CLINICAL NOTES AND CASES.

Cases of Choreic Convulsions in Persons of Advanced Age.

By M. D. Macleod, M.B. Edin., &c., Assist. Med. Supt. Cumberland and Westmorland Asylum, Garlands, Carlisle.*

The three cases here reported present all the characteristic symptoms of chorea as it is known as a clinical entity—viz., incessant inco-ordinated muscular twitchings beyond the voluntary control of the patient, ceasing during sleep, and usually accompanied by a mental disturbance more or less marked. They present, however, some unusual features. The subjects were of advanced age (one a male patient aged 52, and two female patients aged 62 and 72 respectively), and in each case on post-mortem examination there was found a lesion affecting the motor area of the surface of the cerebral hemispheres. It is also remarkable that the two females were sisters.

I will first describe the symptoms during life, and then the morbid appearances, for the sake of readier comparison.

Case I.—James F., aged 52, married, from Bootle, Chandler. Admitted 3rd September, 1873.

Is stated to have been a steady, sober man. His father died of "palsy." Had been known to be affected for six months, but he himself told me his complaint had been gradually coming on for about three years. He had been irritable, restless, violent, and refractory before admission.

On admission his mental state was one of slight dementia, with a great degree of irritability. He was very talkative, and as his speech was uttered in a jerking manner, it gave an idea of considerable incoherence to his conversation. The contortions of his body and the grimaces he made when he spoke rendered his appearance and conduct more insane than really was the case.

His memory seemed fairly good, and, if talked to patiently, he could answer ordinary questions. When, however, anything vexed him his chorea became more violent—so affected his speech, and, seemingly, to a certain extent, his ideas, that his conversation was quite unintelligible.

Bodily he was markedly choreic. His trunk, limbs, muscles of expression, his tongue, and his muscles of articulation, were the seat of constant inco-ordinate spasms or twitchings. These

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twitchings were intensified by attempts at voluntary motion by emotional states and by any sensory irritation. It was subsequently found they ceased during sleep. There was some paresis of his legs, but he could walk about fairly well, except that his limbs were jerked about as he moved them. The right leg was weaker than the left. There was no apparent loss of sensation, and the special senses were normal, only that speech was jerking.

His temperature was 97°·6, and his pulse 72. His lungs and heart, as far as could be made out, were normal. He was much emaciated, as he had been poorly fed for some time past. He had a ravenous appetite.

In a few weeks he got very fat, became quieter in conduct and less talkative. He, however, always remained irritable and childish.

No marked change was noticed in his case until the beginning of 1875, when his chorea increased, and he began to get thin. At this time also he became more demented, and some stupor was noted. Various remedies had been tried—the bromide and iodide of potassium, arsenic, iron, quinine, &c.—but without any benefit.

Towards the end of 1875 he got very feeble, and gradually lost the power of walking or standing. The general choreic spasms continued, and, along with the advance of the paresis, seemed to increase. It is recorded on the 1st February, 1876, that there was no apparent loss of sensibility. His memory and speech gradually failed. The stupor increased, and for some months before his death he passed his motions involuntarily. He died on the 18th February, 1876.

Up to the time of his death the choreic convulsions continued; only the day or two before he died, as he lay almost quite comatose, they decreased in intensity and frequency.

The other two cases are so remarkably like the foregoing that I will state them more briefly:—

Case II.—Isabella S., aged 62, married, from Carlisle, housewife. Admitted 5th December, 1876. Had been an industrious woman, but "peculiar" in disposition. Her father had some disease which caused him to "shake all over" before he died. She had been ill for two years of her present convulsive disease, and lately had been very irritable and violent.

On admission—mentally she was childish, irritable, and fractious.

Bodily she was choreic all over. The spasms ceased during sleep. Senses (except "choreic" speech) and sensation normal. Considerable paresis of the legs. She was well nourished, and her other organs were healthy.

No change took place till January, 1880, when she got weaker and paralysed. Although both sides of her body were paralysed, her right arm, leg, and face were distinctly more powerless.

The choreic twitchings continued till death, but less intense and frequent on the right side than the left. Her mental powers

gradually failed, and for several weeks she lay in a state of great stupor.

She died 19th August, 1880.

Case III.—Mary B., aged 72, widow, from Carlisle, housewife. Admitted 22nd September, 1879.

A sister of Isabella S. Stated to have been a quiet, steady woman. She had been affected for more than two years. Had been noisy at night before admission.

In this case the chorea was well marked and general. The convulsions ceased during sleep. There was no affection of sensibility. She was very feeble, and was paralysed in both her lower extremities.

Mentally there was great stupor. She was much emaciated. She gradually got weaker, more powerless over her whole body, and died 19th January, 1880. The choreic movements continued till death.

Post-mortem Appearances.

Case I.—James T.

Nervous System.—Skull-cap thick, dense; diploë obliterated. The dura mater was thick and tough. Under the dura mater (which was incorporated with its upper wall) and over the vertex of each hemisphere, on each side, was a cyst lined with a dark—in some parts semi-purulent—layer of false membrane, evidently of old formation. In other parts this membrane was organised, and had vessels ramifying in it. These cysts contained ten to twelve ounces of dark bloody fluid, in which was suspended flocculent particles. The cysts extended in each hemisphere over the parietal lobes, the posterior part of the frontal lobes and down to the fissure of Sylvius. They touched each other at the falx, but did not communicate. Bands extended from wall to wall of the cysts. The vessels of the meninges were enlarged and varicose. The pia mater underneath was intensely congested and adherent to the parietal convolutions. The arteries of the brain were atheromatous. There were gliomatous bodies on choroid plexus. The pacchionian bodies were very numerous, especially on the left side. The brain looked small (weighed 42·5 oz.). The convolutions were atrophied, especially in the superior parietal lobes, the grey matter of which was much thinned. The brain on section was firm and congested. The peri-vascular spaces were well marked. The floors of the lateral ventricles were rough and granular. These cavities contained little fluid. The fourth ventricle was dry, its floor covered with large coarse granulations.

The cerebellum was firm, its white substance, pink in colour, full of puncta. The pons and medulla were firm on section, and somewhat congested.

The membranes of the spinal cord were congested. There were some lymphy-looking opaque patches, about the size of a threepenny piece, on the anterior surface of its lumbar portion. The spinal pia

mater was congested and closely adherent. The cord, as a whole, was soft, and there were some minute greyish soft spots on its right anterior columns.

The other organs of the body were healthy.

CASE II.—Isabella S.

Nervous System.—Scalp and skull-cap normal. On removing the skull-cap the left side of the brain was seen to bulge, and there was a sense of fluctuation over the left hemisphere. On cutting into the dura mater of the vertex and sides of the left hemisphere seven to ten ounces of grumous bloody fluid escaped. Under the dura mater of the vertex and sides of the left hemisphere there was a cyst lined with a thick, dark-coloured false membrane. This cyst extended over an area of brain comprising the posterior part of the frontal and the whole of the parietal lobes of the left side. The dura mater over it was much thickened and congested, and the pia mater under this membrane, over the marginal part of the superior parietal lobule, was thickened and adherent to that convolution. The membranes of the right side were normal. The surface of the convolutions at the upper part of the ascending frontal, ascending parietal convolution, and the superior parietal lobule of the left side was depressed and flattened. The vessels of the brain were normal; the lateral ventricles were much dilated, and filled with straw-coloured fluid, especially the right. There were large gliomatous bodies on the choroid plexuses. The brain on section was firm, somewhat congested. The cerebellum and pons were firm, stained pink, and congested. The spinal cord was apparently normal.

The other organs of the body were healthy.

CASE III.—Mary B.

Nervous System.—The scalp and calvarium were normal. In the texture of the dura mater were several tumours. The largest, about the size of a chesnut, was situated on the upper part of the left hemisphere. There were clustered round it several small tumours, about the size of large peas. They pressed on the roots of the first and second frontal convolutions, and on the upper part of the ascending frontal and ascending parietal convolutions, causing a distinct depression in them—a thinning but not entire destruction of the underlying grey matter. In the anterior part of the falx cerebri was a small tumour, about the size of a pea. These tumours were of a hard fibrous scirrhous nature. The arteries of the brain were atheromatous. The brain on section was firm, pale, and seemed otherwise normal. The spinal cord presented no abnormality.

The other organs were normal.

In noticing the salient points of these cases, the age of the patients calls for remark. Convulsive diseases are common enough at all ages, but here we have convulsive diseases of a special clinical type which are rarely found after the age of

twenty years. Many such cases are recorded, but, so far as I could find, few of the reports give details.

Niemeyer remarks, in his chapter on Chorea, that "even the most advanced old age, however, is not entirely secure from it, and in such cases the affection exhibits a peculiar intractability." In Trousseau's "Clinical Medicine" mention is made of reports of cases in people in the sixties and seventies. A case is reported at some length from Camillo Golgi in Von Ziemssen's article on Chorea, in his "Cyclopædia of Medicine," of a man attacked in his 32nd year. The disease lasted for ten years, and at its commencement was accompanied by maniacal excitement. On post-mortem examination an affection of the corpus striatum and the convolutions in connection with it was found.

In the recent volumes of the "Journal of Mental Science" there are reports of three cases—one of a male patient in the Royal Edinburgh Asylum, aged 36, by Dr. James Maclaren, in Volume xx., p. 97. The disease was of six years' duration, and had been preceded by severe frontal headache. A brother of the patient had suffered from "brain fever." Amelioration of the symptoms had ensued under treatment.

Dr. Clouston has very kindly given me notes of the termination of this case. The patient remained unchanged for a year after the report (April, 1874), when all the mental and bodily symptoms became worse. By the end of 1875 he was decidedly more facile and childish, and the choreic movements more constant—very distressing and exhaustive to his strength. He died of exhaustion from chorea and from gangrene of the right lung on the 10th July, 1876.

The chief appearances observed on post-mortem examination were a thick, dense skull-cap; large pacchionian bodies; dura mater adherent and thickened; pia mater thickened and milky, and extremely tough; a slight extravasation of blood to the extent of 2½ inches under the arachnoid, involving the middle lobe of the right side, also the left, but less extensive; atrophy of convolutions, which were easily separable from the membranes; fluid under the membranes, obscuring the form of the convolutions. The atrophy was general over the surface of the brain. The base of the brain was healthy. An abnormal quantity of fluid in the ventricles; the brain substance bloodless, the grey matter pale and distinctly divided in two layers; the white matter pale. In the centre of the optic thalamus of the right side was a softened spot, about the size of a pea, of dark colour, but with no distinct clot. Gorging of the vessels

of the spinal membranes; congestion and staining red of the anterior columns of the cord.

Dr. Sutherland, in Vol. xxv., p. 308, puts on record a case in a woman aged 53. It was the second attack, and seemed to be of a somewhat hysterical nature.

Dr. Mackenzie Bacon records a case under his care, in Vol. xxvi., p. 203. The subject was a female, aged 58. This case closely adheres to the type of Dr. Maclaren's and my cases, with (as Dr. Bacon courteously informs me) the characteristic cessation of spasm during sleep. After death, "chronic arachnitis, a shrunken brain, and a large collection of serum in the ventricles" were the morbid appearances observed.

I am indebted to the kindness of Dr. Henry Barnes for the following notes of a case he showed me at the Border Counties Home for Incurables, at Stanwix, near Carlisle:—A woman, aged 47, had been the subject of chorea or choreic spasms for 38 years. At the age of nine she received an injury to the lumbar portion of her spine, which was followed by partial loss of power in her legs; spasmodic involuntary twitchings of the muscles of the calf of the left leg then appeared. These never ceased, but spread to the other limbs, and during the last nine years the spasms have become almost general over the whole body. The tongue can be protruded straight and kept out though there are spasms of the arms. She can put out her hands readily and directly to grasp any object.

The legs, face, and trunk are, however, markedly choreic, and there is considerable paresis of her legs. Voluntary movements or attempts to move increase the jerks. There is a cessation of convulsions during sleep. There is no loss of tactile or ordinary sensation, and the sensibility to heat is normal. With regard to the mental state, she is decidedly childish. The patient had been subjected to a hard life, and was ill-used by her step-mother. Latterly she was addicted to alcohol and opium.

The question of heredity as having some effect in the causation of the chorea, or, at least, in bringing about the type of convulsion, presents itself. In my three cases hereditary predisposition existed markedly in the two females, who were sisters, and whose father had some "shaking" disease before he died.* In the case of the male it is not so apparent. His father, he told me, had "palsy." A brother of Dr. Maclaren's case had "brain fever"—probably some meningitic disease.

* Since writing the above I have been informed that two brothers of these women are choreic.

Coming now to the morbid anatomy, we have in those cases a distinct coarse lesion affecting, and limited to, the motor area of the brain surface. These lesions began as irritative lesions, but became in the end destructive lesions, and progressively so, as may be seen by the gradual advance of motor paralysis in each case.

The patients not having been seen till the disease was advanced and the convulsion general, it can only be a matter of conjecture where the irritation commenced. In Case I. the lesion was large, with arachnoid cysts, and bilateral; in Case II. the lesion presented the same appearance, but was confined to one hemisphere; while in Case III. tumours pressing on one side only of the brain were found. In the latter cases the disease was of the left hemisphere, yet the chorea was on both sides of the body. Had the earlier history of the disease been accurately obtainable, it might have been found that the convulsions were at first confined to one side of the body or to one limb, in all probability the right side and the right leg. The origin and progress of the chorea in Dr. Barnes' case is more or less traceable, as commencing in the left leg, proceeding up the limb; then attacking the other leg from below on to both sides of the body and face, the arms being still comparatively unaffected. Bilateral convulsions, where the lesion is one-sided, may be explained by the views of Dr. Broadbent, which go to show that centres giving rise to movements which are associated together are bilaterally coordinated through communicating fibres between spinal nuclei or some higher centres. An irritative lesion of the one side may then, through organic connection, gradually produce perversion of function of the associated centre on the other side.

In Dr. Maclaren's and Dr. Bacon's cases the lesion of the motor area is not so apparent; yet the extravasation of blood over the middle lobes, and the marked traces of inflammatory diseases in the membranes, in the one case, the chronic arachnitis in the other, point strongly to irritation of the surface of the hemisphere.

I feel that these reports are not so definite or complete as to warrant any conclusion relative to the pathology of chorea being drawn from them. They tend to show, however, that in chorea neither a specific morbid appearance will be found, nor that the disease is likely to be the result of any one pathological state, but that chorea and choreic convulsions are due to irritation of the motor centres of the cerebrum.