CASE OF GLOSSO-LABIAL PARALYSIS, WITH PROGRESSIVE MUSCULAR ATROPHY AND LATERAL SCLEROSIS.

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THE following case was admitted into the National Hospital for the Paralysed and Epileptic, Queen Square, on September 18, 1879, under the care of Dr. Ramskill, to whom I am indebted for permission to publish the case.

The early part of the notes were taken by my friend and predecessor, Mr. A. E. Broster.

R. R., a waiter, aged 36, married, had no children, wife had one miscarriage. Patient had lived well and drunk freely, and had smoked much. Had scarlet fever when aged 10, but no rheumatic fever, and no syphilis.

Hereditary History.—A first cousin was insane; father's sister paralysed, and also her daughter; his father and two paternal uncles died of phthisis.

Present illness began in March 1879. Six months before admission he had had much worry, and three months previous to onset of illness he had walked through some water in the winter time, after which he never seemed free from rheumatism. He first noticed in March 1879 that his hands began to feel weak, a month later his speech began to be affected, and about the same time he had some difficulty in swallowing.

Present state in September 1879, as taken by Mr. Broster, was as follows:—He is very low-spirited, and complains of pain in the back of his head; he has the characteristic sad inanimate expression. *Face.*—In closing his eyes, the left side of face moves more than the right; has some difficulty on both sides in showing the upper teeth; there is much tremor about his chin and upper lip. Masseters act well. Complains of food collecting between his teeth and gums. Movements of eyes and eyelids normal, pupils equal.

Tongue protruded about $\frac{3}{4}$ inch beyond margin of teeth, and

deviates to the left, very tremulous, and much wasting on both sides.

Speech is an indistinct monotone, no inflexion, and not affected by closing anterior nares.

Swallowing is difficult, fluids sometimes regurgitate through nares. *Soft palate* seems wasted, uvula points to left.

Neck.—Head tends to fall forwards, and he moves it back stiffly; he can, however, turn it horizontally very well.

Trapezii weak, and left worse than right.

Supra- and infra-spinati wasted. *Serrati* and muscles of back not affected.

Upper limbs both very much wasted, and the muscles are very flaccid. The interossei and balls of thumbs are almost gone, and on the right more than the left; marked wasting of extensors of wrists, right also more than left; the right deltoid is, however, less wasted than the left, and both pectoral-maj. are somewhat affected. He can extend the left fingers, but not the right; can flex and extend both elbows, but cannot raise the arm higher than on a level with the shoulder. Has constant fibrillar tremor of the muscles of the arms and of the sterno-mastoids, and also especially in the pectoral-maj. and the deltoids.

Lower limbs feel feeble to him, and he thinks he walks stiffly, and his legs give way under him; the knees shake much and seem to "shoot him up;" cannot run, but can walk uphill better than down. He has some twitching in the muscles of his legs.

Sensibility and organs of special sense normal, and heart not affected.

He was discharged on January 15, 1880, but was re-admitted on October 14, 1880, when I found that he was generally weaker; his tongue was more wasted, and the mucous membrane thrown into folds; could protrude it only just beyond the teeth; the soft palate contracted fairly well in phonation. The upper limbs were more wasted, especially the small muscles of the thumbs and interosseous spaces, which were very hollow. He could not move the thumbs by their small intrinsic muscles, nor separate the fingers.

Both hands were flexed at the proximal phalangeal joints.

Forearms are also very thin, but he can just flex and extend the wrist-joints. Grasp of either hand by the dynamometer = 0. Can pronate fairly well, but not supinate forearms. Can extend the elbows, but not flex them so well; both upper arms wasted. Cannot quite flex the elbows, owing to the forearms being fixed in the pronation position. Deltoids are so wasted that he cannot abduct either arm from the trunk; he can, however, put the hand on to the opposite shoulder by action of

the pectoral maj. Can shrug his shoulders well. Supra- and infra-spinous fossæ are very marked and hollow. He can put his hands behind his back by the latissimus dorsi, and draw his scapulæ forwards by the serrati.

The fibrillar tremor is so marked that the fingers are continually being involuntarily moved. The muscles most affected are those which are not absolutely but only somewhat paralysed, the movement being most visible in the muscles of the arm, forearm, the pectorales majores, the sternomastoids and trapezii; while the muscles of the palm of the hand and the deltoid are not affected, or perhaps the latter are to a very slight degree; the quivering is more marked when the muscles are slack, and after they have been used.

He cannot lift the head forwards when lying on his back.

Lower limbs are somewhat wasted, the calves measuring 11½ inches in diameter. He has all the movements of the joints of the legs, but with diminished power. In walking he scuffs very much, dragging his feet along the ground. All the muscles of lower limbs, except the gluteus maximus, have fibrillar tremors, but less than in the arms. The muscles of the back are also affected in the same way.

The joints of lower limbs are rather rigid.

All the muscles, and especially those of upper limbs, are very sensitive to direct percussion, but the contraction is fibrillar, and not of the whole muscle as in health.

Knee-phenomenon (patellar tendon reflex) is very excessive; right and left ankle-clonus present on both sides.

Plantar reflex readily obtained on either side; also the *abdominal* and *epigastric* reflexes.

Electrical reaction to faradisation; the intrinsic muscles of the left thumb do not react to the strongest current of a Stohrer's battery; the left extensor digitorum and right and left interossei require a strong current; the muscles of the right thumb, the right biceps and extensor digitorum, react to one degree of the scale weaker; both flexor digitorum muscles and right deltoid respond a little better; while the triceps of both sides and the left deltoid and biceps are somewhat below normal. The pectoralis major and the trapezius on both sides, however, react readily to a normal stimulus.

Patient was discharged from the hospital in about the same condition, on January 1, 1881.

In July 1881 he was again readmitted, and said he had not been able to walk for three months—since April.

Swallowing is now more difficult, and speech worse; soft palate moves only very slightly in phonation, and he says "ngrum" for "grub;" but pronounces better when anterior nares are closed. Cannot blow with his mouth unless the

nostrils are closed. Soft palate moves better to reflex tickling with a pen.

Left forearm weaker than right. He cannot extend the fingers or wrists, though he can flex them a little; can just flex distal phalanx of thumbs, but cannot extend them. Can just flex the left but not the right elbow. Can just put either hand to opposite shoulder, but the left with more difficulty than the right.

Can use trapezii and serrati much less; otherwise the movements are as before, only weaker. Movements of thorax very slight, breathing being chiefly diaphragmatic. He cannot bring the head forwards when it is thrown back, without bending the whole body forwards. Quivering of muscles more marked.

Legs much more wasted; calves measure only $10\frac{1}{2}$ inches. He cannot walk, and can only just stand by holding on to a rail; while lying down can lift the heels off the bed, but moves the knees and ankles very slowly and with very little power.

Rigidity, which is absent in the arms, is very marked in the joints of the legs.

Knee-phenomenon is very excessive, and *ankle-clonus* very well marked.

The plantar, abdominal, and epigastric reflexes are all well marked, the first two being excessive, the epigastric less pronounced.

The cremasteric reflex is also slightly obtained.

Oct. 10th.—Patient to-day, while eating, suddenly choked from impaction of food in his larynx, and before medical assistance could be given, he was dead.

For the following examination I am indebted to my friend Mr. Horsley, Surgical Registrar at University College Hospital.

EXAMINATION OF THE SPINAL CORD.

BY VICTOR H. HORSLEY, M.B., B.A. LONDON.

The cord was not in a good state of preservation, proving so brittle that it was unfortunately impossible to examine the dorsal region; this apparently was not due to any pathological softening.

Spinal Cord.—*Membranes* apparently normal, but the pia mater unusually adherent; microscopic examination showed, however, that the pia mater was considerably sclerosed, presenting on section an almost homogeneous appearance, from

the dense fibrous tissue of which it was composed, and containing very few nuclei corpuscles.

Vessels.—The middle coat of the smaller arterioles presented the same change, only much less marked, and the tunica adventitia had an unusually close texture; the internal coat was quite normal in every way. In one section a concentric calcareous mass of "brain sand" was found.

Nerve-roots.—Not diminished in size, nor altered in appearance (latter after hardening).

On microscopical examination, some sections of the nerve-roots showed slight destructive changes in the myelin sheaths; but these changes were post-mortem, rather than ante-mortem and pathological. The axis-cylinders showed no alteration at all. The perineurium partook of the general thickening of the fibrous tissues.

Spinal Cord. Neuroglia.—The trabeculae passing in from the pia mater were everywhere strongly marked, and the neuroglia throughout the cord was increased, and stained readily with anilin black; but in the whole motor region of the right side and left descending lateral tract in the cervical enlargement, and in both descending lateral tracts in the lumbar enlargement, the increase of the neuroglia amounted to well-marked sclerosis, and in some places consisted of dense fibrous tissue. This increase in the connective tissue was simply an excessive fibrosis of the neuroglia, which (much thickened) showed a differentiation into fibres in those places where the lesions were most marked. The capillaries were surrounded by a dense ring of the same tissue, its thickness generally being equal to the diameter of the vessel, and here and there the lumen of the perivascular lymphatics was occupied by a delicate nucleated connective tissue. There was no trace of any inflammatory process, in fact the cord presented a markedly anæmic appearance. The neuroglia in the cornua of grey matter was also condensed, and filling up the gaps in the tissue formed as the cells degenerated (*vide infra*). The walls of the vessels in the grey matter were especially thickened, and connective tissue was noticed in the perivascular lymphatics, just as described above. In addition to these changes there was found a bright substance which stained moderately with anilin blue-black, and was of homogeneous appearance, with here and there a nucleus of a leucocyte. This substance was formed in the posterior commissure between the fibres, and forming a long mass stretching the length of it; it was also formed in the perivascular lymph-spaces and scattered in the grey substance, but most usually in the lymph-spaces. Besides these situations, it was present in the cervical region at the bottom of the anterior fissure, and resting on the pia mater. It

probably was of albuminous nature, it differed from myelin in amount of refractive power, and is perhaps identical with that substance lately noted in a case of tetanus (Doran).

NERVE STRUCTURES.—1. *Corpuscles.*—The large motor cells in the anterior cornua had undergone extensive degeneration and destruction. The degeneration was not so marked, however, as simple atrophy of the whole corpuscle, which was very obvious; the cells remaining having but few processes, and the protoplasm of the whole corpuscle not staining readily with anilin. The degeneration was pigmentary, and in the right anterior cornu of the upper third of lumbar enlargement two corpuscles were completely degenerated, appearing as masses of orange-yellow granular pigment. No vacuolation was observed in any of the remaining corpuscles. The whole process resulted in the total destruction of the majority of the nerve corpuscles; thus in the cervical region there were left scarcely any (opposite the level of the 6th cervical nerves), and the surviving cells were only scattered remnants of corpuscles. In the lumbar region, however, there were more to be seen, and in the upper third of the lumbar enlargement there were three or four partly degenerated corpuscles in the left anterior cornu, the cells belonging to the large infero-external group. On the opposite side there were three cells with two or three different processes, and belonging to the upper half of the similar group. This condition varied, so that at a point a few millimetres lower there was only to be seen one corpuscle of the left anterior cornu, and then at another point the large group of cells was fairly well represented, especially on the left side.

2. *Fibres.*—The fibres throughout the "healthy" parts of the cord appeared to be fairly normal, and there was no swelling or atrophy of the axis-cylinders, which, however, did not stain readily with anilin. In the sclerosed portions the fibres simply had undergone atrophy, and there was no increase in the axis-cylinder here at all; the fibres evidently had wasted and disappeared. There were of course all degrees of the process to be seen.

Central Canal in the cervical region was almost obliterated, and the epithelium multiplied. In the lumbar region it was flattened from before back, and filled with epithelium degenerated to a considerable extent.

Nothing abnormal was found in the medulla oblongata. The rest of the brain was not examined.

Remarks.—The above case was under observation from a few months after its commencement until the death of the patient, a period extending over two years and seven months, and it

was therefore possible to make clinical records throughout its course.

The disease began with "rheumatic pains" following exposure to wet and cold, and the first marked symptom consisted in weakness of the hands.

A month after the hands were first affected, he began to suffer from well-marked symptoms of disease in the medulla oblongata, such as: difficulty in speaking and swallowing; and when he first came under observation—six months after the first symptoms showed themselves—the bulbar symptoms were very characteristic, as: pain in the back of the head; difficulty in showing the upper teeth; a sad expression, from depression of the angles of the mouth (it may here be noted that the temperament of patients suffering from this disease is usually remarkably cheerful and sanguine); tremor of chin; food collecting between the cheeks and gums; tongue tremulous and wasted; speech, a drawling monotone, and uvula wasted and deviating to the left.

The muscles of the neck were wasted and weak, and there was atrophy with paralysis of certain groups of muscles in the upper limbs; but this was unattended by any rigidity.

The lower limbs were stiff about their joints and feeble, so that he walked with difficulty.

There was also marked fibrillar tremor in the affected muscles, which was powerful enough to cause involuntary extension of the finger; this tremor was subsequently noticed as being most marked in those muscles which were considerably affected, but to be absent in those which were so much destroyed as to be powerless. The tremor was also produced by direct percussion, and took the place of the contraction of the muscle as a whole, as occurs in health to direct percussion.

The subsequent course of the disease was progressively worse.

The soft palate failed to act in deglutition, but still reacted reflexly to tickling with a quill-pen, and the patient was thereby unable to inflate his cheeks or pronounce explosive consonants, unless the anterior nares were first closed.

The muscles of the neck also lost power, so that he could not bring the head forwards by the sterno-mastoids. The groups of muscles in the upper limbs became more affected and the legs more rigid, so that two years after the onset of the illness he was unable to walk, and the knee-phenomenon (patellar tendon reflex) became very excessive, ankle-clonus also being very readily obtained in both legs.

At the end of his illness respiration became chiefly diaphragmatic. The sphincters were unaffected throughout.

This case would seem to be one of progressive muscular

atrophy, followed by lateral sclerosis—*amyotrophie spinale déutéro-pathique* of Professor Charcot—and not one of amyotrophic lateral sclerosis, in which the first symptoms are due to primary lateral sclerosis, and consist of weakness, usually of the arms, with spasmodic permanent contraction and rigidity of their joints; atrophy *en masse*, and not muscle by muscle—the patient being, however, able to raise the arm, but with difficulty, due to the rigid condition.

In this case, on the other hand, the atrophy was primary and attacked individual muscles; loss of power followed the atrophy, and there was no rigidity in the arms. The rigidity of the legs, however, occurred after the wasting of the arms, and was the first symptom noticed in the lower limbs. In cases of progressive muscular atrophy the cells of the anterior cornua are affected most in the cervical and least in the lumbar enlargements, and this was found in the microscopical examination of the present case. Sclerosis of the lateral columns was, however, very well marked in the lumbar regions; so that though the joints of the legs were noticed to be somewhat stiff six months after the onset of the illness, inability to walk, coupled with increased knee-phenomenon, and well-marked ankle-clonus, did not supervene till two years after the onset. The legs, though small, did not present excessive atrophy of individual muscles, the appearance being probably due to lateral sclerosis more than to atrophy of the anterior cornua; and when unable to walk, he would still move all the joints of the legs, though with great difficulty, owing to descending sclerosis, which was perhaps antecedent to the wasting of the cells of the anterior cornua of the lumbar enlargement (?).

The absence of rigidity in the arms, notwithstanding the well-marked sclerosis in the lateral columns, was perhaps due to the extreme destruction of the motor cells in the cervical region.

The group of muscles most affected in the upper limbs comprised the intrinsic muscles of the hands, the extensors, and, to a rather less degree, the flexors of the fingers and wrists, the supinator longus, the flexors of the elbows, the deltoids and the supra- and infra-spinati; while the pectoralis major, latissimus dorsi, triceps, teres major, and subscapularis were less affected.

According to the experimental researches of Professors Ferrier and Yeo,¹ the segment of the spinal cord at the level of the 4th and 5th cervical nerves supplies the muscles contained in the first-named group, with the exception of the flexors of the fingers and the intrinsic muscles of the hand, which are supplied by the 8th cervical and first dorsal segment respect-

¹ Proc. Roy. Soc., No. 212, 1881.

ively; while the second group is supplied by the 6th, except *teres major* and *subscapularis*, which are supplied by the 7th cervical. The flexors of the fingers are also supplied by the 6th and 8th, and the triceps by the 8th cervical nerves, and the flexors have not suffered to the same extent as the extensors. The pronators supplied by the 8th cervical were also much less affected than the supinators. The above grouping is also borne out by the faradic reaction, where the flexor digitorum reacts better than the extensor; and, on the right side, the triceps better than the biceps and deltoid; while the muscles of the hand and the interossei react very feebly or not at all; the pectoralis major and trapezius acting about normally. Clinically, the above grouping is illustrated by the fact, that in January 1880, the patient could pronate, but not supinate the forearms; could extend, but not flex elbows; and though he could not abduct either arm from the trunk, he could put the hands to the opposite shoulders, and place the arms behind the back.

From the above grouping of the affected muscles, one might expect to find that the motor cells at the level of the 6th and 7th and perhaps the 8th cervical nerves would be less affected than the other parts of the cervical enlargement; but post-mortem examination, made three months after the patient was last examined, failed to reveal any difference.

The relative affection of the various muscles is in favour of the disease being progressive muscular atrophy and not a primary sclerosis of the lateral columns. With regard to the former disease there are two theories; the one, maintained by Friedreich and Lichtheim, that muscular atrophy is primarily a myositis; and the other, that it has a primary spinal origin. According to Dr. Ferrier,¹ the atrophy of the groups of muscles in accordance with the functional relations of the segments of the cord, as is also probable in this case, is strongly in favour of the spinal origin of primary muscular atrophy, and this is also borne out here by the bulbar paralysis and the attendant lateral sclerosis.

This case agrees with Dr. Ferrier's observations,² in that the disease commences from one end—in this case the lower—of the cervical enlargement, afterwards involving the upper end, while the middle part, represented by the 6th and 7th cervical, is much less affected. The 'main à griffe,' which is produced by invasion of the 1st dorsal and 8th cervical roots, did not occur, owing to the extensors of the wrists being involved in the lesion of the 4th and 5th cervical.

¹ 'BRAIN,' Part XV.

² Ibid.