Different Faces of Hemifacial Spasm: Etiological Classification

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ABSTRACT

OBJECTIVE: To propose an etiological classification of hemifacial spasm (HFS). **BACKGROUND:** HFS is characterized by unilateral, involuntary, irregular, clonic or tonic movements of muscles innervated by the seventh cranial nerve. Usually without any identifiable etiology, HFS has been most frequently attributed to vascular loop compression at the root exit zone of the facial nerve. Other etiologies are rare but facial nerve injury, Bell's palsy, brain tumor, multiple sclerosis, and genetic causes of HFS have been reported. Treatment with botulinum toxin (BTX) effectively induces sustained relief from symptoms of HFS in the long term, with only minimal and transient adverse reactions. **METHODS:** Medical records and videos of consecutive patients referred to the Movement Disorders Clinic, Baylor College of Medicine, during the period 2000 to 2006 were reviewed and all relevant data, including demographic and clinical information, was entered into a database and analyzed with a specific focus on etiology. **RESULT:** Among 140 patients with HFS we found the following etiologies: 1) 88 (63 %) presumably due to vascular compression of the facial nerve; 2) 13 (9 %) synkinesis following Bell's palsy; 3) 10 (7%) facial nerve injury; 4) 1 facial myoclonus caused by Rasmussen encephalitis; 5) 1 facial dystonia; 6) 11 (8%) facial tics; 7) 1 multiple sclerosis, 8) 2 vascular insults; 9) 1 familial HFS; and 10) 11 (8 %) psychogenic. Video of illustrative cases in each group will be shown. **CONCLUSION:** Although most cases of HFS are probably caused by vascular compression of the facial nerve, other etiologies should be considered, particularly when atypical features are present.

INTRODUCTION

1. Hemifacial spasm (HFS) is characterized by involuntary, irregular, clonic or tonic movements of muscles innervated by the seventh cranial nerve on one side of the face.
2. Usually without any identifiable etiology, HFS has been most frequently attributed to compression of the facial nerve at the root exit zone by an ectopic anatomical or pathological structure resulting in ephaptic transmission [7].
3. Other reported causes of facial nerve compression resulting in HFS include vascular abnormalities[6, 10], tumors[6], structural abnormalities[1], otitis media with effusion[4], vacular headache[2] and lacunar pontine infarction[11].
4. Although nearly always unilateral, rare cases of bilateral HFS have been reported[8].
5. Several families with HFS have been reported, suggesting that some patients are genetically predisposed to develop this disorder[5].
6. HFS can also be the initial or only manifestation of a psychogenic movement disorder[9].
7. Peripheral facial nerve injury or prior Bell's palsy can also result in HFS as a result of aberrant regeneration of the facial nerve.
8. HFS may be confused with other facial movement disorders, such as blepharospasm, oromandibular dystonia, facial tic, hemimasticatory spasm and facial myokymia.

METHODS

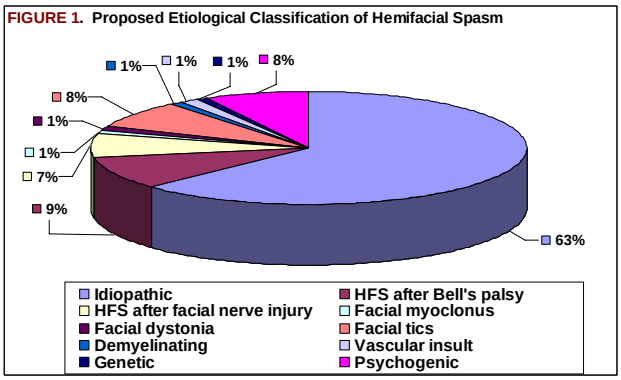
During the period from 2000 to 2006, 135 patients with the diagnosis of HFS were evaluated at the Movement Disorders Clinic, Baylor College of Medicine. Medical records and videos of all patients with the diagnosis of HFS were carefully reviewed.

RESULTS

Table1.

Classification	N (%)	Sex (M/F)	Age at onset (years)	Duration (years)
1. Idiopathic (presumably vascular)	88 (65%)	52/83	61.15±15.2	13.21±7.20
2. Synkinesis after Bell's palsy	13 (10%)	1/12	56.5±18.1	3.75±8.45
3. Facial nerve injury	10 (8%)	2/8	44.9±9.56	3.48±6.72
4. Facial myoclonus	1	F	20	6
5 .Facial dystonia	1	M	63	2
6. Facial tics	11 (8%)	7/4	38.45±22.29	12.9±10
7.Demyelinating	1	F	64	1.5
8. Vascular insult	2	1/1	72/72	7years/4 months
9.Familial	1	F	81	30
10.Psychogenic	11 (8%)	1/10	35.66±18.68	2.22±2.28

FIGURE 1. Proposed Etiological Classification of Hemifacial Spasm



DISCUSSION

1. We summarized our case series and reviewed the literatures and proposed an etiological classification of HFS as following:
   i. Compression of seventh cranial nerve
   ii. Synkinesis after Bell's palsy and HFS after facial nerve injury
   iii. Facial myoclonus
   iv. Facial Tics
   v. Demyelination
   vi. Genetic
   vii. Psychogenic
   viii. Cortical origin
   ix. Hypertension and ventrolateral medulla compression
   x. Hyperactive facial motor nucleus
2. In our patient series, 9% had HFS after Bell's palsy and 7 % patients had HFS secondary to nerve injury. Those patients did not differ from idiopathic HFS patients in terms of age at onset and clinical presentations as previous report[12]. The mean interval between Bell's palsy and nerve injury are similar as well (3.75±8.45 years v.s.3.48±6.72 years).
3. Facial myoclonus can mimic HFS. The case we presented here share some common features with HFS with constant contraction of the orbicularis oculi and zygomaticus muscle on the left side of face which were exacerbated by speech or smile.
4. Facial tics can be confused with HFS if they occur in isolation, but many patients frequently describe a premonitory sensation prior to the tics and, in contrast to HFS, tics are often suppressible [3]. In our cases, facial tics can be part of multiple motor tics (64 %) or the only single tic when patient fist sought for medical help (36 %). A detail history should be obtained for previous history of tics or other comorbid disorder such as obsessive-compulsive disorder, attention deficit with/without hyperactivity.
5. Several familial with HFS have been reported suggesting that some patients are genetically predisposed to develop this disorder. In our case, an 81-year-old Caucasian female presents with bilateral HFS with age at onset around 50. Two of her sisters also showed HFS. There was no clear difference in her clinical manifestations with other sporadic patients with HFS. There may be a pattern of autosomal-dominant inheritance with low penetrance and possible younger age of onset.
6. In previous report, psychogenic facial spasm was diagnosed in 2.4% of patients evaluated for HFS with a mean age of 34.6 years, which was considerable younger than that of patients with organic HFS, most of whom usually present after age 40 [9]. In the present study, 8% of our patients referred for evaluation for HFS were diagnosed with psychogenic facial spasm. The mean age of onset is 35.66±18.68 years, and female account for most of those cases (10 out of 11).
7. 7 patients (8%) in this group were Asian, in contrast to 131 patients of oro-mandibular dystonia in our data base in the same period of time, none of them was Asian. This finding may further support previous observation that HFS is more frequent in Asian population

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