Hemifacial spasm secondary to vascular compression of the facial nerve

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Idiopathic hemifacial spasm is an uncommon, benign, but distressing condition characterized by the insidious onset of paroxysmal, repetitive, clonic contractions of the facial musculature. The condition usually begins with contractions of the orbicularis oculi and spreads first through contiguous muscles to involve all the muscles innervated by the seventh nerve on one side of the face.[1]

In a review of 106 cases of cryptogenic hemifacial spasm in 1945, Ehni and Woltman[1] pointed out several characteristics of the condition. They found that the disorder occurred only in adults and that the mean age of onset was 45 years, although recently hemifacial spasm has been reported in an 8-year-old child.[2] Ehni and Woltman[1] observed a sex distribution of six women to four men. Either side of the face was affected in either sex with equal frequency. In six of their patients the spasms were bilateral, but not synchronous nor symmetric. The spasms consisted of intermittent, irregular series of single muscle twitches occurring in rapid sequence. In 12 patients the spasms were observed in sleep.

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Psychic upsets, fatigue, and voluntary movement of the face were the most common factors that either precipitated or increased the severity of the spasms. None of the patients could voluntarily stop the spasms, although nine patients experienced periods of spontaneous remission lasting from a few weeks to 3 years. In three patients the spasms were accompanied by trigeminal neuralgia on the ipsilateral side, a condition referred to previously by Cushing[3] as "tic convulsif."

The failure of the spasms to spread beyond the distribution of the facial nerve and to be affected by a capsular infarct led Ehni and Woltman[1] to conclude that the lesion causing the disorder must be located distal to the corticobulbar pathway. Regeneration following division of the facial nerve at the stylomastoid foramen is usually accompanied by recurrence of spasm. Division of the nerve at this point followed by anastomosis with another cranial nerve is accompanied by reinnervation of the face, but not by recurrence of spasm.[4, 5] These observations led Ehni and Woltman to conclude that the lesion responsible for the spasm must lie within the segment of the nerve between the stylomastoid foramen and the facial nucleus.

Although most agree that compression of the nerve by some pathologic process causes the spasms, the nature of the pathologic process is variable. Woltman et al,[6] Proud,[7] and Pulec,[8] on decompressing the facial nerve in its canal through the temporal bone, observed edema and thickening of the sheath surrounding this portion of the nerve. They concluded that intratemporal pathology is the most common cause of idiopathic hemifacial spasm.

Other authors have pointed to an association between posterior fossa neoplasms and hemifacial spasm. Revilla[9, 10] observed spasms in patients with cerebellopontine angle neurinomas, meningiomas, and cholesteatomas. Gardner and Sava[11] reported the case of a patient with hemifacial spasm in whom the seventh nerve was squeezed between petrous bone and pons. The brainstem had been displaced, presumably by an expanding lesion on the opposite side. In 1966 Gardner and Dohn[12] reported a case of bilateral hemifacial spasm in a patient with basilar impression due to Paget's disease.

Some authors have emphasized that hemifacial spasm can be the result of compression of the seventh nerve by vascular structures in the posterior fossa. Campbell and Keedy[13] observed a cirsoid aneurysm of the basilar artery in two patients in whom hemifacial spasm was accompanied by trigeminal neuralgia. Laine and Nayrac[14] described a similar lesion in a patient with hemifacial spasm. Of 19 patients in whom posterior fossa exploration was performed for hemifacial spasm, Gardner and Sava[11] found three in whom the seventh nerve was compressed by a cirsoid aneurysm of the basilar artery. In three other cases, the nerve was compressed by an arteriovenous malformation and in seven others by a redundant anterior inferior cerebellar or auditory artery. Jannetta[15] reported eight cases of hemifacial spasm in which he performed microsurgical exploration and found the facial nerve compressed by vascular structures in the cerebellopontine angle. Decompression was followed by

relief of the spasms in each of his patients.

Gardner and Dohn[12] presented a vertebral angiogram which demonstrated a redundant loop of basilar artery projecting into the left cerebellopontine angle in a patient with both left hemifacial spasm and left tic douloureux. The cerebellopontine angle was not explored in this case, however. More recently Eckman et al[16] presented angiographic evidence of elongated and tortuous vertebral and basilar arteries invading the cerebellopontine angle on the appropriate side in four patients with hemifacial spasm. Similar aberrant vessels believed to be the cause of hemifacial spasm have been demonstrated angiographically by others.[17-19]

We agree that idiopathic hemifacial spasm is most often caused by anomalous vascular structures compressing the seventh nerve in the cerebellopontine angle. In recent years 45 patients have undergone exploration of the cerebellopontine angle for hemifacial spasm at the Cleveland Clinic. In 14 of these patients, the facial nerve was found to be compressed by an elongated and tortuous vertebral or basilar artery. Preoperative vertebral angiography in four of these 14 patients clearly demonstrated the abnormality in each. The purpose of this paper is to analyze briefly these 14 cases and to demonstrate the aberrant blood vessels observed in two of the four abnormal angiograms.

Analysis of cases (*see Table*)

At the time of operation, the 14 patients ranged in age from 44 to 71 years. There were five men and nine women. All had typical unilateral hemifacial spasm which, in most, was reported as having begun in the orbicularis oculi. It had spread and involved all the facial muscles on one side at the time of surgery. The left side was affected in eight patients and the right side in six. The duration of symptoms varied between 16 months and 20 years. Ipsilateral facial weakness was reported in five of the patients. The spasms were accompanied by tic douloureux on the ipsilateral side in one patient. In another, constant dull pain was associated with the spasms. Audiometry disclosed some deterioration in auditory acuity compatible with neurologic disease in four patients. The deficit was bilateral, although most severe on the side of the lesion in three and unilateral on the same side as the hemifacial spasm in one. Additional abnormal neurological findings in one patient included a spastic, ataxic gait, positive Romberg sign, and fasciculations in both thighs.

Angiographic findings

Of the four vertebrobasilar angiograms which demonstrated the aberrant blood vessels preoperatively we have selected two (*Figs. 1 and 2*). The changes observed in these are representative of those seen also in the other two. They consist of elongation and an anomalous curvature of the distal segment of the vertebral and proximal basilar artery with a convexity directed toward the right cerebellopontine angle in case 1 (*Fig. 1 A,B*). In case 2 the distal segment of vertebral artery is elongated and doubled over on itself, forming a loop which extends into the left cerebellopontine angle (*Fig. 2 A,B*). The terminal segment of the artery pursues a transverse course

Table. Hemifacial spasm due to compression of facial nerve by aberrant vertebral or basilar artery

Case no.	Age, duration	Sex, side	Additional symptoms and signs	Angio	Operative findings	Treatment	Result	Follow-up	Complications
1	58/4 yr	F/R	Constant, dull R facial pain	Yes	7th nerve compressed by anomalous vertebral and anterior inferior cerebellar arteries; excessive amount of choroid plexus beneath 7th nerve	Neurolysis, Gelfoam interposed, nervus intermedius sectioned, choroid plexus removed	Immediate complete relief; recurrence in 8 mo	8 mo	Dizziness and deterioration of auditory acuity on R
2	54/16 mo	M/L	Bilateral hearing loss worse on L compatible with neurologic disease	Yes	Ectatic loop of vertebral artery impinging on 7th and 8th nerve complex	Neurolysis, Gelfoam interposed, nervus intermedius sectioned	Immediate complete relief	2 yr	Nystagmus on lateral gaze, contralateral 4th nerve palsy, diminished ipsilateral corneal reflex, and pallanesthesia over contralateral side of body
3	61/5 yr	F/L	Hearing loss due to 8th nerve damage on L	Yes	Thickened petrous ridge; ectatic loop of vertebral artery contiguous with 7th nerve	Neurolysis, Gelfoam interposed, nervus intermedius sectioned	Partial relief; involuntary closure of L eye on chewing; recurrence of mild spasms 9 mo postoperatively	2 yr	Transient L facial weakness
4	64/4 yr	F/R	Bilateral central hearing loss worse on R	Yes	Internal auditory artery coursing between 7th and 8th nerves gave rise to arterial plexus overlying 7th nerve; facial nerve contiguous anteriorly with segment of vertebral artery	Neurolysis	Immediate complete relief; recurrence at 1 yr	1 yr	Mild transient R facial weakness
5	64/5 yr	F/R	R facial weakness	No	Facial nerve displaced by large atheromatous loop of basilar artery	Neurolysis, Gelfoam interposed, nervus intermedius sectioned	Immediate complete relief	5 wk	Transient increase in R facial weakness
6	68/4 yr	M/L	L facial weakness	No	Loop of basilar artery contiguous with 7th nerve anteriorly; branch of basilar artery coursed cephalad between 7th and 8th nerves	Neurolysis, Gelfoam interposed	Immediate complete relief; recurrence at 18 mo	18 mo	Complete 8th nerve deafness on L; increase in L facial weakness
7	64/4 yr	F/L	None	No	Facial nerve compressed by loop of basilar artery	Neurolysis, nervus intermedius sectioned	Immediate complete relief	1 yr	Occasional dizziness; mild hearing loss on L
8	71/4 yr	F/L	L facial weakness	No	Facial nerve displaced to L by distended segment of basilar artery	Neurolysis	HFS unchanged initially; complete relief by 10th postoperative day	3 mo	None
9	69/4 yr	M/L	Tic douloureux on L; L facial weakness, moderate bilateral central hearing loss worse on L, spastic gait, positive Romberg test, fasciculations in both thighs	No	Brainstem rotated and displaced to L by atherosclerotic aneurysm of basilar artery; 5th nerve compressed against side of brainstem; 7th and 8th nerves displaced dorsolaterally by aberrant basilar artery	Neurolysis, 5th nerve root partially sectioned	Complete relief	14 mo	Transient L facial paralysis followed by partial recovery; complete nerve deafness on L
10	53/20 mo	M/R	R facial weakness	No	Pons shifted to R by tortuous and distended basilar artery; 7th and 8th nerves separated by large internal auditory artery	Neurolysis, Gelfoam interposed	Complete relief	1 mo	Transient increase in R facial weakness; mild hearing loss on R
11	44/2 yr	F/L	Clicking sound L ear	No	7th nerve compressed by loop of basilar artery	Neuroloysis, Gelfoam interposed, nervus intermedius avulsed	Immediate complete relief	1 mo	Mild L facial weakness mixed hearing loss L ear

12	57/20 yr	M/L	None	No	Facial nerve displaced cephalad and posteriorly by tortuous loop of atheromatous basilar artery	Neurolysis, Gelfoam interposed	Immediate complete relief	4 mo	Mild L facial weakness
13	66/4 yr	F/R	None	No	Facial nerve displaced cephalad and posteriorly by elongated loop of basilar artery and compressed by anterior inferior cerebellar artery	Neurolysis, nervus intermedius divided	Immediate complete relief	2 mo	Mild R facial weakness
14	58/4 yr	F/R	None	No	Elongated, dilated loop of basilar artery displacing the 7th nerve just distal to its point of emergence from the pons	Neurolysis, nervus intermedius divided	50% to 75% relief initially; return to original level of intensity 5 mo postoperatively	8 yr	Transient hearing loss R ear

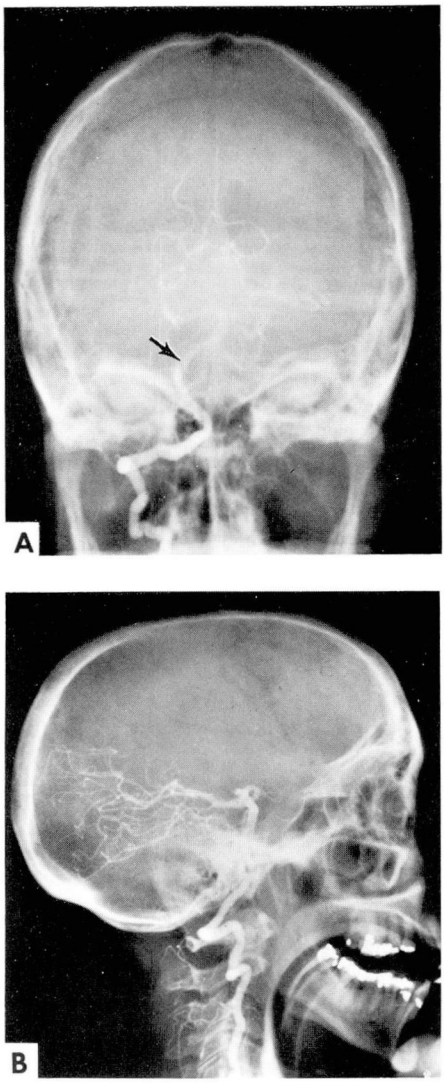

Fig. 1. A, B, Case 1. Right vertebral angiogram, arterial phase, half-axial and lateral projections. Elongation resulting in an arch of vertebrobasilar artery projecting into the right cerebellopontine angle (arrow).

toward the point of origin of the basilar artery.

Operative findings

Cerebellopontine angle exploration through a suboccipital craniectomy disclosed an elongated and tortuous vertebral or basilar artery distorting and compressing the seventh nerve in all 14 patients. The operating microscope was utilized in four of the dissections. The operative findings of the

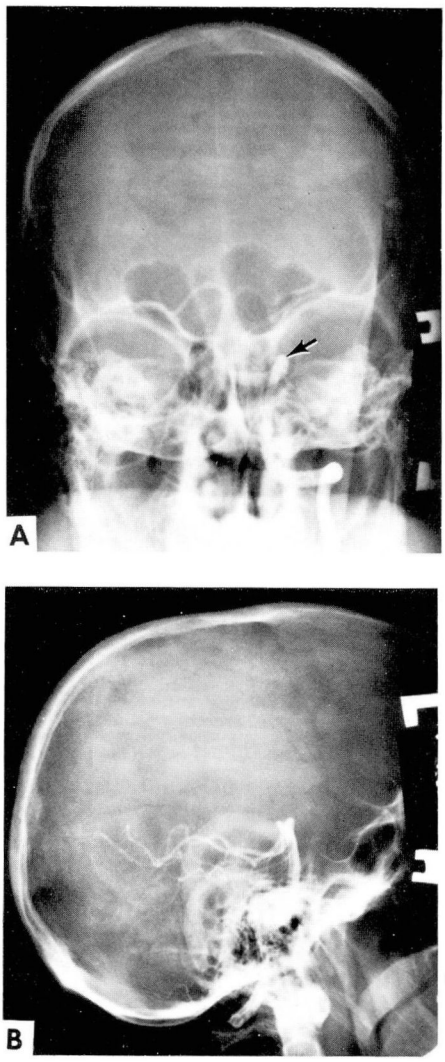

Fig. 2. A, B, Case 2. Left vertebral angiogram, arterial phase, anteroposterior and lateral projections. Redundant loop of vertebral artery extends into left cerebellopontine angle (arrow).

four cases in which angiography was performed will be discussed in detail.

In case 1 the seventh nerve was found to be compressed on its anteromedial aspect by an anomalous vertebral artery. Beneath the nerve was seen an extraordinary amount of choroid plexus. It was believed that by removing the choroid plexus, pressure on the nerve from the artery could be eliminated. The choroid plexus was therefore partially removed by coagulation. The seventh nerve was believed also to be compressed by the anterior inferior cerebellar artery. These two structures were separated and a piece of absorbable gelatin sponge (Gelfoam) was interposed between them. A segment of the superior aspect of the seventh nerve thought to be nervus intermedius was divided. A neurolysis was performed on the intact portion of the seventh nerve according to the method of Gardner.[20]

In case 2 a similar ectatic loop of vertebral artery was found impinging on the seventh and eighth nerve complex near the brainstem in the left cerebellopontine angle. The seventh and eighth nerves together were separated from the artery by interposing a piece of Gelfoam. The nervus intermedius was isolated and sectioned. A neurolysis was performed on the intact portion of the seventh nerve.

In case 3 there was an unusual bony thickening of the petrous ridge, particularly in the region of the internal auditory meatus. An aberrant ectatic loop of vertebral artery projected laterally to meet the seventh nerve at its point of emergence from the brainstem. The artery could easily be displaced with a probe, but immediately resumed its original position when released. A pledget of Gelfoam was inserted to separate the two structures after a neurolysis was performed on the nerve. The nervus intermedius was identified and divided.

In the fourth case an elongated and tortuous internal auditory artery coursed laterally between the facial and auditory nerves and gave rise to a small arterial plexus which overlay the facial nerve as it entered the internal auditory meatus. Anteriorly, the seventh nerve was contiguous with a distended and tortuous segment of vertebral artery. The seventh nerve was dissected away from the eighth nerve and a neurolysis was performed on the portion of seventh nerve proximal to the arterial plexus.

In case 9 the offending lesion, an atherosclerotic basilar aneurysm, was more extensive than those seen in most of our other cases. It was directly compressing three cranial nerves and distorting the brainstem as well. The segment of abnormal vessel compressing the seventh nerve is shown in *Figure 3*.

Results of surgery

Surgery was considered successful in 13 of the 14 cases. In one it was considered a failure because the patient ini-

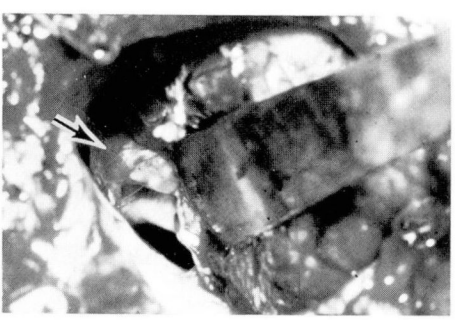

Fig. 3. Case 9. Operative photograph showing loop of tortuous aberrant basilar artery (arrow) adjacent to seventh and eighth nerve complex.

tially had only 50% to 75% relief and the spasms progressed to their original level of intensity by the 5th month following surgery.

Of the 13 successful cases, surgery in 12 was followed by complete relief and, in one, by partial relief of the facial spasms. Patient 3 is categorized as having only partial relief because chewing was occasionally accompanied by involuntary closure of the left eye. Occasional mild facial twitching began recurring about 9 months postoperatively in this patient. Three others were symptom free before signs of recurrence at intervals ranging from 8 to 18 months. Eight patients showed no signs of recurrence when last seen 4 weeks to 2 years postoperatively.

Five patients had ipsilateral facial weakness for the first time after surgery. Four others in whom facial weakness was noted before surgery experienced worsening of the weakness afterward. One of the four patients with preoperative facial weakness had complete paralysis of facial musculature following surgery. In most cases the deficit was transient and the strength returned to preoperative levels. The patient in whom complete paralysis developed experienced only partial recovery of the deficit.

Seven of the patients also experienced some deterioration of their hearing. The deficit was bilateral but worse on the ipsilateral side in two. Most patients recovered at least partially.

In addition to dizziness, of which two patients complained, other complications were observed in only one patient. These included nystagmus on lateral gaze, diplopia due to a contralateral fourth nerve palsy, a diminished ipsilateral corneal reflex, and a decrease in sensitivity to light touch and vibration over the contralateral side of the body and extremities.

Discussion

We have presented 14 patients with hemifacial spasm in whom the facial nerve was distorted by an elongated and ectatic vertebral or basilar artery. We are convinced that this was the cause of the spasms in these patients. In all cases compression of the facial nerve was actually observed during cerebellopontine angle exploration. Neurolysis of the facial nerve was performed in all cases, sectioning of the nervus intermedius in nine, and separation of the aberrant vessel from the nerve in seven. These procedures were followed by complete relief in 12 cases and partial relief in one.

We want to emphasize that the point of compression is most often located proximally along the nerve just after it emerges from the brainstem. If this segment is not explored carefully, preferably with the operating microscope, the lesion may easily be overlooked.

The 14 patients reported in this study constitute 0.30% of all the patients who have undergone cerebellopontine angle exploration for hemifacial spasm at the Cleveland Clinic. In another large percentage of our patients, not included in this study, the facial nerve was compressed by a less well defined vessel, such as the anterior inferior cerebellar or internal auditory artery. These findings corroborate our contention that compression of the facial nerve by an aberrant vascular structure in the cerebellopontine angle is the most common cause of hemifacial spasm.

Others have postulated that disturbances of nerve conduction could arise

as a consequence of compression by an abnormal vascular structure. Sunderland,[21] in his study of the neurovascular relationships of the internal auditory meatus, was one of the first. In a subsequent study of 210 autopsy specimens he found that tortuosity and deviation of the vertebral and basilar arteries from their normal location occurred frequently.[22] It was of a degree sufficient to deform the pontomedullary junction and compress the seventh and eighth nerves in 27 instances. Unfortunately, the clinical records of the cases examined were not available so that he could not correlate the anatomical changes with the symptomatology that had existed during life.

In a recent study of the pathogenesis of elongation and distention of intracranial arteries at the base of the brain in 34 autopsy specimens, Sacks and Lindenburg[23] observed atrophy caused by pressure exerted on adjacent cranial nerves and brain in 28 cases. They emphasized that arterial elongation and distention must be considered in the differential diagnosis of any patient with neurologic signs and symptoms suggestive of a space occupying lesion.

An important point should be learned from the present study: in all patients with hemifacial spasm, whether or not accompanied by other neurologic signs, angiography of the vertebrobasilar system should be performed preoperatively. Of the four patients evaluated in our series, this technique revealed the nature of the pathology by demonstrating an anomalous vascular structure projecting into the region of the cerebellopontine angle in each.

Just how pressure on a nerve produces paroxysmal repetitive muscle contractions has been the subject of considerable discussion. Woltman et al[6] were among the first to postulate that local irritation of the nerve by compression and ischemia might cause spontaneous discharge of impulses or facilitate the activation of inactive fibers by impulses traveling through adjacent active fibers in patients with hemifacial spasm.

Hering[24] was the first to show in his laboratory experiments of 1882 that such cross stimulation does indeed occur between adjacent nerve fibers. He created an "artificial synapse" in frogs by dividing the sciatic plexus in the pelvis. He was able to demonstrate that impulses traveling in ascending sensory afferent fibers would cross over to stimulate efferent motor fibers at the level of the interrupted plexus.

In a somewhat more sophisticated experiment, Granit et al[25] created an artificial synapse by placing a ligature around the sciatic nerve in frogs so loosely that it did not block conduction of the original impulse. By recording with electrodes from the roots of the sciatic nerve which they had divided intraspinally, they demonstrated that impulses descending in the motor fibers would cross stimulate afferent sensory fibers at the level of the ligature. A response could also be produced in motor fibers by stimulating the cut ends of the afferent sensory fibers. Moreover, they were able to abolish the response by removing the ligature and irrigating the compressed segment of nerve with Ringer's solution much as is done today during an intracranial neurolysis.

In recent years Gardner[26, 27] has been one of the strongest proponents of the theory that hemifacial spasm is caused by cross stimulation at a point of nerve

compression. He, in fact, suggests that the direction of cross stimulation is probably afferent to efferent and contends that this explains why, when other intracranial procedures fail, sectioning of the nervus intermedius is usually curative. The observations made in our series of patients add further support to this theory.

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