

CASE OF SYMMETRICAL SYPHILITIC DISEASE OF THE THIRD NERVES, WITH ARTERIAL AND OTHER LESIONS.

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[HEADACHE; attack of somnolence with delirium, lasting three days. Nine months later, left hemiplegia, drooping of both eyeballs and of left eyelid; extension of weakness to right leg; drowsy condition. Four months later, increase of coma; death with febrile symptoms.]

Frederick W., æt. 50, commercial traveller, was brought by his wife to Queen's Square Hospital on August 31st, 1881. His condition was then as follows: He can just walk when supported, but appears to have little control over his legs. The grasp of the hands is feeble, equal on both sides. The tendon reflexes at the knees are exaggerated, equal on both sides; at the carpal end of the radii present, equal on both sides. There is slight nystagmus. Neither eyeball moves freely in the upward direction; on the left side, movement in that direction seems quite lost, and that eye is constantly directed downwards. There is said to be failure of vision in the left eye. The right fundus oculi is normal; of the left I cannot obtain a satisfactory view. He is in a drowsy semi-comatose condition; puts out his tongue; moves any of his limbs. There is no actual paralysis, except that mentioned of the ocular movements.

The history was obtained from his wife, who appeared intelligent and trustworthy. His illness began in November, 1880 (nine months ago); before that he had had good health, with the exception of much headache. He woke one morning looking very ill, rambled "about trains and coaches," and for three days slept constantly, waking only to take his meals. He had no fits or paralysis, but after the disappearance of the somnolence his memory and mental faculties did not perfectly return for three months.

After that he remained fairly well, but seemed "weak," and did not go back to work. His employers dismissed him in July, 1881, and this was a great shock to him. A week afterwards, he returned home from a walk, dragging the left leg, and weak in the left arm; the left eyelid drooped, and the eyeballs were directed downwards, as now. The right leg became weak in a few days, though less so than the left. There was no affection of sensation. He seemed always drowsy and sleepy. His wife further stated that he had always been a healthy man; she knew of no nervous disorders, and of no consumption in his family. She had been married to him twenty-two years, had five healthy children, and had never miscarried. His occupation had entailed much mental work and anxiety.

The hospital being closed for repairs, he could not be then taken in. Mercury and iodide of potassium (the latter in increasing doses) were administered. No change of importance was reported till

December 28th.—He has been weaker and more drowsy during the last fortnight, and during the last two days much worse; always sleeping, perfectly helpless, unable to speak or understand when spoken to. As seen now in the outpatient-room he is in a semi-comatose condition, but can be roused sufficiently to enable him to put out his tongue or open his eyes. He cannot stand or walk; the eyes are still directed downwards; the tongue is protruded slightly to the right side; the patellar tendon reflex is absent on the right side, and obtained once only on the left; the plantar (cutaneous) reflex present on both sides.

He was admitted, and after being taken to the ward became cold and comatose; temperature (rectal) $95\cdot6^{\circ}$; pulse feeble; respirations shallow; deglutition almost impossible; pupils small, inactive to light, but conjunctival reflex normal.

Brandy was administered freely.

December 29th.—Temperature normal. Has spoken and taken food, but easily goes off to sleep.

Evening temperature, $102\cdot6^{\circ}$.

December 30th.—Much worse; breathing rapidly; quite unconscious; apparently moribund. Has vomited frequently in the night.

Temperature (10 A.M.), $102\cdot2^{\circ}$; (2.30 P.M.), $103\cdot6^{\circ}$.

Died at 3.30 P.M.

I am indebted to Dr. Beevor for the notes taken after admission, including the details of the history, and also for making the post-mortem, the account of which follows.

[Post-mortem nineteen hours after death; small tumours on cervical dura mater; thickening of the other membranes,

and softening of the cord in that region ; disease of the basilar artery ; symmetrical enlargement of the third nerves.]

Calvaria and dura mater of skull normal ; arachnoid and pia show slight thickening and opacity ; and in the sulci about the ascending parietal convolutions, a few opaque bodies the size of a pin's head.

(Subsequent microscopical examination showed that there was here no tubercle, that the vessels of the pia mater were quite normal, and that the cortical substance of the brain beneath was natural.)

Cerebrum and Cerebellum, both surface and interior, normal.

Basilar Artery decidedly thickened, the thickening being not uniform, but taking the form of nodules, projecting from the sides of the artery. No thrombosis found anywhere.

The third Nerves of both sides present a fusiform enlargement, soft, reddish-gray in colour, commencing a few lines beyond the superficial origin of the nerves, and extending nearly $\frac{3}{4}$ inch along them.

Pons and Medulla.—On the anterior surface of the pons, in its left half, are two small reddish-gray elevations, extending a little way into the substance of the pons, each about the size of half a pea : the one situated just to the left of the middle line, near the origin of the sixth nerves, the other about the level of origin of the left fifth nerve, half-way between it and the middle line. All the nerve roots, except the third, appear normal. Posteriorly the membranes covering the back of the medulla, from the point of the calamus downwards, are thickened, granular, and adherent.

Spinal Cord.—On opening the bony canal, nothing abnormal ; on opening the dura mater its inner surface is seen to be studded with warty excrescences, flattened, reddish-gray, and soft-looking, size of half a pea and smaller ; there is no effusion, and the membrane, except in the immediate neighbourhood of the tumours, is smooth ; the tumours do not extend below the cervical region, and are limited to the posterior aspect of the cord. The arachnoid and pia mater seem thickened, and cut harshly. The cord itself is swollen and soft. In the dorsal region the arachnoid and pia still seem hard ; in the lumbar region normal.

Right Lung.—Lower lobe congested ; upper lobe adherent to the chest wall, containing near its apex an old quiescent cavity, about the diameter of a threepenny piece, filled with caveous putty-like material.

All the other viscera quite normal.

I have made a microscopical examination of the diseased tissues, with the following results—

Third Nerves.—Sections through the thickest part of the

swelling consist of small cells or nuclei, aggregated together without definite stroma, staining deeply. Examined in glycerine under a high power, they are seen to be irregularly rounded, granular, without definite nucleus, having the size and appearance of lymph corpuscles. The tissue is permeated by numerous pervious blood-vessels, whose walls seem to be formed simply by an aggregation of the cells. No part of the section is free from disease, though in some places the cells are less thickly distributed, and here a very few nerve fibres are visible. Sections made where the nerve is regaining its normal size, show much less of the cell infiltration, and further, that it affects principally the periphery of the nerve. Thus the periphery, consisting principally of cells, takes up logwood or carmine: the central part, where a large proportion of healthy nerve fibres are seen, takes up osmic acid.

Basilar Artery.—Transverse sections, not passing through the nodules mentioned above, show that the adventitia is normal for the most part, though in some places infiltrated by the small round cells: the muscular coat is quite normal; the elastic layer of the intima (fenestrated coat of Henle) is well marked and normal. Between this and the endothelium intervenes a layer of new formation, attaining perhaps twice the thickness of the external coats, and consisting of a fibrillated ground substance, interspersed with numerous nuclei; many of these nuclei are spindle-shaped, others rounded (possibly spindle-shaped, but seen in transverse section): they stain less deeply, and are more irregular in shape and distribution than the nuclei of the muscular tissue in the media. In the most internal part of this layer rounded and more deeply staining nuclei become more abundant, till it merges into the endothelium. The lumen of the artery remains of a very fair size.

Sections involving a nodule have a different appearance. The nodule projects from the outer part of the adventitia with which it blends, and in which it sends out prolongations extending round the vessel, so that the disposition of the disease is in the shape of a signet ring. The structure is exactly that of the diseased part of the third nerve. The media and intima are normal.

The first of these descriptions tallies with the account given by Heubner¹ of syphilitic arterial disease (viz. a spindle-celled growth in the tunica intima between the fenestrated membrane of Henle and the endothelial layer; originating in his view from the endothelium). The second implies a different lesion, viz. an inflammation or growth limited to the adventitia.

Cervical Dura Mater.—The warty excrescences have just the

¹ 'Die Luetische Erkrankungen der Hirn-Arterien.' Ch. iii. and fig. 1 of the illustrations.

same structure as that described in connection with the third nerve, except that the blood-channels are less abundant, and a fibrillar stroma is visible in some parts, especially at the base of the tumour, where it blends with the normal fibrous structure of the dura mater. The tumour sends out spurs along the inner surface of the membrane, which correspond with the roughenings noted post-mortem.

Cord and Pia Mater.—Throughout the cervical region, there is slight infiltration of the pia mater in its whole circumference. The periphery of the cord, in its posterior parts especially, is unduly vascular. In the upper cervical region patches of infiltration extend from the pia mater into the cord in the region of the posterior median fissure and of one posterior nerve root. Again, opposite the sixth cervical nerve there is a patch the size of a pea, situated at the mouth of the posterior median fissure, and sending prolongations into it along the coats of a vessel. The central parts of the cord easily fall away from the sections.

In the upper dorsal region, the cord between the posterior nerve roots was unduly vascular; in the lower dorsal region, the white matter round the central gray portion was opaque, structureless and ill-stained; and there was a patch of infiltration extending from the pia up to one posterior cornu. In the lumbar region again there is a similar, but even larger patch, though the rest of the cord is here healthy.

To summarise: the lesions in the spinal cord were limited to, or originated from, the pia mater, and affected principally, and in the lower parts of the cord entirely, its posterior aspect; lastly, there was no systematic degeneration.

Pons and Medulla.—Of the two superficial tumours, that near the sixth nerve had the structure already detailed. I could not decide whether it interfered with the sixth nerves within the medulla, but externally they seemed healthy. A third still smaller superficial tumour lay just behind one of the olives, it differed in that it contained, in the centre of the cell growth, dimly staining fibrillated tissue; i.e. it approached more nearly to the ordinary type of a gumma.

At the point of the calamus scriptorius there was a patch of disease, distinct but very limited in extent, and not involving the adjacent nuclei. No other disease was found in the medulla or pons, and it was specially noted that the oculomotor nucleus was normal.

The diagnosis made during life, and sustained post-mortem, was syphilitic disease of the cerebral arteries. It was based chiefly on the peculiar semi-comatose state of the patient and the history of transient hemiplegia. The absence of thrombosis does not preclude us from referring these symptoms

to the arterial disease; for, as Heubner observes, the infiltration of the intima deprives the arteries of their elasticity, and thereby interferes with the cerebral circulation apart from actual obstruction. The immediate cause of death must be sought, I imagine, in the cervical region of the cord where the pia mater was most uniformly infiltrated, and the cord swollen and soft.

There can be little doubt that the disease was syphilitic. The wide distribution of it and the symmetrical invasion of the third nerve (putting aside the arterial disease) would scarcely be found in a simple inflammation. Looking to the character of the cells, and to the fact that in the third nerve and pia mater the disease was diffuse, it would perhaps be better called a syphilitic inflammation than a new growth; but in the dura mater and the pons there were distinctly isolated tumours, one of which at least was becoming fibrous in the centre; which indicates that the formation of typical gummata might have followed.

As regards the localities of the lesions there are some points of interest: First the symmetrical character of the lesion in the third nerves, to which I shall allude presently. Secondly, with regard to the cord and its envelopes: Though in the cervical region the pia mater was to a certain extent thickened all round, yet both here and down to the lumbar region definite patches of infiltration, extending from it into the cord, were found only on the posterior aspect of the cord (chiefly involving the posterior nerve roots and the posterior median fissure). Now although there was no systematic sclerosis, it seems probable that, had the patient lived, secondary ascending degeneration of the posterior columns would have set in. The possibility of a locomotor ataxy thus originating from a syphilitic meningitis is a matter of some interest.

Considerations of interest are also raised by the lesion of the third nerves in connection with the loss of the upward movement of the eyes. Paralyses in general may be roughly divided into three classes according as the lesion is situated—(1) on the nerve after it leaves the cord or medulla; (2) in the lowest (spinal or medullary) nucleus of the nerve; (3) in some higher (mostly cerebral) centre. In the first class of cases the paralysis follows the anatomical distribution of the nerve; the second class, as a rule¹, differs little in this respect from the first; in the third class the paralysis need not follow the distribution of any one nerve, but may affect movements presided over by individual branches of one or more nerves—

¹ For exceptions see Buzzard, 'BRAIN,' Vol. V., 'On Ophthalmo-plegia Externa in Conjunction with Tabes dorsalis;' and Sturge, 'Transactions of Ophthalmological Society,' vol. i. p. 176.

movements connected by a functional rather than by an anatomical association. In no case is the functional association of movements better exemplified than in that of the eyes, and for the classification of paralyses affecting these movements I would refer to a paper by Dr. Sturge.¹

Now, seeing that in the present case the paralysis of the eyeballs was symmetrical, and that it affected the upward movement chiefly,² it was not unnatural to diagnose a central lesion. I remember to have seen, when casualty physician to St. Bartholomew's Hospital, a woman with fits of an uncertain nature, who could not raise her eyes above the horizontal plane, the other movements of the eyes and eyelids being perfect. Dr. Gowers³ showed to the Ophthalmological Society a woman with ocular paralysis, limited to the upward movement of the eyes, associated with optic neuritis and other symptoms of intracranial disease, in which case he suggested a cerebellar lesion. Nevertheless, in the case now under consideration, the lesion turned out to be peripheral; symmetrical indeed, and therefore explaining the bilateral character of the paralysis, but leaving in some doubt the question why the upward movement of the eyes should have been so particularly affected. From the fact that the disease seemed to spread along the peripheral parts of the nerve, it might be surmised that the fibres, which go to form the branch to the levator palpebræ and superior rectus,⁴ run in the periphery of the nerve trunk; but I do not insist upon this supposition, seeing that at the thickest part of the enlargement of the nerve there were very few fibres left at all. It seems better to fall back upon the analogy of paralyses in another organ, viz. the larynx. Dr. Felix Semon has shown⁵ that here, whether the seat of the lesion be central or peripheral, the *abductor* fibres of the recurrent laryngeal nerve are most likely to be affected. Why this should be so, when the lesion is upon the nerve trunk, does not appear easy of explanation. Dr. Ferrier⁶ surmises that in general "the extensor and abductor nerves and muscles have less vital resistance, and are sooner exhausted than the flexors," and that "a generally enfeebling cause will show itself first in the extensors." He instances the paralysis

¹ 'Ophthalmological Society's Transactions,' vol. i., "Two Cases of Simultaneous Paralysis of both Third Nerves."

² Originally there had been ptosis of the left eyelid, sufficiently marked to attract his wife's attention. The droop of the eyelids when he came to the hospital was less marked than the downward direction of the eyes.

³ 'Ophthalmological Society's Transactions,' vol. i. p. 117.

⁴ Unfortunately the subdivisions of the third nerves were not specially dissected out. But the nerves had regained their normal size and appearance before entering the orbit.

⁵ 'Archives of Laryngology,' vol. ii. No. 3.

⁶ 'BRAIN,' Vol. IV. p. 311, "The Localisation of Atrophic Paralyses."

of the extensors in lead poisoning, and the experiments of Onimus upon the electro-irritability of muscles post-mortem. But whatever the explanation, the present case shows, I think, that a lesion of the third nerve trunk so complete as to leave (where at its worst) very few nerve fibres visible, may be expressed by a paralysis of the upward movement of the eyes, or at most by that and an incomplete ptosis.