Continuous muscle spasm in intramedullary tumours of the neuraxis

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There are four descriptions in the literature of a brain-stem glioma causing a characteristic continuous hemifacial spasm with paresis of the affected muscles (Rasdolsky, 1935; Sogg, Hoyt, and Boldrey, 1963). We recount a fifth case and record the electromyographic studies which have not been fully described before. In the literature this type of facial spasm has only been described in intramedullary lesions.

The electromyogram suggests that the hemifacial spasm is due to the motor neurones firing continuously at an excessively high rate, presumably due to damage to the mechanisms which normally inhibit such a high rate of firing. The mechanism seems to be similar to that affecting the shoulder and arm muscles in a case of cervical cord tumour described by Rushworth, Lishman, Trevor Hughes, and Oppenheimer (1961). Penry, Hoefnagel, Van Den Noort, and Denny-Brown (1960) described a similar spasm in traumatic lesions of the cord. This type of spasm may be diagnostic of an intramedullary lesion of the neuraxis.

CASE REPORT

R.R.P., aged 22, complained in December 1959 that for the previous six months his right eye had tended to close spontaneously. By March 1962 he had developed a progressive, continuous, right facial hemispasm and there was now double vision on looking to the right. He had morning headaches for two months but no loss of vision; he was aware of some deafness in the right ear but admitted to no tinnitus or vertigo.

Neurological examination showed gross nystagmus on looking to the right, with the rapid movement to the right; to the left the nystagmus was of smaller amplitude and pendular; there was vertical nystagmus also. The right corneal sensation was blunted and there was minimal impairment of all modalities of sensation over the right half of the face and gums. The jaw appeared to deviate to the right on opening the mouth although the right facial spasm made it difficult to be certain of this. The right lateral rectus muscle was weak. The right face was continuously screwed up so that the tissues of the right cheek felt thickened (Fig. 1). There was continuous faint rippling of the contracted muscles of the right face which were paretic. The spasm persisted unchanged during sleep and was equally evident in repose as during animated conversation. There was no fasciculation and no associated movements of the type common in facial nerve injuries. Taste was diminished over the right side of the tongue. There was slight perceptive deafness and diminished caloric response on the right. Posterior pharyngeal sensation was diminished on the right and there was curtain movement of the pharynx to the left. The right half of the soft palate was weak and the right vocal cord immobile. The sternomastoid and trapezius muscles were somewhat thinner and weaker on the right. In the limbs there was dysdiadochokinesia and diminished tone on the right: sensation, reflexes, and sphincters were

FIG. 1. Appearance on 14 March 1962, from left to right: face at rest, showing teeth, showing tongue.
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not affected. The cerebrospinal fluid was under normal pressure and contained 37 mg. of protein per 100 ml., less than 1 cell per c.mm., and normal serology. The electroencephalogram showed no localized intracranial disturbance but it was marred by a continuous right electromyogram due to spasm in the right facial muscles.

R.R.P. was admitted to the National Hospital, Queen Square, where a Myodil ventriculogram showed that the Sylvian aqueduct was deviated to the left and there was a large filling defect in the right side of the fourth ventricle (Fig. 2). On March 23, 1962 Mr. Wylie McKissock explored the posterior fossa. The right half of the pons and medulla was smooth, white, and grossly distended due to an intramedullary lesion 'almost certainly a glioma'. A median bony decompression was made by nibbling the posterior margin of the foramen magnum. He was given a course of deep x-ray therapy. Treatment resulted in a temporary arrest of the symptoms and some subjective improvement. He continued in modified employment until May 1965 when he went downhill rapidly, with increasing intracranial pressure, morning headaches, and papilloedema. He died on 29 May 1965.

ELECTROMYOGRAPHY

The patient was first seen for electromyography on 5 June 1961. At this stage there was continuous involuntary spasm of the right side of the face. A strength duration curve in the right frontalis muscle showed partial denervation. Needle electrodes were inserted in the right frontalis and the levator anguli oris muscles. There was continuous, spontaneous activity of a remarkable nature (Fig. 3). There were almost continuous high-frequency discharges of low voltage units at a frequency of approximately 100 per second. Against this background higher amplitude units fired at a frequency of approximately 20 per second. Attempted voluntary contraction of the paretic muscles did not significantly change the pattern of discharge. Conduction latencies in the facial nerve were normal. It was not possible to affect the discharge by stretching the facial muscles manually or by moving the head and neck.

Electromyography was carried out again in March 1962 on both the frontalis and levator anguli oris muscles and the findings were exactly the same as those of June 1961.

At a final examination in May 1965 there was still a good deal of spontaneous activity but the rate of firing
FIG. 4. Section of brain-stem showing right pontine tumour.

FIG. 5a. Transverse section of pons showing tumour cells surrounding motor neurones (haematoxylin and eosin × 100).

FIG. 5b. Rounding of contour of motor neurones and absence of dendrites (Nissl × 400).
of the low voltage units had decreased to approximately 60/second. In addition, units of larger amplitude fired regularly at a frequency of two to three per second. Only a few extra motor unit discharges appeared on voluntary effort. The conduction latency was still normal. Again there was no reflex response to stretch. Stimulation of the supraorbital nerve induced a reflex response on the normal side but no response was detected on the affected side.

POST-MORTEM REPORT

BRAIN CUTTING Cutting the brain merely showed some slight symmetrical dilatation of the lateral and third ventricles. Cutting the brain-stem showed a large, white, very poorly demarcated tumour grossly expanding and obliterating all anatomical features in the right midbrain, pons, and medulla. This extended up all three cerebellar peduncles on the right to involve the cerebellar white matter on that side and to displace the dentate nucleus backwards. The fourth ventricle was flattened and distorted from the right antero-posterior direction, being reduced to a mere slit (Fig. 4).

HISTOLOGY Sections of the pons showed that part of the tumour was a low grade polar astrocytoma, especially in the right side of the pontine tegmentum. This is presumably where it started, but its nature had elsewhere changed into that of an astrocytoma grade III containing large pleomorphic astrocytic forms and mitotic figures. The rounded contour of the motor neurones and the absence of dendrites (Fig. 5a and b) is similar to that described by Rushworth et al. (1961).

DISCUSSION

Our case is the fifth described in the literature where a brain-stem glioma was associated with this unusual type of continuous facial spasm with weakness and it is worth considering if this sign is pathognomonic of an intramedullary lesion of the brain-stem.

This sign is easily distinguished from cryptogenic hemifacial spasm in which intermittent, shock-like spasms occur in the facial muscles of one side: the muscles are of normal power and the nervous system is otherwise intact. The facial spasms which sometimes follow injury to the facial nerve are confined to muscles supplied by the damaged branch of the nerve: the affected muscles are usually weak but the history of injury makes the diagnosis evident. A Bell’s palsy may be followed by spasms of the weak muscles but here the history indentifies the condition and the fasciculation is intermittent. Other extramedullary lesions which might affect the facial nerve are the group of cerebello-pontine angle ‘tumours’.

This group includes acoustic neuromas, posterior fossa meningiomas, ependymomas, choroid plexus papillomas, cholesteatomas, neuromas of the fifth, ninth, or tenth cranial nerves, abscess, aneurysm or cyst of the lateral recess.

In Cushing’s (1917) very large series of acoustic neuromas there were only two cases which showed facial hemispasm: in both the spasms were intermittent. Revilla (1948) reviewed the records of the Johns Hopkins Hospital and in 205 cases of cerebello-pontine angle tumour there was none with continuous facial spasm and weakness. Therefore it seems that this syndrome only occurs in cases of intramedullary brain-stem tumour.

Even in pontine gliomas, which are the common tumours in this region, continuous facial spasm with weakness is rare. Bucy and Keplinger (1959) reviewed details of 105 brain-stem gliomas: they describe no case of facial involvement resembling this condition. It should be noted that the mean age of the five patients who had a pontine glioma and showed this sign was 37 years. This is a striking finding as pontine gliomas usually occur in children. Of the 105 cases of pontine glioma collected from the literature by Bucy and Keplinger (1959), 44% were in the age group 5-9 years and only 14 patients were over 20 years of age when diagnosed. This suggests that continuous facial spasm with weakness is a rare but possibly pathognomonic sign of intramedullary brain-stem tumours which is most likely to be seen when the tumour arises in the third or later decades.

The facial spasm shown by this patient was unusual: it was continuous throughout the day and persisted during sleep. To the naked eye the spasm was not affected by efforts to contract or relax the facial muscles which were paretic. Electromyogram of the facial muscle showed unusually high frequency continuous discharges (100 per sec.) which were not influenced by reflex mechanisms and were scarcely affected by voluntary contraction. This electromyographic pattern is quite distinct from that seen in pyramidal or extrapyramidal spasticity, both of which are caused by overactivity of the muscle spindle stretch mechanisms and in neither of which conditions is there such a high rate of motor neurone firing. A similar electromyographic picture was described by Rushworth et al. (1961) in a man who had spasm of the shoulder and arm muscles due to a tumour of the cervical cord. Rushworth did elaborate investigations of the reflex mechanisms which normally affect the rate of motor neurone firing and showed that the motor cells in his case were uninfluenced thereby. In Rushworth’s case (Rushworth et al., 1961), as in ours, the onset of the spasm was insidious. Penry et al. (1960) described similar continuous spasms in relation to traumatic cord lesions, and here, as might be expected, the onset of the spasm was sudden. In Rushworth’s, Penry’s and our case, the spasm, once it developed, was continuous and was associated with an exceptionally high rate of motor neurone firing. Rushworth et al. (1961) pointed out that their case showed the anomaly that
the motor cells which were unable to respond to reflex mechanisms or to volition continued to fire involuntarily and at an excessive rate. These authors suggested that the essential lesion in such continuous spasms is isolation of the anterior horn cells from the interneurones which convey the excitatory and inhibitory potentials that determine the rate of firing of these cells. Such isolation of the anterior horn cells would explain the loss of voluntary control and why it was not possible to influence the rate of firing reflexly. Penry et al. (1960) considered that the continuous and excessively high rate of firing of the anterior horn cells in his cases might be due to isolation of the motor neurones from their Renshaw cells.

The Renshaw cells are situated close to the anterior horn motor cells in the pathway of the efferent motor fibres. These cells are activated by collaterals from the motor axons and are stimulated when an action current passes down the motor fibres (Eccles, Fatt, and Koketsu, 1954). The axons of the Renshaw cells terminate on the anterior horn motor cell membranes. Action potentials from the Renshaw cells increase the inhibitory potential of the anterior horn motor cells after contraction and prevent motor neurones from firing at excessively fast rates. This mechanism may act as an ‘anticonvulsive’ device and it has been said that tetanus toxin and strychnine act by interfering with the Renshaw cells. Penry et al. (1960) added evidence that in their cases the Renshaw cells had been selectively damaged. They consider from experimental evidence that motor cells which are cut off from voluntary and reflex controls and from the inhibitory effect of the Renshaw cells do fire continuously.

Rushworth et al. (1961) thought that the excessive rate of firing of the anterior horn cells in his case was more likely due to isolation of the motor cells from all of their interneurones than from interference with the Renshaw mechanism particularly. He suggested that damage to the dendrites of the motor cells similar to that shown in Fig. 5b might be a factor in isolating the motor neurones. Clinically and electromyographically the spasm in our case is similar to that described by Penry et al. (1960) and Rushworth et al. (1961). In favour of Rushworth’s view that damage to the Renshaw mechanism is not essential in order to produce continuous spasm, there is evidence that there are no Renshaw cells in the brain-stem. Cajal (1909) described the recurrent collaterals which we now recognize as Renshaw fibres in the spinal cord but could find no recurrent collaterals in the brain-stem. Porter (1965) explored the hypoglossal motor neurones with microelectrodes and found no positive evidence of a Renshaw cell mechanism in this part of the brain-stem.

If there are no Renshaw cells in the brain-stem and if the spasm in our case is similar to that described by Rushworth et al. (1961) and Penry et al. (1960) we agree with Rushworth that damage to the Renshaw cells is not the essential cause of this type of spasm. It may be that motor neurones isolated from their excitatory and inhibitory interneurones will fire continuously.

In conclusion, continuous spasm of paretic muscles which is associated with electromyographic evidence that the motor cells are firing excessively fast and are uninfluenced by reflex mechanisms appears to be found only in cases of intramedullary lesions of the neuraxis.

**SUMMARY**

Continuous hemifacial spasm and weakness in association with a pontine glioma is described. Similar spasm and weakness in upper limb muscles has been described in cervical cord intramedullary lesions. The spasm is of a peculiar type due to a very high rate of firing of the motor neurones which may be due to isolation of the anterior horn cells from the control of the excitatory and inhibitory interneurones. This sign appears to be diagnostic of intramedullary lesions of the neuraxis.

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