CLINICAL SECTION

DEMONSTRATION OF NEUROLOGICAL CASES

Held at The West End Hospital for Nervous Diseases

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Ladies and Gentlemen,

I propose to show you four cases of organic disease of the nervous system.

Case I

Syphilitic meningo-myelitis; 37 years since the date of onset.

As the first patient walks in, just observe his gait to determine its type. This man is now aged 67, and a very old friend of mine; one I met in the first world war. You notice that he walks with a somewhat spastic gait, showing some adductor spasm and a tendency to catch the toes on the floor. When I first saw him in 1915, at the age of 30, he could not walk at all and was admitted to my ward in a military hospital with a provisional diagnosis of 'hysteria' or, as he himself put it, he was believed to be 'swinging the lead.' Nevertheless, he showed very definite signs of organic disease which even in those days it was difficult to overlook. There was a more or less total paralysis of the legs, with a certain amount of spasticity. In addition, he showed loss of sensation of a dissociated type, that is loss to pin-prick and thermal stimuli, over the left leg and extending up to about the eighth dorsal segment in rather an irregular fashion; there was no hard and fast line of demarcation between the anaesthesia and normal sensation, but from above downwards the latter gradually faded away. Motor power was rather less in the opposite limb—the right leg. In other words, he showed what is known as a 'Brown-Séquard syndrome,' in which the dissociated loss of sensation is on one side of the body with signs of upper motor neurone disturbance predominating on the opposite side. Thus, on the left side dissociated anaesthesia existed over the leg and upwards to about the eighth dorsal segment, while the right leg showed reduced motor power as compared with the left, increased knee and ankle jerks, ankle clonus and a pronounced extensor plantar reflex.

As a result of investigation and treatment, his condition gradually improved and he was able again to walk. Although he did not become fit enough to resume Army service he has done very well, as you see, during the intervening 37 years, and I doubt if he has deteriorated appreciably over this long period. He suffers occasionally from bladder frequency, but has good control, and the spasticity of the legs remains at very much the same level; he can get about quite well and is able to drive a car.

On examination, you observe that the pupils are slightly unequal, the right a little larger than the left, but they react to light and accommodation and convergence. There is no disability of the upper limbs; the arm jerks are not abnormal and the co-ordination of his arms is quite good. The knee jerks are much exaggerated, the right being more so than the left. There is now no ankle clonus on either side. Both plantar reflexes are extensor but the right is more definitely so than the left. Consequently the signs of motor disturbance still predominate in the right leg. Further, this limb remains slightly more spastic than the left leg and he does not flex the hip quite as well as he flexes the left hip. Dorsi-flexion at the ankle is fairly good although better on the left side. He is quite steady in the Romberg position and any tendency to sway is accounted for by the reduced motor power in the lower limbs. The former left-sided dissociated anaesthesia has entirely disappeared.

Bearing in mind that the onset occurred 35 years ago, what is the diagnosis? It is not likely to be one of disseminated sclerosis with such a long period of quiescence. Originally the presence of a Brown-Séquard syndrome would suggest the possibility of an intra-medullary neoplasm involving mainly the right side of the cord, or of a similarly placed syringomyelia. Actually the condition is one now seen with increasing rarity. On investigation his cerebrospinal fluid showed only 5 lymphocytes per c.mm. with a slightly increased

protein content of 40 mg., a Lange reaction of luetic type and a negative Wassermann reaction. The blood W.R., however, was strongly positive. Consequently the condition was one of syphilitic meningo-myelitis predominating more on the right side of the cord than the left.

As you will recall, the spinal cord is very closely invested by the pia mater; not only does it surround the cord but it sends pial septa into the medullary substance. What happens in syphilitic meningo-myelitis is a gummatous infiltration of the pia which leads to thickening not only of the investing pia but also of the pial septa. As the gummatous formation of the pia organizes and contracts, it produces pressure and torsion on the subjacent long tracts of nerve fibres and particularly upon the somewhat sensitive pyramidal tract in the lateral columns; in this case there was also interference with the adjacent grey matter on the right side. Thus the lateral columns were involved by the gummatous infiltration, the predominant motor signs on the right being due to more involvement on this side of the cord than on the left; similarly the nerve-fibres subserving heat, cold and pain, having crossed from the opposite (left) side, were caught up in the predominant gummatous mass in the right side and so led to the dissociated anaesthesia on the left side of the body.

In those days, long before the introduction of penicillin, we treated neurosyphilis with successive courses of an arsenical, usually neoarsphenamine, and bismuth, with potassium iodide by the mouth. This treatment was given in the present case.

The patient rapidly improved on treatment and within a few weeks he was beginning to walk. After the lapse of three years his blood Wassermann reaction became entirely negative and has so remained until this day. His persisting degree of paraplegia, of course, is due to permanent scarring of the cord. While the original inflammatory lesion has healed long ago, a portion of the pyramidal tract has without doubt been destroyed, especially on the right side.

The particular pathological class of neurosyphilis to which this case belongs—being a manifestation of the tertiary stage—is that of ‘meningo-vascular neurosyphilis’ as distinct from ‘parenchymatous neurosyphilis,’ of which tabes dorsalis and general paresis are the most typical examples. Although a useful classification from both a clinical and pathological standpoint, there is really no hard and fast line between meningo-vascular and parenchymatous forms of neurosyphilis. For instance, secondary ‘parenchymatous’ degeneration may occur in meningo-myelitis and other forms of meningo-vascular neurosyphilis as the result of pressure on and torsion of the long tracts of the spinal cord, and

gummatous lesions may exist in the central nervous system in cases of tabes dorsalis and general paresis.

This case illustrates the arrest of syphilitic meningo-myelitis as a result of treatment even with arsenicals, bismuth and iodides, with comparative longevity, the patient still surviving and able to walk nearly 38 years later, at the age of 67. As far as present experience goes, syphilitic meningo-myelitis now being rare, treatment with penicillin offers only a slightly better prognosis, initial improvement being rather more rapid than with arsenicals and bismuth.

Case 2

Intracranial rifle bullet, in situ for 37 years, causing unilateral cerebellar symptoms.

The second case is of rather different type, a cerebellar disturbance.

First of all we will see how the patient walks. You notice very little disturbance of gait. He tends to turn his left foot inwards but that is about the only abnormality. Slight unsteadiness is noticed when he turns. He has a full range of movement of the legs, a little hesitation in ‘kicking off,’ as it were, with the left leg, and what you might call a slight intention tremor of the leg in so doing. He does not walk on a widened base, and he is almost, but not quite, steady in the Romberg position. He shows a slight tremor of the outstretched hands, and you will observe a slight intention tremor of the right arm. The left arm, however, is grossly inco-ordinate, and dysidiadokinesia is present to a pronounced degree. There is no sensory disturbance. At one time he showed some nystagmus, but this has entirely disappeared. All deep reflexes are about equal on the two sides. He shows gross inco-ordination of the left leg on applying the heel-knee test, together with a marked intention tremor. Consequently we have pronounced inco-ordination of left arm and of the left leg; slight inco-ordination of the right arm but no inco-ordination of the right leg. If anything, the left arm and left leg are slightly hypotonic as compared with the limbs of the right side.

What is the nature of the lesion? The answer will probably surprise you, because the condition is almost of the nature of a physiological experiment. The history is that during the first world war, in 1915, during the Dardanelles campaign, he was shot by a Turkish sniper; the bullet entered just behind the left ear and remained in the cranial cavity, coming to rest in front of the cerebellum almost in the mid-line, where it has remained for 37 years. During its passage the bullet evidently damaged the left inferior cerebellar peduncle and consequently the afferent nerve tracts passing from

FIG. 1.—Case 2. Lateral view of intracranial bullet.

FIG. 2.—Case 2. View of intracranial bullet in Towne's position.

the spinal cord to the cerebellum, with resulting inco-ordination of the left arm and left leg.

The patient vaguely remembers being hit and feeling faint and dizzy. He has no recollection of any event following this until he reached England about three weeks later. When he regained consciousness he recalls that his left arm and leg appeared partially paralyzed. Within six months, however, he was walking about and using his left hand.

The first X-ray examination was made in October 1915, shortly after the injury, when the entire rifle bullet was seen precisely where it still remains—just above the basilar process in front of the cerebellum. Here it lies obliquely across the mid-line in the antero-posterior view, and with its nose pointing downwards and backwards in the lateral view. Further X-ray examinations have been carried out at intervals of a few years, the last being in 1951 (Figs. 1 and 2). There has been no alteration in its position beyond a slight increase in downward tilting in the antero-posterior view. It is noted that just above the base of the bullet an irregular area of calcification has formed; this has slowly increased in density since the original X-ray examination. As to the cause of this calcification, it is interesting to speculate whether it is due to calcification of damaged nervous tissue or to the proliferation of small fragments of bone carried into the cranial cavity at the time of the original wound.

The case was shown before the clinical section of the Royal Society of Medicine in 1928. Apart from the inco-ordination demonstrated, the only symptoms from which the patient has suffered are occasional headaches, occipital in distribution. He is able to get about quite well and even to ride a bicycle.

Case 3

Friedreich's ataxia

The next patient illustrates an entirely different type of cerebellar disorder. His age is now 26 and he has considerable difficulty in walking. He first noticed unsteadiness of gait and some weakness of the legs 12 years ago—at the age of 14—and these symptoms have gradually become worse. You observe that he is too ataxic to walk without the support of my arm and that although the right foot is plantigrade, the left foot becomes strongly inverted and plantar-flexed, apparently due to spasm of the tibialis posticus muscle. On attempting to stand, even on a widened base, he is very unsteady. He shows slight nystagmus on lateral deviation of the eyes, but the pupils react normally and the optic discs are also normal. Speech is slightly jerky and punctuated. There is no tremor

of the hands and arms at rest nor even when outstretched, but you see considerable inco-ordination with the finger-nose test and pronounced dysidiado-kokinesia of the same type as we saw in the left arm of Case 2. All arm jerks are absent and the knee jerks cannot be obtained even on reinforcement; the ankle jerks are similarly absent. You note the highly-arched foot—pes cavus—and the bilateral extensor plantar reflexes. There is no loss of superficial sensation but vibration sense, although present, is considerably diminished as well as tendo Achillis sensation.

From the physical signs we have elicited, the diagnosis is fairly straightforward, the case being one of Friedreich's ataxia. Curiously enough, no family history of the disorder can be traced. There is one younger child in the family, aged 18, who is quite healthy and his mother and father are both alive and well, but little is known of other relatives.

Of several varieties of heredo-familial ataxia that have been described, Friedreich's ataxia is the most typical of the group. It is, in fact, doubtful if the other forms of heredo-familial ataxia that have been differentiated are true clinical entities. They probably represent mere variants of the same disorder, as there is considerable overlapping of symptoms and signs between the varieties as well as gradations from one to the other. Marie's ataxia, for instance, in its pure form is characterized by cerebellar ataxia with optic atrophy, the pyramidal tracts and posterior columns of the spinal cord being relatively little affected; consequently the knee and ankle jerks are retained and the plantar reflexes flexor. One meets with an occasional case, however, showing all the signs of typical Friedreich's ataxia with the addition of optic atrophy. The Sanger-Brown type, a rare form, is characterized by the presence of oculomotor palsies, optic atrophy, exaggerated tendon reflexes but no nystagmus. Similarly, cases of more or less typical Friedreich's ataxia are sometimes encountered showing ptosis and other palsies of the extrinsic ocular muscles.

Case 4

Cerebral arterio-venous angioma

The last case is that of a little girl, aged six years, who was brought up to hospital on account of her dragging the right leg. The onset of the disorder occurred 15 months previously and the weakness of the leg had gradually increased. At an earlier age she was taken to a certain hospital where the parents were told that the trouble was due to a birth injury. This statement annoyed the local doctor who had looked after the mother during her confinement and he reassured her that the labour and birth had been perfectly straightforward, and

FIG. 3.—Case 4. Angiogram; antero-posterior view

FIG. 4.—Case 4. Angiogram from lateral aspect.

that there had been no question of any head injury.

As one noticed at the first examination, there is an angiomatic formation on the conjunctivae and a faint naevus, as you can see, on the left side of the forehead, that is, on the same side as the weak leg, not on the opposite side. On these signs the possibility of an intracranial angioma was suspected. This provisional diagnosis was rendered practically certain by the bruit you can hear on auscultation of the skull in the right temporal region and which is also audible in lesser degree on the left side; further, over the right common carotid artery you can feel a definite thrill.

The diagnosis of intracranial angioma was finally confirmed by angiography which shows a diffuse angioma of arterio-venous type on the right side of the brain in the parietal region (Figs. 3 and 4). A straight X-ray on this little patient was negative. Some angiomatica, however, show progressive calcification which renders the lesion visible on ordinary X-ray examination.

I have to thank Dr. Colin Edwards for kindly allowing me to demonstrate this patient.

Intracranial angiomatica are divisible into two main classes:

1. Malformations of the blood vessels of congenital origin.
2. Haemangioblastomata, or true neoplasms, of the elements forming the blood vessels.

The first class—angiomatic malformations—may be subdivided as follows:

(a) Telangiectatic angiomatica, which are small capillary lesions of the same type as occur in the skin and which may be present in the central nervous system. These are of no clinical importance as they cause no symptoms and are discovered only on pathological examination.

(b) Capillary angiomatica which are formed mainly of capillaries and involve the meninges or are spread out on the cortex of the brain. Very often they co-exist with similar naevi in the skin, particularly on the face of the same side as the intra-cranial angiomatica. In such cases there is usually a contralateral hemiplegia and Jacksonian epilepsy; the condition is then known as the Sturge-Kalischer-Weber syndrome. Such capillary angiomatica are very liable to calcify and they can then be detected on straight X-ray examination.

(c) Venous angiomatica. Intracranial angiomatica consisting solely of veins also occur but are comparatively rare. The veins constituting the angioma are large and very thin-walled; consequently they are extremely liable to rupture. As a rule the angiomatic formation involves the parietal area of one cerebral hemisphere and is usually wedge-shaped, the broad part of the wedge

reaching the cerebral cortex and the point penetrating deeply into the brain. Hemiplegia with Jacksonian epilepsy are frequent symptoms. Unlike the intracranial capillary angiomatica, the purely venous angioma does not tend to calcify and is not, therefore, demonstrable by radiography. Sooner or later one of the thin-walled veins ruptures with resulting fatal haemorrhage.

(d) Arterio-venous angiomatica. This type of angioma consists of a tangle of pulsating blood vessels of varying sizes through which arterial blood flows from enlarged arteries of entry and escapes by means of several, usually enlarged, emissary veins. The angioma is almost invariably aneurysmal in character, showing arterio-venous communications either from the first or developing later, and is usually very extensive. Occasionally the angioma has extensive vascular connections with dura and scalp when the blood vessels of the latter, especially in the temporal region, as well as the carotid artery, may be enlarged.

The arterio-venous angioma usually becomes symptomatic in childhood—as in the present case—or early adult life. Gradually increasing hemiplegia is an almost constant symptom while Jacksonian epilepsy is very frequent. Some cases show exophthalmos, usually on the side of the lesion but occasionally bilateral, with or without swelling of the optic discs, a condition rarely seen in purely venous angiomatica. Unfortunately arterio-venous angiomatica tend to cause gradually increasing symptoms, and rupture is by no means infrequent.

As regards treatment, in some cases, following craniotomy, it is possible to clip one or more arteries of supply to the angioma. If this is not possible, ligation of the common carotid is worth while as this procedure reduces the liability to rupture. Deep X-ray therapy has also given fairly good results in reducing the size and extent of the angioma.

The second class of blood vessel tumour—the haemangioblastomata—are true neoplasms formed from angioblasts, that is, the primitive structures which form blood vessels. These neoplasms occur particularly in the cerebellum, especially of children and young adults, and they also assume different forms as follows:

(a) Capillary, consisting of a collection of capillaries and small vascular spaces, with a stroma of endothelial cells, and surrounded by a hyalinized zone.

(b) Cellular, which, in addition to the collection of vascular channels, contain large epithelial cells with prominent nuclei lying between the vessels.

(c) Cavernous, composed of large vascular sinuses lined with endothelial cells and showing in the surrounding tissue epithelial cells with a fine

reticulum. These neoplasms are especially liable to form cysts of varying size containing yellowish fluid, in which the angioblastoma is represented by a mural nodule. In the cerebellum, such cysts may be multiple, causing the organ, in section, to present the appearance of Gruyère cheese.

Cerebellar haemangioblastoma should be treated whenever possible by surgical extirpation. In operating, however, it is not sufficient merely to

empty the cyst; if a satisfactory result is to be obtained, it is essential to excise the mural nodule, otherwise a recurrence is certain.

Sometimes cerebellar haemangiomas are associated with a similar condition of the retina, the combination being known as von Hippel's disease. In other cases haemangiomas occur not only in the cerebellum but also in the spinal cord, kidneys, suprarenals, liver and pancreas when the syndrome is termed Lindau's disease.

LUPUS ERYTHEMATOSUS, PERIARTERITIS NODOSA AND MILIARY TUBERCULOSIS

Report on an Unusual Case

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Cases of miliary tuberculosis following ACTH and cortisone therapy for disseminated lupus erythematosus have been reported in the literature, although the direct proof that the hormonal therapy is responsible for the development of miliary infection is lacking, circumstantial evidence has laid the blame for the mishap on these new drugs. The following case history of a patient who received no hormonal therapy is therefore of great interest as it started as a rheumatoid arthritis and chronic discoid 'buffer-fly' lupus of face and later, after a pelvic operation, developed cervical adenitis followed by periarteritis nodosa affecting the small intestine and at autopsy showed miliary tuberculosis of kidneys and spleen. This combination of events has not hitherto been observed:

Case Report

W.S., aged 45 years, housewife. First attended as an out-patient in May 1948 for recurrent attacks of sore throat and fleeting joint pains affecting ankles, elbows and hands. She had been investigated for rheumatism at another hospital about six years before, when nothing abnormal was found. She came of a family of 20, otherwise there was nothing relevant in her family or past history.

On examination her general health was good. The appearance of the finger joints was suggestive of an early rheumatoid type of arthritis. No other abnormal signs were found. The following investigations were done.

Urine: N.A.D. Blood count: Normocytic anaemia and leucopenia. E.S.R.: Normal. Blood W.R. and Kahn: Negative. Serum calcium, uric acid, serum cholesterol: Normal. X-ray of chest and knees: No abnormality noted. X-ray of hands: 'Incipient rheumatoid change with multiple subcortical cyst involving each carpus and the head of the metacarpal in left minimus.'

She was treated with wax baths to hands, fersolate tablets, two t.d.s., and calcium aspirin, gr. 10, t.d.s. In March 1949 she developed a rash on the face which was diagnosed by Dr. W. Tillman as lupus erythematosus. She was given a course of bismuth injections which made the rash worse but temporarily improved the joint pains. In May she had acute exacerbation of her arthritis, again preceded by sore throat and fever, when she was referred to the ear, nose and throat surgeon who thought that there might be a possibility of tonsillar infection and prescribed a course of penicillin for a week. This again improved the joint pains and as the rash was not getting better she was admitted in July 1949 under Dr. Tillman. The above investigations were repeated with the same result except that the blood Wassermann was at first strongly positive, but subsequent blood tests were progressively less strong and the reaction was therefore regarded as false positive, which is occasionally found in lupus erythematosus. The throat swab was negative and she was again seen by the ear, nose and throat surgeon, who now