

CEREBELLAR FITS OF THE HUGHLINGS JACKSON TYPE.

BY DOUGLAS FIRTH, LONDON.

THE association of fits with cerebellar tumours is not uncommon. Two varieties have been described, in both of which the spasms are tonic in character and not clonic. The first variety is seen in unilateral cerebellar disease,¹ the homolateral limbs being most affected and adducted to the body, while the contralateral limbs are abducted. Limbs, trunk, and head show a 'screw-like' rotation from the side of the lesion to the healthy side, towards which the eyes are deviated. The second variety, described by Hughlings Jackson as occurring in lesions of the middle lobe, consists of "head retraction with arching of the back, flexion of the elbows, supination of the hands, and rigid extension of the legs with pointing of the toes".² In regard to this variety, however, Sir David Ferrier thinks that it is questionable if these phenomena are the direct result of cerebellar irritation proper.³ In the following account it will be noted that the fits had mainly the characters of the Hughlings Jackson type, but there were some additional features which may be ascribed to the other cerebral lesions.

History.—The patient, a boy, age $8\frac{3}{4}$ years, was admitted to hospital, Aug. 22, 1919, suffering from 'fits'. For eight months past he had been failing in health and losing flesh; but the first definite indication of illness was noted at the beginning of August, when he complained of nausea and was sick once. A week later he became somewhat lethargic; only rarely playing about like a normal child, and complained of headache, which was followed in a day or two by giddiness. On Aug. 20 and 21 a fit occurred each evening, and on the 22nd a fit at 11.30 a.m. was followed by three more before 3.30 p.m.; the boy was then brought to hospital. Vomiting had been noted each evening for a week. The past history was uneventful, the child having been confined to bed only once in his life, and then for measles. There was no history of tuberculosis in the family of either parent; but a brother, age 10, had died two and a half years previously from tuberculous peritonitis.

Examination.—The boy was lying comfortably in bed, fully conscious, and, when questioned, complained of frontal headache and nausea. The body and limbs were somewhat wasted, the skin dry

and harsh, and the eyes a little sunken. The tongue was coated with brownish fur, but nothing else abnormal was discovered in the digestive, circulatory, or respiratory system.

Examination of the nervous system revealed very little. The mental condition appeared normal, vision was not impaired, the pupils were active and equal in size, all ocular movements were good, and no nystagmus was present. The optic discs were white and the edges indistinct; no choroidal tubercles were found. The other cranial nerves were normal. Both hands showed a fine tremor, but no inco-ordination was noted, and there was no paralysis or weakness of the four limbs.

Of the reflexes, both knee-jerks were present, but weak; the ankle-jerks could not be elicited; the plantar reflex was flexor on both sides; and the abdominal reflexes were active and equal. There was some ataxia of the usual cerebellar type, but the boy did not tend to fall to either side in particular.

The urine contained no albumin or sugar. The temperature was normal and the pulse 88. The Wassermann reaction was negative; a slight reaction was given to P.T. by von Pirquet's test.

The first fit in hospital was reported on Aug. 24; and as time went on, the fits increased, not only in frequency, but also in severity and duration, and the vomiting became a pronounced symptom.

On Aug. 23, a fit lasting a few seconds was reported, and on the 30th, one lasting eight minutes was seen. At the onset of this fit the child cried out; the head was drawn to the right and tonically retracted throughout the fit. Twitching of the right facial muscles was followed by tonic contraction. The eyes were directed to the right, the pupils were equal and three-quarters dilated, and for the last two minutes of the fit there was regular nystagmus to the right. Tonic extension of all four limbs was present, the right arm relaxing first at the termination of the fit. The trunk showed a slight degree of opisthotonos throughout. After the fit, the knee- and ankle-jerks were absent, and there was slight rigidity of the right lower limb.

A day or two later another fit was observed. At the commencement, the head and eyes were turned to the right, with nystagmus to the right. The mouth was also drawn over to the right, and the right orbicularis palpebrarum tonically contracted; these phenomena continued throughout the fit. At the onset the right arm was flexed in tonic contraction and held close to the side, but after a few seconds it began to extend, this extension being accompanied by that of the other three limbs in tonic contraction. After five minutes, opisthotonos appeared, and, as it developed, the boy rolled over on to his right side, with marked head retraction; the feet were then

plantar-flexed with the toes pointed, this action being immediately followed by general relaxation.

A third observed fit commenced with rigidity of the right arm and leg in tonic extension, and over-action of the right side of the face. Extension of the left leg followed rapidly, and, in two minutes, extension of the left arm. Next, head retraction appeared, and was followed by opisthotonos. The left arm and leg did not remain tonically extended, but were slowly flexed from time to time, and after three minutes became flaccid. The divergence of the head and eyes to the right persisted throughout the fit, which lasted nine minutes.

The description of these three fits gives the salient features of the rest. In all those of which notes were made, nystagmus to the right was present; the termination was heralded by rolling over on to the right side; and the boy was unconscious during the convulsions. On Sept. 3 weakness of the right face and grip was noted; and by this time the boy had become more drowsy and the mental condition was deteriorating. On the next day a definite reaction was given to the subcutaneous injection of $\frac{1}{10}$ mgrm. of O.T.

On Sept. 10 it was seen that a slight change had taken place in the onset of the fits. The head was turned towards the left side, while the eyes continued to turn to the right, and the right arm and leg were more affected by the tonic spasm than the left. In the intervals between the fits the right leg was markedly spastic, with greatly increased knee-jerk and positive Babinski's sign.

The fits showed a curious tendency to appear in groups; thus between Aug. 23 and Sept. 7, a period of sixteen days, fourteen fits were reported, one on Aug. 23, two on Aug. 30, two on Sept. 3, and nine on Sept. 5. Between Sept. 8 and 12 sixteen fits occurred, in groups of five, four, one, and six per diem, with only one day (the eleventh) free. As the number of fits was obviously increasing, the vomiting and headache persistent and distressing, and the general condition deteriorating rapidly, a decision was made to have recourse to operative measures with a view to relieving the intracranial tension. The risk of a subsequent tuberculous meningitis was appreciated, but it was evident that, if nothing was done, the boy would quickly succumb.

Operation.—On Sept. 13, Mr. John Everidge performed a right subtentorial decompression. On removal of the bone, no cerebral pulsation was seen, but after incision of the dura the brain herniated through the opening, and pulsation became normal. A finger introduced into the cerebellum did not detect any tumour.

After the operation the fits ceased, although the vomiting continued about once daily. In all other respects the results of the operation were most satisfactory until Oct. 4, when the temperature

began to rise and the child was drowsy ; as time went on the vomiting became more and more persistent, coma gradually ensued, and death took place on Oct. 15.

Post-mortem Report (for which I am indebted to Dr. Stanley Wyard).—On the skull being opened, the membranes were found congested, and about the base they were much thickened, and showed considerable lymphoid exudate. The brain substance presented the signs of increased pressure—flattening of the convolutions and diminished depth of the sulci. Along the vessels lying in both Sylvian fissures were numerous minute tubercles. On section of the encephalon, three tuberculomata were discovered. One, about one-third of an inch in diameter, was situated in the anterior pole of the left centrum ovale, involving fibres from the frontal lobe only ; those from the sensory motor area were unaffected. Two were in the cerebellum ; one of these, about the size of a cob-nut, lay free in an abscess cavity in the right lobe of this organ, the other occupying its middle lobe. It was not visible on the surface, but when the cerebellum was removed from the pons, the mass was found bulging into the fourth ventricle and pressing on the upper and middle cerebellar peduncles. This tumour was about three-quarters of an inch in diameter, and was almost exactly in the middle line. Dr. Wyard was of opinion that the cavity in the right lobe of the cerebellum was recent in origin and was probably formed after the operation.

The lungs showed numerous scattered tubercles throughout. The pericardium was firmly adherent everywhere, and could not be stripped off the heart, at the base of which was a mass of caseous tuberculous material. The valves were normal.

The contents of the abdominal cavity, except for a few small tubercles on the surface of the liver, a small caseous mass in that organ, and a smaller mass in the right kidney, presented no pathological appearances.

Pathogenesis of the Symptoms.—Explanation of the symptoms is difficult owing to the multiple tumours present in the brain and cerebellum—especially as tumours of the frontal region often give rise to symptoms which suggest cerebellar lesions. The head retraction, opisthotonos, extension of the limbs, and pointing of the toes during the fits, as described by Hughlings Jackson, were undoubtedly associated with the tumour of the middle lobe of the cerebellum ; but as this tumour was not directly affected by the operation, which only relieved pressure under the tentorium, and as the fits ceased immediately the intracranial tension was released, it would appear, as Ferrier has suggested, that such fits were due to the pressure of the tumour on the subjacent structures, of which there was no doubt, and not to any direct irritation of the middle lobe.

The tumours in the frontal and right cerebellar lobes may afford an explanation of the other features of the fits—the nystagmus and turning the head to the right, the facial symptoms, and the rolling over on to the right side at the end of the fit. These tumours did not give rise to fits *per se*; but when fits, originated by the pressure on the hind-brain, were in evidence, they exhibited additional stimuli, and produced ‘complicating’ symptoms.

Some of these symptoms would seem to be contrary to those usually described. For instance, in Stewart and Holmes’ case,¹ where a hæmorrhage affected Deiters’ nucleus, the patient turned on to the sound side. Now Ferrier and Turner⁴ have pointed out that rotation from the side of the lesion and the taking up of a forced position on the side of the lesion are essentially the same thing; so it is possible to regard the attitude assumed here at the end of the fits as a ‘forced position’. The nystagmus—only present during the fits, a rare occurrence—was to the right. Nystagmus is present in the large proportion of cerebellar lesions, both to right and left, but the swing towards the side of the lesion is more ample and regular than to the unaffected side. Had the boy lived, nystagmus would probably have developed in the intervals between the fits, and to the left, that which showed being an early incomplete form of the common type.

“The symptomatology of the prefrontal lobes is essentially true of lesions of the prefrontal section⁵ of the centrum ovale”;⁶ so the presence of the frontal tumour suggests an explanation of the facial symptoms, and the turning of the head and eyes to the right. Similar phenomena have been described by Bruns⁷ and Chouppe⁸—by the former in a case of hæmatoma indenting the left frontal lobe where the patient had slight paralysis of the right hand and face; and by the latter in one of tuberculous meningitis in which there was rotation of the head and eyes to the right. Here a superficial lesion, the size of a shilling, was found on the superior part of the middle frontal convolution in the left hemisphere.

When multiple tumours of the brain are present, it must always be a matter of great difficulty to allot the part each has played in the production of symptoms. While the boy was alive, the main group of symptoms during the fits was that described by Hughlings Jackson, and was recognized as such. After the post-mortem findings had been considered, the attempt was made, with some diffidence, to correlate the remaining symptoms with the other cerebral lesions.

Another instance of tonic spasms due to intracranial disease has been brought to my notice by Miss Noel Olivier, house physician at the Victoria Hospital for Children. They occurred at the termination of a case of tuberculous meningitis in a boy, age 5 years, who

was admitted with a history of five days' drowsiness and died twelve days after admission.

The record of the case shows nothing noteworthy in the course of the disease until twelve hours before death, when the onset of tonic spasms was noted. The boy was then deeply comatose, with increasing pulse and respiration rates, and, once the convulsions commenced, so frequent were the attacks and so short the intervals of relaxation that the boy was hardly ever at rest during the twelve hours during which they lasted. All the convulsions were of the same type, and were ushered in by a paroxysm of rapid, deep, rhythmic, grunting respiration, accompanied by sweating, and followed at once by rigid tonic extension of the upper and lower limbs.

The arms were held close to the side with the elbows extended, the forearms everted, the wrists palmar-flexed, and the fingers flexed. The lower limbs were rigidly extended; the ankle on the left side was extended and the toes were pointed, while on the right side dorsiflexion of the ankle was followed by extension. As the convulsion subsided it was possible to obtain marked exaggeration of both kneejerks, but that on the left side was especially active. Ankle-clonus was also more marked on this side. No clonic movements at all were observed. The trunk was extended, with slight opisthotonos, but the head was neither retracted nor drawn to either side. The face was very little affected except for over-action of the occipitofrontalis, causing wrinkling of the brow.

Each convulsion came on very rapidly, lasted five to ten minutes, and subsided gradually, the complete relaxation between the attacks only lasting three to five minutes. No definite position of the body was assumed during or between the attacks, although there seemed to be a tendency to lie on the left side.

Post mortem, the brain showed a well-marked tuberculous meningitis, with much dilatation of the lateral ventricles, which contained a quantity of turbid fluid.

REFERENCES.

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- ⁴ FERRIER AND TURNER, *Phil. Trans.*, 1894, B. clxxxv, 719.
- ⁵ PITRÉS, *Lésions du Centrum Ovale*, Paris, 1877.
- ⁶ FERRIER, *Allbutt and Rolleston's System of Medicine*, 2nd ed., viii, 101.
- ⁷ BRUNS, *Deut. med. Woch.*, 1892, xviii, 138.
- ⁸ CHOUPE, *Bull. Soc. Anat.*, 1871, 2nd s., xvi, 380.