

# ภาวะกล้ามเนื้อใบหน้ากระตุกครึ่งซีกที่รักษาควบคุมอาการยากจากภาวะหลอดเลือดแดงเวอร์ทีบรัลขยายตัวผิดปกติ

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## บทคัดย่อ

**วัตถุประสงค์:** เพื่อรายงานผู้ป่วยที่มีภาวะกล้ามเนื้อใบหน้ากระตุกครึ่งซีกที่มีสาเหตุจากภาวะหลอดเลือดแดงเวอร์ทีบรัลขยายตัวผิดปกติกดเบียดเส้นประสาทสมองคู่ที่ 7

**วิธีการศึกษา:** ศึกษาและนำเสนอ อาการนำ อาการแสดง ผลตรวจภาพถ่ายทางรังสีวิทยาและการรักษาของผู้ป่วยชาย อายุ 44 ปี ที่มีปัญหากล้ามเนื้อใบหน้ากระตุกครึ่งซีกเรื้อรัง ที่รักษาควบคุมอาการยาก

**ผลการศึกษา:** การตรวจตาแร็บบพบการกระตุกของกล้ามเนื้อที่ใบหน้าครึ่งซีกซ้ายแบบหดรัดเกร็ง ควบคุมไม่ได้เป็นระยะ การตรวจทางระบบประสาทไม่พบอาการแสดงของก้านสมองหรือหูอื้อ ผู้ป่วยได้รับการฉีดโบทูลินัมทอกซินเพื่อลดการกระตุก รวมทั้งหมด 5 ครั้งภายในระยะเวลา 2 ปี ภายหลังอาการไม่ดีขึ้นจึงได้แนะนำเรื่องการตรวจเพื่อวินิจฉัยเพิ่มเติม ผลตรวจเอกซเรย์แม่เหล็กไฟฟ้าสมองพบหลอดเลือดแดงเวอร์ทีบรัลฝั่งซ้ายขยายตัวผิดปกติและอยู่ใกล้ชิดเส้นประสาทสมองคู่ที่ 7 ฝั่งซ้ายซึ่งน่าจะเป็นสาเหตุของอาการกระตุก ผู้ป่วยกำลังจะได้รับการผ่าตัดวางวัสดุเพื่อแยกหลอดเลือดแดงออกจากเส้นประสาท (microvascular decompression)

**สรุป:** ภาวะหลอดเลือดในสมองกดเบียดต่อเส้นประสาทสมองคู่ที่ 7 พบเป็นสาเหตุของภาวะกล้ามเนื้อใบหน้ากระตุกครึ่งซีกได้น้อย การตรวจภาพถ่ายทางรังสีวิทยาควรพิจารณาทำในผู้ป่วยที่มีหูอื้อ, อาการกระตุกที่รักษาควบคุมอาการยาก หรือในผู้ที่มีอาการแสดงทางระบบประสาท **จักษุเวชสาร 2018; กรกฎาคม-ธันวาคม 32(2): 103-108.**

**คำสำคัญ:** ภาวะกล้ามเนื้อใบหน้ากระตุกครึ่งซีก, โบทูลินัมทอกซิน, ภาวะหลอดเลือดแดงเวอร์ทีบรัลขยายตัวผิดปกติ

ผู้พิมพ์ทั้งหมดไม่มีส่วนเกี่ยวข้องหรือผลประโยชน์ใดๆ กับผลิตภัณฑ์ที่ได้กล่าวอ้างถึงในงานวิจัยนี้

# Intractable hemifacial spasm from vertebral dolichoectasia: A case report



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## Abstract

**Purpose:** To report a case of intractable hemifacial spasm (HFS) from vertebral dolichoectasia.

**Methods:** The author reported a case of 44-year-old male who presented with chronic involuntary eyelid twitching. The patient's symptoms and signs, radiographic findings and management were described.

**Results:** Eye examinations revealed clonic movements of facial muscles innervated by the left facial nerve. Neurological examinations revealed no signs of brainstem pathology or tinnitus. The patient received 5 botulinum neurotoxin injections for spasm over a 2-year period, after which his symptoms did not improve. Magnetic resonance imaging (MRI) brain demonstrated the V4 portion of the left vertebral artery located close to the left facial nerve. The patient underwent suboccipital craniotomy with microvascular decompression.

**Conclusion:** Vascular compression of the facial nerve is a rare cause of hemifacial spasm (HFS). Neuroimaging should be considered in patients presenting with intractable spasm or associated neurological signs. **Thai J Ophthalmol 2018; July-December 32(2): 103-108.**

**Keywords:** Hemifacial spasm, Botulinum neurotoxin, Vertebral dolichoectasia

*No Author has a financial or proprietary interest in material or method mentioned*

## Introduction

Hemifacial spasm (HFS) is characterized by unilateral, involuntary brief, tonic or clonic movements of facial muscles innervated by the facial cranial nerve.<sup>1-3</sup> Vertebrobasilar dolichoectasia is an abnormal distension and malposition of the vertebral or basilar arteries.<sup>4</sup> Dolichoectasia may compress the dorsal root entry zone of the facial cranial nerve and is a rare cause of HFS. The author reported a case of vertebral artery dolichoectasia presented with intractable hemifacial spasm.

## Case report

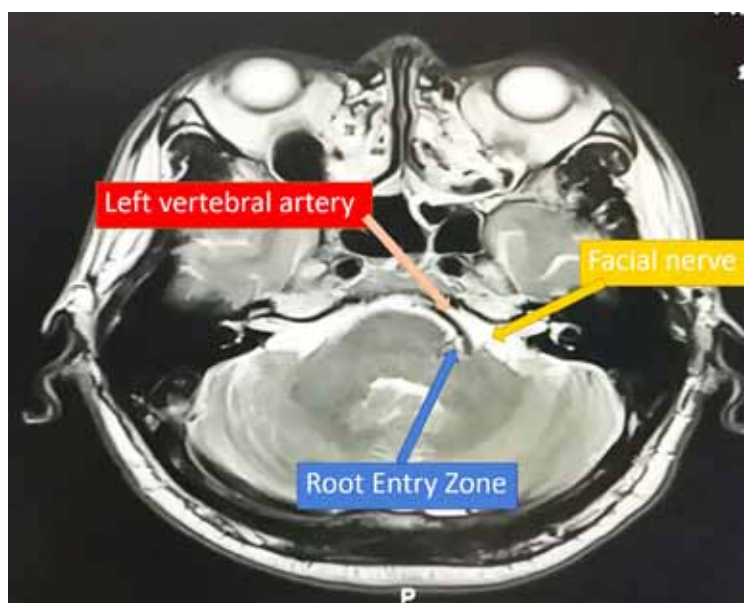
A 44-year-old healthy Thai male complained of involuntary eyelid twitching for approximately 4 years prior to presentation. He did not have tinnitus or a previous history of facial palsy although, he went to the secondary care hospital and was diagnosed with

blepharospasm. Initially, the patient received benzodiazepine (clonazepam) agents. His symptoms did not improve, hence the patient was referred to our hospital. Initial visual acuities were 20/20 OU. The anterior segment and posterior segment examinations were normal both eyes. Neurological examinations revealed involuntary brief, clonic movements of facial muscles innervated by the left facial nerve without facial nerve palsy (Figure 1). The other cranial nerves examinations did not reveal abnormalities.

His twitching occur about 2-3 episodes per day and each episode lasts only a few minutes, but sometimes an eyelid twitch lasts about 20 minutes. The patient was initially diagnosed with idiopathic hemifacial spasm. At each visit, botulinum neurotoxin (2.5 units per 0.1 ml of concentration) was injected into 8-10 sites in the orbicularis oculi and orbicularis oris muscles. The patient completes the 2-year course of



**Figure 1** Shows patient during the episode of hemifacial spasm (See Color figure on page 116)



**Figure 2** MRI brain shows the V4 portion of the left vertebral artery located very close to the left facial nerve. (See Color figure on page 116)

continuous botulinum neurotoxin treatment received a total of 5 treatments of 4 months apart. The mean dose of botulinum neurotoxin was 24 units (range 20-25 units). The response rate according to patient's subjective improvement was 70%. The adverse reaction was weakness of lower facial muscle (House-Brackmann grade III left side) without ptosis or diplopia. However, in later visit, when the dose was fixed, the patient reported the symptom persisted. Magnetic resonance imaging (MRI) brain demonstrated marked tortuous bilateral vertebral arteries with the V4 portion of the left vertebral artery located very close to the left facial nerve (Figure 2). Neuroimaging suggested a left HFS caused by the left vertebral artery dolichoectasia. The patient was referred to the neurosurgeon and underwent suboccipital craniotomy with microvascular decompression.

## Discussion

Disorders of overactivity of the facial nerve included hemifacial spasm (HFS), benign essential

blepharospasm (BEB), facial myokymia, and eyelid myokymia. Any lesions of facial nucleus, facial fascicle, facial nerve, or extra-axial brainstem pathology may produce hyperexcitable states of facial nerve. HFS is unilateral episodic spasm, often begin around the orbicularis oculi muscles, and then spread to ipsilateral side of all facial muscles. Some patients begin as twitching of orbicularis oculi muscles only, but over the course of several years, spreads to involve of all facial muscles on ipsilateral side. The episodes of spasms may last seconds to minutes, persist during sleep and have unknown factors that may exacerbate the spasm<sup>5</sup>. Facial muscle examinations revealed intact facial nerve function in HFS.

BEB is bilateral episodic spasm of orbicularis oculi muscles. The episodes of spasms are similar to HFS, but unlike HFS, the spasms in BEB are related to exacerbating factors such as sunlight exposure, stress or physical activities and BEB is better during sleep.<sup>6</sup> The exact cause of BEM is unknown, but may associated with basal ganglia dysfunction.<sup>7</sup> Neuroimaging is

not required in BEB. Facial myokymia is unilateral undulating contraction of facial muscle bundles. The episodes of abnormal movements often begin within a portion of orbicularis oculi muscles and may spread to involve most of facial muscles. The cause of facial myokymia is an intramedullary pontine lesion involving the facial nucleus or its fascicle.<sup>8</sup> Multiple sclerosis and brainstem tumors are the typical etiologies. Conversely, eyelid myokymia is a unilateral or bilateral benign fasciculation of orbicularis oculi muscles.<sup>9</sup> Most of patients will spontaneously resolve within days or weeks.

Intracranial dolichoectasia is an abnormal increased diameter of the cerebral artery (ectasis) with or without a long and tortuous course (dolichosis) of at least one cerebral artery. Dolichoectasia occurs in about 0.08-6.5% in the normal population.<sup>10</sup> Dolichoectasia may associated with advanced age, hypertension and arteriosclerosis, but most cases are sporadic. Dolichoectasia may compress the surrounding intracranial structures either by the trunk vessel or its branches. Han et al. reported HFS 1642 patients had vertebrobasilar dolichoectasia about 0.7%.<sup>11</sup> Advanced neuroimaging technique of the posterior fossa reveals identifiable causes of HFS include compression of the facial nerve at as its exit from the brainstem by dolichoectasia, cerebellopontine angle tumor, or temporal bone tumor.<sup>12,13</sup> MRI and/or MR angiography (MRA) skull base have a sensitivity of 77.27% and a specificity of 75% to detect the vascular compression.<sup>14</sup>

Medical treatment in HFS include anticonvulsive agents (clonazepam, carbamazepine, baclofen).<sup>15</sup> Oral medications have been used for patients with mild and infrequent HFS. Oral medications are unsatisfactory in those with moderate to severe HFS disrupt daily activities. Botulinum neurotoxin acts on the presynap-

tic cholinergic nerve terminal to inhibit the release of acetylcholine, resulting in muscle paralysis. Although the effect of botulinum is not sustained, the injections are typically repeated at intervals of 3-4 months. Nevertheless, botulinum injection alleviates symptoms of HFS in about 85-95% of patients. Furthermore, injections are minimally invasive procedure and has become the treatment of choice.<sup>16</sup> In our report, the patient was unresponsive to initial treatment with clonazepam and botulinum injections. The author postulated that HFS may be caused by the extra-axial brainstem lesion including tumors and vascular compression of the facial nerve. MRI brain has demonstrated vertebral dolichoectasia which is the most likely cause of intractable HFS. Surgical treatment in HFS include microvascular decompression to remove the vascular compression of the facial nerve.<sup>17</sup> Rosenstengel et al. reported the surgical success rate of 83% and 87% at postoperative 6 months and 18 months, respectively.<sup>18</sup>

Abdel Hamid et al. reported a HFS patient who had previously receive botulinum neurotoxin injection for one time which provided relief of symptoms for months before recurrence. MRI and computerized tomography angiogram (CTA) show vertebrobasilar dolichoectasia causes compression at root entry zone of facial nerve. In the present case, because one-time botulinum neurotoxin injection relieved the most of symptoms, so the authors delay performing neuroimaging. However, when the last adequate botulinum neurotoxin injection shows no improvement, the authors discuss about further investigation for the case. In many countries, nearly all cases with HFS should be perform neuroimaging to exclude the compressive lesion of the facial nerve by tortuous blood vessels or brainstem tumors. In Thailand, some cases have financial problems, so we did not perform neuroimaging

except the patients have neurological signs or botulinum neurotoxin treatment failure as in the present case.<sup>19</sup>

In conclusion, the present case demonstrated that neuroimaging should be considered in patients presenting with intractable hemifacial spasm in spite

of adequate botulinum neurotoxin injection, or those with associated neurological signs to exclude extra-axial brainstem lesions such as vertebral dolichoectasia.

**Potential conflicts of interest:** None

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