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 Clare,5,6 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.,6 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,12 one was associated with necrotizing myelopathy,6 six resulted from damage to the spinal cord of the newborn,8 one occurred after cervical trauma,6 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 expressed 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"8 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-