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CASE REPORTS

Hemimasticatory spasm misdiagnosed as other mimickers: Report of two cases

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Abstract

Hemimasticatory spasm is a rare neurologic disorder characterized by unilateral, paroxysmal involuntary contraction of the masticatory muscles. It can be confused with other unilateral facial spasms. In this report, we present two patients with hemimasticatory spasm who were initially misdiagnosed with other disorders. The first patient was a 54-year-old man with a 9-year history of right facial spasm. He underwent microvascular decompression, without benefit, at another hospital with the diagnosis of hemifacial spasm two years prior to presenting at our hospital.On examination, right masseter and temporalis muscles had irregular contractions with twitches and prolonged spasms. His brain magnetic resonance imaging was normal.He showed a good response to carbamazepine. The second case was a 60-year-old man presented with a 10-year history of jaw-closing movement. Previous treatment for temporomandibular joint disorder was ineffective. He was previously diagnosed as oromandibular dystonia at another hospital, and biperiden and diazepam treatment resulted in slight improvement. There was tonic contraction and hypertrophy of the left masseter. He improved with carbamazepine. These cases illustrate the importance of hemimasticatory spasm as differential diagnosis, and the good response to carbamazepine.

INTRODUCTION

Hemimasticatory spasm (HMS), which is characterized by unilateral, paroxysmal, involuntary contractions of one or more masticatory muscles, is a rare disorder of the trigeminal nerve motor branch. Gowers[1] first described HMS in 1897, and only a few cases have been reported since then. Because of its rarity, HMS may be confused with other facial movement disorders, such as hemifacial spasm (HFS), temporomandibular joint disorder (TMD), oromandibular dystonia (OMD), facial myoclonus, facial tics, or other causes of abnormal facial movements. To alert clinicians of this disorder, we present here two Korean patients with HMS that were initially misdiagnosed as HFS and OMD.

CASE REPORTS

Case 1

A 54-year-old man with a 9-year history of paroxysmal spasms on the right side of the face was referred to Seoul National University Hospital(SNUH). Spasm episodes occurred several times a day and often precipitated by chewing. By maintaining the mouthslightly open, the contractions could be prevented. However, occasionally the spasms were too severe for him to open his mouth. Two years prior to visiting SNUH, he underwent microvascular decompression (MVD) at another hospital with the diagnosis of HFS without any improvement. He had also been treated with botulinum toxin injections in the facial nerve innervated muscles without any benefit.

Physical examination showed symmetry of facial muscles without hypertrophy of the masseter and temporalis muscles. The right masseter and temporalis muscles showed irregular contractions, ranging from brief twitches to prolonged spasms that could be interrupted by voluntary mouth opening. Facial sensation and muscle power were normal, and other neurological examination was

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also normal. Brain magnetic resonance imaging (MRI) along the path of the trigeminal nerve was unremarkable. Based on the typical clinical features, he was diagnosed with HMS. He showed excellent response to carbamazepine, with initial dose of 200mg tid, which was gradually increased to 400mg tid. The patient remained well on carbamazepine during the 2 years follow up.

Case 2

This was a 60-year-old man who had involuntary, intermittent spasm in his left jaw for 10 years. Initially, his involuntary clenching of jaw was often provoked by chewing. The episodes of spasm occurred during mastication, and occasionally were accompanied by pain. He went to a dentist and was diagnosed as TMD. He was given splint therapy, which was ineffective. Biperiden and diazepam were given at another clinic with diagnosis of OMD, which resulted in slight improvement of symptom.

He presented to SNUH due to progressive worsening of symptoms with increasing spasm frequency and intensity. By then, speaking also precipitated muscle spasms (Video1). Sudden clenching of the teeth would occasionally cause injuries to his tongue and oral mucosa. The spasms occurred many times a day. Neurological examination revealed hypertrophy of the left masseter muscle and a mild hollowing of the left cheek. Facial sensation and muscle strength were normal. Other clinical findings were also normal. Needle electromyography (EMG) showed spasm of the left masseter muscle without co-contraction of left temporalis muscle and right masseter muscles. There were no abnormality in blink reflex, facial nerve excitability, and facial nerve stimulation tests. Carbamazepine (200mg tid) resulted in dramatic reduction of the intensity and frequency of the spasm. The good response to carbamazepine was maintained during the 1-year follow up.

DISCUSSION

HMS is a rare movement disorder characterized by paroxysmal contractions of unilateral jaw-closing muscles, such as the masseter, temporalis, and medial pterygoid muscles.[2]To our knowledge, only 38 cases of HMS have been reported in the literature. In our review of the 38 cases reported (Table 1), women appear to be affected more frequently than men (female:male = 2.8:1.0). The mean age at onset was 39 years (range, 15-63 years). The involuntary contractions usually last from a few seconds to several minutes. The spasm episodes may occur many times a day, often precipitated by chewing, speaking or other voluntary jaw-closing, and may be interrupted by voluntary jaw opening. Occasionally, sudden and severe spasms may result in biting of tongue or oral mucosa, breaking of teeth, or even dislocation of the temporomandibular joint.[2-5] Hypertrophy of the involved muscle commonly occurs and may be accompanied by atrophy of the subcutaneous tissue or localized scleroderma.[6-12] With detailed history, careful observation and awareness of the condition, the diagnosis of HMS is not difficult. However, as shown in our patients, misdiagnosis is not uncommon, including that of HFS (Case 1), TMD (Case 2), and OMD (Case 2).

In contrast to HMS, HFSis an involuntary clonic or tonic movement of muscles innervated by the facial nerve, and the distribution of the involved muscles varies with the affected branches.[13] Vascular contact with thefacial nerve is the most common attributed etiology, which is the theoretical basis of undertaking MVD. The success rate of MVD is said to be up to 90%.[14] A common symptom in both HMS and HFS is the unilateral involvement of muscles of the head. However, HMS involved the muscles of mastication, whereas HFS involved the facial muscles and the spasm usually begins in the region of the orbicularis oculi muscle, which then gradually spreads to other muscles on the ipsilateral face.

As for TMD, it is due to dysfunction of the temporomandibular joint and the adjacent orofacial structures.[15]Common symptoms of TMD include chronic pain or tenderness in the orofacial area during speaking or chewing, with restriction of jaw opening. Clicking or popping noise sounds in the temporomandibular joint may occur during jaw movement. However, unlike HMS, jaw muscle spasm is not a dominant feature of TMD.

OMD usually presents with repeated involuntary sustained contractions that affect mainly the muscles of the lower part of the face, including the mouth, jaw, tongue, and pharynx.[16] OMD is usually bilateral, but unilateral involvement can also be occasionally seen.[7] Differentiation of HMS from unilateral OMD may be difficult, but the brief and paroxysmal spasms of HMS is unlike that of OMD. In contrast, sensory tricks can be effective in patients with OMD, but not in patients HMS.

Tic and facial myoclonus can also mimic HFS or HMS.[17] Patients with tic have premonitory

Table 1: Cases of hemimasticatoryspasm in the literature

Author, Year (Ref)	Sex/ Age at onset	Involved muscles	Carbamazepine
1. Auger, 1992[2]	F/20	R masseter and temporalis	-
	F/17	R medial pterygoid	Beneficial
	F/20	R masseter and temporalis	No benefit
2. Kaufman, 1980[3]	F/25	L masseter	No benefit
3. Esteban, 2002[4]	F/47	L masseter	-
4. Christie, 2014[5]	F/32	R masseter	-
5. Lapresle, 1982[a6]	F/15	R masseter	-
6. Thompson, 1986[7]	F/31	R masseter	Beneficial
7. Parisi, 1987[8]	F/38	R masseter	Not mentioned
8. Cruccu, 1994[9]	M/18	L temporalis	Beneficial
	F/44	R masseter and temporalis	Beneficial
9. Ebersbach,1995[10]	F/26	L masseter and temporalis	-
	F/30	R masseter and temporalis	-
10. Kim, 2000[11]	F/34	R masseter	-
11. Kumar, 2008[12]	F/49	L masseter temporalis and lateral pterygoid	No benefit
12. Sinha, 2011[16]	M/38	R masseter and temporalis	-
13. Yaltho, 2011[17]	F/63	L masseter	-
14. Gunduz,2007[20]	F/62	R masseter and temporalis	-
15. Jimenez, 2008[21]	M/40	R masseter and temporalis	-
16. Gopalakrishnan,2011[22]	F/56	L masseter and temporalis	-
17. Mir, 2006[23]	M/26	L masseter and temporalis	Beneficial
18. Teive,2002[24]	F/44	R masseter and temporalis	No benefit
19. Cersosimo, 2004[25]	F/29	R masseter and temporalis	-
20. Chon,2012[26]	M/40	R masseter and temporalis	Not mentioned
21. Wang,2013[27]	F/50	L masseter	-
	F/42	R masseter and temporalis	-
	M/38	R masseter	-
	F/48	R masseter	-
	F/57	L masseter and temporalis	-
	F/53	R masseter and temporalis	-
22. Dou, 2014[28]	F/45	L masseter	-
23. Thompson, 1983[29]	F/57	L masseter and temporalis	-
24. Yoshii, 1989[a30]	M/44	L masseter, lateral and medial pterygoid	-
25. Kim,1994[b31]	M/44	R masseter	No benefit
26. Wang,2004[c32]	F/39	L masseter	-
	M/32	R masseter and temporalis	-
	M/53	R temporal	-
	F/52	L masseter and temporalis	-

[a]Only the abstract is available in English.
[b]The article was published in Korean.
[c]The article was published in Chinese.
Ref, reference; L, left; R, right; EMG, electromyography; F, female; M, male; CT, computed tomography; MRI, magnetic resonance imaging; MRA, magnetic resonance angiography; MVD, microvascular decompression; CPA, cerebellopontine angle; SCA, superior cerebellar artery.

sensation, bizarre and purposeful movement, and can suppress the movements temporarily.[18] Facial myoclonus is characterized by brief, sudden, shock-like muscle jerks.[19] These are different from the paroxysmal tonic contractions of masticatory muscles innervated by the trigeminal motor nerve in HMS.

EMG and MRI can be helpful in localizing the pathological locationand help clarify the diagnosis. Characteristic EMG results for HMS are normal motor unit potentials with high frequencies that correlate with the involuntary spasms of masticatory muscles, mostly affecting the masseter, and then the temporalis muscles[2,9,11], which is seen in our Case 2. MRI helps to determine secondary causes of HMS from brain lesions including pontine infarction[20], biopercular syndrome[21], and cerebellopontine angle hematoma[22] or along the trigerminal nerve. Subcutaneous tissue atrophy and muscle hypertrophy can be easily identified through coronal MRI.[11] Magnetic resonance angiography may help to elucidate vascular pathology associated with the trigeminal nerve.

An initial trial with oral drugs such as carbamazepine should be attempted[7,9,23], which was very beneficialin both of our cases. A review of reported cases shows that carbamazepine was effective in 5/12 cases (42%) that the drug was used (Table 1). Botulinum toxin can also attenuate the spasms.[5,10-12,20,21,23-26] It can be tried if carbamazepine fails. Although there are several reports that MVD of trigeminal nerve root was effective in the treatment of HMS, its exact role in the management of HMS is still uncertain.[26-28]

The mechanisms of HMS remain unclear. The electrophysiological similarity between HMS and HFS suggest that same mechanism may play a role in both conditions.[3,29] The pathogenesis of HMS in most cases is speculated to be compression of the trigeminal motor nerve or nerve branch followed by demyelination. In electrophysiological investigations, an absent or reduced masseter silent period and an impaired inhibition of masseter reflex during muscle spasm may be related to ectopic excitation that is secondary to focal demyelination of trigeminal motor fibers.[2] Compared with medial and lateral pterygoid muscles, masseter and temporalis muscles are more likely to be involved in HMS. It has been speculated that the trigeminal nerve branch to the masseter and temporalis muscles is compressed between the lateral pterygoid muscle and skull surface as a result of change in the adjacent tissue.[9,11] Under surgical exploration, vascular contact along the motor root of the trigeminal nerve has been demonstrated in some cases of HMS, and such contact can be relieved by MVD.[26-28] Other than a peripheral mechanism, HMS secondary to pontine infarction has been reported and may be associated with hyperactivity of the trigeminal motor nucleus.[20]

Since involuntary movements of the jaw may be due to a variety of conditions and because HMS is a rare disorder, early diagnosis may be difficult. Although neurophysiological and neuroimaging studies may be helpful, awareness of the condition and careful observation are crucial to avoid delay in diagnosis.

DISCLOSURE

Conflict of interest: None

Legends to the video

Video 1. http://www.neurology-asia.org/content/20/1/neuroasia-2015-20(1)-73-v1.avi The video shows brief clenching of the jaw induced severe repetitive spasms of the left masseter, which is visibly hypertrophied. There was difficulty in opening the mouth. The right side of his face is normal.

REFERENCES

1. Gowers WR. A manual of diseases of the nervous system. 2nd ed. Philadelphia:Blakiston, 1897: 585-7.
2. Auger RG, Litchy WJ, Cascino TL, Ahlskog JE. Hemimasticatory spasm: clinical and electrophysiologic observations. *Neurology* 1992; 42(12):2263-6.
3. Kaufman MD. Masticatory spasm in facial hemiatrophy. *Ann Neurol* 1980; 7(6):585-7.
4. Esteban A, Traba A, Prieto J, Grandas F. Long term follow up of a hemimasticatory spasm. *Acta Neurol Scand* 2002; 105(1):67-72.
5. Christie C, Rodriguez-Quiroga SA, Arakaki T, Rey RD, Garretto NS. Hemimasticatory spasm: report of a case and review of the literaturedagger. *Tremor Other Hyperkinet Mov (N Y)* 2014; 4:210.
6. Lapresle J, Desi M. Scleroderma with progressive facial hemiatrophy and atrophy of the other side of the body. *Rev Neurol (Paris)* 1982; 138(11):815-25.
7. Thompson PD, Obeso JA, Delgado G, Gallego J, Marsden CD. Focal dystonia of the jaw and the differential diagnosis of unilateral jaw and masticatory spasm. *J Neurol Neurosurg Psychiatry* 1986; 49(6):651-6.
8. Parisi L, Valente G, Dell'Anna C, Mariorenzi R, Amabile G. A case of facial hemiatrophy associated with linear scleroderma and homolateral masseter spasm. *Ital J Neurol Sci* 1987; 8(1):63-5.
9. Cruccu G, Inghilleri M, Berardelli A, *et al*. Pathophysiology of hemimasticatory spasm. *J Neurol Neurosurg Psychiatry* 1994; 57(1):43-50.

10. Ebersbach G, Kabus C, Schelosky L, Terstegge L, Poewe W. Hemimasticatory spasm in hemifacial atrophy: diagnostic and therapeutic aspects in two patients. *Mov Disord* 1995; 10(4):504-7.
11. Kim HJ, Jeon BS, Lee KW. Hemimasticatory spasm associated with localized scleroderma and facial hemiatrophy. *Arch Neurol* 2000; 57(4):576-80.
12. Kumar N, Krueger BR, Ahlskog JE. Hemimasticatory spasm with lateral jaw deviations. *Mov Disord* 2008; 23(15):2265-6.
13. Wang A, Jankovic J. Hemifacial spasm: clinical findings and treatment. *Muscle Nerve* 1998; 21(12):1740-7.
14. Miller LE, Miller VM. Safety and effectiveness of microvascular decompression for treatment of hemifacial spasm: a systematic review. *Br J Neurosurg* 2012; 26(4):438-44.
15. Dym H, Israel H. Diagnosis and treatment of temporomandibular disorders. *Dent Clin North Am* 2012; 56(1):149-61.
16. Sinclair CF, Gurey LE, Blitzer A. Oromandibular dystonia: long-term management with botulinum toxin. *Laryngoscope* 2013; 123(12):3078-83.
17. Yaltho TC, Jankovic J. The many faces of hemifacial spasm: differential diagnosis of unilateral facial spasms. *Mov Disord* 2011; 26(9):1582-92.
18. Martino D, Mink JW. Tic disorders. *Continuum (Minneap Minn)* 2013; 19(5):1287-311.
19. Espay AJ, Chen R. Myoclonus. *Continuum (Minneap Minn)* 2013; 19(5):1264-86.
20. Gunduz A, Karaali-Savrun F, Uluduz D. Hemimasticatory spasm following pontine infarction. *Mov Disord* 2007; 22(11):1674-5.
21. Jimenez-Jimenez FJ, Puertas I, Alonso-Navarro H. Hemimasticatory spasm secondary to biopercular syndrome. *Eur Neurol* 2008; 59(5):276-9.
22. Gopalakrishnan CV, Dhakoji A, Nair S. Hemimasticatory spasm following surgery for vestibular schwannoma. *Mov Disord* 2011; 26(14):2481-2.
23. Mir P, Gilio F, Edwards M, *et al*. Alteration of central motor excitability in a patient with hemimasticatory spasm after treatment with botulinum toxin injections. *Mov Disord* 2006; 21(1):73-8.
24. Teive HA, Piovesan EJ, Germiniani FM, *et al*. Hemimasticatory spasm treated with botulinum toxin: case report. *Arq Neuropsiquiatr* 2002; 60:288-9.
25. Cersosimo MG, Bertoti A, Roca CU, Micheli F. Botulinum toxin in a case of hemimasticatory spasm with severe worsening during pregnancy. *Clin Neuropharmacol* 2004; 27(1):6-8.
26. Chon KH, Lee JM, Koh EJ, Choi HY. Hemimasticatory spasm treated with microvascular decompression of the trigeminal nerve. *Acta Neurochir (Wien)* 2012; 154(9):1635-9.
27. Wang YN, Dou NN, Zhou QM, *et al*. Treatment of hemimasticatory spasm with microvascular decompression. *J Craniofac Surg* 2013; 24(5):1753-5.
28. Dou NN, Zhong J, Zhou QM, Zhu J, Wang YN, Li ST. Microvascular decompression of trigeminal nerve root for treatment of a patient with hemimasticatory spasm. *J Craniofac Surg* 2014; 25(3):916-8.
29. Thompson PD, Carroll WM. Hemimasticatory spasm-a peripheral paroxysmal cranial neuropathy? *J Neurol Neurosurg Psychiatry* 1983; 46(3):274-6.
30. Yoshii K, Seki Y, Aiba T. A case of unilateral masticatory spasm without hemifacial atrophy. *No To Shinkei* 1989; 41(4):343-6.
31. Kim YJ, Lee KS, Na JH, Kim BS, Ko YJ. A Case of Hemimasticatory spasm. *J Korean Neurol Assoc* 1994; 12(1):175-8.
32. Wang YW, Ma XC, Zhang ZK, Shen DG, Su FX, Fu KY. Hemimasticatory muscle spasm: an electromyogram analysis. *Zhonghua Kou Qiang Yi Xue Za Zhi* 2004; 39(2):155-7.