Dandy-Walker Syndrome

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SUMMARY
The problem of a patient with hydrocephalus, a posterior fossa 'cyst' and a cerebellar malformation is rather rare. Four cases of the syndrome are presented. The clinical manifestations, pathology, radiological investigations and management of the condition are briefly discussed.

The Dandy-Walker syndrome, an eponym introduced by Benda, is an embryological abnormality. It is characterized by an anomaly of the cerebellum, associated with hydrocephalic dilatation of the lateral, 3rd and 4th ventricles.

CASE REPORTS

Case 1
A premature male infant presented at the age of 1 month with an enlarging head. Lumbar air encephalography was performed and dilated lateral ventricles were demonstrated, but the 3rd and 4th ventricles were not visualized. Dimer-X ventriculography was then performed for further elucidation, and this demonstrated dilatation of the lateral and 3rd ventricles, and a cyst occupying the whole of the posterior fossa. The cerebellum and the 4th ventricle were not visualized. The posterior fossa was explored through a midline incision, and a large 'cyst' occupying the whole of the posterior fossa was found. As much as possible was excised, but numerous large venous channels hindered excision. The 'cerebellar hemispheres' could be identified as small nodules, about 8 mm in diameter, lying adjacent to the midbrain structures. The child made an uneventful recovery and had no neurological deficit. The posterior decompression area then began to swell. It was decided to insert a ventricular shunt for treatment of the hydrocephalus, but the child died suddenly before this could be carried out. A postmortem examination confirmed the diagnosis of Dandy-Walker syndrome (see discussion).

No neural tissue was found in the cyst on histological examination. The tissue adjacent to the midbrain contained no tissue resembling cerebellum.

Case 2
A 2-week-old male infant was referred because he had a progressively enlarging head. Lumbar air encephalography was performed and dilated lateral ventricles were demonstrated, but the 3rd and 4th ventricles were not visualized. Dimer-X ventriculography was then performed for further elucidation, and this demonstrated dilatation of the lateral and 3rd ventricles, and a cyst occupying the whole of the posterior fossa. The cerebellum and the 4th ventricle were not visualized. The posterior fossa was explored through a midline incision, and a large 'cyst' occupying the whole of the posterior fossa was found. As much as possible was excised, but numerous large venous channels hindered excision. The 'cerebellar hemispheres' could be identified as small nodules, about 8 mm in diameter, lying adjacent to the midbrain structures. The child made an uneventful recovery and had no neurological deficit. The posterior decompression area then began to swell. It was decided to insert a ventricular shunt for treatment of the hydrocephalus, but the child died suddenly before this could be carried out. A postmortem examination confirmed the diagnosis of Dandy-Walker syndrome (see discussion).

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Case 3
A child aged 2½ months was brought to hospital because of vomiting and restlessness. She had a minor cleft palate, tended to retract her neck, and had a bulging fontanelle and a rapidly enlarging head. Skull radiographs showed splayed sutures. Ventriculography with Dimer-X³ and Myodil demonstrated large ventricles, but no contrast medium passed from the 3rd ventricle into the aqueduct. A tentative diagnosis of aqueductal stenosis was made, and a ventriculoperitoneal shunt was inserted. The cleft palate was repaired 2 months later. The child was examined again at the age of 2 years. After an upper respiratory
tract infection, she became irritable and tended to retract her neck when stimulated. The shunt appeared to be functioning. The lumbar pressure was 130 mm CSF, but she was restless during the investigation. Computer-assisted tomography demonstrated a posterior fossa cyst and enlarged ventricles (Fig. 2). The child suddenly collapsed and died a few hours after having been transferred to the Neurosurgical Unit, before any further investigations could be carried out. Postmortem examination demonstrated the classic features of the anomaly (see discussion), and the shunt appeared to have been functioning.

Fig. 2. Computer-assisted tomogram of case 3, demonstrating ventricular dilatation and the position of the ventricular catheter in the posterior horn of the right lateral ventricle. The fluid-filled dilated 4th ventricle is well demonstrated.

Case 4

A 10-year-old boy was referred to the Neurosurgical Department with a septic ventriculoperitoneal shunt. He had been hydrocephalic for many years. He had been given a shunt as an infant, and the shunt had had to be revised on numerous occasions. Computerized axial tomography revealed a posterior fossa cyst. Because of the sepsis involving the peritoneal end of the shunt, it was decided to remove the shunt tubing, but the child died suddenly a few hours later. Postmortem examination confirmed the diagnosis of Dandy-Walker syndrome. A 4-cm plastic tube was found embedded in the right hemisphere, near the roof of the inferior horn. There was a small abscess surrounding the embedded tube, a remnant of a previous shunt revision.

DISCUSSION

Various theories on the aetiology of the condition have been proposed. Taggart and Walker believe that the condition is caused by failure of the foramina of Luschka and Magendie to appear, causing dilatation of the 4th ventricle (commonly, but erroneously, referred to as a 'cyst'). Brodal and Hauglie-Hanssen suggest that increased intraventricular pressure of unknown cause produces the changes. They state that the anomalies arise considerably before formation of the foramina. Benda agrees that foraminal atresia is only part of the syndrome, and is not always present.

The major abnormality appears to be in the development of the rhombencephalic roof, predominantly the posterior portion of the vermis. It may be absent, attenuated, or heterotopic and grossly abnormal. This is associated with maldevelopment of the adjacent choroid plexus. The cerebellar hemispheres may be separated widely, or reduced to small nodules lying anteriorly against the petrous bone. The tentorium cerebelli is situated in an abnormally high position, probably because the posterior migration of the transverse and confluent sinuses is prevented.

The child may be stillborn or may die shortly after birth. In survivors, there may be signs and symptoms of raised intracranial pressure. Occasionally, the symptoms are intermittent. Patients may live for many years without problems, but it appears that at some stage cerebrospinal fluid circulation is impaired, and sequelae develop. There may be no cerebellar signs. A fairly high proportion of these patients are mentally retarded. There may be additional congenital abnormalities involving the central nervous system and other systems. A familial incidence has also been reported. The only clinical finding which may differentiate these cases from other cases of hydrocephalus may be a prominent posterior fossa 'shelf', or elongated occiput, but this is not often present. An abnormally high situation of the transverse sinus marking on the skull radiograph, indicating a raised tentorium cerebelli, should arouse suspicion of the condition. This finding, however, may not be present, and, in addition, may not be visible until the child is almost, or more than, 2 years old.

The diagnosis can be made by lumbar air encephalography, provided a communication exists, which appears to be the case in 30% of these patients. Ventriculography is the alternative form of investigation.

With both procedures, a large posterior fossa 'cyst' can be demonstrated, with probable dilatation of the lateral and 3rd ventricles. (The latter finding is not always present.)

Recently, computer-assisted tomography has aided diagnosis. A large midline defect is demonstrated in the cerebellum. A large, low-density 'cyst' occupies most of the posterior fossa, and the cerebellum may be identified lying against the petrous bones. The posterior fossa is visualized on much higher cuts than normal because of the abnormally high situation of the tentorium cerebelli.

Angiography can be used to demonstrate the abnormality. The vermian branches are seldom present and other vessels, apart from the anterior inferior cerebellar artery which supplies the remaining cerebellum, may be abnormal.

The use of cisternography and radio-isotope studies to determine cerebrospinal fluid flow has been suggested as another method of investigation.

The management of the Dandy-Walker syndrome remains controversial; opinions are divided, some favouring direct surgical attack on the dilated 4th ventricle, and
some believing in cerebrospinal fluid diversionary procedures. The treatment of choice seems to be shunting of the lateral ventricles. In the majority of cases there is free communication between the dilated 4th ventricle and the rest of the ventricular system. In the case of normal lateral and 3rd ventricles, the dilated 4th ventricle alone can be shunted. Combined shunts are suggested when a lack of communication is suspected or demonstrated, e.g. when there is an associated aqueductal stenosis.

The best surgical results obtained to date are those of Carmel et al. who reported a mortality of 27%. Most other series, as evidenced by our 4 cases, have a bleak outcome.

REFERENCES

Metastatic Hepatocellular Carcinoma of the Heart
A Case Report

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SUMMARY

A metastatic tumour growing in the heart more than 2 years after total removal of the primary hepatic growth is a rare event. Jaundice as a result of bile production by the metastasis is most unusual. The primary tumour did not recur in the regenerated liver. Features which may prove useful in making a clinical diagnosis are briefly considered. Death resulted from a thrombo-embolus from the right atrium lodging in the opening of the tricuspid valve.


Metastatic cardiac tumours are not often diagnosed clinically and are infrequently seen at necropsy. In this article we report a case of metastatic hepatocellular carcinoma growing in the right ventricle of the heart. The primary liver tumour had been excised 2½ years previ

vously, and no recurrence was evident at postmortem examination. The cardiac metastasis and the pulmonary emboli from this source produced a large amount of bile, resulting in icterus. We are not aware of a similar case in the literature.

CASE REPORT

A 59-year-old Black man was admitted to hospital on 9 February 1977 with complaints of progressive dyspnoea on exertion for about 3 months, swelling of the lower legs, and progressive, painful distension of the abdomen. In 1974 a partial right-sided hepatectomy had been performed for a well-circumscribed liver cell carcinoma. A cholecystectomy had been done simultaneously.

On examination the patient was found to be fairly well nourished. He was slightly jaundiced, and had signs of right-sided heart failure. The left lobe of the liver was palpable and no right lobe was evident. The old operation scar was unremarkable.

Laboratory investigations showed a haemoglobin concentration of 14.9 g/100 ml; a red blood cell count of 4.53 million/µl; a white cell count of 4,400/µl with a differential leucocyte count of neutrophils 51%, lymphocytes 37%, monocytes 9%, eosinophils 2% and basophils 1%. The erythrocyte sedimentation rate was 8 mm/1st h (Westergren); total protein 70 g/l, albumin 31 g/l and

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