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, 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) 8 (6%) presumably attributable to vascular compression of the facial nerve; 2) 13 (9%) synkinesias 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, 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[1].
3. Other reported causes of facial nerve compression resulting in HFS include vascular abnormalities[2,10], tumors[11], structural abnormalities[12], otitis media with effusion[13], vascular headache[14] and lacunar pontine infarction[15]. Although nearly always unilateral, rare cases of bilateral HFS have been reported[8].
4. Several families with HFS have been reported, suggesting that some patients are genetically predisposed to develop this disorder[16].
5. HFS can also be the initial or only manifestation of a psychogenic movement disorder[17].
6. Peripheral facial nerve injury or prior Bell’s palsy can also result in HFS as a result of aberrant regeneration of the facial nerve.
7. HFS may be confused with other facial movement disorders, such as blepharospasm, oromandibular dystonia, facial tic, hemimasticatory spasm and facial myokymia.

RESULTS

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.

FIGURE 1. Proposed Etiological Classification of Hemifacial Spasm

<table>
<thead>
<tr>
<th>Classification</th>
<th>N (%)</th>
<th>Sex (M/F)</th>
<th>Age at onset (years)</th>
<th>Duration (years)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. Idiopathic (presumably vascular)</td>
<td>88 (65%)</td>
<td>52/83</td>
<td>61.15±15.2</td>
<td>13.21±7.20</td>
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<tr>
<td>2. Synkinesia after Bell’s palsy</td>
<td>13 (10%)</td>
<td>1/1</td>
<td>56.5±18.1</td>
<td>3.75±4.54</td>
</tr>
<tr>
<td>3. Facial nerve injury</td>
<td>10 (8%)</td>
<td>2/8</td>
<td>44.9±9.56</td>
<td>3.48±6.72</td>
</tr>
<tr>
<td>4. Facial myoclonus</td>
<td>1</td>
<td>F</td>
<td>20</td>
<td>6</td>
</tr>
<tr>
<td>5. Facial dystonia</td>
<td>1</td>
<td>M</td>
<td>63</td>
<td>2</td>
</tr>
<tr>
<td>6. Facial tics</td>
<td>11 (8%)</td>
<td>7/4</td>
<td>38.45±22.29</td>
<td>12.9±10</td>
</tr>
<tr>
<td>7. Demyelinating</td>
<td>1</td>
<td>F</td>
<td>64</td>
<td>1.5</td>
</tr>
<tr>
<td>8. Vascular insult</td>
<td>2</td>
<td>1/1</td>
<td>72/72</td>
<td>7 years/4 months</td>
</tr>
<tr>
<td>9. Familial</td>
<td>1</td>
<td>F</td>
<td>81</td>
<td>30</td>
</tr>
<tr>
<td>10. Psychogenic</td>
<td>11 (8%)</td>
<td>1/10</td>
<td>35.66±18.68</td>
<td>2.22±2.28</td>
</tr>
</tbody>
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REFERENCES