HEMIFACIAL SPASM—A REVERSIBLE PATHOPHYSIOLOGIC STATE*

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In 19 patients having hemifacial spasm, surgical exposure and neurolysis of the 7th nerve was performed in the cerebellopontine angle. In 7 cases the nerve was found to be compressed by a definite pathologic process, and in an additional 7 by a redundant loop of the anterior inferior cerebellar or internal auditory artery. In 5 there was no apparent compression of the nerve. Gentle manipulation of the exposed nerve reproduced the hemifacial spasm. The operation was followed by complete relief of spasm in 12 patients, by satisfactory relief in 5, by no relief in 2 and by recurrence in 2. This article, however, concerns mechanism rather than treatment.

REVIEW OF LITERATURE

Wilson introduced the subject of hemifacial spasm as follows:

"Facial spasm may be cryptogenic or symptomatic, non- or postparalytic, uni- or bilateral, partial or total, tonic, clonic, tonicoclonic, or fibrillar. Common though it is, both causation and pathogenesis are obscure, while pathological data are scanty and ambiguous."

In 1945 Ehni and Woltman analyzed 106 cases of cryptogenic hemifacial spasm selected from the records of 663 patients with various "unwonted" movements of the face. They pointed out that the spasm was limited to the muscles supplied by the 7th nerve and characterized by contractions resembling a response to intermittent faradization of the nerve. They found that the spasm usually originated in the orbicularis oculi which was the muscle involved most frequently, that the patient felt no compulsion to make the movement and was unable to stop it by exercise of the will. It occurred predominantly in women, only in adults, and with equal frequency on either side. Psychic upsets, fatigue, and voluntary movements made the spasms worse, but in 12 patients spasms also were observed during sleep. In 6 of their cases the spasms were bilateral but were neither synchronous nor symmetric. Nine of their patients had experienced a spontaneous remission. There was trigeminal neuralgia on the side of the spasm in 3 patients. In 2 the hemifacial spasm had been preceded by facial paralysis although in neither was the twitching of an associated-movement type.

Ehni and Woltman confirmed the previous observations of others, that the spasm never spreads beyond the distribution of the 7th nerve, is not affected by a stroke, and therefore cannot be the result of a cortical discharge. They found that regeneration following interruption of the facial nerve at the stylomastoid foramen almost always was accompanied by recurrence of the spasm, whereas, as had been shown previously, if the facial nerve was divided at this foramen and anastomosed with the 11th or 12th cranial nerve, the reinnervation of the face was not accompanied by recurrence of the spasm. These demonstrations, they pointed out, proved that the lesion causing this condition must lie in the portion of the nerve between the stylomastoid foramen and the nucleus. They found that little or no benefit was to be expected from the use of drugs including Dilantin Sodium.

Although Ehni and Woltman concluded that gross lesions of the facial nerve can and do cause symptomatic twitching that is very
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like if not indistinguishable from true or cryptogenic hemifacial spasm, they nevertheless excluded from their series every case in which there was evidence of a gross pathologic lesion in the posterior cranial fossa. They apparently did not consider the associated neurologic abnormalities in 41 patients as evidence of such a lesion.

Wartenberg\textsuperscript{20} in 1952 stressed the fact that in hemifacial spasm there frequently is definite though mild weakness of the affected facial muscles as well as the intrafacial associated movements (synkinesis) that sometimes develop after recovery from Bell’s palsy. He pointed out the many similarities between cryptogenic and postparalytic facial spasm and concluded that both are caused by a lesion in the facial nucleus. A study of the illustrations of facial asymmetry that he interpreted as weakness, however, shows it more likely to be the result of the associated contraction (synkinesis) of antagonistic muscles. He agrees with previous authors that:

“Gross organic lesions directly affecting the nucleus or trunk of the facial nerve can certainly produce the clinical picture of hemifacial spasm. . . . But practically speaking, in the genesis of hemifacial spasm such gross organic lesions are so rare as to be negligible.”

In 1917 Cushing\textsuperscript{1} reported 4 instances of facial twitching in 30 patients having tumors of the 8th nerve: an illustration of 1 of these patients discloses classic hemifacial spasm. In 1920 he\textsuperscript{3} described the association of hemifacial spasm with tic douloureux in 3 patients and applied the term tic convulsif to this combination. Revilla, reporting Dandy’s experience with tumors in the cerebellopontine angle, described hemifacial spasm in 4 of 160 patients with neurinomas,\textsuperscript{16} in 1 of 13 with meningiomas,\textsuperscript{17} and in 1 of 13 with cholesteatomas. Laine\textsuperscript{14} described hemifacial spasm in a patient having cistoid aneurysm of the basilar artery, while Campbell and Keedy\textsuperscript{2} found the same lesion in 2 patients with the combination of hemifacial spasm and trigeminal neuralgia.

Woltman \textit{et al.}\textsuperscript{21} carried out a neurolysis by uncovering the facial nerve in its bony canal but abandoned the procedure\textsuperscript{22} when analysis of the results showed that only 2 of 10 patients had lasting benefit. They were surprised to find that their patients also were relieved of their synkinesis, since this form of associated movement generally is attributed to anatomic misdirection of regenerating nerve fibers. As a possible explanation for hemifacial spasm and its accompanying synkinesis they suggested that local irritation of the nerve causes spontaneous activity and facilitates the initiation of impulses in inactive fibers by impulses traveling over adjacent fibers.

Varying degrees of relief of hemifacial spasm have been described following various types of partial interruption of the extracranial portion of the facial nerve\textsuperscript{3,4,11,15,18} but perhaps the first planned attempt to relieve cryptogenic hemifacial spasm by an intracranial approach was by Bradgon.\textsuperscript{1} He crushed the nerve with a hemostat in the cerebellopontine angle in 6 patients, producing complete facial paralysis that subsequently recovered with no return of the hemifacial spasm. The first of his patients so treated has been relieved for 11 years. We believe it significant that while this procedure produced temporary paralysis of efferent fibers, it probably resulted in permanent interruption of afferent fibers. Bradgon’s results thus lend support to Hunt’s\textsuperscript{19} concept that hemifacial spasm may result from irritation of the sensory portion of the 7th nerve “conveyed directly to its motor nucleus.”

\textbf{ANALYSIS OF CASES}

In our 19 patients the ages at operation varied from 38 to 75 years, and the duration of the hemifacial spasm from 5 months to 18 years (Table 1). Fifteen of the patients were women and 4 were men. In 9 cases the right side was involved, in 8 the left. Only 1 of these patients (Case 3B) had experienced a spontaneous remission. The spasm was entirely unilateral with the exception of Case 6B in whom there were observed occasional mild fasciculations in the orbicularis oculi of the opposite side. In most, if not all cases, the spasm had appeared first in the orbicularis oculi and this muscle was involved in the spasm in every instance at the time of the
### TABLE 1

Summary of 19 cases of patients operated upon for hemifacial spasm*

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Age Duration (Years)</th>
<th>Sex</th>
<th>Side</th>
<th>Other Neurologic Abnormality</th>
<th>Operative Findings</th>
<th>Result</th>
<th>Follow-Up</th>
</tr>
</thead>
<tbody>
<tr>
<td>1A</td>
<td>66</td>
<td>F</td>
<td>R</td>
<td>None</td>
<td>7th nerve compressed by cirsoid aneurysm basilar artery</td>
<td>Marked</td>
<td>Immediate complete relief. Recurrence in 5 mo.</td>
</tr>
<tr>
<td>2A</td>
<td>57</td>
<td>M</td>
<td>L</td>
<td>None</td>
<td>7th nerve compressed by cirsoid aneurysm basilar artery</td>
<td>Marked</td>
<td>Immediate complete relief. Mild facial weakness</td>
</tr>
<tr>
<td>3A</td>
<td>58</td>
<td>F</td>
<td>R</td>
<td>None</td>
<td>7th nerve compressed by cirsoid aneurysm basilar artery. Nervus intermedius divided?</td>
<td>Mild</td>
<td>After 4 days satisfactory relief. Sense of taste preserved</td>
</tr>
<tr>
<td>4A</td>
<td>73</td>
<td>F</td>
<td>R</td>
<td>Trigeminal neuralgia rt.</td>
<td>Arteriovenous malformation C.P.A. Large artery transfixed trigeminal root</td>
<td>Not tested</td>
<td>Immediate complete paralysis of face. Recovery in 3 mo. with no recurrence. T.N. relieved by alcohol block</td>
</tr>
<tr>
<td>7A</td>
<td>49</td>
<td>M</td>
<td>R</td>
<td>Dysphagia, ataxia and numbness rt. face</td>
<td>Pons displaced to rt. compressing 5th, 7th and 8th nerves</td>
<td>Marked</td>
<td>Immediate complete relief. Very mild facial weakness. Recurrence in 4 mo.</td>
</tr>
<tr>
<td>1B</td>
<td>38</td>
<td>F</td>
<td>L</td>
<td>Complete nerve deafness lt. Mixed deafness rt. Virtually absent caloric responses bilaterally. Vertigo on eating</td>
<td>Loop of i.a.a. (possibly a.i.c.a.) displaced 8th nerve forward against 7th</td>
<td>Not tested</td>
<td>Immediate, almost complete relief</td>
</tr>
<tr>
<td>2B</td>
<td>55</td>
<td>F</td>
<td>R</td>
<td>None</td>
<td>7th nerve dislocated posteriorly by loop of a.i.c.a.</td>
<td>Marked</td>
<td>Immediate satisfactory relief</td>
</tr>
<tr>
<td>3B</td>
<td>54</td>
<td>F</td>
<td>R</td>
<td>None</td>
<td>7th nerve transfixed by a.i.c.a.</td>
<td>Not tested</td>
<td>Immediate complete relief</td>
</tr>
</tbody>
</table>

* In Group A the 7th nerve was compressed by a pathologic process. C.P.A. = cerebellopontine angle.

In Group B the 7th nerve was compressed by the anterior inferior cerebellar artery (a.i.c.a.) or internal auditory artery (i.a.a.).

In Group C the 7th nerve was not compressed by any visible structure.
<table>
<thead>
<tr>
<th>Case No.</th>
<th>Age Duration (Years)</th>
<th>Sex</th>
<th>Side</th>
<th>Other Neurologic Abnormality</th>
<th>Operative Findings</th>
<th>Irritability</th>
<th>Result</th>
<th>Follow-up</th>
</tr>
</thead>
<tbody>
<tr>
<td>4B</td>
<td>69 3</td>
<td>F</td>
<td>R</td>
<td>None</td>
<td>7th nerve compressed by loop of a.i.c.a. lying anterior to it</td>
<td>Marked</td>
<td>Immediate satisfactory relief</td>
<td>6 mo.</td>
</tr>
<tr>
<td>5B</td>
<td>61 8</td>
<td>M</td>
<td>L</td>
<td>None</td>
<td>7th nerve displaced and separated from nervus intermedius by large i.a.a. Nervus intermedius divided</td>
<td>Mild</td>
<td>After 5 days complete relief. Mild facial weakness, cleared completely by 10th day. Sense of taste lost</td>
<td>4 mo.</td>
</tr>
<tr>
<td>6B</td>
<td>75 6</td>
<td>F</td>
<td>L</td>
<td>None</td>
<td>7th nerve distorted by both i.a.a. and a.i.c.a. Nervus intermedius divided</td>
<td>Mild</td>
<td>After 3 days, complete relief. Sense of taste lost</td>
<td>3½ mo.</td>
</tr>
<tr>
<td>7B</td>
<td>62 6½</td>
<td>F</td>
<td>L</td>
<td>None</td>
<td>7th nerve distorted by very large a.i.c.a. Nervus intermedius divided?</td>
<td>Mild</td>
<td>Immediate complete relief. Sense of taste preserved</td>
<td>1 mo.</td>
</tr>
<tr>
<td>1C</td>
<td>44 8/12</td>
<td>F</td>
<td>L</td>
<td>None</td>
<td>7th nerve uninvolved</td>
<td>Not tested</td>
<td>After 2 days, satisfactory relief. Transient mild exacerbation at 2 yrs.</td>
<td>4 yrs.</td>
</tr>
<tr>
<td>2C</td>
<td>67 5</td>
<td>M</td>
<td>L</td>
<td>Bilateral nerve deafness of equal degree. Caloric responses normal</td>
<td>7th nerve uninvolved</td>
<td>Not tested</td>
<td>No relief</td>
<td>3 yrs.</td>
</tr>
<tr>
<td>3C</td>
<td>44 4</td>
<td>F</td>
<td>L</td>
<td>None</td>
<td>7th nerve uninvolved</td>
<td>Not tested</td>
<td>Immediate complete relief</td>
<td>90 mo.</td>
</tr>
<tr>
<td>4C</td>
<td>62 5/12</td>
<td>F</td>
<td>L</td>
<td>None</td>
<td>7th nerve uninvolved</td>
<td>Marked</td>
<td>After 3 wks. complete relief</td>
<td>9 mo.</td>
</tr>
<tr>
<td>5C</td>
<td>44 18</td>
<td>F</td>
<td>R</td>
<td>None</td>
<td>7th nerve uninvolved</td>
<td>Mild</td>
<td>No relief</td>
<td>4 mo.</td>
</tr>
</tbody>
</table>

examination. In no case was the spasm preceded by Bell's palsy. Pressure or tapping over facial muscles or nerve did not induce contractions.

The cases have been divided into three groups according to the operative findings (Table 1). Group A consists of 7 cases in which the 7th nerve was compressed by an obvious pathologic process. This consisted of a cirsoid aneurysm of the basilar artery (Fig. 1) in 3 cases, an arteriovenous malformation in the cerebellopontine angle in 3 cases, while in the remaining case the 7th nerve was squeezed between petrous bone and pons because of displacement of the latter structure ascribed to an expanding lesion on the opposite side. Two of the 7 patients had trigeminal neuralgia, 2 had cerebellar symptoms, and 3 had no neurologic symptom or sign other than the hemifacial spasm.

Group B consists of 7 cases in which the 7th nerve was compressed and distorted by a redundant anterior inferior cerebellar or internal auditory artery. One of these patients (Case 1B), whose case has been reported previously, had other signs, consist-
ing of bilateral nerve deafness, complete on the side of the spasm, with impaired caloric responses bilaterally and in addition was subject to episodes of vertigo when she began to eat.

Group C consists of the remaining 5 cases, in 1 of which (Case 1C) there was bilateral nerve deafness of equal degree with preservation of the caloric responses. In this case a U-shaped loop of the anterior, inferior cerebellar artery accompanied the 7th and 8th nerves to the internal meatus but did not compress either one.

Preoperative motion-picture studies were obtained in all cases except Case 1B, and postoperative studies in all except Case 2A. A careful review of these films discloses that facial weakness was present before operation only in Case 5A. In several cases, facial weakness had been described by the original examiner but careful study of the films demonstrated that the facial asymmetry in these instances was the result of associated contraction (synkinesis) of antagonistic muscles that interfered with wrinkling of the forehead or elevation of the angle of the mouth. This observation is confirmed further by the fact that in these cases weakness was not present after the hemifacial spasm had been relieved. The same thing was found to be true of contracture. In no patient in whom contracture was described prior to operation was it observed after the hemifacial spasm had been relieved. The appearance suggestive of contracture is caused by mild tonic spasm since organic shortening of muscles could not disappear immediately with the relief of hemifacial spasm.

The hemifacial spasm appeared to be purely clonic in 1 case, purely tonic in 1 case, and a combination of clonic and tonic spasm in the remaining cases. It is obvious from a study of these films that tonic spasm represents merely clonic spasms occurring so close together that the interval is too short for relaxation to occur. Although the severity and extent of the spasm varied from case to case, there was no consistent difference in the three groups. The duration of the individual attacks varied among the patients, and in the same patient at various times. A refractory period during which an attack could not be precipitated did not appear to be present.

From a review of these films it is obvious
that voluntary movement such as talking, showing the teeth, smiling and closing the eyes precipitated the hemifacial spasm in 13 cases and probably precipitated it in 4: in only 1 case did the spasm shown on the film appear to be entirely independent of voluntary movement. Involuntary blinking frequently was observed to precipitate the hemifacial spasm, but in 4 cases involuntary blinking at times was accompanied by a single twitch at the corner of the mouth as in the post-Bell’s phenomenon. These observations suggest that hemifacial spasm should be considered a severe form of synkinesis. It differs from the post-Bell’s variety in that it tends to be self-perpetuating and in that it may accompany a particular voluntary movement on one occasion and not on another.

**RESULTS OF OPERATION**

The 19 operations were all performed under thiopental-sodium or halothane anesthesia. In each, except Case 6A, in which the facial nerve was obscured completely by an arteriovenous malformation, the nerve was manipulated gently with a nerve hook as has been advocated by Taarnhøj in the treatment of trigeminal neuralgia. In several instances the nerve also was Irrigated by a forceful stream of Ringer’s solution delivered by a 10 cc. syringe through a 22 gauge needle. This was done in an effort to separate the nerve fibers as was suggested by the work of Frankenhaeuser and Nyström. These authors showed that a severed portion of a peripheral nerve with an intact sheath did not swell appreciably when soaked in Ringer’s solution but when the sheath was split or stripped off, the nerve increased in diameter from 18 to 52 per cent because of accumulation of fluid between the fibers.

In 12 patients the face was observed during the manipulation of the 7th nerve. In 7 of these 12 patients the gentlest stroking of the nerve, and in some even the retraction of the cerebellum preliminary to exposure of the nerve, was accompanied by severe reproduction of the hemifacial spasm. It was observed that as gentle stroking of the nerve was continued, the paroxysms produced became less severe, indicating that a rise in threshold of irritability was taking place. Each of these patients was relieved of spasm following operation. In the remaining 5 of the 12 patients, in whom the face was observed during manipulation, a more pronounced stimulus was required to reproduce an attack. In 1 of these patients (Case 5C) the operation was not followed by relief. Similar stimulation of the 7th nerve in 6 patients who did not have hemifacial spasm did not cause facial twitching.

In this series of 19 patients, the operation was followed by immediate relief of the hemifacial spasm in 12, by delayed (2 to 21 days) relief in 5, and by no relief in 2. The degree of relief was complete in 12 and satisfactory in 5. The 2 failures were both in Group C, in which the 7th nerve was not compressed by any visible structure. There was almost complete recurrence of the hemifacial spasm in 2 cases, both of which were in Group A. There were no failures and no recurrences to date in Group B.

Weakness of the facial muscles followed operation in 4 cases in Group A. In Cases 2A and 6A there was mild facial paresis persisting until the time of discharge. The follow-up reports describe no recurrence of hemifacial spasm and fail to mention weakness. There was severe facial paralysis in Case 4A which began to improve after 3 months and has cleared almost completely. In Case 7A weakness of the face was demonstrable after operation only by a diminution of the blink reflex that returned to normal within a few weeks. In Case 5B there was some slight weakness of the facial muscles observed in the motion-picture film made 5 days after operation which had cleared entirely in the film made 5 days later.

**DISCUSSION**

In Group A only a confirmed skeptic witnessing the operation would question that compression of the nerve by the pathologic process was the cause of the hemifacial spasm. In Group B, in which the nerve was compressed and distorted by a loop of a
normal artery, the causal relationship may be questioned in view of Sunderland's finding that a large loop of the anterior inferior cerebellar artery was related intimately to the 7th and 8th nerves in 64 per cent of routine autopsies.* Despite the incidence of such vessels, it is difficult to argue with the fact that, in these 7 cases of hemifacial spasm, freeing of the nerve from the vessel and the interposition of a bit of Gelfoam where feasible, was followed by relief in every case and at the cost of mild and transient weakness in only 1 instance. It must be noted however that in the last 3 cases, in addition to this manipulation, a bundle of fibers, believed to be nervus intermedius, was divided. This was done in an attempt to interrupt the afferent limb of the presumed reflex arc. Distortion of the 7th nerve by the aberrant vessel made identification of this structure difficult. Though each of these 3 patients exhibited decreased lacrimation postoperatively, in Case 7B sense of taste was not impaired on the anterior portion of the tongue.

The poorest results were obtained in the 5 cases (Group C) in which the 7th nerve was not compressed by any structure in the cerebellopontine angle. There were 2 complete failures and probably a third since the relief occurring 3 weeks after operation in Case 4C could represent a spontaneous remission. Furthermore in this group the benefit was immediate in only 1 of 5 as compared with 6 of 7 in Group A and 5 of 7 in Group B. The poorer results in Group C suggest that when there is no apparent involvement of the intracranial portion of the nerve, the responsible lesion may be located beyond the internal meatus as was the case in the 2 patients of Woltman et al.,24 who were relieved following neurolysis within the bony canal. It is planned to divide the nervus intermedius in future cases, particularly those in which the 7th nerve is not compressed.

CONCLUSIONS

Hemifacial spasm is a severe form of synkinesis which differs from that developing after recovery from Bell's palsy in that it is self-perpetrating and not produced invariably by repetition of the same voluntary or involuntary movement. The facial asymmetry that in these cases frequently has been interpreted as facial weakness usually is the result of simultaneous contraction (synkinesis) of antagonistic muscles. The relief of synkinesis in these cases shows that it is not caused by anatomic misdirection of nerve fibers.

The operative findings and results indicate that hemifacial spasm is the expression of a reversible, pathophysiologic state commonly (13 of 19 cases) produced by mild, long-lasting compression of the 7th nerve by a vascular structure in the cerebellopontine angle. Reports in the literature show that it also may be produced by the pressure of a slowly growing tumor in this location.

How can pressure on the nerve produce a pathophysiologic state that is manifested by synkinesis and by repetitive twitchings that resemble the effect of intermittent electrical stimulation of the nerve? The most obvious explanation is that the resulting squeezing together of nerve fibers and the resulting reduction in thickness of their insulating myelin sheaths, permit transaxonal "short circuiting" of the action current. Such an effect was produced experimentally by Granit et al.10 These authors created an "artificial synapse" between afferent and efferent fibers, by placing a ligature about the sciatic nerve so gently that it did not interrupt the passage of the nerve impulse. They subsequently abolished this response by removing the ligature and irrigating the compressed portion of the nerve with Ringer's solution.

If hemifacial spasm is produced by such transaxonal stimulation, the short circuit may be either from efferent to efferent or from afferent to efferent fiber. A simple short circuit from efferent to efferent fiber would explain the nonrepetitive post-Bell's type of

* We reviewed the operative notes of the last 20 cases of vestibular-nerve section done at the Cleveland Clinic, and found that an artery was mentioned in only 4 instances. The vessel wound about the 8th nerve in 1 case, it transfixed the nerve in another, while in 2 instances it separated the 7th from the 8th nerve.
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synkinesis while a reverberating short circuit between afferent and efferent would be compatible with the repetitive twitches of hemifacial spasm. In the latter case, interruption of proprioceptive fibers should be curative.

REFERENCES