

Clinical Cases.

A CASE OF PROGRESSIVE MUSCULAR ATROPHY WITH UNILATERAL ATROPHY OF THE TONGUE.

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IN the course of progressive muscular atrophy, and especially towards its later stages, it not unfrequently happens, as we all know, that the degenerative process which primarily had invaded the large nerve cells of the anterior cornua of the spinal cord, extends upwards and involves the motor nerve nuclei of the pons and medulla.

In such cases the characteristic symptoms of glosso-labial paralysis are added to those of the original affection.

In the following case of progressive muscular atrophy the degenerative process had extended to the medulla and pons, and had seriously affected the nerve nucleus of the 9th (hypoglossal), and to a less extent that of the 7th (facial), on the right side, the result being most marked atrophy of the right half of the tongue, and some wasting of the right cheek. There was no affection of speech, no difficulty in swallowing, in short, no symptoms of glosso-labial paralysis. All the movements of the tongue could be accurately performed; indeed, until I directed the patient's attention to the condition of the organ he was not aware that there was anything the matter with it. This fact, and the circumstance that the lesion in the medulla was so limited and so well defined, must be my excuse for reporting the case in what may perhaps be thought somewhat wearisome detail.

M. B., æt. 55, labourer, living in the Cowgate, Edinburgh, was sent to me by my friend Dr. Wilson, at the beginning of March of the present year, and was subsequently, at my request, admitted to the Royal Infirmary, Ward 23, where through the kindness of Professor Sanders I had repeated opportunities of examining him. I have also to thank Professor Sanders for allowing me to make use of his

notes, and Mr. Frank Rand, the clinical clerk, for valuable help in the examination of the case.

The patient's chief complaints were muscular weakness, most marked in the upper extremities, especially in the right; numbness and increased sensitiveness to cold in the right half of the body; and giddiness.

Previous History.—Ten years ago he began to feel his right hand and arm "numb and weak." The weakness slowly but gradually increased. After a time (the exact date he was unable to fix) the left arm and hand became similarly affected. Until two years ago he was able to follow his ordinary occupation, that of a labourer. The weakness then (two years ago) became so great that he had to take to chopping sticks. For the past year he has not been able to follow any employment. Ever since the muscle weakness was first noticed he has been subject to attacks of "lightness in the head." These attacks have been much worse of late, and he has in consequence had several falls.

He knows no cause for his complaint. His general health until lately has been particularly good. He has been fairly temperate. Has not had syphilis. Like most labourers, he has been a good deal exposed to wet and cold, but has never suffered from rheumatism. His surroundings until lately have been fairly comfortable.

Family History.—Good. His father and mother died at an advanced age. His brothers and sisters (five in number), and his own children, are all healthy. So far as he knows, he has no hereditary tendency to nerve complaints.

Present Condition.—He is a small man, and looks much older than his years. Is very feeble, and walks with difficulty by the help of a stick, the gait being tottering and senile. He has a marked stoop.

There is a recent scar over the right eyebrow, the result, he says, of a fall during an attack of giddiness.

Nervous System; Motor Functions.—The muscular power generally is very much below par. The weakness is extreme in the upper extremities, especially in the right. The patient is unable to "spread" the fingers of the right hand. The movements of the phalangeal joints of the fingers and thumb of the right hand are imperfect. This is in great part owing to joint stiffness. All the other movements of the right upper extremity can be performed, though with extreme feebleness, the slightest passive resistance being sufficient to prevent their execution. The grasping power of both hands, as measured by the dynamometer, is much below par, that of the right being much weaker than that of the left.

The right hand presents to some extent the "bird-claw"

character, but the appearance is not typical, owing to the fact that the fingers and thumb are rigid at their first phalangeal joints.

The rigidity of the little finger dates from childhood, and is the result of a fracture; that of the ring-finger was caused by an abscess some few years ago; that of the middle finger resulted from an injury thirteen years ago; that of the thumb dates back twenty years, and resulted from an injury; that of the forefinger came on of itself, without any obvious cause. (? Could these injuries have been in any way the cause of the disease? Could they have been the exciting cause of an ascending degeneration, which primarily affected the nerve nuclei supplying the hand muscles?)

The fingers of the left hand are slightly flexed at their first phalangeal joints. There is some stiffness and rigidity on passive movement at the same joints.

All the small muscles of the right hand and thumb are in an extreme state of emaciation. The muscles of the right forearm and the muscles attaching the right upper extremity to the trunk are also much wasted. The atrophy is more marked in some muscles and in some parts of muscles than in others; the upper part of the trapezius, for example, is much more affected than the lower.

The small muscles of the left hand and thumb, and of the left upper extremity, are also atrophied, though in a less degree than those of the right.

The right half of the tongue is much wasted; it is soft and flabby, marked with furrows and wrinkles, and presents a striking contrast to its plump fellow of the opposite side. The atrophy is accurately limited to the middle line. The tip of the organ when fully protruded is slightly turned to the right (atrophied) side. All the movements of the tongue can be accurately performed; in fact, as I have already remarked, until the patient's attention was directed to the condition of the organ he was not aware that there was anything the matter with it. The soft palate and uvula are natural. There is no difficulty in speech, mastication, or deglutition.

The right side of the face is somewhat atrophied, the wasting being most marked in the cheek, which feels thinner and less firm than the left.

The other muscles supplied by nerves coming off from the medulla and pons seem normal. The muscles of the trunk, with the exception of those before mentioned, and the muscles of the lower extremities, are soft and flabby, but do not present any appearance of special atrophy.

The *mechanical irritability* of the atrophied muscles seems natural. *Electrical irritability*, a careful examination of the

atrophied muscles, including the tongue, show a simple diminution, especially in the strength of the contraction produced, to both forms of current. There is no trace of "reaction of degeneration;" even in the muscles which seem most atrophied a contraction can be obtained by a strong faradic current. Very marked fibrillary twitchings frequently occur in the atrophied muscles, including the right half of the tongue.

The *reflexes* (superficial, deep, and organic) all normal.

Co-ordination seems normal.

The *muscular sense*, as tested by weights, appears to be normal.

Sensory Functions.—He complains of feeling cold. Says that the left side of his head and the right side of his body are numb.

Tactile sensibility and the *power of localising impressions* are natural.

Sensibility to pain is natural.

Sensibility to temperature is imperfect. He can always distinguish cold objects, indeed sensibility to cold is increased, but he not unfrequently says warm objects are cold. At other times a pretty prolonged contact of a warm object is necessary before it is recognised as such (delayed impression).

Sight.—The left eye is blind, the result of an old injury. Sight in the right eye is fairly good. Nothing of importance was detected on ophthalmoscopic examination.

Taste is natural.

Smell is natural.

Hearing.—Aërial sounds are very imperfectly heard in the left ear; the skull sounds, as tested by the tuning-fork, are well heard on both sides.

Intelligence is good. The patient sleeps well, and with the exception of occasional attacks of giddiness feels nothing wrong with his head. The attacks of giddiness are of short duration, and generally occur when he gets up or turns round quickly. He does not vomit, and has never had a fit.

The *temperature* is normal or *subnormal*.

The *radials* are somewhat hard and rigid; the heart is weak; and there is slight bronchitis. But with these exceptions the other organs and systems are normal.

The patient remained for some time under observation in hospital. His general health improved considerably. The muscular condition underwent no obvious change. He used to like faradisation, thinking it did him good. When last seen (Oct. 7th) he was much *in statu quo*.