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STUDY OF

A CASE OF BULBAR PARALYSIS,

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Notes on the Origin of Certain Cranial Nerves.

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STUDY OF A CASE OF BULBAR PARALYSIS, WITH NOTES ON THE ORIGIN OF CERTAIN CRANIAL NERVES.

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Bibliographical.—There is much literature on the anatomical facts of this disease, mainly from the pens of German and other Continental writers. Notwithstanding the large amount of attention which has been given to the subject, much ambiguity still exists regarding the exact anatomical lesions which occur. The intention of this short biographical account is to submit a statement of the facts observed by others, so that a comparison may be readily made between these and our own observations.

1. *Distribution of the disease in the white matter of the central nervous system.*—The various statements concerning the highest limit of the disease are of profound importance. Usually the sclerosis, which involves the pyramidal tracts alone, has not been found higher than the pons Varolii. But certain observers have described conditions which are believed to signify degenerative processes in the higher parts of the cerebrum. Kahler and Pick[1] traced the disease into the crura cerebri, and observed further, an atrophy of the cerebral convolutions of the Rolandic area. Kojewnikoff[2] noted that the sclerosis stopped at the level of the pons, but he was able to trace "granular bodies," which he had also found in the sclerosed area, through the middle third of the crura cerebri, the internal capsule and the corona radiata. He also noted

[1] Kahler and Pick, *Prager Vierteljahrsschrift*, 1879.
[2] Kojewnikoff, *Arch. de Neurologie*, No. 18, 1883.

that the grey matter of the convolutions was of normal appearance. Charcot and Marie[1] showed, in addition to the "granular bodies," a disappearance of the pyramidal cells of the motor cortex. Lumbroso[2] saw these "granular bodies" in the internal capsule and in the cortical motor region.

In opposition to these observations, and of equal importance with them, are the negative results of competent observers, who have carefully looked for these cerebral changes and found them absent. Thus Debove and Gombault[3] searched in vain for the "granular bodies." Marie[4] failed to find any change higher than the pons Varolii. Rovigli and Melotti[5] noted the integrity of the cerebral peduncles and the internal capsule, and most recently Joffroy and Achard[6] confirm the intact condition of the pons and peduncles. In the spinal cord the sclerosis is limited to the tracts which are motor in function, chiefly the crossed pyramidal tracts, but also in some cases the direct. The degeneration is usually most pronounced in the cervical region, and there it has been found not to be sharply limited to the crossed pyramidal tract, but to extend some distance forwards into the antero-lateral area. Degeneration of the anterior ground bundle has been noted by many observers, *e.g.*, Moeli, Leyden, Flechzig, Charcot, and others.[7] Muratoff[8] found that in all cases where the anterior ground bundle was implicated the posterior longitudinal fasciculus was atrophied. This seems to favour Flechzig's view, that these two tracts are functionally analogous. Kronthal[9] noted slight degeneration of some of the fibres of the fillet, accompanied by some increase of the neuroglia. This also was observed by Muratoff.[10] A slight non-essential sclerosis of the posterior columns has been noted in some cases.[11]

[1] Charcot and Marie, *Arch. de Neurolgie*, 1885.
[2] Lumbroso, *Lo Sperimentale*, 1888.
[3] Debove and Gombault, *Arch. de Physiologie*, 1879.
[4] Marie, *Arch. de Neurolgie*, 1887.
[5] Rovigli and Melotti, *Riv. Sperimental*, 1888.
[6] Joffroy and Achard, *Arch. de Med. Experimental*, No. 3, 1890.
[7] Quoted by Muratoff, *Vide infra*.
[8] Muratoff, *Neurol. Centralblatt.*, 1891, p. 513.
[9] Kronthal, *Neurol. Centralblatt.*, 1891, p. 133.
[10] *Loc. supra cit.*
[11] Mentioned by Dr. Ormerod in a critical digest of Amyotrophic Lateral Sclerosis, BRAIN, vol. ix., p. 245, to which we are indebted for much valuable information.

2. *Distribution of the disease in the central grey matter.*—This consists of atrophy and disappearance of the multipolar nerve cells, which are included in Dr. Hughlings Jackson's "lowest level" (the bulbo-spinal centres), with the important exception that the oculo-motor nuclei are usually not degenerated. The greatest amount of atrophy is in the hypoglossal nucleus. The cells of the corpus dentatum of the inferior olive have been found affected by only one observer.[1] The nucleus ambiguus was found atrophied by Kronthal;[2] usually it is unaffected, as also is Clarke's column.

3. *Condition of the peripheral nervous system.*—Degenerative changes have been observed in the peripheral nerves in some cases. Joffroy and Achard[3] noted a parenchymatous degeneration of the nerves of the extremities in a case of this nature. Kronthal (*loc. cit.*) describes degeneration of several cranial nerves, while the peripheral spinal nerves were unaffected. Oppenheim,[4] in a case of chronic anterior poliomyelitis, found slight degenerative change in the mixed nerves, while the anterior roots were healthy. But it must be borne in mind that in many instances no change has been found in the peripheral nervous system.

4. *Nature of the change in the central nervous system.*—The sclerosis is in appearance similar to that of descending degeneration resulting from a cerebral lesion. That it is not due to a diffuse chronic myelitis is evidenced by the fact that the periphery of the cord, which is usually affected by the inflammatory condition, here completely escapes, and also the posterior columns. Whether the change be originally parenchymatous or interstitial is difficult to determine, as in the later stages the resulting sclerosis is the same; and early stages of this disease do not, as a rule, admit of pathological examination. Leyden observed that the axis cylinders were never hypertrophied, as occurs in disseminated sclerosis.

[1] Maier: *Virch. Archiv.*, vol. 61. (Quoted by Ormerod, *loc. cit.*)
[2] *Loc. supra cit.*
[3] *Loc. supra cit.*
[4] Oppenheim: *Arch. fur Psych.* Bd. xix.

Much uncertainty exists regarding the starting point of this disease. In "pseudo-bulbar paralysis" double lesions of a hæmorrhagic nature have been found implicating the pyramidal tracts above the pons, and giving rise to secondary degenerations on both sides. In the chronic progressive form under discussion, we have been unable to find any primary lesion recorded which would account for the more or less symmetrical nature of the sclerosis. Although some observers have noted changes of a degenerative nature in the large cells of the motor cortex, many cases have been recorded where no such alterations have been detected. The view that the disease is a primary affection of cells and fibres, motor in function, seems the only explanation which our present knowledge can justify.

We are indebted to Dr. Hughlings Jackson for permission to record the following case:—

CLINICAL SUMMARY.—*Onset with "loss of voice," shortly succeeded by hemiplegic weakness of right side and later on difficulty in swallowing. Admitted with paralysis of tongue, lips, palate, and vocal cords, weakness and atrophy of right arm and leg, and inability to articulate. Later on paralysis of right side and paresis of left, paralysis of the neck muscles and muscles of deglutition. Finally, paralysis of diaphragm. No affection of eye muscles and sphincters. Duration, about sixteen months.*

G. W. V—n, ætat. 43, was admitted into the National Hospital for the Paralysed and Epileptic, Queen Square, on October 8th, 1888, and owing to his inability to articulate, and difficulty in writing, a very brief history was obtained. In December, 1887, he began to "lose his voice," but shortly after this he noted a gradually increasing stiffness and weakness of the right arm and leg; he does not think that the left side of the body was affected at this time. Some five months later, in April, 1888, he complained of considerable difficulty in swallowing and inability to expectorate effectually. He has suffered no pain during his illness. He denies syphilis. He said he had rheumatic fever nine years ago.

Condition on admission.—The body is emaciated, the

expression is characteristic of the disease. This has been well described as "sad inanimate" while the temperament is buoyant and expectant. The movements of the face are weak on both sides, the orbicularis oris being especially affected, so that he is quite unable to whistle or put his mouth into the position for kissing. The upper facial group, frontalis, and corrugator supercilii is in a state of over-action, so that the forehead is crossed by deep wrinkles; the orbiculares palperbrarum act well and efficiently; the naso-labial fold is prominent on both sides. The lower facial group is weak; the lower lip hangs down and the mouth is slightly open. The tongue lies flaccid on the floor of the mouth and cannot be protruded beyond the dental arch: he is quite unable to articulate, an endeavour ends merely in the production of a noise. He understands everything that is said to him, he answers questions in writing, and he reads and appreciates the daily papers. Mentally he is facile, laughing and sobbing on slight provocation. The vision is good, the fundus of the eye is normal and there is no defect in the movements of the eyes. Hearing also is good. Dr. Semon examined the larynx and reported that there was bilateral and symmetrical paresis of the abductors; the glottis on inspiration did not open to more than three mm., on phonation there remained a small oblong gap between the cords (paresis of the internal tensors). There is paresis of the soft palate. He swallows pulps more readily than fluids or solids, but the taking of food usually starts a troublesome cough: this difficulty in swallowing appears to be due to an inability to get food into the lower pharynx. The vomiting and swallowing reflexes are not obtained on tickling the back of the tongue. The cardiac and respiratory functions are normal.

He walks with a feeble, hemiplegic gait. There is considerable wasting of the muscles of the right arm and hand, especially of the interossei and thenar groups. All movements are effected but feebly, except that he is unable to dorsiflex the right foot. In the right leg there is no marked wasting. The tendo Achillis is rigid. Both knee jerks are exaggerated, the right being more marked than the left.

An ankle clonus is not obtained: the plantar reflexes are active, the left more than the right. Defæcation and micturition are normal. The urine contains neither albumen nor sugar. The arteries are not atheromatous. *Electrical reactions.*—All the muscles react to the faradic current; but there is a quantitatively diminished reaction of the muscles of the right side in comparison with those of the left.

Progress.—Only a summary of the notes taken during the remainder of the patient's life is given here. By December the atrophy and paralysis had considerably advanced, the small muscles of the left hand being more atrophied than on admission. The faradic excitability of the right hand muscles had also correspondingly diminished; he could barely hold a pen in his right hand. In January, 1889, it was noted that the whole of the left side was becoming weaker and that he had great difficulty in chewing owing to paresis of the jaw muscles. At the same time a slight ankle clonus was observed on the left side, there was none as yet on the right. The condition of the hands was as follows:—Atrophy of the dorsal interossei and abductor indicis as well as of the thenar and hypothenar muscles, the position of the fingers is that seen in ulnar paralysis: the muscles all presented increased mechanical irritability. No fibrillary tremors were observed. At Dr. Jackson's suggestion the urine was repeatedly tested for the presence of sugar, but at no time, not even to within a few days of death was any observed. At no time during the course of the disease did the saliva trickle from the angles of the mouth.

In February the movements of the neck muscles were found to be feeble, flexion of the head backwards and forwards was effected for a short distance by muscular action, beyond this the head fell backwards or forwards by its own weight; rotation was, however, well carried out. Movement of the head occasioned considerable pain over the position of the Atlas. During the remainder of this month and the following the patient was unable to move in bed without assistance. Towards the end of March, the following condition was noted:—

Olfactory Nerve.—Sense of smell perfectly normal.

Optic Nerve.– Sight unimpaired ; discs normal.

Oculo-Motor Nerves.—All ocular movements well carried out, but some coarse nystagmoid jerkings were occasionally observed on extreme lateral deviation ; pupils medium and equal, react to light and accommodation ; no ptosis.

Trigeminus (Motor branch).—Paresis, almost amounting to paralysis of masticatory muscles.

(Sensory branch.)—No impairment of sensation. Taste normal.

Portio dura.—Lower facial muscles completely paralysed, the upper group is in a tonic state of overaction.

Portio mollis.—Hearing is unimpaired.

Glossopharyngeus.—Vomiting reflex is in abeyance.

Vagus and accessorius vagi.—Paralysis of the abductors of the vocal cords. Paralysis of articulation. Paresis of the soft palate.

Hypoglossus.—Glossoplegia with wasting.

Spinal accessory.—Paresis, with wasting of sterno-mastoid muscles and upper parts of both trapezii.

Cervical nerves.—Paralysis of the muscles which move the head ; paralysis of the diaphragm ; atrophy of the infra-hyoid muscles.

Brachial plexus.—Paralysis and atrophy of the muscles of the right shoulder, arm, and hand ; rigidity and increased tendon jerks ; paresis and atrophy of the left arm and hand ; paralysis of the right serratus magnus, paresis of the left ; atrophy and paresis of both pectorales. No affection of sensation.

Dorsal nerves.—Paresis of the abdominal and back muscles.

Lumbar plexus.—Paralysis, atrophy, and rigidity of the muscles of the right leg ; paresis of the left leg.

Sphincters.—Unaffected.

April 1st, 1889. *Exitus letalis.*

Autopsy.—Dura mater slightly adherent to calvarium ; considerable opacity of the arachnoid over the hemispheres, but not over the base ; no excess of cerebro-spinal fluid ; no *naked eye* changes in brain or spinal cord.

Lungs, liver, and kidneys were of normal appearance.

Heart: slight, easily torn adhesions between epi- and pericardium.

The muscles were all of good colour, slightly soft, but not friable; they were all more or less atrophied. There was complete absence of subcutaneous fat. The platysma myoides were pale, but well developed on both sides.

MICROSCOPICAL EXAMINATION OF BRAIN, MEDULLA OBLONGATA, AND SPINAL CORD.

Vertical transverse sections of the brain were made at levels, which included part of the motor region, and carefully examined with a view to finding some alteration of the cortical cells, but no trustworthy results were obtained. Allowing for the influence of *post-mortem* decomposition, the appearance did not seem to differ much from the normal, but it was remarked that the cortex generally was very vascular.

The internal capsule is, unfortunately, affected by *post-mortem* softening in places, but those regions of it which have escaped seem to be natural.

The crura cerebri at the level of the third nucleus show signs of decomposition, which make it difficult to say whether, or not, there is any degeneration of fibres in the motor tract.

The third nucleus is not deficient in ganglion cells, though the cells themselves seem to be highly pigmented. The posterior longitudinal bundles are normal. Some sections were made of the upper pons, which showed marked degeneration of the pyramidal tracts, but not so complete as in the next section.

Unfortunately the region intervening between the last section and that at the level of the fifth nerve is missing, so that our next section is taken through the pons at the lowest level of the fifth nucleus. Here sections were made, which include the middle peduncle of the cerebellum, and, in fact, all the white matter of the cerebellum, and also the nucleus dentatus. The white fibres of the central parts of the section show the beaded appearance of *post-mortem*

decomposition, but the peripheral parts are in good histological preservation. Only the lowest part of the motor nucleus of the fifth nerve can be examined in these sections, and in it can be seen only two or three ganglion cells, but several carmine-stained patches indicating degenerated cells. What can be seen of the sensory nucleus is natural. The other nuclei to be seen in this section are the superior olive, the nucleus trapezoides, and the nucleus dentatus of the cerebellum, all of which appear to be natural. The condition of the pyramidal bundles is very striking. To the naked eye, after Weigert-Pal treatment, the pyramidal regions appear very lightly stained, while under the microscope there can be seen no normal large or medium medullated fibres, and much fewer than normal small fibres. It is possible that decomposition may be accountable for a certain amount of the myeline *débris* lying about, but the transversely crossing fibres, which are numerous, show little or no decomposition. Under a high power double stained sections of the pyramidal bundles show a loose granular ground substance, plentifully besprinkled with nuclei, a few rather large droplets of blue-staining myeline matter, and a number of small black dots indicating small medullated nerve fibres or their remains. In some parts the granular ground substance is more condensed, forming a true sclerosis, but this is more noticeable in the cord than in the medulla. We may say, therefore, that the whole of the pyramidal tract is completely degenerated on both sides. There appears to be no affection of the transverse fibres forming the trapezoid body so-called. The fillet appears to be unaffected by disease, but its fibres are much beaded by decomposition, owing to delayed penetration of the hardening reagent. The posterior longitudinal fibres present quite normal appearances.

The next section includes the nuclei of the sixth and seventh nerves, and the intra-medullary course of these nerves, and the eighth. The sixth nucleus, Fig. 1, VI. n., is perfectly natural in appearance. The fibres of the sixth nerve are also natural.

The seventh nucleus, VII. n., shows very few ganglion

cells, and these are abnormally rounded, atrophied or granular. The individual fibres of the seventh root in its course through the medulla seem to be quite natural, that is, none are to be seen in the process of degeneration, but the bundle, as a whole, is decidedly smaller than natural. On the other hand, the compact little bundle of transversely-cut medullated fibres, the ascending loop of the seventh Fig. I., VII. a., which is usually so conspicuous above the posterior longitudinal bundles, is here invisible by Weigert-Pal staining. On carefully examining it with higher power we find that normal nerve fibres are completely absent from it, and that it consists of loose meshwork of neuroglia and

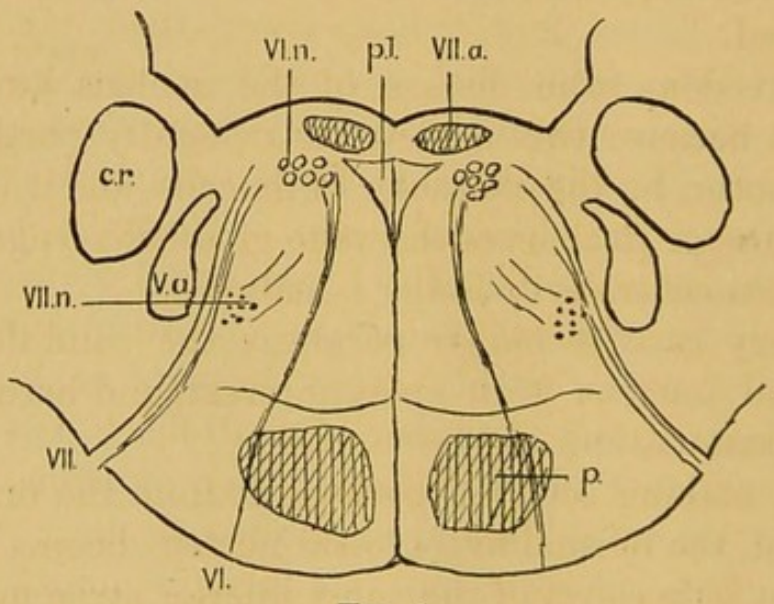

FIG. 1.

Diagram of lower pons, showing sixth and seventh nerves and nuclei, p. l., posterior longitudinal bundles, p. pyramidal tract, VII. n., seventh nucleus degenerated, VII. a., ascending loop of seventh root, VI. n., nucleus of sixth nerve, c.r., restiform body, V.a., ascending root of fifth nerve. In this and the succeeding diagrams, normal white and grey matter is drawn in outline without shading. Degenerated tracts are cross-hatched. Nerve nuclei containing healthy ganglion cells are represented by groups of small circles, those containing degenerated cells by groups of black dots.

nuclei. This taken with the much less atrophied condition of the issuing seventh root is remarkable, and will be considered further on. The degeneration of the pyramidal tracts is as in the preceding section.

At the junction of the pyramids with the pons the seventh nucleus is still visible, and at this level, though much affected, presents more nearly normal appearances than it

did higher up, that is, there are a few normal ganglion cells. The pyramidal tracts are totally degenerated, contrasting with the healthy arciform fibres surrounding them. The eighth nucleus and nerve are natural. The restiform body and the ascending root of the fifth and eighth nerves are here, as elsewhere, normal.

The next cranial nerve to be examined is the ninth. The nucleus seems to be perfectly natural, and the same may be said of the ascending root. At this level also the nucleus ambiguus, a group of cells lying on the inner side of the ascending root of the fifth, presents quite natural appearances, except for the pigmentation of its cells, which is so common an appearance that it can scarcely be considered pathological.

The freedom from disease of the nucleus ambiguus is important because this nucleus is evidently partly, if not entirely motor, by the character of its cells, and it is believed to be the motor nucleus of the vago-glossopharyngeal group. The nucleus centralis of Roller is natural.

In every case of bulbar paralysis the condition of the hypoglossal nucleus is of great interest, and here demands detailed examination.

When making sections backwards from the brain to the spinal cord, the normal hypoglossal nucleus begins to appear at just about the level of the most inferior striæ medullares. It is at first only indicated by a few large multipolar cells scattered among the small round cells which belong, probably, to the eighth and ninth nuclei on the one side, and to the nucleus funiculi teretis on the other. As we proceed spinal-wards, however, the nucleus becomes more and more defined, lying close under the floor of the fourth ventricle, and consisting exclusively of large multipolar ganglion cells. Ventrally to the nucleus pass out the fibres of the hypoglossal nerve and close under the nucleus among the root-fibres lies another diffuse collection of small cells, the small-celled nucleus of the hypoglossal (Roller). In well-stained sections there is a close network of medullated fibres all through the chief nucleus. In each section of the nucleus when at its fullest size may be counted from thirty to

forty ganglion cells, diminishing to about twenty at the level of the calamus scriptorius, and still further back to five or ten, that is, at the level of the lower or spinal extremity of the olive, just below which the nucleus disappears. The small-celled nucleus is most distinct in the cerebral or upper third of the chief nucleus, and becomes less marked as we go backwards till it disappears. It has several large cells scattered about in it.

Fig. 2 shows the relations of the nucleus in the medulla at a level at which it is usually best seen, that is, about its upper third. At the most cerebral end the nucleus contains a considerable number of normal cells. A little further back, however, this is not the case. Here, where there

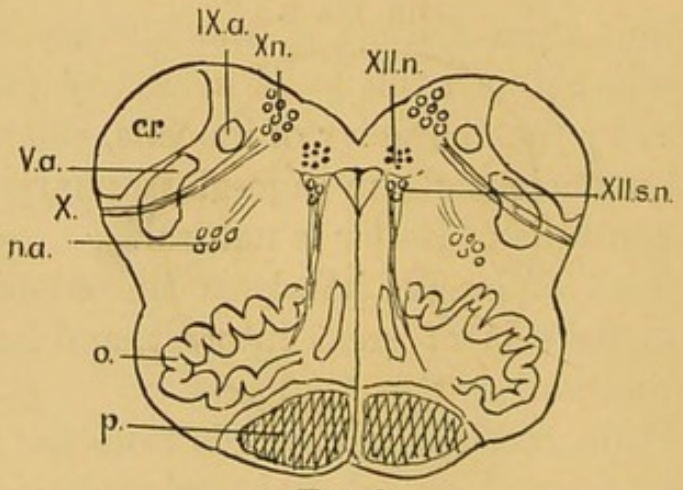

FIG. 2.

Medulla at middle olive, or lower part of floor of fourth ventricle, showing relative position of vagus and hypoglossal nuclei. IX.a. so-called ascending root of glossopharyngeal. X.n. vagus nucleus. XII.n. hypoglossal nucleus degenerated. O. olive. XII.s.n. so-called small-celled nucleus of hypoglossal (Roller) n.a. nucleus ambiguus. Other lettering as in Fig. 1.

should be counted from thirty to forty well-formed, large multipolar ganglion cells, there are to be found in some sections perhaps as many as ten at the most, generally four or five, and in some even none at all.

The ground substance of the normal nucleus should be fairly compact, finely granular neuroglia supporting numberless interlacing white fibres which come out very distinctly by Weigert-Pal staining. In the affected nucleus, however, the supporting tissue consists of a loose meshwork of fine fibrils with many nuclei. There are some white medullated

fibres but not so many as in the normal nucleus. The root bundles of the nerve contain decidedly fewer fibres than natural, in fact the hypoglossal roots are not the conspicuous objects that they are in sections of the normal medulla oblongata.

The small-celled nucleus does not appear to be affected in any way, and among its cells may be found many normal large cells.

The vagus nucleus (X.n.) is normal.

Below the calamus scriptorius, where the central canal is completely surrounded by the central grey tube, the nucleus lying dorsally to the hypoglossal is that of the eleventh nerve or accessorius vagi. This nucleus is quite

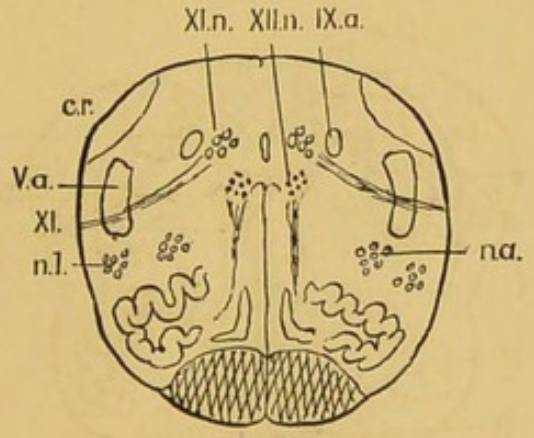

FIG. 3.

Medulla at lower olive below calamus scriptorius showing the relative position of accessorius and hypoglossal nuclei. XI.n. accessorius nucleus. n.l. Nucleus lateralis.

normal in appearance. The nucleus lateralis presents no sign of degeneration.

In the lowest part of the medulla (Fig. 4) just above the decussation, we find a great degeneration of the anterior horns (c.a.) which here first make their appearance. The hypoglossal nucleus is still visible and degenerated. The nucleus funiculi gracilis, and nucleus cuneatus internus (n.c.) and externus (seen in sections higher up than this) are quite natural. The pyramids at this level show considerable numbers of natural nerve fibres both large and small.

In the spinal cord at the first cervical nerve the grey

matter of the anterior horn is almost devoid of ganglion cells, in some of the sections none are to be seen at all, in others at most two or three. The lowest fibres of the anterior pyramids are still to be seen crossing to form the crossed pyramidal tract which is now seen to be distinctly affected. It contains very few large nerve fibres compared with the healthy cord, so that by Weigert-Pal staining it appears as a lightly-stained patch, but by carmine staining it comes out a dark patch owing to a slight condensation of the neuroglia.

At cervical ii., besides disappearance of ganglion cells, there is also marked wasting of the horns, rather more marked on the left side. The lateral horns share in this wasting. There are a few natural bipolar cells in the base

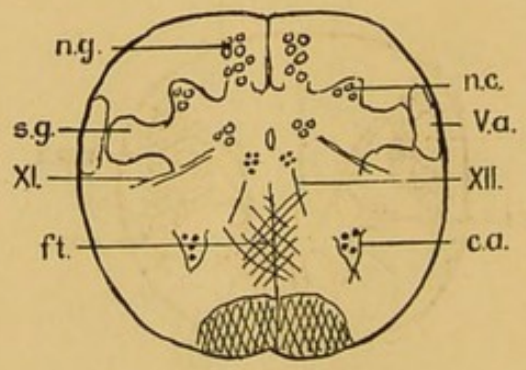

FIG. 4.

Medulla at level of first appearance of anterior horns of cord. c.a. Anterior horns, ganglion cells degenerated or absent. s.g. Substantia gelatinosa. n.c. Nucleus cuneatus internus. n.g. Nucleus funiculi gracilis. f.t. Decussation of fillet, natural.

of the posterior horn, the position taken lower down by the posterior vesicular column. The degeneration of the crossed pyramidal tract reaches to the periphery of the section in the region of the posterior horns. There is well marked affection of the anterior median column and the direct pyramidal tract.

At the level of cervical iv. (Fig. 5), the shrinking of the anterior horn brings into prominence the lateral horn, particularly on the left side. Here are a few cells, but there are the remains of many degenerated cells lying about.

There is still a very small strip of degeneration on each side of the anterior fissure (d.p.t.).

At cervical vii. the attenuation of the anterior horns is very marked, especially on the left side. The left lateral horn stands out in marked prominence, and is seen to contain many natural ganglion cells.

At dorsal ii. we have the first appearance in this series of the posterior vesicular column. This appears to be perfectly natural here and in all the sections below this level. The wasting of the anterior horns is slightly more pronounced on the left side. The crossed pyramidal tract is extensively affected, but it shows a fair sprinkling of normal large white nerve fibres scattered through it.

At dorsal vi. the wasting of the anterior horns is extreme. The degeneration of the crossed pyramidal tract now reaches the edge of this section. It has here the

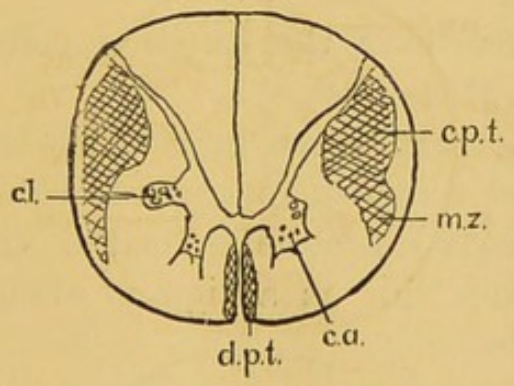

FIG. 5.

Transverse section of spinal cord at level of cervical iv.; c.p.t., crossed pyramidal tract degenerated; d.p.t., direct pyramidal tract degenerated; m.z., descending fibres in the "mixed zone" degenerated; c.l., lateral horn with some normal cells, but many degenerated.

appearance of a sclerosis, but has in it many quite normal large nerve fibres, many more than in the last section. These fibres are, many of them, probably ascending from the posterior vesicular column to gain the direct cerebellar tract.

At dorsal viii. (Fig. 6), the thinning of the anterior horns is, perhaps, at its maximum. In some of the sections a ganglion cell or two may be seen, but in others none at all. The posterior vesicular column is unaffected, contrasting strongly with the wasted anterior horns.

At lumbar i., though the anterior horns are smaller than

usual, they contain a considerable number of healthy ganglion cells. The sclerosis of the crossed pyramidal tract is most pronounced on the left side. The posterior vesicular column is fully developed and perfectly normal.

At lumbar iii. (Fig. 7), the grey matter presents more nearly normal appearances than in any of the preceding

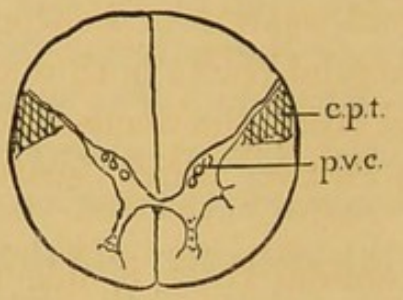

FIG. 6.

Transverse section of cord at level of dorsal viii.; p.v.c., posterior vesicular column.

sections, but even here the ganglion cells are below the normal in point of number.

The anterior root fibres proceeding from the anterior horns in all these sections appeared to be rather thin and to retain the Weigert-Pal staining badly. We hesitate, however, to pronounce this as a sign of atrophy of the fibres,

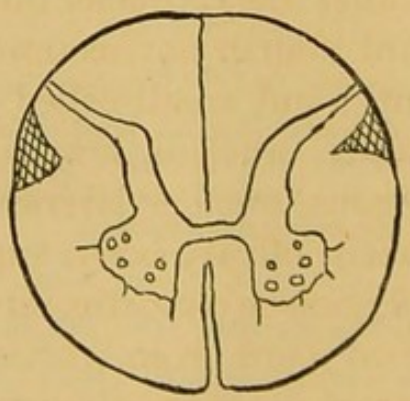

FIG. 7.

Transverse section of cord at level of lumbar iii.

because it is an appearance common in lightly stained sections of normal spinal cord. The transverse sections of the anterior roots themselves, which could be seen in most of the sections, were critically examined with absolutely negative results. It is difficult to believe that any considerable disappearance of white fibres could fail to be discovered in such a large number of sections.

Summary of Microscopical Appearances.

1. Motor cortex apparently normal.

2. White fibres of internal capsules and crura cerebri also normal.

3. Pyramidal tracts of upper and middle pons deeply degenerated, showing complete absence of white medullated fibres, and some condensation of neuroglia base.

4. Crossed pyramidal tracts both degenerated all the way down the cord, but from the decussation downwards there are some healthy white fibres scattered about in the sclerosed area.

The direct pyramidal tract is degenerated as far down as cervical iv.

5. Cranial nerve nuclei affected are :—

(*a*) Motor nucleus of fifth.

(*b*) Nucleus of seventh extensively, issuing roots attenuated only, but ascending loop deeply degenerated, being quite devoid of medullated nerve fibres.

(*c*) Hypoglossal nucleus extensively, roots much thinned.

6. Cranial and other nerve nuclei not affected by disease are those of the third (fourth not examined), sixth, seventh, ninth, tenth, eleventh, and small-celled nucleus of twelfth. Also nucleus dentatus, nucleus trapezoides, nucleus centralis, nucleus ambiguus, nucleus lateralis, nucleus arcuatus, nucleus funiculi teretis, olivary body, superior olive, nucleus funiculi gracilis, and nucleus cuneatus internus, and externus.

7. Grey matter of cord from cervical i. to lumbar i. shows extreme atrophy, with almost complete loss of ganglion cells, in the anterior horns especially, and to a very great extent in the lateral horns also. The issuing anterior root fibres appear rather thin, but the anterior roots outside the cord are normal.

8. The posterior vesicular column is everywhere normal.

The following points appear to us to be worthy of special remark :—

Distribution of disease in motor grey matter.—All the ganglionic cell groups innervating the somatic muscles, that is those nervous centres included in Dr. Hughlings Jackson's "lowest level," are deeply degenerated, with the exception of the oculo-motor nuclei, and those which control the sphincters ("escape of eyes and perinæum").

Distribution of disease in motor white matter.—There is intense degeneration of the pyramidal tracts in the middle pons, and to a lesser degree of the upper, but lower down, that is at the level of the olive, these tracts show considerable numbers of what may be healthy fibres, and also numbers of small medullated fibres. Below the decussation, the area of sclerosis differs in no respect from an ordinary descending secondary degeneration. The extension forwards of the degeneration into the region of the "mixed zone" is not due to diffuse myelitis, but as has been shown by one of us,[1] marks the normal position of certain of the descending motor fibres.

These facts suggest the possibility of the existence of a primary lesion of the pyramidal tracts in the pons with corresponding secondary descending lesions.

Condition of the peripheral nervous system.—As pointed out in the pathological report, though the ganglion cells of the anterior horns of the cord and medulla are deeply degenerated, yet the anterior nerve roots are perfectly healthy. On the other hand, the roots of the hypoglossal and facial nerves are very much thinned by the disappearance of many nerve fibres, particularly in the case of the former, but those which are present are natural in appearance. We can offer no explanation of this apparent anomaly.

Relation of facial nerve to seventh nucleus.—We are enabled by the microscopical examination of this case to produce anatomical evidence of the possible innervation of the orbicularis palpebrarum by the oculo-motor nuclei. As above described, the oculo-motor nuclei (third and sixth, fourth not examined) are quite healthy, while the seventh nucleus, and the ascending loop of the nerve (Fig. 1, vii. a.)

[1] *Tooth.*—Secondary degeneration of the spinal cord, 1889, p. 36. Figured also by Hadden and Sherrington in BRAIN, Vol. VIII. p. 502.

lying dorsally to the posterior longitudinal bundles are deeply degenerated. Now it is probable that this ascending limb includes all the fibres from the nucleus proper. But the nerve root, on the other hand, passing through the medulla to issue from the side as the seventh nerve, though not presenting the size and compactness of the normal root, yet contains many perfectly normal fibres. We must suppose that these have come from some other source than the highly degenerated nucleus, and we believe that they come from the oculo-motor nuclei by way of the posterior longitudinal bundles, and are passing in the trunk of the facial nerve to supply the unaffected muscles of the upper face, or, as we may call them, the oculo-facial group (frontalis, corrugator supercilii and orbicularis palpebrarum).

In support of this view we refer to a suggestive paper by Mendel,[1] who performed experiments after Gudden's method on rabbits and guinea pigs. The upper and lower eyelids were stitched together, and the frontalis extirpated, in newborn animals. After six to ten months they were killed. Careful examination of the abducens and seventh nuclei failed to show any change in them. The facial trunk on the side operated upon appeared thinner than on the other side. Marked changes were found in the hinder parts of the third nucleus, namely, great diminution in the number of the ganglion cells, so as to cause the nucleus, as a whole, to be much smaller on the same than on the opposite side. The cells were atrophied and their protoplasm stained less vigorously than normal. He suggests that the path of connection between the oculo-motor nuclei and the facial nerve is the posterior longitudinal bundle.

There is reason to suppose that another important set of fibres, which reach their destination by way of the facial nerve, namely, those which supply the orbicularis oris, have their nucleus of origin in other than the seventh nucleus, probably the hypoglossal. The clinical facts of all cases of bulbar paralysis lend support to this view; they all show

[1] *Mendel.—Ueber den Kernursprung des Augen-Facialis; Neurolog. Centralbl.*, 1887, p. 537. Mendel refers to a discussion on the subject in the *Berlin. Klin. Wochenschr.*, 1887, No. 48.

the very close relations which exist between the lips and the tongue. But in order to demonstrate the separation between the motor innervation of the lips from the other facial muscles, we refer to a case, mentioned by Dr. Gowers,[1] of poliomyelitis affecting the seventh nucleus only, where the orbicularis oris escaped. Our case does not add any anatomical evidence in solution of the problem as to how the fibres which supply the lips reach the facial nerve trunk from their nucleus of origin. The path is possibly the lower part of the posterior longitudinal bundles, but our case throws no light on this point.

Relation of eleventh nerve (accessorius vagi) to hypoglossal nucleus.—This nerve is usually described as consisting of two parts, that accessory to the vagus, accessorius vagi, or eleventh nerve, and a spinal division, or accessorius spinalis. About the latter we have nothing to say, its origin, distribution, and function being fairly clear. But as to the former we find very little information, even in the latest text books. According to Lockhart Clarke, and Schwalbe, the accessorius vagi nucleus is the lowest of the three nuclei contained in one cylinder of grey matter, which nuclei are from above downwards the glossopharyngeal or ninth, the vagus or tenth, and the accessorius vagi or eleventh. As it is impossible to differentiate the lower fibres of the vagus nerve roots from those of the accessorius vagi, as they issue from the medulla between the restiform body and the olive, except inferentially by their position, so no hard and fast line can be drawn between the nuclei of origin of these nerves. But for the purpose of description it may be convenient to state that so long as the central canal is surrounded by grey matter, the ganglionic cell group lying *dorsally* to the hypoglossal nucleus, may be regarded as that of the accessorius vagi. Where, however, the canal has opened out into the fourth ventricle, the cell group lying *externally* to the hypoglossal nucleus, and thus forming part of the floor of the ventricle, is that of the vagus. This, so far as we can make out from authors, is the present position of anatomical knowledge on the subject.

[1] Gowers' "Diseases of the Nervous System," 1888, vol. ii., p. 222.

In the case of V——n, we can say that the common nucleus of the ninth, tenth, and eleventh nerves is quite healthy. (This appears to be the condition in all recorded cases in which the state of the nucleus is specially noted, with one exception.[1]) This is at variance with the fact that he had paresis of the soft palate, and paralysis of the abductors, and adductors of the vocal cords, which muscles are regarded as being supplied through the vagus by its accessory branch. That the eleventh nerve is the path by which motor fibres reach the palate and vocal cords was long ago demonstrated clinically by Dr. Hughlings Jackson, and has been recently corroborated by experiments of stimulation and section of that nerve within the skull.[2] As we have just said, the eleventh nucleus was in our case healthy, we must therefore look elsewhere for the nucleus of origin of its motor fibres. The nucleus ambiguus from its position and connection would, at first sight, appear likely to be the motor nucleus in question, and indeed has been called by some writers (Obersteiner) the motor vago-glossopharyngeal nucleus. In the present case its cells, however, are perfectly healthy. By a process of exclusion, then, we are driven to localise the nucleus of origin of the motor fibres of the eleventh nerve in the sphere of the hypoglossal nucleus.

Anatomical evidence is hitherto wanting, but the clinical facts of nearly all cases of bulbar paralysis point to this conclusion.

Conclusions summarised.—1. Our conclusions with regard to the innervation of the facial muscles may be briefly summarised as follows, and are diagrammatically represented by Fig. 8.

The facial muscles may be divided into three divisions. First, the oculo-facial group, frontalis, orbicularis palpebrarum, and corrugator supercilii. Second, the middle group, elevators and depressors of the angle of the mouth, zygomatics, risorius, and buccinators. Third, the oro-facial

[1] Kronthal, *op. cit.*
[2] Beevor and Horsley, *Proc. Roy. Soc.*, vol. xliv., 1888.

group, or orbicularis oris. All these muscles are innervated by fibres included in the facial trunk, and are all paralysed, when the *nerve* is affected, as is shown by any ordinary case of Bell's paralysis. When, however, the facial *nucleus* only is diseased there results paralysis of the middle group only.[1] The upper group is paralysed when the oculo-motor nucleus is affected, the course of the fibres being probably the posterior longitudinal bundle.[2]

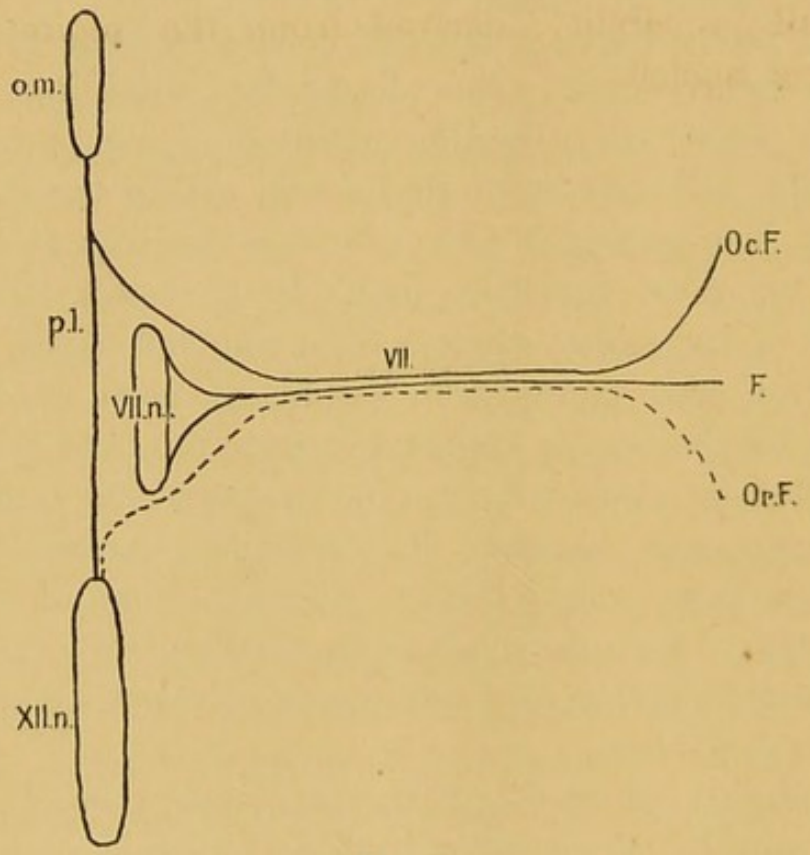

FIG. 8.

Diagram showing the origin and distribution of the fibres contained in the facial nerve trunk; o.m., oculo-motor nucleus including for diagrammatic purposes, the third, fourth, and sixth nuclei; p.l., posterior longitudinal bundle conveying fibres from the oculo-motor nucleus to Oc.F., the oculo-facial group; VII.n., the nucleus of the seventh nerve supplying fibres to F., the middle facial and lower muscles of expression; XII.n., the hypoglossal nucleus supplying fibres to Or.F., the oro-facial, or lip muscles represented by a broken line.

Paralysis of the orbicularis oris is associated with that of the hypoglossal group of muscles, and is therefore presumably innervated by that nucleus. The course of the fibres is at

[1] Gowers, *Loc. cit.*

[2] We have reason to suspect that in total nuclear ophthalmoplegia this does occur. See also a case mentioned by Uhthoff, *Berl. Klin. Wochenschr.*, 1887, p. 913.

present obscure, but is possibly again the posterior longitudinal bundle.

2. The eleventh nerve or accessorius vagi is known to contain motor fibres for the palate and vocal cords. The apparent nucleus of this nerve is indistinguishable from that of the vagus with which it is continuous. Its nerve roots may be regarded as the lowest fibres of the vagus, and its nucleus as the lowest part of the vagus nucleus. Its motor fibres which innervate the palate and larynx are, in all probability, derived from the region of the hypoglossal nucleus.