of the left eyelid concomitantly decreased with laughter after low-dose botulinum toxin type A injection. In addition, resection of preseptal orbicularis ocular muscle has been reported to be successful. It is important that Marin-Amat syndrome, a rare synkinesis, be considered in patients with involuntary closure of the eyelid with jaw opening or laughter developing after peripheral paralysis.

Conflicts of interest: No conflict of interest was declared by the authors.

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Authors' contributions to the article

Z.Y. has designed the main idea of the study. The discussion section of the article was written by Z.Y. and A.D.M. S.T. has reviewed and made the necessary corrections and confirmed them. In addition, all authors discussed the entire study and confirmed its final version.

Informed consent: Written informed consent was obtained from the patient.