HEMIFACIAL SPASM: A NOTE ON THE ETIOLOGY
IN TWO CASES*

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(Received for publication March 8, 1947)

While the etiology of hemifacial spasm is unknown, it is generally held that the lesion lies either in the facial nucleus or in the proximal portions of the nerve.\textsuperscript{3,7} Since the disorder is never fatal, pathological examinations have thus far been very rare. It so happened that two patients with typical facial motor tic came to be operated upon by the suboccipital route for an associated homolateral tic douloureux. Since each was found to have a cirsoïd aneurysm of the basilar artery which compressed the facial nerve on the affected side, these observations appear worthy of record.

Case 1. Mrs. L. B. (U. #53170), a 62-year-old white woman, was admitted to the Albany Hospital Nov. 26, 1938 with the history of twitching in the left side of the face of 4\textfrac{1}{2} years' duration and of intermittent pains in the left jaw and left side of the tongue for the previous 18 months. The movements had involved chiefly the muscles about the left eyelids and gradually spread to the cheek, and were rapid, painless and largely uncontrollable. They were worse when she was with people, particularly strangers. They were present during sleep. There had been no involvement of any muscles other than those of the left face, no vertigo, loss of consciousness, nor any mental disorder.

The pains, which were confined to the left mandibular nerve area, were sharp, severe and came in groups of several short "jabbings" rapidly repeated. After some attacks a burning sensation would persist in the involved area or occasionally this latter sensation might be experienced alone. Talking and chewing often precipitated these episodes. The pain bore no consistent relationship to the facial twitchings. The past and family histories were non-contributory.

Examination. The patient was a rather slender woman of good color and general condition. Principal interest lay in the left face, the upper portion of which was frequently distorted by twitchings. These were unpredictable in onset, and could not be volitionally abolished even temporarily. They tended to involve the orbicularis oculi muscles principally, and the zygomaticus caninus group to a lesser extent. At times a series of contractions would all but close the left eye for several seconds. Often the movements appeared to be so small and so sharply limited as to suggest involvement of but a portion of a muscle. The patient felt no compulsions nor was there any suggestion of this type of disorder. No weakness of the facial muscles was discerned. The left corneal reflex was very slightly diminished, although the patient was not conscious of any difference in sensation in the two eyes. The remainder of the neurological examination was normal. The otological consultant, Dr. Benjamin Volk, found the hearing to be normal in each ear; both labyrinths were mildly hypoactive. The rest of the physical findings were not remarkable. The fundi appeared normal. The B.P. was 170/80.

Laboratory Findings. RBC: 4,100,000; WBC: 4,800; Wassermann reaction negative. Free hydrochloric acid was present in the gastric juice and the stool contained no occult blood.

The left mandibular nerve was injected with alcohol at the foramen ovale. Resulting anesthesia was incomplete and tic pains continued.

Operation. On Dec. 9, 1938 a left suboccipital approach was carried out under avertin and

\textsuperscript{*} Presented at the meeting of the Harvey Cushing Society, October 10, 1946, Boston, Massachusetts.
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intratracheal anesthesia. When the cerebellum was retracted medially the facial and acoustic nerves were seen to be pushed laterally and forward by a canary-yellow, oval-shaped mass, approximately 1 cm. in diameter. The internal auditory artery was 1 to 2 mm. in thickness and formed a loop which tended to obscure both the 8th nerve and the mass in question. In order to obtain a satisfactory view of the latter it was believed necessary to divide this arterial loop. After this had been done and the cerebellum retracted further it was clearly seen that the yellow mass was but a xanthochromatous area in the wall of a cirsoid aneurysm of the basilar artery.

After a large petrosal vein had been thrombosed and divided, the sensory root of the trigeminal nerve came into view. The aneurysm curved forward, passing under the root which it had pushed dorsolaterally against the edge of the tentorium. By means of a nerve hook approximately the lateral 1/3 of the sensory root was divided.

She recovered promptly from the operation, had complete anesthesia in the mandibular nerve area and mild hypesthesia in the 1st and 2nd divisions. Apparently as the result of division of the internal auditory artery there was immediate and complete paralysis of the left 7th and 8th cranial nerves.

Subsequent Course. She was permanently relieved of pain and was so happy to be free from the facial twitching that she decided not to return for a spinofacial anastomosis. She reported by letter 4 months after operation that the facial paralysis was clearing up but that the left ear remained deaf. A neurologist who examined her several times at her home in another city reported that there was partial recovery of movement in the left face, that it became almost symmetrical and that occasional fine twitching movements were observed about the left orbit. She remained deaf in the left ear. There was no return of pain and she continued in good health for the next 8 years. She dropped dead July 16, 1946. No autopsy was performed.

Case 2. Mrs. Ida P. (U. #8789), a 64-year-old white woman, was admitted to the Albany Hospital June 26, 1946 complaining of left facial pains typical of tic douloureux, which had begun 5 years before, and of twitching of the left side of the face of 3 years' duration.

The pains had appeared spontaneously in the left brow in May 1941. They were described as sudden, sharp and severe, each lasting but a matter of seconds. Within a few weeks they spread to involve the left cheek, side of the nose and occasionally the left temple. Upon the advice of her dentist, several left upper teeth were removed, but without relief. A trigger zone appeared just lateral to the nose, making it difficult to wash the face. In December 1941 the left supra- and infraorbital nerves were injected with alcohol; complete relief from pain followed for 8 months. Again an alcohol injection of the supraorbital nerve was performed which provided relief for 6 months. At the end of this time the pains recurred with increased severity in both the 1st and 2nd division areas. In our absence overseas, she consulted a neurosurgeon in another city, who, in December 1942, performed a subtotal division of the sensory root of the left trigeminal nerve by the temporal approach. Complete anesthesia resulted over the lower jaw and she was unable to chew on that side. The pains recurred in the left cheek and side of the nose 3 years later, and were then unaffected by reinjection of the infraorbital nerve.

Approximately 2½ years after the onset of the tic douloureux, a twitching set in about the left orbit. This soon spread to involve the cheek and the corner of the mouth, but never to any other muscles. These movements were not completely inhibitable voluntarily and were worse when she was with people or was nervous. She felt no compulsion to make the movements and was quite puzzled by them. They occasionally awoke her from sleep. She had observed no relation between the twitching and the pain.

Examination. She was a small well nourished woman, the left side of whose face was frequently contorted by twitchings about the left orbit, cheek and corner of the mouth (Fig. 1). The onset, duration, and intensity were quite variable although at times a rough pattern appeared. On such occasions, a series of small contractions beginning in the orbicularis oculi muscles would culminate in a closure of the left eye and the drawing upward and outward of the left corner of the mouth, lasting 5 to 15 seconds. She could not tell when a contraction would start, could not voluntarily inaugurate one, nor having begun, could she do more than
temporarily and slightly lessen it. The movements were never observed to spread to muscles other than those supplied by the left facial nerve.

The left temporal and masseter muscles were markedly atrophic and the jaw on opening deviated to the left. The incisional scar in the left temple was well healed and the scalp somewhat sunken in over the decompression.

_Laboratory Data._ RBC: 4,900,000; WBC: 7,900; blood non-protein nitrogen: 41 mgm. per cent; blood chlorides: 450 mgm. per cent.

_Operation._ On June 27, 1946 the patient was anesthetized with intratracheal ether and placed upon her right side in the Mount position. A lumbar puncture was then performed with the stylette temporarily left in place. A small curved incision was then made in the angle between the left mastoid and lateral sinus. As soon as the dura was opened, the stylette was removed from the lumbar puncture needle. With the escape of fluid the cerebellum fell away from the lateral wall of the posterior fossa. The internal auditory artery formed a loop which lay on the posterior surface of the acoustic nerve. After a large petrosal vein had been divided the sensory root of the trigeminal nerve, which was in this instance bifid, became clearly visible. A large cirsoid aneurysm of the basilar artery (about 1 cm. in diameter) coursed forward (Fig. 2) pushing the facial and acoustic nerves laterally, then curved medially just behind and barely touching the trigeminal at its junction with the pons. Its pulsations, while readily apparent and easily palpable through the forceps, were not extensive. In one portion of its wall was a bright yellow area.

The sensory root of the trigeminal nerve was then completely divided with a nerve hook and the wound closed in the usual manner with silk.

_Course._ Convalescence was uneventful. Complete anesthesia resulted in the left trigeminal
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field and there has been no return of pain in 3 months' time. The facial twitching remains practically unchanged after 7 months.

DISCUSSION

Hemifacial spasm* is a benign disorder, readily distinguishable from compulsive tic and usually from Jacksonian and focal epilepsy. Ehni and Woltman2 have summarized the outstanding characteristics as follows:

(1) The spasms are of an intermittent twitching nature, such as might be encountered in intermittent faradization of the facial nerve. (2) The eyelids on the side are almost always involved. (3) The spasms are usually unilateral, and when they are bilateral they are not synchronous or equal in extent or severity. (4) The spasms may persist in sleep. (5) The patient does not feel any compulsion to make the movement. (6) The patient is unable to stop the movement by exercise of the will. (7) The patient cannot reproduce the movements voluntarily—especially is he unable to approach the speed with which the fine twitchings occur. (8) Psychic upsets of any sort, fatigue and voluntary movements of the face make the spasms worse. (9) Children do not have hemifacial spasm. (10) The spasms are limited to the muscles innervated by the facial nerve.

But one autopsy study of this malady has come to our attention. Schultze8 in 1875 recorded the case of a man with typical hemifacial spasm of approximately a year's duration who died as a result of pulmonary tuberculosis. He had not been deaf in either ear. Autopsy examination of the brain disclosed a small cherry-sized aneurysm of the left vertebral artery which lay against

* The variety that sometimes follows paralysis of the facial nerve is not included in the group of cases under discussion.
the left 7th and 8th nerves, but which did not compress the 9th (Fig. 8). Microscopic examination of the left facial and auditory nerves was reported to have disclosed no abnormality.

![Fig. 3. Aneurysm of the left vertebral artery compressing the 7th cranial nerve as recorded by F. Schultze.](image)

The not infrequently observed association of hemifacial spasm with certain neurologic changes is noteworthy. Ehni and Woltman report anatomic evidence of arteriosclerosis in 41 of their 106 cases of hemifacial spasm. They recorded that 15 of their 106 patients had weakness of the involved side of the face, while 14 had impaired hearing on the affected side, 3 on the opposite side and 1 bilaterally. Instances of corneal anesthesia, hypesthesia in the homolateral face, homolateral or contralaterally abnormal plantar reflexes, superior rectus paralysis, homolateral diminution of taste, weakness of that side of the tongue and diminution of the ankle jerk and of vibratory sensibility in the contralateral side have been recorded.

Harris and Wright and others have called attention to the occurrence of an associated homolateral tic douloureux.

Two cases have been reported (Habel; Ehni and Woltman) which indicated that the irritative lesion lay in the lower motor neurone. In each instance the hemifacial spasm, which had been established for several years, was not affected by a supervening homolateral hemiplegia which included the face.

While the exact incidence of aneurysms of the posterior fossa is unknown, it is recognized that they are not uncommon. Thus McDonald and Korb found that in 202 of 1023 recorded cases the lesion involved the basilar or vertebral arteries. Dandy reported 21 in this locality among 108 cases. Of these, 2 were large sacculations, 8 small sacculations and 11 were cirroid.
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Such an abnormality he described as an “S-shaped elongation of the basilar and/or vertebral arteries, with consequent lateral bulging that compresses either the fifth or eighth nerve or both.” Most of these were operative findings, having been associated with tic douloureux or Ménière’s syndrome. (He recorded no instance of hemifacial spasm.) In 9 of the 11 the bulge of the artery had been to the left. In each of our cases the disorder was left-sided.

It is thus suggested that vascular disorders at the base of the posterior fossa may account for a proportion of the cases of the so-called idiopathic hemifacial spasm. More observations in this connection are highly desirable.

SUMMARY AND CONCLUSIONS

Two cases of hemifacial spasm associated with homolateral tic douloureux are reported. Each patient was operated upon by the suboccipital approach and was observed to have a cirrroid aneurysm of the basilar artery.

While but one autopsy study is available, the literature yields suggestive evidence that such vascular abnormalities in the posterior fossa may account for at least a certain number of these maladies.

REFERENCES