Spontaneous cerebral ventriculostium: two cases

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SUMMARY Two cases of 'spontaneous cerebral ventriculostium' are reported. In one case the diagnosis was made during life, and the hydrocephalus was not relieved by the ostium. The unique feature of the other has been the presence of a large defect in the skull bones, posteriorly, which has been a 'nature's help' towards maintaining the hydrocephalus stationary, after an initial period of progression.

The spontaneous formation of an opening between a dilated cerebral ventricle and the brain surface has rarely been recorded. Sweet (1940) described two such cases and found only two previously recorded examples (MacGillavry, 1910; de Lange, 1929). Leslie and Alker (1964) reported a further case, and while reviewing the literature on the subject maintained that the rupture of an obstructed cerebral ventricle directly into the subarachnoid space with consequent bypassing of the block appears to be exceedingly rare. They stated that before their case report a search of the literature had yielded only four such cases (MacGillavry, 1910; de Lange, 1929; Sweet, 1940; Tandon and Harkmark, 1959) in contrast with over 20 ventricular diverticula. Another case of spontaneous ventriculostomy has been reported in the Hungarian literature by Scheda and Csanadi (1963). Liss and Mervis (1965) reported a case of spontaneous ventriculostomy through the lamina terminalis and Lavender and Du Bouley (1965) described three cases of aqueduct stenosis exhibiting a cystic expansion of the posterior end of the third ventricle into the posterior fossa.

The two cases reported below are interesting in that they are in the rare group of only six cases reported so far, and, in addition, show ventricular diverticula as well. The second case is also a good clinical example of multiple complications of hydrocephalus in one individual. These are the only two cases seen in the Pathology laboratory, North Staffordshire Royal Infirmary, over the last 30 years, and this also emphasizes the rarity of this condition.

CASE 1

P.S., a female aged 29 years, had been vaguely unwell for three months, and had complained of a stiff neck, occipital headache, and difficulty in walking for six weeks. She had been deaf for several years and was stated always to have been mentally subnormal. Amenorrhoea had been noted for the past year. She was a well-nourished woman of masculine build, with heavy jaw, moderate hair development on the upper lip, and a male distribution of pubic hair. The head was bent forward while walking and there was some ataxia. Rombergism was present. Fundi showed bilateral papilloedema, more marked on the right. The right pupil was smaller than the left, and did not react to light but both reacted normally on accommodation. Ocular movements were full. Nystagmus was induced by looking upwards or to either side. The left soft palate showed slight paresis. Labyrinthine tests and tests for auditory function were not possible due to lack of cooperation. There were bilateral extensor plantar reflexes; abdominal reflexes were absent. The right knee jerk was normal, the left absent. Some dysdiadokokinesia of the arms was noted but none of legs. The WR was negative in blood and cerebrospinal fluid (CSF). The CSF contained protein 30 mg/100 ml., cells 1 per c.mm., pressure 350 mm water. Blood urea was 31 mg/100 ml. Ventriculography (Fig. 1 a, b) revealed the presence of a big cavity in the arachnoid space below the posterior horn of the lateral ventricle. The presence of air in the cavity confirmed that there was an abnormal communication between this and the ventricular system.

An exploratory operation was proposed, but death occurred during induction of anaesthesia. It should be stressed that this operation was carried out in 1941, when the risks of giving general anaesthetics to this type of case were not appreciated. From what is

FIG. 1 a and b. Ventriculograms showing the presence of a big cavity in the arachnoid space below the posterior horn of the lateral ventricle. The presence of air in this cavity signifies an abnormal communication with the ventricular system.

FIG. 2. Sagittal section of the cerebral hemisphere showing the presence of an abnormal cavity above the cerebellum, the latter being considerably deformed as a result. The opening in the anterior wall through which the third ventricle is communicating with this cavity is arrowed.

now known about the rises in CSF pressure which occur under all forms of general anaesthesia, it seems likely that death was inevitable.

NECROPSY FINDINGS *Skull and brain* The skull was thin with prominent vascular markings. There was compression of the pituitary body and erosion of the posterior clinoid processes by the distended third ventricle.

The gyri were flattened; the distended third ventricle bulged into the interpeduncular fossa, and there was a moderately well-developed cerebellar pressure cone. Between the tentorium cerebelli and the cerebellum was a fluid-containing cavity formed by arachnoid membrane, and situated in the mid-line.

A sagittal section showed extreme dilatation of the first three ventricles and enlargement of both foramina of Monro (left 7×5 mm, right 9×4 mm). The corpus callosum was raised and stretched so that its posterior portion passed forward to terminate in the splenium. The cause of the hydrocephalus was obliteration of the cerebral aqueduct by a mass situated ventral to the quadrigeminal plate, measuring $16 \times 21 \times 18$ mm. It was firm, sharply demarcated from the surrounding brain, and its cut surface showed grey and white interlacing fibres. The normal site of commencement of the cerebral aqueduct was marked by a U-shaped depression, but no remnant of the canal itself could be found.

Situated in the posterior commissure was a roughly circular opening 8 mm in diameter, and through this the third ventricle communicated with the fluid-containing cavity already noted. The superior colliculi lay below this opening, and the pineal gland and tela chorioideae of the third ventricle above it. The cavity with which the ventricle communicated, was situated in the mid-line between the cerebellum and the tentorium cerebelli, and was triangular in the sagittal section, measuring 45 mm anteroposteriorly and 30 mm transversely. Compression by this cavity had resulted in considerable deformity of the cerebellum (Fig. 2). Its floor was formed posteriorly by the superior surface of the cerebellum, and anteriorly by the quadrigeminal plate and anterior medullary vellum. The anterior wall contained the opening already noted and the attenuated posterior commissure. The roof was formed by arachnoid mater running from the anterior wall to the posterior limit of the floor.

Other organs Both lungs were conspicuously small, the right weighing 230 g and the left 213 g. The pulmonary artery was correspondingly small in size. The heart weighed 284 g and appeared normal. The adrenals together weighed 20 g. The thymic tissue

was scanty. The cervix and body of the uterus were of approximately equal length and the greater part of the cervix was supravaginal. Both ovaries showed thick white capsules with numerous cysts 0.625–1.25 cm in diameter in the cortical zone; the medulla consisted of homogeneous white connective tissue.

Microscopy The mass in the brain-stem was an astrocytoma consisting of interwoven glial fibres and numerous piloid astrocytes, and in its anterior portion there were fairly numerous multinucleated large-bodied astrocytes. The destruction of the aqueduct appeared to be complete and no structure suggesting aqueductal tissue was found.

The adrenals showed slight convolution of the cortex (within normal limits). No fuchsinophil material was demonstrated by Vine's method. The pituitary gland appeared normal.

COMMENTS The clinical and pathological findings in this case suggest a very slowly progressive lesion. The deafness which had been present for years was possibly central in origin, and caused by the tumour. The symptoms which brought the patient under observation, and among which stiff neck and occipital headache were prominent, were produced by a rapid increase in the sub-tentorial pressure resulting from obstruction of the tentorium, mainly by the expanding arachnoid cavity. The masculine characteristics and the regressive changes in the ovaries and uterus were considered a consequence of disturbance of the pituitary gland or related neural centres resulting from distension of the third ventricle.

CASE 2

P.A., a male aged 15 years had been an inpatient of Stallington Hospital since 4 months old. He had been admitted from a local general hospital (City General Hospital) where he was admitted soon after birth, and diagnosed as a case of severe rapidly increasing hydrocephalus, his head circumference having grown by 12.5 cm during two and a half months stay there. The patient was crying day and night, moving his head frequently as if in pain, rubbing it on the pillow and needing frequent sedation. This, together with the psychological effect of the patient's appearance on the parents and the disturbing effect on other siblings, precipitated his admission to Stallington Hospital.

At his birth the labour was protracted, lasting 18 hours; the child had a pressure mark on the back of his head, and his head began to grow rapidly a week after birth. An examination on admission to hospital

FIG. 3. Demonstrates the presence of a large swelling on the back of the head at the junction of the occipital and sagittal sutures.

FIG. 4. Section of the base of the skull showing deeply excavated pituitary fossa.

revealed a healthy child with well-marked deformity of the skull, the head circumference measuring 60 cm. The pupils reacted normally and the fundi were normal. No other abnormality was noted.

Over a period of years the patient developed many complications of hydrocephalus, and a recent examination showed the following features. The patient was severely subnormal intellectually. He was an epileptic, having both major and minor fits since 8 years of age. He had been in status epilepticus a number of times but had responded to treatment.

He was a bed-ridden patient, well developed for his age, and adipose. His appetite had been voracious. He had periods of somnolence, was rather lethargic, and had been complaining of tiredness and headaches (frontal and occipital) off and on. He showed features of precocious puberty, having a heavy growth of axillary and pubic hair since 11 years of age. He weighed 61 kg and his height was 165 cm. After admission to hospital his head circumference had increased to 71.25 cm in four years and remained stationary for the rest of his life. The skull had greatly enlarged both in circumference and height. In the occipital region there was a large soft cystic swelling (Fig. 3). A large defect in the skull bones was palpable at the junction of the occipital and sagittal sutures (the posterior fontanelle was wide open), and the swelling appeared to be part of brain and meninges protruding through this defect; the sac appeared to be adherent to the scalp. A plane radiograph of skull confirmed this. The swelling became more tense at times, and the patient complained of posterior headaches.

He had acneiform rashes on his face. His vision was poor because of marked bilateral corneal scarring. Nystagmus was present and there was evidence of former bilateral tarsorrhaphies. The proptosis of his eyeballs had caused extensive corneal ulceration, resulting in marked scarring. Tarsorrhaphy was done to prevent further damage to the eyes without much success. No papilloedema was noted.

Marked spasticity of all four limbs was noted, the left upper limb being least affected. The patient was bed-ridden as a result. Bilateral brisk deep reflexes, ankle clonus, and extensor plantar reflexes were present.

The urinary 17-ketosteroid excretion was 6.4 mg in 24 hours and excretion of 17-hydroxycorticosteroids was 5.5 mg in 24 hours.

The patient developed terminal bronchopneumonia, and died in three days.

NECROPSY FINDINGS Skull and cranial cavity: the skull was thin. There was a large defect in the skull bones which appeared to be situated at the junction of the occipital and sagittal sutures. This sac was ad-

FIG. 5. Sagittal section of the cerebral hemisphere showing the presence of a large cavity (arrowed) between the tentorium and the cerebellum. This is communicating with the third ventricle through a hole in the posterior wall.

herent to the scalp and was opened inadvertently on reflecting the scalp. From it issued a large quantity of brown fluid. The brain was finally removed in continuity with the cervical cord. No abnormality was found in the cervical vertebrae, and there was no Arnold-Chiari malformation. Examination of the skull base showed that the pituitary fossa was deeply excavated (Fig. 4). The brain had to be fixed for examination but it was noted that the cavity described was either formed by the brain or adherent to it. The opening into the cavity opened into the ventricular system on the left side. The ventricles were enormous and through the hole a view of the entire lateral ventricular system on both sides could be obtained. The whole of the ependyma was stained brown from old blood pigment. Further examination showed that the corpus callosum was raised and stretched to the point of non-existence. There was no trace of the aqueduct in the mid-brain. There was a large fluid-containing cavity between the tentorium and cerebellum (Fig. 5). This communicated with the third ventricle through a hole in the thin posterior wall. It had forced the medulla and cerebellum into the foramen magnum giving rise to spontaneous rupture of the lateral ventricle and attachment to the scalp. A spontaneous third ventriculostium had

formed a large subtentorial sac compressing cerebellum.

Microscopy There were a few ependymal lined clefts only in the aqueductal region. Heavy subependymal haemosiderin deposits were seen in the ventricles. The aqueduct was forked. It was difficult to exclude an inflammatory occlusion after this interval. The adenohypophysis of the pituitary gland appeared normal.

Other organs Both lungs showed a well developed confluent pneumonia in the lower lobes of recent onset. The right lung weighed 700 g, and the left 500 g. The heart weighed 200 g, and was quite normally formed, as were the great vessels. The foramen ovale was closed. The abdominal organs appeared entirely normal, apart from the spleen which was soft and septic in appearance. The liver weighed 1,230 g, the spleen was 180 g, the right kidney was 150 g, and the left kidney was 130 g.

The primary cause of the hydrocephalus was forking or atresia of the aqueduct. A spontaneous third ventriculostium had formed a large subtentorial sac, and had acted as a fluid plug, thereby adding to the already existing obstruction (Pennybacker and

Russell, 1943). The medulla and cerebellum had also been forced into the foramen magnum and this must have further embarrassed the return of the cerebrospinal fluid.

COMMENTS The head circumference had been increasing progressively up to the age of 4, and remained stationary for the last 11 years. Bilateral proptosis of the eyeballs was seen but no papilloedema was noted; the development of marked keratomalacia and corneal ulceration leading to gross corneal scarring had made fundal examination virtually impossible. The presence of a defect in the skull bones situated at the junction of the occipital and sagittal sutures had provided temporary relief in that the increased CSF pressure could force the brain and the meninges into another direction as well. The rupture of the left lateral ventricle leading to the formation of a sac attached to the scalp through the defect was a result.

The precocious puberty, voracious appetite, somnolence, and adiposity denoted disturbance of the pituitary-hypothalamic centres, resulting from the third ventricular distension.

DISCUSSION

Two cases of spontaneous cerebral ventriculostium are described. In case 1, the diagnosis of ventriculostium was made during life by visualizing the arachnoid cavity in the ventriculogram. Only one previous case has been reported so far (Leslie and Alker, 1964). The unique feature of case 2 was the presence of a large defect in the skull bones at the junction of the occipital and sagittal sutures. The left lateral ventricle had

ruptured and formed a sac with its attachment to the scalp through this defect, this being demonstrable clinically as a fixed fluctuant swelling. No such case has been reported previously. Forcing of the medulla and the cerebellum into the foramen magnum had aggravated the hydrocephalus.

My thanks are due to Mr. Harvey Jackson for permission to publish the first case; to Dr. A. J. McCall, consultant pathologist, North Staffordshire Royal Infirmary, for his invaluable help in the preparation of the article, post-mortem findings, and the photographs of the first case; and to Dr. L. Bowcock, Department of Illustration, North Staffs, Hospital Centre, for the photographs of the second case.

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