CASE 7


The patient was a 61-year-old diabetic Negro man who had developed progressive muscular weakness over the preceding seven to ten years. The weakness had been confined to the lower limbs until the last year of his life, when the upper extremities became involved. He experienced dyspnea and dysphagia a few weeks before hospitalization.

His mother and one sibling had died with diabetes mellitus; another sibling and a maternal aunt were alive with this condition. There was no family history of neuromuscular disease.

There was marked atrophy of the muscles of the extremities, particularly evident among the thenar and interosseous muscles. The deep tendon reflexes were hypoactive, but no sensory deficits were found, and pyramidal tract signs were absent. The functions of cranial nerves I to XII were intact, but there were prominent fasciculations of the tongue.

The serum creatinine was 0.4 mg/100 ml, SGOT 24 units, LDH 340 and 580 units, CPK 23 and 15 units, and PBI 3.8 mcg/100 ml. The spinal fluid was clear and contained only two lymphocytes per mm³, the total protein was 25 mg/100 ml, and the sugar was 95 mg/100 ml.

Intraneuronal conglomerate masses were observed in the majority of the anterior horn cells and in some of the motor neurons in the brain stem.

Submitted are: 1 stained slide, 1 unstained slide, and 1 composite photograph of the light and electron microscopic observations.

For discussion: Nature of the neuronal inclusions.