THE DANDY-WALKER SYNDROME

A REPORT OF THREE CASES

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Most cases of hydrocephalus in infancy and childhood are due to an obstruction; nearly half of these are the result of malformations. The well-known causes of this condition are the Arnold-Chiari malformation, herniation of the cerebellum and medulla through the foramen magnum, often associated with spina bifida or meningomyelocele, and obstruction of the aqueduct of Sylvius — a congenital lesion often aggravated by inflammatory factors. A further well-documented syndrome consists of stenosis of the foramina of the roof of the fourth ventricle, massive dilatation of that chamber, hydrocephalus, and an anomaly of the rostral portion of the vermis. Benda[1] in 1954 proposed the term 'Dandy-Walker syndrome' for this condition. The syndrome is of particular interest to those concerned with the investigation and treatment of hydrocephalus in infancy and childhood, since early definitive operation may obviate the need for the currently-fashionable shunting procedures. This paper concerns the case reports of 3 patients with this syndrome, 2 of which were confirmed at autopsy.

CASE 1

Clinical History

I.G., Coloured male, aged 3 years. Until the age of 2 years and 10 months the patient had been quite well. On 12 February 1959 he was hit on the head with a cricket bat, but did not lose consciousness. He was first seen a week later, complaining of headache dating from the time of the injury. Clinical examination was negative apart from congenital right ptosis. The skull X-ray showed no fracture. On 3 March 1959

headache was still the main complaint, but the mother had noted that in the past 2 weeks the child had become drowsy. On 5 March 1959 he had a convulsion and fell out of bed. The following day he became drowsier and vomited. He had another convulsion, which was probably mainly left-sided on 8 March 1959, the day of his admission.

On examination he was irritable and uncooperative when woken. He had a right ptosis, a left external rectus muscle palsy and mild papilloedema, more marked on the left side. The left plantar response was extensor. X-rays of the skull and chest showed no abnormality. A lumbar puncture showed a normal cerebrospinal-fluid pressure with normal biochemical findings and 8 lymphocytes per c.mm. A pneumo-encephalogram was done. Slight dilatation of the lateral ventricles which were not displaced and a large shadow of air in the posterior fossa over the posterior surface of the cerebellum were not recognized as evidence of the Dandy-Walker syndrome (Figs. 1 and 2).

The sixth-nerve palsy and the mild papilloedema improved rapidly. At the time of his discharge on 18 March 1959 there were no abnormal signs apart from the right ptosis. In view of this clinical improvement a presumptive diagnosis of cortical venous thrombosis with temporary intracranial hypertension, due to trauma, was made.

When seen on 25 March 1959 he had no complaints.

On 6 May 1959 he was readmitted with headache and vomiting and increase in the size of his head. In addition to the previously-noted ptosis he now had bilateral papilloedema and a crackpot sound on skull percussion. X-rays of the skull showed widening of the sutures. On 8 May 1959 a ventriculogram through frontal burrholes showed considerable enlargement of the lateral ventricles and a large air-filled space in the posterior fossa which was noted previously on encephalography (Fig. 5). This air was demonstrated to pass beyond the arch of the atlas (Fig. 6).

Ventricular drainage was established and on 11 May 1959 a myodil ventriculogram showed that 'myodil' flowed into the third ventricle, some remaining in the posterior fossa, whilst drops of myodil were seen high in the posterior fossa. No myodil entered the spinal canal (Figs. 3 and 4).

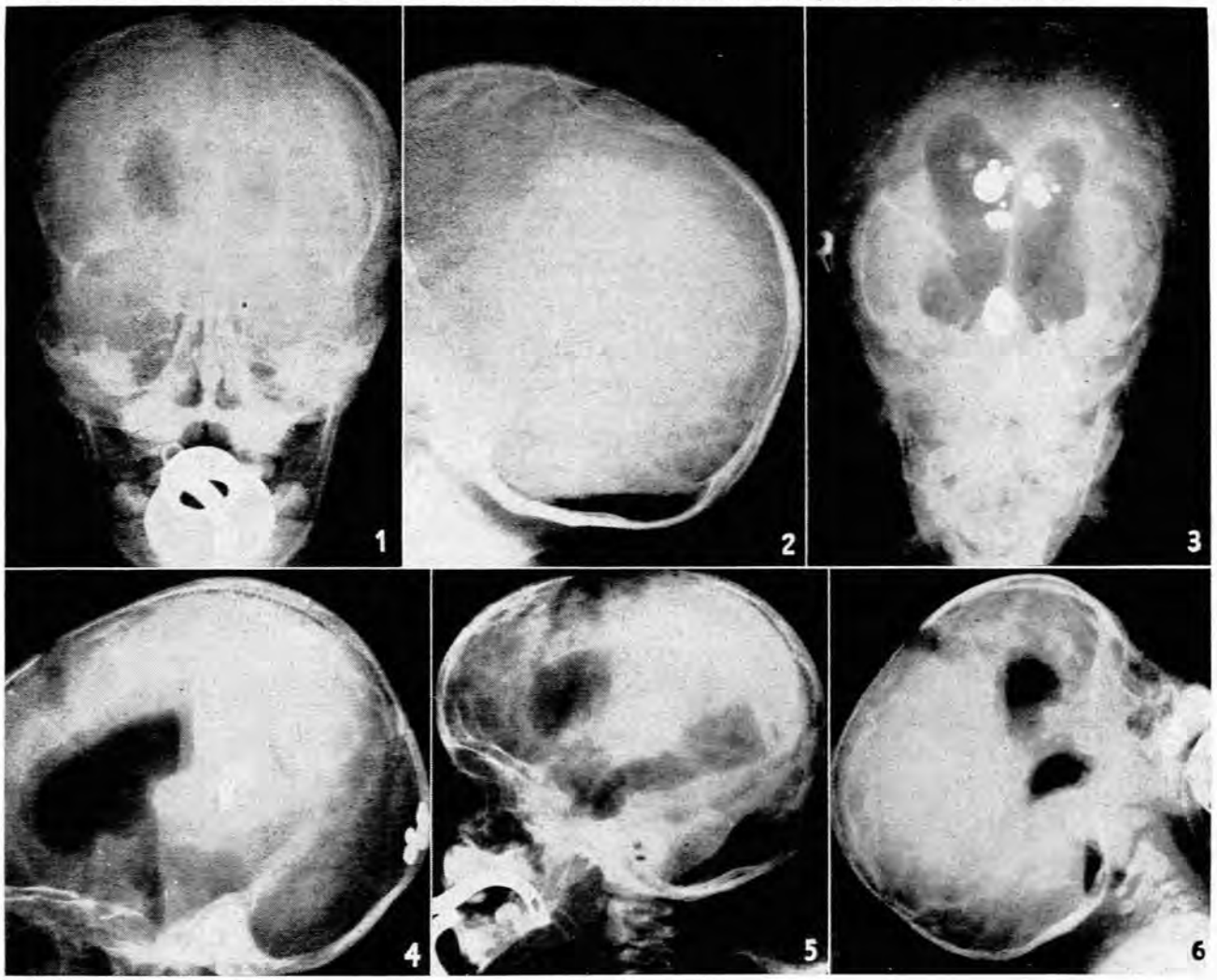

Fig. 1. Case 1. Pneumo-encephalogram; antero-posterior view showing mildly dilated lateral ventricles.
Fig. 2. Case 1. Pneumo-encephalogram; lateral view showing collection of air in the posterior fossa extending higher than usual and representing the large 'cystic' fourth ventricle.
Fig. 3. Case 1. Myodil ventriculogram; antero-posterior view with 30° tilt shows a single globule of myodil in the third ventricle with myodil droplets in the posterior fossa. The latter are in an unusual position in the 'cystic' fourth ventricle.
Fig. 4. Case 1. Myodil ventriculogram; lateral view showing the single globule of myodil in the third ventricle and droplets in the posterior fossa.
Fig. 5. Case 1. Ventriculogram; lateral view showing dilated lateral ventricles with air in the posterior fossa extending beyond the usual limits of the cisterna magna and representing the 'cystic' fourth ventricle.
Fig. 6. Case 1. Ventriculogram; lateral view showing dilated frontal and temporal horns of lateral ventricles, large third ventricle and air—extending beyond the level of the arch of the atlas in the posterior fossa. This is the most inferior part of the 'cystic' fourth ventricle.

A ventriculo-peritoneostomy was performed on 12 May 1959 and the patient was well the following morning. His respirations then became periodic and failed at 4.45 that afternoon. The lateral ventricle was needled, but the patient died.

Postmortem Findings

The body was that of a well-nourished Coloured male child. The head circumference was 48·3 cm.

A left ventriculo-peritoneostomy had been performed, connecting the left lateral ventricle with the peritoneal cavity. The tube was patent.

The lungs were pale and distended with moderate pulmonary oedema. Throughout the interior of both lungs were multiple fresh intrapulmonary haemorrhages up to 2 cm. in diameter.

Apart from congestion of the abdominal viscera the main findings were confined to the brain. The brain weighed 1,264 g. The vault and base of the skull appeared normal. The cranial nerves were all present and intact as far as could be judged macroscopically. The venous sinuses were patent and showed no evidence of thrombosis, old or recent.

After removal of the cerebellar hemispheres from the posterior cranial fossa, an excess of moderately blood-stained cerebrospinal fluid was released. The meninges overlying and surrounding the site of the burrhole transmitting the rubber catheter showed congestion and a slight fibrinous exudate, rather more than would be consistent with the recent operative trauma. This localized exudate was confined to the supratentorial compartment on the left side, and the meninges overlying the rest of the brain surface showed slight thickening and opacity only.

On section of the brain the dilatation of the lateral and third ventricles was confirmed. The aqueduct and foramina of Monro were dilated and the aqueduct measured 0·2 cm. in diameter at its narrowest part. The caudal end of the aqueduct opened into an enormously dilated fourth ventricle.

Viewed posteriorly, the roof of the fourth ventricle (Fig. 7) was represented rostrally by a thin membrane which had been partially torn during removal. On lifting this a cavity which was an abnormally dilated fourth ventricle was seen. The displaced cerebellar hemispheres formed its lateral walls and the floor or anterior wall (medulla) had a relatively normal appearance. Above, the chamber tapered to the dilated aqueduct, through which a thick probe could easily be passed into the third ventricle. Below, the chamber narrowed as the attachment of the membrane approached the midline. The membrane was transparent but showed occasional strands of white tissue running between the lateral recesses. The vermis was at the rostral end of the membrane and measured 5×3×2 cm. Normal arborizations were present. Posteriorly the membrane had been torn through the midline during removal and it was consequently impossible to state whether the foramen of Magendie was patent or not.

The membrane overlying the lateral recesses of the dilated fourth ventricle was slightly thickened and no foramina could be demonstrated.

The cranial nerves of the pons and medulla showed no obvious abnormality. Multiple sections of the left side of the brain and brain stem failed to reveal any focal lesions.

Histological Examination

Sections of the lung confirmed the presence of fresh pulmonary haemorrhages. There was no evidence of infection. Sections of cerebral cortex and midbrain showed no abnormality of note, but the overlying pia-arachnoid was congested and infiltrated by acute inflammatory exudate of moderate degree. No organisms could be demonstrated. The cerebellar hemispheres showed normal structure and arborizations.

The inner or medial surface of the lateral cerebellar hemispheres, which formed the lateral walls of the cystic fourth ventricle, were lined by ependymal cells, which showed non-specific reactive proliferation. The outer layer of the membrane was composed of pia-arachnoid continuous with that covering the cerebellum. Between these two was a layer of connective tissue which varied in density from that of the loose areolar type to dense collagenous fibres, usually in clumps, but occasionally isolated. Glial fibres and cells could be identified amongst these (Fig. 8). No medullated nerve fibres could be demonstrated. The membrane was also the site of a recent acute inflammatory reaction.

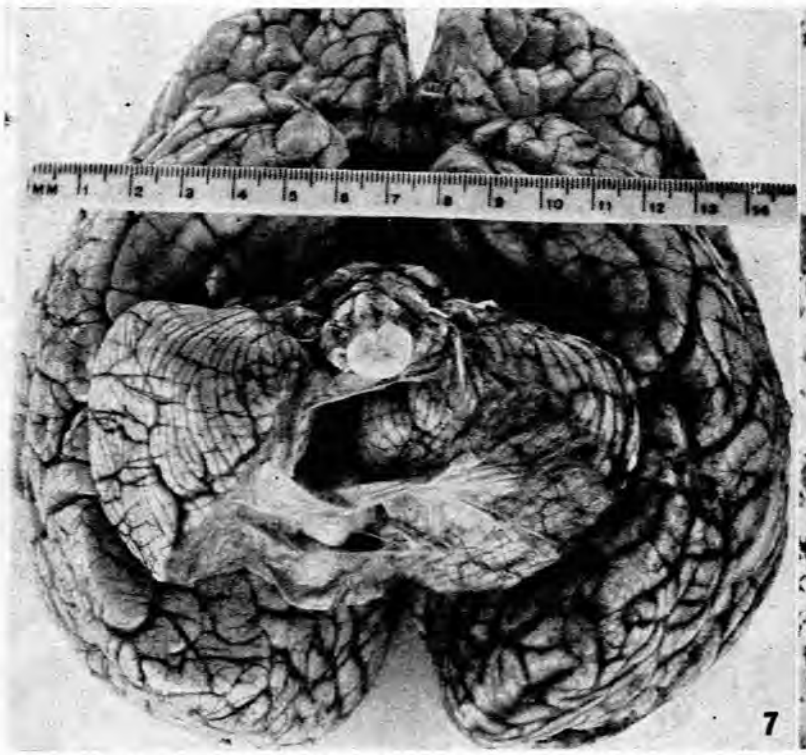

Fig. 7. Case 1. Photograph of the base of the brain showing the dilated fourth ventricle covered by the partly torn membrane.
Fig. 8. Case 1. Photomicrograph (× 100) of the membrane covering the fourth ventricle, showing pia arachnoid above and glial cells and fibres arranged in clumps.

CASE 2

Clinical History

J.D., European male, aged 25 years. This man was born with a large head and shortening of the left arm and leg. Despite this he had led a normal life and worked until November 1959, when, following a mild head injury, he awoke with severe headache. He vomited and was delirious. He was admitted to hospital. A lumbar puncture showed normal fluid.

He was referred to a neurologist because the headache and vomiting had persisted. He had bilateral early papilloedema, but no other abnormal neurological signs. An air encephalogram showed large ventricles.

On 24 December 1959 a posterior fossa exploration showed a large fourth ventricle cyst containing yellowish fluid. The arachnoid over the posterior surface of the ventricle was excised and a ventriculo-cisternostomy performed.

Microscopically the cyst walls showed irregular bands of collagenous tissue with gliosis, not suggesting a parasitic cyst.

Excellent recovery occurred, but on 25 January 1960 he complained of pain in his elbows and inability to feed himself. Examination was negative; his cerebrospinal fluid was normal.

He was kept under observation and became progressively weaker, developing inability to swallow and to cough. It was then found that he had a right foot drop and soon afterwards all reflexes disappeared.

On 5 February 1960 ventriculostomy and drainage was instituted. The cerebrospinal fluid contained 60 mg. per 100 ml. of protein and 18 lymphocytes. His condition deteriorated further, he developed an intercostal paralysis and died on 16 February 1960.

Postmortem Findings

This confirmed the uniformly enlarged head, especially the vault, and the other external features. The lungs were moderately oedematous and had two focal areas of haemorrhage, one well-circumscribed and the other of more recent origin. The liver had focal fatty change, most marked in the left lobe. There was persistence of the foetal lobular pattern in the kidneys and an aberrant renal artery entered the upper pole of the right kidney.

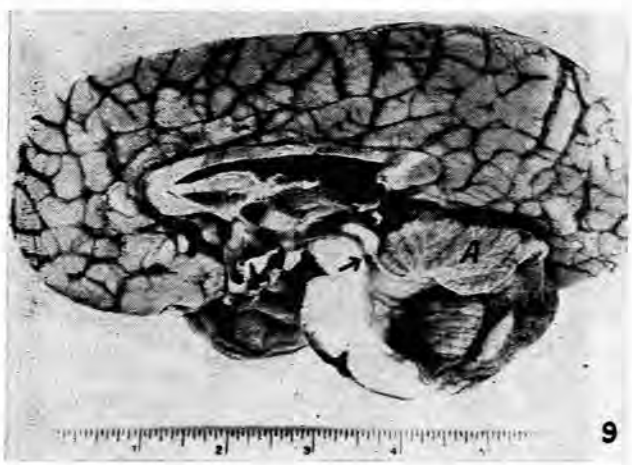

Fig. 9. Case 2. Sagittal section of the brain. Note the generalized hydrocephalus with dilatation of the aqueduct (arrow) and upward displacement of the malformed vermis (A) by the dilated fourth ventricle.

The bones forming the cranial cavity were normal. The meninges were normal except at the operative sites where there were scanty adhesions and some thickening. The brain was large and showed features of a moderately severe hydrocephalus. In particular the floor of the third ventricle was paper-thin and had ruptured during removal of the cranial contents. The fourth ventricle showed enormous cystic dilatation with a roof composed of membrane only. The posterior portion of the vermis was absent and the cerebellar hemispheres were displaced laterally. The features were identical with those of case 1 and will not be repeated in detail (sagittal section — see Fig. 9).

Histological Examination

The histology of the membrane showed the same structure as in case 1. There was a moderate amount of glial tissue in the intermediate layer but no medullated fibres were present. The membrane was oedematous and infiltrated by a scanty acute inflammatory exudate, compatible with a postoperative origin. Sections of the cerebellum were normal.

CASE 3

Clinical History

H.V., European female, aged 7 months. The child was born normally. At the age of 2 months the mother suspected that the child's head was abnormally large. The patient began vomiting and would not hold up her head from 1 week before admission on 24 August 1959.

On examination she appeared hydrocephalic with less frontal bossing and more occipital prominence than usual. The head circumference was 49·6 cm. and the anterior fontanelle was widely open and tense. There were no abnormal neurological signs and the other systems appeared normal on examination.

On 27 August 1959 percutaneous punctures showed lateral ventricles 1·5 cm. from the skin. Twenty-five c.c. of air were injected into each lateral ventricle. On X-ray a symmetrical dilatation of the lateral ventricles with filling of a large posterior fossa 'cyst' extending beyond the usual position of the internal occipital protruberance was shown (Fig. 10).

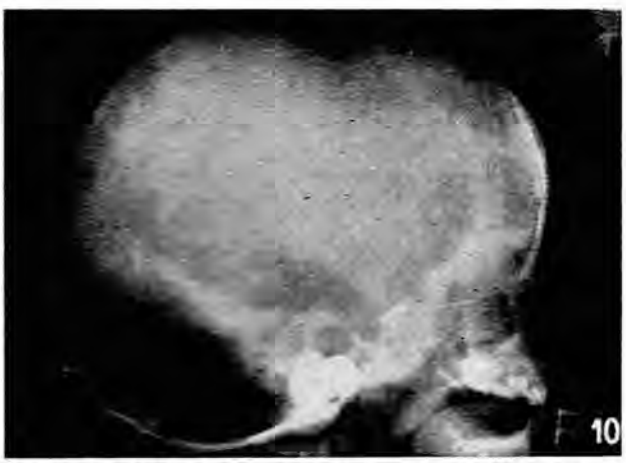

Fig. 10. Case 3. Ventriculogram: lateral views showing the enormous dilatation of the fourth ventricle.

On 4 September 1959 a suboccipital craniectomy was performed. On opening the dura a large cyst was seen. This was the dilated fourth ventricle. At its rostral end, the aqueduct of Sylvius with a diameter of 1 cm. was observed. Rudimentary cerebellar hemispheres, but no cerebellar vermis, were present. On stripping the cyst wall bleeding occurred from misplaced choroid plexus in the right inferior angle. The dura was left open and the wound was closed.

Postoperatively the child made repetitive jerking movements of the left hand and leg. The rectal temperature rose intermittently to 105°F. during the first and third postoperative days. Apart from this and initial vomiting after feeds, the course was uneventful. The patient was discharged on 19 September 1959 on the 18th postoperative day.

On 23 September 1959 her fontanelle was slack and she held up her head better than previously, but the head circumference remained unchanged. She was taking her feeds satisfactorily.

Five months later the patient became ill, vomited frequently, lapsed into coma, and died. Autopsy done elsewhere showed

gross hydrocephalus and confirmed the operative findings. No results of histological examination are available.

DISCUSSION

John Hilton[9] in 1863 in his *Lectures on Rest and Pain* described a case of obstruction at the foramen of Magendie. According to Gibson,[5] the first authentic case was reported by Dandy and Blackfan[3] in 1914. Dandy[4] in 1921 described the operative treatment of the condition. He considered that the condition was due to lack of development of the foramina. This view was held by Taggart and Walker,[13] and histological confirmation of the congenital origin was obtained in 2 of their 3 cases, as Dorothy Russell[12] has pointed out.

Benda[1] proposed the term 'Dandy-Walker syndrome' for the condition, which he considered was a developmental anomaly of the area of the fourth ventricle belonging to the category of cleft formation or rachischisis. He thought that although atresia of the foramen of Magendie may be present it is not the essential feature of the syndrome. However, Gibson[5] presented 2 cases, and maintained that atresia of the foramen of Magendie leads to hydrocephalus in later foetal life and is the crucial maldevelopment in the condition. Both these theories have been disputed by Brodal and Haughlie-Hanssen[2] who have used comparative anatomical methods to show that some factor responsible for the development of the hydrocephalus acts before the foramina of Magendie and Lushka have developed. They maintain that since the vermis of the cerebellum is normally completely developed before the foramina are formed, the causative factor must precede foraminal obstruction in the condition where agenesis of the vermis is invariable. From embryological studies it has been found that the foramen of Magendie commences to form at about the 3½-month stage of foetal development (100 mm. crown-rump length). An evagination of the tela choroidea into the pia-arachnoid forms a diverticulum which subsequently atrophies resulting in a patent foramen between the fourth and fifth months. The development of the foramina of Luschka appear to parallel that of the foramen of Magendie. Hochstetter (1929) (quoted by Brodal and Haughlie-Hanssen[2]) estimated that they appear at a foetal stage of 129 mm. crown-rump length. This author, during the same study, set the date of fusion of the cerebellar hemisphere anlage at a stage of 40 - 45 mm.

The crucial point in the pathogenesis is whether the primary defect is that of atresia of the foramina with subsequent dilatation of the ventricles (a pure mechanical effect), or whether factors operating during foetal life delay or prevent cerebellar development and also interfere with opening of the foramina.

There are reports of at least 1 patent foramen in examples of this condition.[2,5] On the other hand, otherwise normal brains have been described in which the foramen of Magendie or one or both foramina of Luschka were lacking (Alexander, quoted by Brodal and Haughlie-Hanssen[2]). Furthermore, in accepted cases of the Dandy-Walker syndrome, the ventricular system has filled by pneumoencephalography *via* the lumbar route and dyes have passed through from the ventricles into the subarachnoid space so that one cannot but suppose that at least 1 foramen has been patent. Matson,[11] and Taggart and Walker,[13] however, found that on occasions dye, but not air, passed through the membrane. They attributed this to dialysis rather than direct communication.

Gardner *et al.*[6,7] claim that failure of outlets of the fourth ventricle to develop in the rhombic roof of the embryo is the cause of the Dandy-Walker syndrome, the Arnold-Chiari malformation, syringomyelia and certain 'arachnoid cysts' of the cerebellum. If this seems to be rather an extravagant theory, so too seems Brodal and Haughlie-Hanssen's theory that the cerebellar malformation of the Dandy-Walker syndrome and the foraminal atresia are due to some unknown factor causing hydrocephalus before the cerebellum has developed fully. The latter theory could be invoked where the cerebellar development is least advanced; it fits in with embryological studies showing that cerebellar development is complete before the foramina open. However, histologically verified remnants of cerebellar tissue in the roof membrane of the dilated fourth ventricle show changes suggesting that the malformation of the cerebellum is due to secondary hypoplasia.[8] We agree with Gibson that atresia of the foramina is the primary cause of the Dandy-Walker syndrome.

Brodal and Haughlie-Hanssen were able to record 30 human cases to which they added 2 of their own. In spite of the relatively few cases reported, there is evidence that the condition is not as rare as this suggests. Laurence[10] reported autopsy findings on 100 cases of hydrocephalus of which 7 were examples of obstruction at the outlet foramina of the fourth ventricle, 12 were cases of Arnold-Chiari malformation, and 48 showed multiple sites of obstruction. Benda[1] found 6 cases of Dandy-Walker syndrome in his last 13 autopsies on hydrocephalic children, 3 of which had Arnold-Chiari malformations. It would seem therefore that the Dandy-Walker syndrome is nearly as common as the Arnold-Chiari malformation.

The brain lesions in our 3 cases do not differ in any detail from the condition under discussion. From the descriptions in the literature, and our own experience, the malformation is fairly uniform in all cases and the morbid anatomical features are unlikely to be confused with those of any other condition. The degree to which the cerebellum is affected varies from case to case. The vermis is always reduced in size and sometimes absent.

Histological examination of the membrane bridging the cerebellar hemispheres has yielded interesting information. The layer between the ependyma and the arachnoid has received particular attention. This space contains predominantly connective tissue, varying from the loose type to dense collagenous fibres. Islands or plates of glial tissue have often been described and, in addition, nerve fibres have been seen. Glial tissue may be separated from the lateral cerebellar hemispheres by 2 - 3 cm., or fibres may form a thin layer reflected from the line of attachment of the membrane passing on the medial surface of the hemispheres to enter their white matter (Gibson ;[5] case 1). These plaques of white matter may be identified in the membrane macroscopically. Traced by serial section, glial fibres are found to be continuous with the rostral attenuated end of the vermis and its nodulus. These observations afford presumptive evidence that the nature of the defective cerebellar structure is secondary rather than primary aplasia.

Clinically the fact that some patients have no signs or symptoms for years (Cases 1 and 2) suggests partial patency of the foramina. We have evidence of ventricular filling by pneumo-encephalography in 2 cases. The late onset of symptoms may be related to some inflammatory process, infective or traumatic, causing complete foraminal closure, as suggested by Laurence,[10] in whose 46 cases with some congenital lesion 32 showed evidence of inflammation of either traumatic or infective origin. However, the inflammatory cellular infiltration observed in our 2 autopsy cases could be ascribed to operative trauma or mild postoperative infection.

The clinical features with both early and late presentation are illustrated in our cases. The third case showed occipital enlargement of the head typical in this form of internal hydrocephalus in infants. The first case illustrated the lack of cerebellar signs characteristic in older patients even when the hydrocephalus is causing severe symptoms of raised intracranial pressure.

X-rays of the skull may be diagnostic when the bulging and thinning of the occipital part of the skull, the high torcula, the obliquity of the transverse sinus, and the greater separation of the lambdoid than other sutures, are seen.[11]

Ventriculography[13] will show air in the posterior fossa under the characteristically high tentorium cerebelli, the large fourth ventricle, huge aqueduct, and symmetrical dilatation of the lateral and third ventricles. Matson[11] noted projection of the cisterna magna to the level of the second cervical laminae and Fig. 6 demonstrates this. He advised views with the brow slightly up or with the occiput slightly up to show these features. We have found the 'backward somersault' used by Ziedses des Plantes[15] most useful in the elucidation of the site of obstruction.

Once it is appreciated that in this condition myodil rapidly passes through the dilated aqueduct of Sylvius into the large fourth ventricle, as demonstrated in our first case, myodil ventriculography can be used to clinch the diagnosis.

Pneumo-encephalographic demonstration of the lesion may be possible if the foramina of Magendie and Luschka are sufficiently patent, as we have shown.

The characteristic high torcula may be demonstrated by dural sinography as Matson[11] has shown.

Treatment of the condition should be operative. Satisfactory results with unroofing of the enormously dilated fourth ventricle have been described. Matson,[11] who gives the largest single series, notes 8 cases of which 3 did well with nothing more than wide excision of the cyst wall; 4 required additional shunting procedures, and 1 died 2 months after operation. At operation the caudal and lateral parts of the membrane must be removed, the abnormal situation of the choroid plexus in this region should be remembered to prevent haemorrhage. Ventriculo-atriostomy should be preferred when the patient's condition does not warrant the more extensive operation for removal of the membrane or when the latter is followed by recurrence of symptoms.

We believe that further investigations of hydrocephalus will reveal a much higher incidence of this condition.

SUMMARY

Three cases of Dandy-Walker syndrome are presented. A review of the literature with a critical account of the theories of origin is given. It is concluded that maldevelopment of the foramina of Luschka and Magendie is the crucial pathogenic lesion. The pathological, clinical, diagnostic, and therapeutic aspects are discussed. An assessment of the incidence of this cause of hydrocephalus is given.

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