Spontaneous Activity of Alpha Motor Neurons in Intramedullary Spinal Cord Tumor*

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THIS report describes an instance of alpha rigidity in man, secondary to an infiltrating spinal cord tumor.

The usual cases of hypertonicity in man have, in recent years, been attributed solely to an abnormality affecting primarily the stretch reflex as modulated by the gamma efferent system. Landau and Clarke,9,10 and others, have cast experimental doubt on that thesis, but there have been only rare descriptions in the literature of instances of muscular hypertonicity thought to result primarily from hyperexcitability in the alpha motor neuron system.

Rushworth, et al.,11 reported a man with rigidity and constant muscular contractions in the arms, which they attributed to isolation of alpha motor neurons by an infiltrating spinal tumor. Penry, et al.,9 reviewed the literature and found six cases of rigidity thought due to alpha motor neuron dysfunction, and added three cases of their own, one of which was the same as that reported by Rushworth, et al. Subsequently Tarlov14 reported two cases of alpha rigidity due to cystic disease of the spinal cord. The above reports comprise 11 cases of rigidity secondary to alpha motor neuron dysfunction. Of these, one case was due to cervical spinal cord tumor,13 one was associated with necrotizing myelopathy,9 six resulted from damage to the spinal cord of the newborn,9 one occurred after cervical trauma,9 and two occurred with cystic disease of the cord.15

The case to be described in this report resulted from a tumor in the thoraco-lumbar cord, and the evidence of spontaneous or isolated alpha motor neuron activity was pressed in the trunk, abdominal, and hip musculature.

The continuous muscular activity and the electromyographic findings are similar to those in the so-called "stiff-man syndrome" and in the syndrome of "continuous muscle fibre activity" described by Isaacs.3,4 However, they differ pharmacologically in that "stiff-man" usually responds to diazepam, and "continuous muscle-fiber activity" to Dilantin, neither of which affected the motor phenomenon in the present case. The pathophysiology of these two non-neoplastic syndromes remains obscure but appears in some way to affect the same inhibitory systems as altered by the tumor in this case.

Case Report

This 55-year-old white man developed "stiffness" in the right hip in March, 1965. In September, 1965, he began to note a feeling of soreness and numbness in the left lower abdomen and back. By January, 1966, he was aware of a limp in the right leg and a peculiar feeling that he variously described as "numbness" and "heaviness" in the left leg. In March, 1966, he was examined by an internist for a complaint of diarrhea; a diagnosis of colitis was made. During that examination the physician noted a board-like abdomen and continuous wave-like contractions in the right paraspinal and right abdominal muscles. The patient was referred for evaluation of these findings in September, 1966, 18 months after onset of the initial neurological complaint.

Examination. He appeared quite healthy and complained only of mild soreness and feelings of heaviness in his body. He was aware of the wave-like contractions of his abdominal muscles but did not seem disturbed about this. Neurological examination of the cranial nerves and arms was normal. The patient's stance was characterized by scoliosis of the lumbar area, convex to the

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right, with a compensatory curve in the thoracic area. His gait was somewhat awkward due to stiffness in the right leg and a "pelvic drop" due to weakness in the right gluteus medius. There was increased resistance to passive movement in both legs, more so on the right. This was not a clasp-knife rigidity, but rather a "plastic" resistance that was uniform throughout the range of movement of the hip and knee. Tonus at the ankle was normal bilaterally. There was no muscular atrophy. Very mild weakness, 4+/5, was present in the proximal muscles of the right leg, but strength was otherwise normal. There was slowness in rapid rhythmic alternating movements at all joints in the legs, again more impaired on the right. Reflexes in the leg were graded at 3+, on a scale of 0 to 4+. There was neither clonus nor a pathologic toe sign.

The abdominal wall had a board-like rigidity at all times. The paraspinal muscles were in a state of chronic contraction, and appeared to be "bunched up." None of the muscles was tender to pressure.

Continuous wave-like contractions were present in the right paraspinal muscles, the lower right intercostal muscles, the right abdominal wall muscles, the adductor muscles of the right hip, and the right gluteus maximus. On the left only the low paraspinal muscles were involved in the spontaneous movements, and these were not nearly as striking as those on the right. Interestingly, neither the right quadriceps nor any of the muscles of the posterior thigh, leg, or foot were involved in these abnormal movements.

Sensory examination revealed a band of hypalgesia and hypesthesia on the right at T-11 through L-2, a band of hyperpathia on the left in the T-9 through T-12 derma-
tomases, and hypalgesia in the left lumbar dermatomes, progressing to analgesia in the upper sacral dermatomes, but with normal sensation in the saddle area. These sensory findings suggested an intramedullary lesion. Vibratory, kinesthetic, and position senses were mildly impaired bilaterally in the legs (Fig. 1).

Films of the thoracolumbar spine revealed scoliosis, but no enlargement of the canal or pedicular erosion was noted. Myelography showed a symmetrical expansion of the cord shadow beginning at the 11th thoracic vertebra and ending with a complete block at the 10th thoracic body. These films were thought diagnostic of an intramedullary lesion (Fig. 2). Spinal fluid protein below the block was 180 mg/100 cc.

**Physiological and Pharmacological Studies.** The wave-like contractions in the axial muscles were seen to be superimposed upon a state of tonic contraction. The contractions appeared to involve the hip adductor muscles much as a whole, and occurred at about 29/min, whereas the external oblique contracted independently in its various individual slips, as did the paraspinal muscles. These abnormal contractions persisted at much the same rate whether the patient was awake or asleep. They persisted under light anesthesia with nitrous oxide and fluothane, but were abolished by a muscle paralyzing agent (anectine). The contractions were not diminished by quinine sulphate 0.9 gm daily, atropine 0.4 mg four times daily, dexamethasone 16 mg daily, Dilantin (sodium hydantoin) 250 mg intravenously and continued thereafter at 100 mg four times daily by mouth, sodium luminal 120 mg daily, and Valium (diazepam) 40 mg daily. The contractions were lessened but not abolished by intravenous Tolserol (mephenesin) given in a single dose to the point of nystagmus, nausea, and drowsiness.

The movements in the abdominal wall were not altered by breath holding at maximum inspiration nor expiration, nor by hyperventilation for 30 sec. Various passive positions in abduction and adduction did not alter the movements in the adductor longus. Forced adduction visibly occluded the phasic contractions in the adductor longus. Forced abduction of the hips did not by reciprocal inhibition appear to lessen the phasic contractions of the adductor longus.

Conventional electromyographic recording of affected muscles, utilizing a concentric needle electrode, indicated continuous discharge of normal appearing motor units, with a voltage range of 300–600 μV, of 4–6 msec duration. No fibrillations, positive sharp potentials, or giant polyphasic units were seen (Fig. 3). Isometric contraction of the sampled muscle would increase the frequency and amplitude of tonic activity; this striking increase of the background activity made it impossible to identify the phasic component with this technique of recording. A similar response was noted with the Jen-drassik (reinforcement) maneuver; the effect, however, was not as gross nor sustained as that seen with direct contraction of the sampled muscle.

Further investigations were carried out by recording simultaneously on an eight channel Grass electroencephalograph the surface potentials over the right adductor longus,
right external oblique, right high paraspinal muscles, right low paraspinal muscles, left low paraspinal muscles, and right rectus femoris muscle. Figure 4 shows the resting pattern. The continuous background activity was admixed with recurring phasic bursts of high voltage activity. The interval between bursts was somewhat variable. There was no consistent relationship of the phasic bursts in the adductor longus to those in the external oblique or in other muscles. There was, as expected, the normal, quiet appearance of the recording from the electrode over the rectus femoris, a muscle not visibly affected by the wave-like contractions.

The effect of a proposed gamma efferent block on the muscular contractions was studied using the method described by Rushworth. In this instance, 1% Xylocaine was injected into the motor point of the adductor longus. The initial 10 cc caused a temporary increase in background activity. Two minutes after the infiltration of a second 10 cc of 1% Xylocaine there was marked reduction of the phasic component of the muscular activity. This is demonstrated in Fig. 5. It is postulated that the dilute Xylocaine was achieving predominantly a gamma block, for during the block, voluntary contraction of the adductor longus was still very strong although its tendon reflex

![Fig. 3. EMG recording of right paraspinal muscle with patient in repose in prone position: Gain: 7 mm = 100 mV, time base: 30 msec per large block.](image)

![Fig. 4. Pattern of electrical activity from muscles in resting state. The spikes in paraspinal leads are EKG artifact.](image)
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was considerably diminished. Presumably therefore the bursts or phasic components of activity in this case are at least maintained, if not generated, by a stretch reflex in the affected muscles.

Figure 6 demonstrates the potentiation of electrical activity during isometric contraction of the adductor longus while the motor point block was still effective for abolishing the phasic or stretch reflex component of the muscular activity. The EMG over the adductor longus during forced adduction was identical before and during the block, and voluntary contraction of the muscle was not impaired. Note the spread of increased activity to the other muscles (Jendrassik phenomenon). The phasic bursts reappeared in the record 18 min after the initial injection (Fig. 7), and simultaneously the muscle visibly began to contract as before.

![Fig. 5. Two minutes after infiltrating motor point of adductor longus with dilute Xylocaine in an attempted selective blockade of gamma efferent fibers. Note marked diminution of the phasic bursts while tonic background activity persists.](image)

![Fig. 6. While block is still effective for gamma efferent fibers, adduction of thighs remains strong and unchanged from pre-block status. Electrical activity shows normal and continuous high spiking.](image)

![Fig. 7. Phasic components return in adductor longus 18 min later as effect of block disappears.](image)
An attempt was made to study isolated motor units by using a bipolar electrode, with the points separated approximately 1 mm. With this technique we were able to study fairly discrete groups of motor units. Once again continuous spontaneous motor activity was recorded from affected muscles at rest (Fig. 8). These discrete units were increased in frequency by isometric contraction, and were apparently not inhibited by contraction of antagonistic muscles. The relatively quiet period between bursts of activity in the isolated group units ranged from 0.5 to 0.8 sec and was not sufficiently constant to be certain of the influence of an induced stretch reflex on the sampled unit.

Clinical Course. The area demonstrated on myelography was explored by laminectomy. The cord appeared swollen and hypervascular (Fig. 9). Under ×4 magnification, an incision was made into the right dorsal column to a depth of 3–4 mm. No visible tumor was seen at this depth, and tissue taken for biopsy was read as showing only neuronal and glial tissue. No fluid was obtained on attempted needle aspiration of the involved area. It was concluded that the lesion was probably a low-grade infiltrating glioma and that further attempts at biopsy might be disastrous. Postoperatively the patient required a urinary catheter for 8 days, but his preoperative strength was little changed. Over the next month he was given a course of deep cobalt radiation. His case has subsequently been followed for 20 months, showing a very gradual increase in his neurological deficit.

Discussion

The clinical, myelographic, and surgical findings leave little doubt that the findings in this case relate to an intramedullary tumor. O'Connor, et al.,7 and Sogg, et al.,13 have described tonic and phasic contractions of the facial muscles in cases with pontine glioma. The histologic descriptions in these cases, and in Rushworth's11 case, indicated

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**Fig. 8.** Bipolar recording of isolated small group of motor units. In lines 2, 3 and 4 the high voltage slow wave indicates contact of reflex hammer with patellar tendon.
isolation of alpha motor neurons (facial nucleus and anterior horn cells respectively) by an infiltrating tumor. These authors also noted a rounding of the contour of the motor neurons, and a relative absence of dendritic endings on the motor cells. The EMG findings in those cases were similar to those in this report.

Gelfan and Tarlov,2 and Tarlov15 have offered experimental and histological evidence that the phenomenon of spinal alpha rigidity in experimental animals and man is due to a loss of the interneuronal influence on alpha motor neurons. In their experimental work a proper ischemic episode in dog was found adequate to destroy the intermediate grey zone, and the central part of the ventral and dorsal grey horns. This lesion produced a state of chronic rigidity in those muscles innervated by the surviving alpha motor neurons in the periphery of the lesion in the ventral grey horn.

A point of anatomical interest in this case related to the predominant involvement of trunk, abdominal, and hip muscles, with sparing of certain muscle groups (quadriceps femoris) seemingly innervated from the same cord segments as were the nearby involved muscles (hip adductors). The topographical arrangement of the anterior horn cell population offers a possible explanation of this phenomenon. The cells projecting to abdominal, trunk, and hip muscles lie in the more ventro-medial aspect of the ventral grey horn and could be isolated by an infiltrating lesion that might spare the more dorso-laterally placed cells in the ventral horn projecting to limb muscles. This clinical evidence for location of the lesion in the ventro-medial aspect of the ventral grey horn fits very well with physiologic studies on inhibitory mechanisms in the cord, for Eccles, et al.,9 and others, have shown that inhibitory mechanisms10 are concentrated in the ventro-medial zone of the ventral horn. The isolation of this inhibitory interneuron population from adjacent motor neurons could contribute to the spontaneous firing of those cells as described in this case.

There are probably at least two factors playing a role in the spontaneous discharge of the alpha motoneuron in the subject of this report; damage to and isolation of alpha motoneurons. Either or both of these adverse factors could account for the asynchronous background or continuous activity recorded in the EMG, but the hypersynchronous bursts that appear to affect simultaneously many muscle bundles, or the entire muscle, would seem to invoke even a third factor, which in this case as shown by the block experiments would appear to implicate the stretch reflex.

Summary

A case of alpha rigidity in man has been reported and the pathophysiological, electrical, and pharmacological features of this state described. The continuous motor activity is thought to represent a release phenomenon, i.e., an isolation of the anterior horn cells from their inhibitory interneuron connections. The wavelike contractions probably represent a mechanism related to

Fig. 9. Picture at surgery, dura open but arachnoid intact. Note obliteration of subarachnoid space by distended cord. At upper end of lesion subarachnoid fluid is again present.
the stretch reflex. The findings of continuous muscle activity in a patient should raise the possibility of an intramedullary lesion, as in this case.

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