A case of bilateral paralysis of facial and auditory nerves, with left hemiplegia, in the secondary stage of syphilis / by James Carslaw, M.A., M.B., C.M., Assistant to the Professor of Medicine and Clinical Medicine, Glasgow University.

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A CASE OF BILATERAL PARALYSIS OF FACIAL AND AUDITORY NERVES, WITH LEFT HEMIPLEGIA, IN THE SECONDARY STAGE OF SYPHILIS, BY JAMES CARSLAW, M.A., M.B., C.M.

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A CASE OF BILATERAL PARALYSIS OF FACIAL AND AUDITORY NERVES,

WITH LEFT HEMIPLEGIA, IN THE SECONDARY STAGE OF SYPHILIS.

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Double facial paralysis is an uncommon condition, but its occurrence in the secondary stage of syphilis along with other nervous lesions is perhaps sufficient justification for a somewhat complete record of this case, especially as opportunities were afforded of investigating the pathological changes.

J. C——,[1] sailor, aged thirty-two, came first under observation at the Western Infirmary Dispensary on April 12th, 1894, and was admitted to Professor Gairdner's ward on the 17th. His main complaint was of the condition of his face, but he mentioned also a certain feeling of weakness in his legs. Apart from occasional attacks of rheumatic pains, never so severe as to cause him to take to bed, he had enjoyed good health up till June, 1893, when he suffered for a short time from a venereal sore. He was well again till November, when he had to go to hospital in New York on account of an eruption over his body, which soon disappeared under mercurial treatment. At the end of December he set sail from New York, and his ship on reaching another port in one of the Northern States was frozen in, necessitating a stay there till the 4th of March, 1894. During that stay several of his shipmates were frost-bitten, and he himself had a swelling of

[1] The clinical report is taken from notes by the late Dr. John H. Carslaw.

his neck. A few days after leaving port (and about nine months after primary syphilis) he was on duty at the wheel, a sharp wind blowing from his right, when he became conscious of a tingling of the right ear, and a feeling of numbness and of something unnatural about the right side of his face. On putting up his hand he found that the eye did not close on his making an effort to shut it. Within the next day or two he was conscious that a similar condition gradually developed on the left side, but it was only after both eyes were distinctly affected that he noticed anything unusual about the mouth, speech and the taking of food becoming difficult. After the numbness passed off, pain was felt in both parotid regions and over both cheeks. The weakness of the limbs was of later and of more gradual development. So far as he could tell, there had been no staggering, and he had no definite sense of giddiness. There was no headache or sickness. As regards his habits, it may be mentioned that he had for many years been in the habit of drinking to excess when ashore. But in the early part of the year his captain had allowed no alcohol to be served on board, and when they were frozen up he had not gone ashore for liquor, nor indulged in it at all. His father and mother both lived to old age, and neither they nor any other member of his family had been rheumatic.

The double facial paralysis was very complete, and at once attracted attention. There was entire absence of expression in the features. The effort to raise the eyebrows was without avail, and on his attempting to close the eyes the lids remained open and the sclerotics were exposed to view, the corneae disappearing as the eyeballs rolled upwards (Fig. 1). The lips could not by any effort be firmly closed, and whistling was impossible. He had considerable difficulty in feeding himself, and his speech, which was very defective, was found to err through his inability to pronounce the labials. The movements of the eyeballs, of the palate, and of the tongue were all normal. The pupils were equal, and reacted normally to light and on accommodation. Sensation was normal on both sides of the face, and the electrical examination of the facial nerves and muscles gave the reaction of degeneration on both sides. There was thus very marked loss of faradic irritability, and

while galvanic irritability of the nerves was lost, that of the muscles was distinctly increased. However, there was no serial change, *KCC* always preceding *ACC* and *AOC*; but, while testing, the contractions of the muscles were noticed to be exceedingly slow and long drawn out.

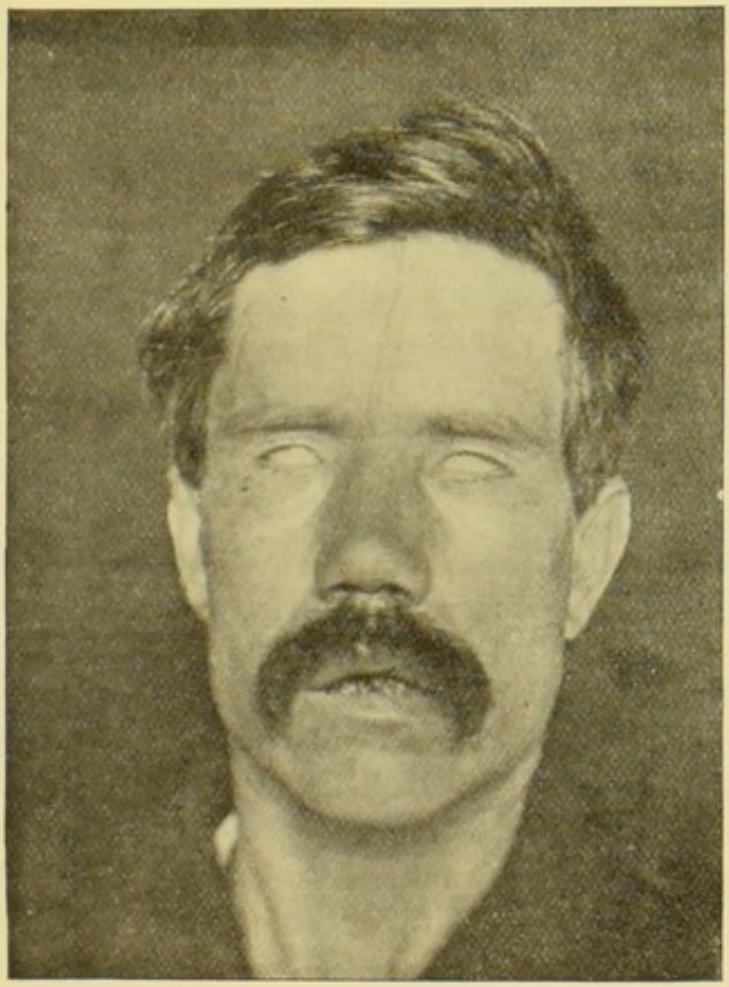

FIG. 1.—BILATERAL FACIAL PARALYSIS (attempting to close the eyes). Photograph of J. C. by Dr. A. W. Russell.—April 19th, 1894.

He was not conscious of being deaf or of any unnatural noises in the ears, and never had had any discharge from his ears. But the result of testing his hearing with the watch was $R=\frac{P}{40'}$, $L=\frac{2}{40'}$, while with the tuning-fork air conduction was in excess of bone conduction, from which Dr. Barr, who examined the patient, inferred that the source of the deafness was in the nervous apparatus. One or two attempts were

made to test the sense of taste, but without very satisfactory results. It may, however, be said that the sense of taste in the anterior part of the tongue was certainly not lost, though probably impaired, and no difference was detected between the two sides. The sense of smell was impaired, but probably from inability to sniff depending on the facial palsy. Examination of the arms and legs gave normal results as to touch, pain, temperature, and electricity. The dynamometer recorded 30 with the right hand and 28 with the left, and the power of the legs seemed fairly natural in spite of the patient's feeling of weakness. The deep reflexes were if anything slightly exaggerated, but there was no tendency to stiffness or spasm. The gait was certainly not very steady, and on the ward floor he required three planks to get along comfortably, but he himself said that he was not any more unsteady than he usually was when ashore. He could turn suddenly in walking without any staggering, and could stand quite steadily with his eyes shut and his heels together.

Physical examination of the chest and abdomen revealed nothing of any importance, and the urine contained neither sugar nor albumen. It should be added that on admission the patient had some slight superficial ulceration of the right tonsil, such as is met with in the secondary stage of syphilis.

The patient improved, both in general health and in regard to some of the special actions of the facial muscles, under antisyphilitic remedies and galvanism. But on May 26th, 1894 (*i.e.* within a year of the primary syphilis), he was found to have left hemiplegia, the left arm being quite powerless, and the left leg less deeply affected, and without sensory disturbance. An interesting point in regard to this hemiplegia was that at first the plantar and knee reflexes on the affected side were distinctly defective, whereas, nine days later, these reflexes were relatively excessive. Again, on June 4th, respiratory difficulty was noticed, with rise of temperature and pulse rate and physicalsigns of pneumonia, a pneumonia which post-mortem was found to be lobular and gangrenous, and probably caused by the passage of food or other foreign substance into the respiratory passages. In this connection a note in the Ward Journal a few days after the onset of the hemiplegia is

of interest, to the effect that "the task of feeding the patient is found to be a very difficult one." The patient died after eight days' acute illness.

Fortunately a post-mortem examination was permitted, and the most important facts made out are here summarized from a report by Professor Coats: Softening of right corpus striatum; lungs in the condition indicating lobular pneumonia with gangrene; heart normal; kidneys and other abdominal viscera normal. The left petrous bone was split open through the tympanum by Dr. Barr, and the course of the Fallopian

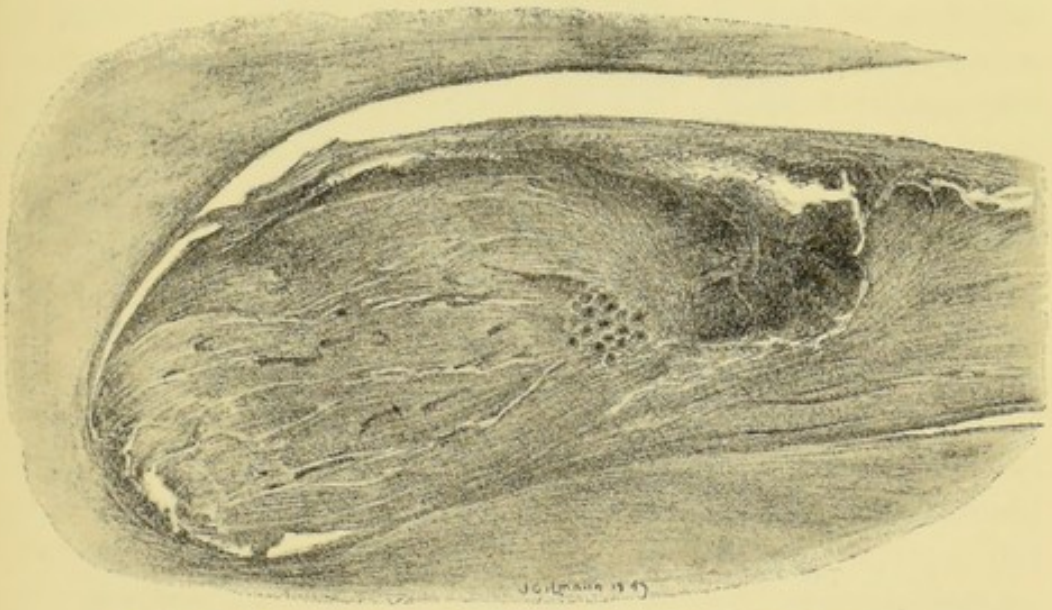

FIG. 2.—FACIAL NERVE IN THE AQUEDUCT OF FALLOPIUS AT THE GENU. Longitudinal section showing a large haemorrhage in the nerve, and a smaller one proximally; also engorged bloodvessels. Part of the geniculate ganglion is shown. (Low Power.)

canal traced out. Dr. Barr reported that "there was no evidence of middle ear disease of any kind, nor of any bony thickening, there being no encroachment on the capacity of the canal. The facial nerve in its course was somewhat thickened, and had a reddish appearance." The right petrous bone was decalcified, and its careful microscopical examination resulted in the discovery of important pathological conditions.[1] There were numerous small haemorrhages, mainly in and under the

[1] I am indebted to Dr. R. M. Buchanan for assistance in the microscopical examination.

sheaths of both the facial and the auditory nerves, compressing the nerves, but also in the substance of the nerves. The haemorrhages were present in the facial nerve in the upper part of its course through the petrous bone, the most distinct one being at the genu (Fig. 2). Besides the haemorrhages, seen in the course of the auditory nerve (Fig. 3), there were also similar small haemorrhages in the membranous cochlea. There was, in addition, apparently degeneration of

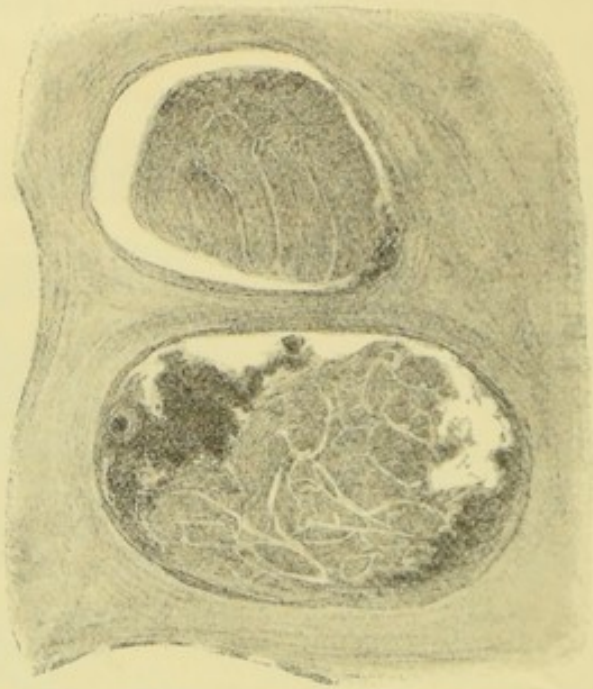

FIG. 3.—THE FACIAL AND AUDITORY NERVES at the outer end of the internal auditory meatus. The facial is slightly oblique, and shows a small haemorrhage. The auditory is transverse, and shows two distinct haemorrhages. (Low Power.)

the facial nerve. Unfortunately, however, the decalcifying process made it impossible to get the nerves stained satisfactorily by Weigert's method, so that the nerve degeneration was not well demonstrated. But specimens from the facial nerve, not far from its exit, showed distinct excess of connective tissue and diminution of nerve fibres. There was marked engorgement of the small bloodvessels of the nerves, and some of them had apparent thickening of the middle coat, and contents so dense and dark-coloured as to suggest thrombosis, but no cellular infiltration was detected. The examination of the brain in the softened area revealed certain localized proliferations of cells in the external coats of the bloodvessels, and also haemorrhages into the perivascular spaces. The vessels of the

pia mater also, examined in this neighbourhood, showed similar conditions of proliferation and haemorrhage, and at several points the cellular proliferation was also seen to extend a short distance into the brain substance. Sections were made of the medulla and pons for the purpose of examining the motor tracts, but these showed no appreciable descending degeneration by Weigert's method. In this connection, however, it should be noted that death occurred only seventeen days after the onset of hemiplegia.

During the patient's illness there was considerable difficulty in determining whether the double facial paralysis was to be regarded as of "rheumatic" (so-called) origin or of syphilitic origin. The patient had a history of previous rheumatic pains, and the onset of paralysis was sudden and distinctly connected in his own mind with exposure to a specially cold wind. Moreover, though simple rheumatic facial paralysis is usually unilateral, genuine cases of bilateral paralysis of the portio dura of this nature have been recorded.[1] However the co-existence of the bilateral nervous deafness, and the subsequent development of hemiplegia from still another nervous lesion indicated the probability of all the nervous symptoms being really syphilitic. The microscopical examination of the tissues has amply confirmed this view, for it demonstrated similar lesions in both the facial and auditory nerves and also in the brain, lesions depending on pathological changes in the smaller bloodvessels and probably characteristic of the secondary stage of syphilis.

It used to be taught that all the syphilitic affections of the nervous system came late in the disease, and were distinctly of the tertiary stage; and no doubt the tertiary period is the common one for such manifestations. Then the lesions are most probably of the gummatous nature, and the facial nerves might be implicated in such lesions, but almost certainly along with some other cranial nerves, and very unlikely both facials alone—still more unlikely only the facial and auditory nerves of both sides. We now know, however, that it is not at all

[1] Among others see an interesting case reported by Prof. Gairdner, *Lancet*, 1861, I. 477, *et seq.*; also a case by Dr. M'Donnell in *Medico-Chirurg. Transactions*, 1875, Vol. LVIII., 369-375.

infrequent for patients to suffer from affections of the cerebro-spinal system in the comparatively early stages of syphilis. At this time various functional disturbances are not uncommon, such as different forms of neuralgia, and sensory disturbances, especially analgesia, to which attention has been specially drawn by Fournier. There are also abundant clinical records of paralysis both central and peripheral occurring early. The tendency has been to regard these as early or precocious tertiary phenomena, but they probably belong as definitely to the secondary period as the skin eruptions or the affections of the throat, eyes, etc., and may be due to lesions similar to those demonstrated in the present case. Dr. Buzzard, in his *Clinical Aspects of Syphilitic Nervous Affections*,[1] discusses the question, quoting the experience of Lancereaux and other continental authorities. Two years ago (Feb. 26 and March 12, 1895), in the London Royal Medical and Chirurgical Society, an interesting discussion took place on this subject: "The affections of the nervous system occurring in the early stages of syphilis,"[2] in which many of the leading British neurologists took part. The general outcome of that discussion was the recognition of the fact that nervous complications of syphilis do occur in the truly secondary stage, and that these are probably dependent on changes in the smaller bloodvessels, not in the nervous elements themselves. Such early affections are as a rule not of serious prognosis, though on account of the destructive processes recovery may be only partial, and opportunity seldom occurs to make pathological examination. Some such examinations have however been reported with apparent absence of anatomical lesions,[3] but at the above discussion Mr. Jonathan Hutchinson related a fatal case of early syphilitic paraplegia, published by M. Lamy, of Paris,[4] in which microscopic changes were found in the spinal cord quite analogous to the changes found in the facial and auditory nerves, and the corpus striatum of the present case. "The stress of the disease had fallen on the perivascular spaces of the vessels, especially the veins, and had respected

[1] Buzzard, *Clinical Aspects of Syphilitic Nervous Affections*, pp. 43 *et seq.*
[2] See *British Medical Journal*, 1895, I., p. 476, etc.
[3] Heubner in Von Ziemssen's *Cyclopaedia*, Vol. XII., pp. 316, 344, 354.
[4] *B. M. J.*, 1895, I., p. 485.

the nervous elements. In the grey matter the perivascular spaces contained colloid matter; in places the veins were thrombosed, and showed microscopic gummata in connection with them. There was some degeneration running in from the pia mater, but most of the nerve elements were normal." Of similar cases of paraplegia, Mr. Jonathan Hutchinson has recorded[1] no less than twenty-one examples, occurring within the first two years after primary syphilis, and regarded by him as secondary. In other cases hemiplegia is the form the disease takes, as in our own case (within one year), and in cases quoted by Dr. Buzzard.[2] Occasionally there is the occurrence of paralysis of single nerves, it may be symmetrically or not. Of such local paralyses, Fournier[3] writes that "Much the most common is facial hemiplegia. Next in order of frequency comes paralysis of the third pair of nerves, which may be either complete or partial. That of the sixth pair is rarer. We may add that of these paralyses the earliest in appearance without the least doubt is facial hemiplegia. It has been observed repeatedly during the fourth, fifth, and sixth months after infection, and one has even seen it (but this is exceptional) along with the first secondary phenomena accompanying the roseola. The ocular paralyses are distinctly later in development." From similar causes, probably, nervous deafness may be rapidly produced at an early stage of acquired syphilis, though this form of deafness is more frequent in the so-called secondary stage of the inherited disease, accompanying interstitial keratitis.

Of these various nervous complications of secondary syphilis, not the least remarkable is the occasional and simultaneous affection of both the divisions of the seventh cranial nerve. These two nerves come into close relations in the temporal bone, and are exposed to similar conditions in passing through dense bony canals, and on this account pathological changes should be rapidly manifested by clinical symptoms. Cases of this kind may be symmetrical like the present, and their early occurrence would explain the symmetry, but that they may

[1] *Archives of Surgery*, Vol. VII., p. 233, etc. (July, 1896).

[2] *Syphilitic Nervous Affections* (Buzzard), p. 44.

[3] Fournier's *Leçons Cliniques sur La Syphilis étudiée plus particulièrement chez la Femme.* Edition, 1881, p. 611, with foot-note.

not be so is illustrated by a case which has recently occurred in the private practice of Dr. J. P. Boyd, of which he has given me the following details. This patient, a young man, contracted syphilis, which was followed by the ordinary secondary affections of the skin and throat. About five or six months after infection he suddenly developed unilateral facial paralysis and profound nervous deafness on the same side. Four months later he had a well-marked syphilitic orchitis, but he has had no new nervous symptoms. Under treatment the facial paralysis has considerably improved, but the nervous deafness remains much the same.

Cases of bilateral affection of both facial and auditory nerves in syphilis are rarely recorded, and in his interesting paper in *Guy's Hospital Reports*[1] on a case of this kind, Dr. Pye Smith has enumerated six examples. To this list, in addition to the present case, there may be added another which has a striking resemblance to it, and of which I have Dr. James Finlayson's kind permission to narrate some details. The patient, J. M., a discharged soldier, now (June, 1897), *aet.* 25, was under Dr. Finlayson's care in the Western Infirmary, Glasgow, in the spring of 1895. His previous health had been good, except for the fact that for some years he had had some discharge from his right ear, though he had not thought he was dull of hearing on that side. In March, 1894, he contracted syphilis, and was in the military hospital under mercurial treatment for five months, during which he developed a bubo, sore throat, skin eruption, and alopecia. At the end of August, 1894 (*i.e.* within six months), he suddenly developed bi-facial paralysis and bilateral deafness, with marked tinnitus and also vertigo. The facial paralysis was well marked though not complete, and R.D. was present on both sides. Though there was evidence of middle ear disease on the right side, the deafness appeared to be essentially due to some nervous lesion, and was very marked. Taste was doubtfully affected. There were no other symptoms of nervous disorder. I have recently seen this patient, and during the last two years he has been quite well except for the condition of his face and his deafness. The facial paralysis

[1] *Guy's Hospital Reports*, 1895. Vol. LI. pp. 234, 235.

is now less marked than formerly, though still quite a distinct feature. The hearing is not any better, and indeed is so bad that he cannot hear the watch on pressure with either ear, and cannot make out conversation, even in loud tones. There is still tinnitus and some vertigo. There have been no fresh complications, nervous or otherwise, and the patient has been able for work.