

Facial myokymia

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The facial musculature may be the site of a great variety of involuntary movements, often of doubtful nature and obscure causation. One form was sharply delineated both by clinical description and probable pathogenesis by Andermann, Cosgrove, Lloyd-Smith, Gloor, and McNaughton (1961). These authors drew attention to the condition of facial myokymia of which they described four cases of their own and found seven reliable reports in the earlier literature. Of these 11 patients, eight almost certainly had disseminated sclerosis; in two this diagnosis seemed probable and in one the myokymia had been the only important symptom at the time of reporting. I have found no subsequent reports of this condition, which might therefore be thought to be exceedingly rare. It is in order to correct this impression that I here report seven episodes of myokymia occurring in five patients observed in the past six years.

Unnecessary repetition in the case histories may be avoided by a general description of the salient and essentially stereotyped clinical features. In the five attacks where the myokymia was the only important symptom the onset was apparently abrupt, the day of onset of abnormal sensations being stated without hesitation. As far as I could determine the whole of the facial musculature on the affected side would be affected from the onset but I was not able to confirm this by personal observation. The onset of the other two attacks coincided with other more serious symptoms and the patients made no complaint of the facial movements. The usual complaint was that one side of the face felt 'screwed up' or 'swollen' and that certain movements were difficult to perform. There was no spontaneous complaint of weakness, movements seeming difficult because of 'stiffness' of the face. Three patients were aware of the flickering movements on the affected side and had observed these in the looking glass.

The appearance of the face when the condition is fully developed is highly characteristic but, like many forms of involuntary movement, not easy to describe. All the muscles of one side of the face appear to be in slight contraction so that the palpebral fissure is narrowed, the angle of the mouth drawn up and the lips slightly pursed. Continuous

flickering can be seen in all the muscles from the frontalis to the platysma. This flickering is quite different from even the most severe fasciculation seen, for example, in the muscles of the upper limbs in motor neurone disease, as it is repeated with extraordinary rapidity. A highly characteristic feature is that the flickering passes over the face in rapid undulating waves, too fast and complex for visual analysis.

In two patients I thought that there was slight weakness of the face on the affected side but in view of the mutually antagonistic action of many of the continuously contracting muscles this impression could well have been wrong. There was certainly no question of a marked facial palsy.

The duration of the phenomenon was highly variable and not easy to assess exactly as usually the movements abated gradually. The condition would initially be obvious even to casual inspection but after a variable period would pass through successive phases in which the continuous contraction became less obvious and the flickering less frequent. Towards the end of an attack the movement might be confined to one or two muscles, particularly those of the lids, and eventually the patient would state that only occasional flickering was experienced and none could be seen during the examination. Of the seven attacks witnessed, the duration varied from three weeks to approximately six months.

Associated symptoms possibly directly related to the disorder of the facial nerve were infrequent. One patient had noticed hyperacusis on the affected side for a period of three days some 12 days before the onset of myokymia. Sense of taste was not disturbed in any of the patients.

CASE REPORTS

CASE 1 J.B. (DRI 181492), a man of 37, of Polish origin, was first seen on 29 July 1959. He had no past history of nervous disease or of any serious illness. The symptoms had been present for six days and affected the right side of the face, all the muscles being in continuous undulating flickering contraction in the manner now recognizable as characteristic of facial myokymia but totally unfamiliar to me at the time. There was no facial weakness. He had no other symptoms and no other abnormal physical signs. The Wassermann reaction was negative. On

4 August the condition was unchanged. When seen on 11 September only occasional fasciculation could be detected in the muscles around the eye and in the platysma on the right but he reported that three days before exactly similar symptoms had appeared on the left. This was indeed so and the fully developed condition was now present on the other side with possibly a little weakness of retraction of the angle of the mouth. This second attack lasted with gradually lessening severity for approximately six months. When examined in June 1960 no fasciculation was observed.

I was able to examine him again in July 1965. He had remained quite free from symptoms in the intervening five years and there were no abnormal physical findings.

CASE 2 B.C. (DRI 229291), a woman of 22, was admitted to hospital under the care of Mr. R. H. Shephard on 1 December 1960. The immediate history was that about two weeks before she had fallen in the street because her right leg had given way and had been unable to get up again. She was helped home but developed rapidly increasing weakness of both legs. On admission she was unable to walk because of spastic paraparesis with marked impairment of all forms of sensation below the tenth dorsal dermatome. The function of the sphincters was not disturbed. At this time a past history was obtained of blurred vision in the right eye and possibly slight difficulty in walking for a few days only some two years earlier. This had been attributed to a blow in the eye but there was an interval of three days between the blow and the visual disturbance. This history could clearly not be accepted as evidence of a disseminated process and a myelogram was carried out. This was normal and the cerebrospinal fluid obtained at the time contained 20 mg. of protein per 100 ml. and no cells were seen.

On 6 December she complained of tingling in both hands and 'ptosis' of the left lid was noted, but the strength of the lower limbs had begun to improve.

On 8 December I found that she had left-sided facial myokymia producing narrowing of the palpebral fissure and responsible for the 'ptosis' but involving all the muscles on that side. There was no weakness of the face. There were widespread abnormalities, positive findings consisting of temporal pallor of the left optic disc, vertical nystagmus, hypalgesia in an ill-defined area on the left cheek and in the left upper limb, incoordination of the left upper limb, absent abdominal reflexes, and spastic paraparesis with a sensory level at the eighth dorsal dermatome. By this time there was, of course, no doubt of the diagnosis of disseminated sclerosis. She was given a course of corticotrophin for two weeks. The facial myokymia recovered in about three weeks. The disease remitted but she was left with a spastic and ataxic gait and considerable euphoria. In April 1964 she experienced diplopia for about a month and during this time nystagmus was again present.

On 3 May 1965 she noticed that it was difficult to whistle and when seen nine days later myokymia was again present on the left side but to a milder degree than in the first attack and accompanied by probably genuine slight facial weakness. The condition again remitted after approximately three weeks.

CASE 3 A.V. (DRI 210343), a boy of 17, was seen on 10 March 1961. He had developed the symptoms of facial myokymia on the left side on 18 February and when seen the condition was characteristic and severe. He had no other symptoms and there were no other abnormal signs. The face did not seem to be weak. There was no past history of nervous disease or of serious illness. The attack subsided gradually over about three months.

I was able to examine him again in July 1965. He had had no further symptoms and there were no abnormal findings.

CASE 4 T.D. (DRI 232363), a postman of 52, developed the first symptoms of a severe and complex neurological disorder at the beginning of December 1964. The onset was with right-sided headache and vomiting, which subsided after two weeks. He then complained of numbness of the right upper lip and of difficulty in focusing his vision. He rapidly became unsteady on his legs and was admitted to hospital under the care of Mr. R. H. Shephard. At that time he was free from headache and the optic discs were normal. Positive findings consisted of a right homonymous hemianopia, nystagmus in all directions of gaze, hypalgesia on the right upper lip, tactile inattention in the limbs on the left, and an ataxic gait. The plantar reflexes were flexor. The E.E.G. contained a delta focus in the left temporal region but a ventriculogram was normal. The ventricular fluid contained 10 mg. of protein per 100 ml. and 4 white cells per c.mm.

When I saw him on 14 January 1965 left-sided facial myokymia was present but the patient did not seem to be aware of it. The probable total duration of the myokymia was about one month and it was no longer present when I examined him on 3 March. By this time the hemianopia was confined to the upper quadrants and the hypalgesia on the face was no longer present but his gait had greatly deteriorated and both plantar reflexes were now extensor.

On 25 May he was again admitted to hospital under my care. He was unable to walk. The visual field defect had persisted and he now had severe spastic weakness of both legs with patchy cutaneous sensory loss below the eighth dorsal dermatome, loss of postural sense in the toes, and of vibration sense in the lower limbs. He was incontinent of urine. A diagnosis of disseminated sclerosis could not be accepted with confidence as the lumbar cerebrospinal fluid contained 175 and 200 mg. of protein per 100 ml. with no increase in cells on the two occasions it was examined. The Wassermann reaction was negative. Because of the persistent left temporal delta focus in the E.E.G. and the right hemianopia, a left carotid angiogram was carried out and was normal.

When last examined on 5 November he had greatly improved. He was walking with two sticks and control of the bladder was normal. He no longer had nystagmus and the field defect was confined to the right upper temporal quadrant. He had mild dysarthria, generalized increase in tendon reflexes, absent abdominal reflexes, extensor plantar responses, and loss of vibration sense at the ankles. The undoubted remission greatly strengthened the probability of the diagnosis of disseminated sclerosis.

CASE 5 J.S. (DRI 293822), a woman of 27, was seen on

12 March 1965. She had no past history of nervous disease or of serious illness. Five weeks before she attended the clinic she began to experience frontal and occipital headache, particularly on waking, and accompanied by nausea. At the same time she noticed that noises sounded unduly loud in the right ear. This symptom only persisted for three days and the headache gradually improved. About two weeks after the onset of these symptoms the right side of her face felt swollen on waking one morning. This feeling had persisted and was accompanied by stiffness of the face and awareness of flickering movement. When seen she had right-sided facial myokymia to a very marked degree, unaccompanied by facial weakness or by any other abnormality. On 23 April the myokymia was just detectable and two weeks later it had stopped after a probable total duration of about six weeks. No further symptoms have so far appeared.

ELECTROMYOGRAPHY

Electromyography (E.M.G.) was carried out in all patients, using fine coaxial needle electrodes. As might be expected in all the muscles sampled there was intense activity in the absence of any attempt at voluntary movement. No attempt could be made to detect fibrillation potentials. With most placements of the needle all that could be recorded was a continuous pattern of potentials not distinguishable from those of normal motor units of the facial musculature. Voluntary movement simply increased the amplitude and density of the pattern.

By adjusting the needle it was sometimes possible

to record something of the rhythmic nature of the spontaneous discharges. For example, Fig. 1 shows that above the continuous pattern of more distant units an apparently double discharge of one unit can be detected as occurring with some regularity approximately nine times a second. In Fig. 2 sampling was more successful in avoiding the confusion of the continuous interference pattern and it is possible to detect at least three different units firing in short bursts. For each unit the rhythm of firing and the number of discharges is distinct, although not absolutely constant. Thus one unit fires in short bursts of from three to eight spikes approximately four times a second, while another fires longer bursts with great regularity every 2.5 to 2.6 seconds. A third unit of lower amplitude can also be seen but its rhythm cannot be detected. The patient (case 3), hearing the rhythmic sound from the microphone, asked what was being recorded as it sounded too fast to be his pulse.

An appearance of synchronization of activity of several units or groups of units was sometimes present, causing brief periods of complete or relative quiescence. The irregularity of these periods suggests that the synchronization was fortuitous. Thus in a continuous recording from the lower lip in case 1 periods of quiescence of from 20 to 50 msec. occurred after successive intervals of 440, 660, 410, 310, 150, 480, and 160 msec. (correct to the nearest 10 msec.).

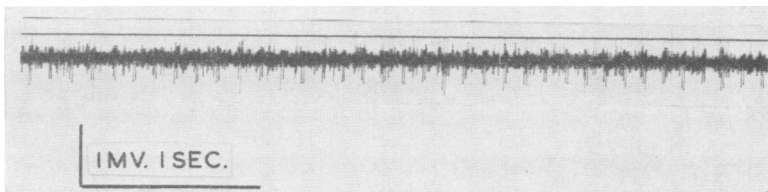


FIG. 1. Recording from the lower lip (case 4).

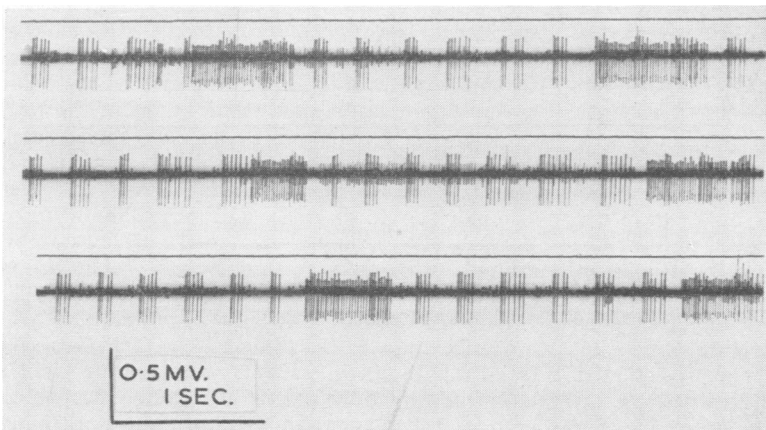


FIG. 2. Recording from the upper lip (case 3). The three strips are a continuous recording.

DISCUSSION

Outside specialized neurological units the various forms of facial 'spasm' are often confused. Thus, in my experience, both clonic facial spasm and the facial dyskinesias that may afflict the elderly are usually diagnosed as a form of tic due to neurosis and treated as such for long periods. The associated movements that may be present after recovery from a Bell's palsy where denervation has occurred are not always recognized as the permanent effects of faulty reinnervation. The clear distinction between the various forms of involuntary movement of the face is of some practical importance for the patient. Thus although Kinnier Wilson (1940) considered that the separation of a myokymic form of facial spasm was 'superfluous', subsequent experience has confirmed its highly distinctive features.

With but minor differences the present report confirms the admirable clinical and E.M.G. description of Andermann *et al.* (1961). The term 'myokymia' seems first to have been used by Schultze (1895) to describe very similar fluctuating fasciculation of the calf muscles. This has different E.M.G. characteristics to those of facial myokymia and appears to be a chronic but entirely benign condition (Denny-Brown and Foley, 1948). Facial myokymia was first described by Bernhardt (1902). I have been able to find one early report not mentioned by Andermann *et al.* (1961), that of Frenkel (1903). This is a typical example of a woman of 22 who suddenly, 'ce jour-là', developed the characteristic movements and persistent contraction on the left side of the face. These persisted for about three weeks. The myokymia had been immediately preceded by iritis on the same side and both conditions were thought to have responded to mercurial treatment although there was no definite evidence of neurosyphilis. The case was reported only a month after the cessation of the movements and no further symptoms had occurred. With the present series the total number of cases described reaches 17. The three case reports of Thiébaud, Isch, Isch-Treussard, and Ebtinger-Jouffroy (1960), while similar in many respects, do not mention the characteristic and unmistakable feature of undulatory fasciculation and must be excluded.

Facial myokymia has never been described as immediately following a facial palsy, although in two patients a transient palsy had occurred on the same side six years before (Newmark, 1903; Alajouanine and Thurel, 1936). The onset is nearly always abrupt, the movements are continuous and never of great amplitude, and the condition invariably remits after a few weeks or months. A confident prediction of recovery can be made even when, as in case 1, the movements promptly appear on the other side. The

spontaneous recovery may be particularly contrasted with the wearisome persistence of the clonic form of spasm. The symptoms are not severe but are alarming and the cosmetic effect can be distressing in a young woman.

Benign myokymia affecting the lower lid is an extremely common condition in normal people and does not resemble the universal affection of the facial muscles on one side described here. The terminal stages of an episode of myokymia might, however, be difficult to distinguish from the banal condition.

I can add nothing to the suggestion of Andermann *et al.* (1961) that the responsible lesion is intramedullary in close relation to the facial nucleus. In two of my patients, as in two of theirs, there was some co-existing sensory loss on the face suggesting involvement of the trigeminal nucleus. In one patient the sensory loss was on the same side as the myokymia and in the other the two conditions were on opposite sides.

Of greater importance is the association with disseminated sclerosis. This was first emphasized by Oppenheim (1917) but I have been unable to read his full case report (Oppenheim, 1916). Kino (1928) established the connexion much more firmly by describing three patients who had episodes of myokymia in the early years of developing disseminated sclerosis. In case 2 of the present series the myokymia occurred at the height of a severe relapse of undoubted disseminated sclerosis and recurred later as an isolated symptom. In case 4 the myokymia was also detected during a complex illness that certainly produced scattered lesions in the central nervous system followed by partial remission but the diagnosis of disseminated sclerosis, although probable, is less certain. In the other three patients the myokymia has been an isolated event. These patients were all in the age group most vulnerable to disseminated sclerosis but no further evidence of the disease has appeared after six years, four and a half years, and five months respectively. Disseminated sclerosis cannot, of course, be excluded on these grounds. The sudden onset and gradual remission greatly resemble the time relationships of the early symptoms of the disease. Of the 17 reported cases, the myokymia has been an isolated event in five; the diagnosis of disseminated sclerosis has been virtually certain in nine and probable in three. Buzzard (1913) thought that his patient, the only example to be reported in this country, had neurosyphilis, but there was no proof of this and disseminated sclerosis is at least as probable.

Lambert, Love, and Mulder (1961) have reported facial myokymia in patients with pontine glioma. Unfortunately only a brief description has so far been published and, although there are many points

of similarity, it is not possible to be certain that they are describing the same condition. Fasciculation of the facial muscles may occur in a variety of affections of the facial nerve and nucleus but, with this possible exception, facial spasm of this precise type does not appear to have been described in any patient who undoubtedly had some condition other than disseminated sclerosis.

CONCLUSION

Seven attacks of facial myokymia affecting five patients are reported and the clinical and E.M.G. features described.

In one patient the diagnosis of disseminated sclerosis appeared certain and in another probable, but in three the myokymia was an isolated event.

The condition is probably much less rare than has been supposed.

I wish to thank Mr. R. H. Shephard who referred two of these patients to me.

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ADDENDUM

Since this paper was submitted a further example of facial myokymia in disseminated sclerosis has been encountered.

A woman of 32 was first seen in 1963. When 19 she had experienced dimness of vision of one eye, she could not remember which, for two months. In 1959, for a period of nine months, she had very frequent brief attacks of weakness of the left leg and after this her walking never quite returned to normal. One month before she was seen her walking markedly deteriorated. The cranial nerves and upper limbs were normal. The gait was ataxic, both plantar reflexes were extensor, vibration sense was lost at the ankles, and the abdominal reflexes were absent. Her symptoms improved and she was able to continue at work. In September 1965 her walking again became more difficult and she had precipitancy of micturition. On 1 December she noticed continual flickering of the whole of the left side of the face and, when seen two weeks later, she had typical facial myokymia with narrowing of the palpebral fissure and elevation of the angle of the mouth. There was no evidence of trigeminal nerve involvement. The myokymia subsided gradually and could not be detected a month after the onset.