

CASE 1

A housewife aged 58 years was referred with the complaint of weakness of her arms and legs. Apart from an illness affecting her legs at the age of 9 years (which had been diagnosed as poliomyelitis, and from which she had apparently made a complete recovery after a few weeks) she had enjoyed good health until about 2 1/2 years before being referred. She had then noticed that her left foot and leg became 'tired and tended to drag' when she had walked for several minutes. After a few weeks she noted a definite weakness in this region even when she was at rest; this weakness progressed, the right leg and foot becoming similarly affected within two or three months. Her hands later became weak so that she experienced difficulty in 'wringing out' wet clothes and in unscrewing bottle tops, and she frequently dropped household objects such as cups and glasses. During the last six months her speech had become less distinct and solid food often tended to stick in her throat when she tried to swallow; there was no nasal regurgitation of liquids when swallowed but at night in bed she frequently had difficulty in clearing the back of her throat of mucus. By this time the patient was severely incapacitated, being able to walk only with considerable difficulty and the weakness of her upper limbs had progressed to such an extent that dressing herself was a long and laboring process; her fingers felt clumsy and weak and undoing and doing up buttons was great trouble to her. During the period of the illness her weight had dropped from 9 st. 2 lb. to 6 st. 12 lb.

Neurological examination

This revealed a generalized loss of muscle bulk. Wasting and fasciculation were conspicuous in the muscles of the shoulder girdle, biceps, triceps, quadriceps and calf muscles. In general the lower limbs were less wasted than the upper limbs. Motor power was generally diminished, being most severely affected where the muscle wasting was most evidence. The limbs were hypotonic, especially the upper limbs. The supinator, biceps and triceps jerks were all either absent or very difficult to elicit, but the knee and ankle jerks were slightly exaggerated and the plantar responses were extensor. No impairment of cerebellar function was demonstrable though muscle weakness occasioned some difficulty with the heel-knee and finger-nose-finger tests. The gait was slow with short shuffling steps and evinced a poverty of knee flexion. The tongue was bilaterally wasted with wrinkling of the overlying mucosa and showed fasciculation when at rest in the floor of the mouth. The jaw jerk was exaggerated and the soft palate moved poorly. There was a nasal intonation of speech and a slight slurring dysarthria. There was no sensory impairment and no impairment of intellectual functions. General physical examination, including a pelvic examination, was otherwise negative. Examination of cerebrospinal fluid revealed no abnormality and serological tests for syphilis were negative in this fluid and in the blood. X-Rays of the chest and cervical spine and further ancillary tests did not disclose any abnormality.

CASE 2

A 59 year old woman was brought to clinic by her husband because of concern about her memory. The couple was interviewed together, with the wife giving most of the history. She started by saying that her family seemed overly concerned about her health. Over the past few weeks, she had some trouble remembering things such as the location of her bank, the post office and occasionally trouble with certain phone numbers..."I've never been good with numbers anyway". Other than such minor problems she felt generally well, although there was some fatigue and she reported "feeling cold most of the time". The husband then added some history. The family had started noticing problems about a year earlier. Initially this was just forgetfulness. Over the past six months, her speech began to noticeably change..."She frequently seems to be searching for words, and sometimes says something that makes no sense, like 'the lawn needs to be cluttered' when she meant 'cut'. Also she gets upset more easily. At the bank last week, she became convinced that the teller was hiding some of her money. It's a new bank and the unfamiliar faces seemed to really bother her. Well...what really got me worried was that last week, we took a walk in our neighborhood and she couldn't find the way back. If I hadn't been there, I'm not sure how she would have gotten home". Her past medical history included hypertension for 15 years. She had also had one period of depression in her 40s that lasted about six months and improved with amitriptyline. Eventually, the medication was discontinued without a recurrence of her depression. Present medications were clonidine and daily vitamins. She did not smoke cigarettes and rarely drank alcohol.

Physical examination

Blood pressure was 150/85. She was neatly dressed. Head and neck exam was normal, as was the general exam. The mental status exam showed her to be alert and oriented x3. Speech was normal, but language was slightly slow. She made frequent pauses to find a word, often with her husband providing it. Several paraphasic errors were made. She could name most objects but when asked what the stem of a watch was she said 'stick', and eyebrow became 'eyeband'. Repetition and comprehension were intact. On memory testing, she could register 3 objects, but retained only 2 at 5 min and none at 15 min. Long term memory seemed normal. She could not do serial 3s, and even serial 2s were slow; also she stopped mid-sequence and couldn't remember the task. Her affect appeared normal, and mood was described as 'good'. She had much difficulty reproducing a drawn figure. Cranial nerves were normal, although both optic nerves were slightly pale. Motor testing showed normal power, but there was definite Geggenghalten; no cogwheeling or spasticity was noted. Reflexes were 2+ and symmetrical with flexor plantar responses; a snout and glabellar sign were present. Sensory exam was normal to pinprick and temperature, but she had trouble with stereognosis. The remainder of the exam was normal.