A CASE SIMULATING INTRACRANIAL TUMOUR

IN WHICH RECOVERY WAS ASSOCIATED WITH PERSISTENT CEREBRO-SPINAL RHINORRHOEA.

By T. R. GLYNN, M.D., F.R.C.P.,

Professor of Medicine, University of Liverpool.

WITH A NOTE BY

E. E. GLYNN, M.A., M.B.Camb., M.R.C.P.,

Pathologist and Assistant Physician, Royal Infirmary, Liverpool;
Lecturer in Clinical Pathology, University of Liverpool.

THE subject of this affection was sent to me early in 1899 by Dr. Owen Evans of Liverpool. The following is my note of his condition at this period:

He was an electrical engineer, aged 21. He had suffered from paroxysmal pain in the head and vomiting for twelve months. His family history was good. His parents and two brothers were alive and well; a third brother and his only sister died at the ages of 2 and 5 from infantile disorders. With the exception of very occasional attacks of migraine, his health, up to the time of the present illness, had been perfect. He had been a well-conducted, industrious youth, and had never had syphilis.

In June, 1898, he suffered from ammonia poisoning, owing to the bursting of a pipe in a refrigerating store; for a day or two he was laid up with headache and vomiting, and was unable to return to work for a fortnight. Two months afterwards, when in a coal mine and walking in a stooping attitude, he struck the top of his head against a beam, temporarily lost consciousness, and vomited. He soon recovered and returned to his lodgings, where mustard was applied to the back of the neck to relieve severe headache; in a few days he went home to Liverpool, as he was too ill to work. He remained at home for a fortnight, and then, feeling better, attempted to resume his duties, but the headache and vomiting returned, and he was henceforth compelled to give up his occupation.

During the twelve months before his first visit to me in 1899 the headache generally occurred about every fourteen days; for the first few months it was limited to the forehead, and subsequently was usually confined to the back of the head; it continued for three to four days, and was associated with vomiting, giddiness, and depression, so that he was obliged to remain in bed. Occasionally also pain in the head and vomiting disturbed him in the night. Dr. Evans noted that during these paroxysms of pain the pupils were widely dilated. He also experienced certain choking sensations, at times attended with much agitation. Double vision on looking to the left was complained of within two months after the onset of the headache. In the intervals between the spells of headache he suffered from want of energy and some failure of memory and giddiness, any attempt to stoop causing him to fall forwards.

He was a well-formed, healthy-looking young man. There was some weakness of the left external rectus, the pupils were dilated, reaction normal, vision normal, with the exception of some concentric restriction of the visual fields; marked optic neuritis, knee-jerks exaggerated, plantar reflexes absent; abdominal and cremasteric reflexes well marked on the right, less marked on the left side; marked left hemianalgesia; gait normal. He usually slept well, and had a good appetite. The thoracic and abdominal organs were normal.

The next note I have of his condition is dated January, 1900. He reported that during the past twelve months he had been better, and sometimes almost fit for work. The paroxysms of headache had been less frequent and less severe; the double vision annoyed him but little. He, however, was still unable to stoop. The optic neuritis persisted, and there was little change elicited on physical examination.

During the next twelve months he distinctly became worse; the headaches never occurred in their usual violent form, but were occasionally troublesome. His memory became very defective, he grew lethargic and childish, laughing loudly on the slightest provocation. Taking little exercise and eating well, he became stout, and his sight failed so that $V. = \frac{1}{12}$. The pupils were dilated and sluggish, there was slight nystagmus, and marked weakness of left external rectus. He reeled in walking, and complained of weakness of the legs and of a tendency to fall forwards, and on several occasions had tumbled on his face. He was unable to walk down hill without assistance owing to an inclination to run. His legs frequently failed suddenly, causing him to collapse to the

ground, and on such occasions he could not rise without help; this accident usually happened on passing from a warm to a cool atmosphere. His arms became weak, and there was volitional tremor of the hands; he frequently dropped articles he happened to be holding.

In the summer of 1901 I found that his pupils were widely dilated. Nystagmus was present. He reeled in walking and was inclined to fall forwards on standing with his eyes closed. There was distinct failure of muscular power of the extremities and tremor of the hands. Slight ankle clonus and absence of the abdominal, cremasteric and plantar reflexes.

During the following twelve months he continued to fail gradually. He no longer complained of headache and rarely vomited, but his intelligence continued to deteriorate; he became apathetic and indifferent to his condition. He could not add two simple figures together and his memory was completely lost. He was nearly blind, and was too weak to stand long without support. On three occasions he suffered from epileptiform attacks, in each of which he remained a long time comatose: one of these convulsions occurred in bed during the night and two during the day. Incontinence of urine set in; unless frequently passed, it ran away during the day and escaped involuntarily in bed at night.

Such was his condition when, in June, 1903, clear fluid commenced to escape in drops from the right nostril and immediately he began to improve, his mental vigour and muscular strength returned, and his sight was restored. The progress toward recovery was rapid, since he was early in August able to walk twenty miles.

I saw him in February, 1904, and found him almost completely restored to health; he complained only of slight failure of memory, some loss of energy, and of the discharge from the nose. The optic neuritis and all the other symptoms of cerebral mischief had disappeared.

I found that some clear fluid escaped from the right nostril. The discharge was collected by my son, Dr. E. E. Glynn, and found by him to present the characteristics of cerebro-spinal fluid.

On a subsequent occasion—October, 1904—he stated that he felt perfectly well, that his memory was as good as ever, and that he had returned to work. He continues to enjoy good health, but the rhinorrhoea persists. Dr. Hunt kindly examined the nasal cavity and found it normal.

NOTE BY DR. ERNEST GLYNN.

The discharge of fluid only occurs when the patient inclines his head slightly forward, and is especially troublesome when reading or writing, he is then obliged to blow his nose frequently, but as the discharge does not stiffen his handkerchief, one easily lasts him a whole day. The rate of the flow when sitting with the chin depressed varies from 1 to 5 drops a minute. When the head is held straight up, as in walking and sitting erect, the discharge ceases, the fluid apparently passing down the back of the nasal fossa into the pharynx, though its passage cannot be seen with a laryngoscope. The quantity of fluid escaping on different occasions, when the patient was sitting with his head forward, was as follows:

February 28th, 1904, 0.86 c.cm. in 30 minutes, at the rate of 41 c.cm., or 1 oz. 3 dr. per diem.

November 7th, 1904, 6.1 c.cm. in 45 minutes, at the rate of 194 c.cm., or 7 oz. per diem.

December 5th, 1904, 4.2 c.cm. in 50 minutes, at the rate of 120 c.cm., or 4 oz. 2 dr. per diem.

January 16th, 1905, 5 c.cm. in 55 minutes, at the rate of 131 c.cm., or 4 oz. 3 dr. per diem.

Examination of Fluid.

1. When fresh it is clear as water, and faintly alkaline. Its specific gravity obtained by weighing 6 c.cm. is 1.007.

2. On acidifying with acetic acid there is a faint opalescence which passes through filter paper. A similar opalescence appeared in specimens of cerebro-spinal fluid obtained by lumbar puncture from three other patients; it is therefore probably not due to mucin washed out of the nose, but to nucleo-albumin.

3. On acidifying with acetic acid and boiling, a faint but distinct cloud appears, which is insoluble in nitric acid, and gives the xantho-proteic reaction for albumin.

4. By saturating the fluid with magnesium sulphate in the usual way and filtering, it was found that both globulin and serum albumin were present, the precipitate of the latter being the denser.

5. By precipitating the proteids in 10 c.cm. of the fluid with absolute alcohol, and drying and weighing the precipitate, the proteids amounted to 0.112 per cent.

6. The fluid when recently secreted reduces freshly prepared Fehling's solution, but if kept for a few days it becomes turbid from the growth of bacteria, and the reduction is then very faint or completely absent.

7. Drops of the fluid inoculated with aseptic precautions into tubes of agar and blood serum always yielded mixed cultures.

8. Fifteen c.cm. of the fluid were examined for choline with a negative result. The absence of choline is consistent with the patient's complete recovery from all cerebral symptoms.

9. Twelve grams of potassium iodide taken by the mouth in solution, or in gelatine capsules, produced the iodine reaction in the saliva in a quarter of an hour, and in the drops from the right nostril in half an hour, and also in a swab of the secretion of the other nostril. The test employed was starch paper and fuming nitric acid.¹

10. Two grains of methylene blue taken as a pill did not appear in the fluid collected one hour afterwards or during the next two days. In case the methylene blue was secreted in a reduced and colourless form, specimens of the fluid were oxidized with peroxide of hydrogen, or by boiling with acetic acid; they were finally shaken up with chloroform with a negative result.

11. The fluid when centrifuged for ten minutes invariably contained a few multinuclear leucocytes and an occasional lymphocyte. The number of these cells was approximately 37,400 per c.cm. or 37.4 per c.mm. This result was obtained in the following manner: Four c.cm. of freshly-secreted fluid were centrifuged for ten minutes, a measured quantity was then poured off, and the remainder shaken up with an equal amount of methylene blue solution. The cells in this mixture were sufficiently numerous to count in a Thoma-Zeiss haemocytometer, thus the number of cells in the original fluid was eventually calculated.

That the fluid is cerebro-spinal is proved by its physical characteristics, its specific gravity, the small percentage of proteid and the presence of a reducing agent formerly called pyrocatechin but now believed to be glucose.

The invariable presence of a few polymorphonuclear leucocytes proves that the production of the cerebro-spinal fluid is associated with inflammation, which must be slight because the number of these cells is small, and the percentage of albumin and the specific gravity is not really higher than normal. In order to make sure that these polymorphonuclear leucocytes were not possibly derived from the normal secretion of the nasal mucosa being washed off by the cerebro-spinal fluid, the other nostril was douched out with water several times. A turbid fluid was obtained. This, when centrifuged, contained many lymphocytes and other cells, but no polymorphonuclear leucocytes.

It has long been known that the subarachnoid fluid sometimes escapes from the cavity of the skull in fractures at its base, but the possibility of its spontaneous discharge through the nose without such serious injury was not suspected, and is not hinted at in textbooks on medicine, or in special works on neurology. To Dr. StClair Thomson belongs the credit of recognizing this curious form of rhinorrhoea and of demonstrating the principles on which the diagnosis of the affection depends.²

Further, Dr. StClair Thomson also merits the distinction of directing attention to the remarkable fact that cerebro-spinal rhinorrhoea is almost invariably accompanied by cerebral symptoms. This conclusion was arrived at as the result of the clinical investigation of a case of the spontaneous escape of cerebro-spinal fluid from the nose under his own care, and of painstaking bibliographical research.

From various sources, and under various names, he collected 20 cases presenting more or less the characters of cerebro-spinal rhinorrhoea; 8 of these being precisely similar to his own case, and 12 closely resembling it, and from the material thus brought together he created a fairly distinct clinical picture of a new affection. Out of the 21 cases, 17 exhibited cerebral symptoms, and in 8 there were retinal troubles. Another example of the disorder—that is, of cerebro-spinal rhinorrhoea with cerebral symptoms—has recently been reported by M. Vigouroux.³

Dr. StClair Thomson suggests that internal hydrocephalus may be the cause of the cerebral symptoms, the retinal troubles and the cerebro-spinal rhinorrhoea, and my case tends to lend support to this opinion; for, as will presently appear, the group of symptoms presented by my patient were characteristic of acquired internal hydrocephalus of the adult.

The quantity of fluid discharged in cerebro-spinal rhinorrhoea varies; in the cases collected by Dr. StClair Thomson the maximum amount was 24 oz. in 24 hours, the minimum 8 oz.; while in M. Vigouroux's patient it was 28 oz. in the same period. In my patient the maximum amount secreted in 24 hours was estimated at 7 oz. less than any hitherto recorded case. The rate of flow varies considerably, for on other occasions it was 1 oz. 3 dr. and 4 oz. 2 dr. A similar variation has been noted in other patients.

In the majority of cases of cerebro-spinal rhinorrhoea the fluid flows from the left nostril; in my patient it escaped from the right. The discharge at night when lying down, the patient believes, passes into his throat.

In many instances headache and other cerebral symptoms

ceased with the onset of the rhinorrhoea and became aggravated when it was suspended. In my patient also the advent of the discharge was associated with rapid improvement and its persistence with recovery.

There can, in my opinion, be no doubt that my patient suffered from acquired chronic internal hydrocephalus and that the spontaneous escape of cerebro-spinal fluid relieved intracranial pressure and led to recovery.

Some of his symptoms were suggestive of a tumour of the middle lobe of the cerebellum; namely, the nystagmus, reeling gait, and ataxia. The protracted character of the cerebral troubles—their duration was nearly five years—is more consistent with the existence of hydrocephalus than with the presence of a tumour. As recovery occurred, the growth, if present, might possibly have been a scrofuloma which cicatrized, but there is no evidence of tubercle or of a tuberculous tendency in the patient or in his family. Or, the growth might have been a gumma, but there is no history of syphilis, and between October, 1899, and January, 1901, he took 84 dr. of potassium iodide without benefit. Moreover, recovery, if it depended on the resolution of a tumour, could hardly have been as rapid as it was.

Other symptoms he exhibited are recognized as frequently present in internal hydrocephalus—giddiness on stooping, leading, if persisted in, to falling, vertigo, and falling on passing from a warm into a cool atmosphere, failure of nervous energy in every direction, associated with optic neuritis and an absence of focal symptoms; there was, indeed, a partial paralysis of the left sixth nerve, but this local paralysis has been noted in other instances of internal hydrocephalus, and was present in two cases of this disease under my care: one of hydrocephalus of the lateral ventricles caused by a very small tumour of the pineal gland pressing on the iter; the other of acquired internal hydrocephalus relieved by operation, presently to be referred to. Moreover, the slight palsy of the sixth nerve did not increase, an unlikely event if due to intracranial tumour. The liability to sudden failure of the legs and collapse to the ground with inability to rise without help, described in the case under consideration, was also present in these patients.

Before considering the pathology of certain cases of acquired internal hydrocephalus, it is necessary to allude to the circulation of the cerebro-spinal fluid. The most probable course is that recently summarized by Mott: "That the cerebro-spinal fluid is for the most part secreted by the choroid plexus in the lateral ventricles, and that it escapes into the third ventricle, thence by the iter into the fourth ventricle, and by the foramina into the subarachnoid space, and thence by the perivascular and perineural lymphatics into the lymphatic system." The "foramina" mentioned are the foramen of Majendie situated in the roof of the fourth ventricle and two smaller foramina, one on each side of it.

An ependymitis, causing an increased secretion of fluid in the ventricles, is said to be a cause of internal hydrocephalus, but it appears to me that so long as the fluid can find its way out of the ventricles over-distension cannot occur. I am, therefore, of opinion that the common cause of acquired internal hydrocephalus is obstruction to the passage of fluid from the ventricles. Among the recognized lesions giving rise to such obstruction are tumours of the middle lobe of the cerebellum, and tumours of the pineal gland, or in the aqueduct of Sylvius.

An exceptional cause of obstruction was found in a patient of mine who died of internal hydrocephalus. Owing to caries of the bodies of the axis and atlas, followed by their collapse and ankylosis, the odontoid process was displaced upwards and backwards, and was maintained in this position, projecting unduly into the cranial cavity and narrowing the foramen magnum; not only was the spinal cord reduced by pressure to about the diameter of a goose quill, but the medulla was dragged backwards and thrust upwards against the cerebellum, thus closing Majendie's foramen. The patient had been under treatment for caries of the cervical vertebrae, and consequent paralysis of the four extremities and respiratory trouble, and had recovered, but died twelve months afterwards from internal hydrocephalus. No other cause of the affection but that described was discovered.

Another cause of internal hydrocephalus, and one I have assumed existed in the case under consideration, is chronic meningitis in the neighbourhood of the roof of the fourth ventricle, forming adhesions between the medulla and cerebellum, and leading to closure of Majendie's foramen and the neighbouring lateral foramina. The presence of the leucocytes in the fluid from my patient may be explained by

the chronic inflammatory process implicating the choroid plexus of the fourth ventricle which surrounds the foramen of Majendie.

I have met with two cases of internal hydrocephalus which apparently originated in retention of fluid in the ventricles induced by such lesion.

A youth was admitted under my care several years ago suffering apparently from a cerebellar tumour, coma being imminent; the skull was trephined over the cerebellum. Mr. Thelwall Thomas, who operated, thrust a finger well under the cerebellum, and in doing so broke down adhesions between this organ and the medulla; a quantity of fluid escaped from the fourth ventricle, the patient's cerebral symptoms were relieved, he recovered, and is to-day a teacher of music.

The other patient, a boy aged 15, was admitted suffering from pulsating headache, persistent vomiting, and giddiness of six weeks' duration. There was no paralysis, the pupils were widely dilated and sluggish, but optic neuritis was absent; gait staggering. Four days after admission respiration suddenly failed, and he died. At the necropsy "all the ventricles were dilated; there was evidence of chronic meningitis about the medulla and cerebellum in adhesions between them, which had blocked up the foramen of Majendie."

A blow on the head is a recognized cause of the internal hydrocephalus of the adult, and it is likely that an accident of this description gave rise to the affection in my patient, and that it did so by exciting a slight partial meningitis in the neighbourhood of the fourth ventricle and cerebellum. He was only going at a walking pace when he collided with the beam, and the injury was not severe, but violent enough to cause slight concussion. His illness distinctly dates from this blow; he never thoroughly resumed his duties, and in two or three weeks the pains in the head became violent.

It may be suggested that the cerebro spinal rhinorrhoea is due to a fracture of the cribriform plate, produced by the blow on the head. I do not believe that such a fracture occurred—first, because the blow was not very severe; and, secondly, because such a fracture would not produce symptoms of intracranial pressure. Lastly, the escape of fluid did not occur till nearly five years after the blow. The patient was quite intelligent enough during the first two or three years of his illness to have noticed persistent dripping from the nose if it had been present.

Opportunities for the pathological investigation of cases of spontaneous cerebro-spinal rhinorrhoea have been rare, and have failed to indicate the course taken by the fluid in its escape from the skull. It is possible that the fluid in the ventricles as it accumulated at last overcame the abnormal resistance I have assumed existed in the thickened roof of the fourth ventricle and in the adhesions between the medulla and cerebellum; that it escaped into the subarachnoid space, and forced its way along the perineural sheath of the olfactory nerves into the lymphatic network of the mucous membrane of the nose, and so to the surface of the epithelium. The fact that there is distinct anosmia on the right side of the nose (the odour of carbon bisulphide being unperceived) suggests interference with the olfactory tract or bulb on that side.

However, it may be asserted, and I think with justice, that if the cerebro-spinal fluid forced its way out of the ventricles, it would, entering the fluid of the subarachnoid space, together with it be gradually dispersed in the ordinary way along the perineural sheaths of the cranial and spinal nerves and the perivascular lymphatics.

It is therefore more probable that adhesive inflammation occurred at the base of the brain adjacent either to the distended third ventricle or the distended descending cornua of a lateral ventricle, that is, in the neighbourhood of the lamina cineria or of some part anterior to it, that a perforation occurred at this spot, and that the fluid, cut off from the general cavity of the arachnoid, found its way under considerable pressure to the perineural sheath of the right olfactory nerve.

It is possible that certain cases of apparent intracranial tumour terminating in recovery are cases of acquired internal hydrocephalus, where the ventricular fluid has after a while forced a way out of the ventricles into the subarachnoid space and been dispersed, and that the relief of symptoms has consequently followed.

The following case was probably one of chronic internal hydrocephalus taking such a course:

A boy, aged 14, was admitted under my care in August, 1878, suffering apparently from cerebellar tumour. He had complained of headache, chiefly occipital, vomiting and giddiness for twelve months. There was slight nystagmus, double optic neuritis and impaired vision, general ataxy and staggering gait. Knee-jerks absent, occasional incontinence, temperature normal. In a few weeks improvement set in, and by October he was perfectly well, and left the hospital.

Although internal hydrocephalus may occasionally terminate by spontaneous evacuation, it is an exceptional occurrence. Hence operative measures should be resorted to in any case with cerebral symptoms pointing to the possible existence of this affection, the surgeon trephining over the lower part of the cerebellum in order to explore the roof of the fourth ventricle.

The prognosis in cerebro-spinal rhinorrhoea is unfavourable, meningitis being as a rule generated from infective organisms finding their way into the cranial cavity; but, as the discharge in my patient is inconsiderable in quantity, the channel of exit is necessarily small and the risk of infection proportionately less.

It is evident that any attempt to relieve the rhinorrhoea by local treatment would be attended with great danger to the patient, and it is not likely that any benefit would follow lumbar puncture.

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ARTHRITIS DEFORMANS AND ITS ALLIES.*

By BERTRAM ABRAHAM, M.B., B.Sc., F.R.C.P.,

Assistant Physician, Westminster Hospital; Examiner, Conjoint Board in England.

I PROPOSE to direct attention to the clinical features of the various affections which are ordinarily grouped under the name of rheumatoid arthritis. At the outset I would venture to say that I count myself among those who think that this name should be discarded as both barbarous and unscientific. This point I have already discussed elsewhere,¹ but I refer to it again now, as I think that in this case, at any rate, it would be well for us to assimilate our nomenclature to that of Continental writers.

At present, however, we have to recognize that the expression, rheumatoid arthritis, is the one in common use in this country, and, furthermore, that it includes at least four different conditions, the relations of which to one another I propose briefly to analyse. These conditions, for convenience sake, we may arrange chronologically, according to the average age of the patient when first attacked. They are (a) the children's disease, described by Dr. Still; (b) the acute process occurring in young women, of which Dr. Hale White, who has previously² reported two typical cases, has given another example to-night; (c) the familiar multi-articular affection of middle-aged women; and (d) the equally well-known chronic malady of the aged.

I do not intend to waste the time of the Society by going over details the knowledge of which is common to all, or to relate at any length the history of illustrative cases. I submit, however, a few skiagraphs and photographs of some of the more characteristic appearances.

Still's Disease.

This is usually referred to as rheumatoid arthritis in children—a most unjustifiable attribution, since in his original paper³ Dr. Still pointed out with the utmost care how it differed from what had previously been described as rheumatoid arthritis. Although the practice of using personal names in connexion with maladies is one generally to be reprehended, I would suggest that until some simple and expressive term be found this should be called "Still's disease."

It is undoubtedly an infective process, and differs from all the other forms which we have under consideration in respect of the enlargement of the spleen and lymphatic glands with which it is usually associated. Dr. Hale White believes that the acute disease which occurs in young women is also infective, and the two are certainly alike in the presence of irregular fever, and the absence of "nervous" symptoms, with the exception of considerable muscular atrophy. But in other respects they differ widely; the one is essentially chronic, is more prevalent in boys, and attacks the fingers very late: the other is acute, has only been described in girls, and usually commences in the fingers. Pathologically each is mainly a synovitis, with only secondary changes in the cartilages, but it is only in the juvenile malady that we find enlargement of the spleen, lymphatic glands and sometimes liver. The condition of the joints in the latter has been carefully studied by Whitman.⁴ On con-

* A paper read before the Clinical Society of London.