Hydrocephalus due to membranous obstruction of the fourth ventricle

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Four patients with hydrocephalus due to membranous obstruction of the fourth ventricle are presented. This rare entity produced radiographic and clinical findings suggestive of posterior fossa tumor. Operative findings included normal cerebellar development and a translucent membrane just above the foramen of Magendie. Etiological possibilities are discussed.

KEY WORDS hydrocephalus membrane fourth ventricle

M embranous obstruction of the fourth ventricle is a rare cause of hydrocephalus. It does not appear to be a partial or abortive Dandy-Walker syndrome, which is characterized by atresia of the foramina of Lushka and Magendie and hypoplasia or absence of the cerebellar vermis. The lesion we are describing is a thin, usually transparent membrane within the fourth ventricle that partially or completely obstructs the flow of cerebrospinal fluid. There is no associated abnormality of the cerebellum. Leptomeningeal inflammation is not seen in the basal cisterns. The membrane is similar in appearance to the web occasionally found at the lower end of the aqueduct of Sylvius.

Only one description of this entity has been found in the English literature.1 We are reporting four examples of membranous obstruction of the fourth ventricle, all verified at operation.

Case Reports

Case 1

A 13-year-old girl was admitted with a 3-year history of progressive ataxic gait accompanied by frontal headache. The parents complained that she was a behavior problem. On examination, the patient was premenarchal, of short stature, with a head circumference of 57 cm (90th percentile). She had bilateral papilledema. There was bilateral dysdiadochokinesia, and left brachial drift. The deep tendon reflexes were exaggerated on the left. She demonstrated ataxia of gait and stance. On the skull films, early suture separation, prominent digital markings, and posterior clinoid erosion were seen. The echoencephalogram (EEG) showed mild, diffuse slowing. Cerebrospinal fluid protein was 9.7 mg%. At ventriculography (Fig. 1), there was gross, symmetrical hydrocephalus above a very large fourth ventricle which was normal in position. No air passed beyond the fourth. At operation the cisterna magna was small and the cerebellum appeared normal. Upon separation of the tonsils, a membrane was found occluding the foramen of Magendie. Wide excision of the membrane established free communication with the subarachnoid space. Postoperatively, the papilledema resolved, the headache and vomiting disappeared, but her
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standing and gait ataxias improved only minimally. Free CSF communication was proven by a phenolsulfonphthalein (PSP) dye test. Histologically, the membrane showed a fibrillar matrix containing a few red blood cells and many fibroblasts. There were no inflammatory cells present. There was no evidence of neoplasia. The family moved to another part of the country, where the girl died 18 months after operation.

Case 2

Between the age of 6 weeks and 3 months, this girl had five tonic seizures, each lasting 1 minute. When she was admitted at 5 months of age, the anterior fontanel was tense with a head circumference of 43.7 cm (70th percentile). The perinatal history was benign. Ventriculography showed severe dilatation of the entire ventricular system (Fig. 2). No air passed out of the fourth ventricle. Protein concentration in the ventricular fluid was 16 mg%. The operative findings included a moderately large cisterna magna containing clear CSF, no evidence of previous infection or hemorrhage, and a thin membrane just above the foramen of Magendie. An opening 1 cm in diameter was created in the obstructing membrane. Free communication between the ventricles and the spinal subarachnoid space was proven by PSP dye tests, but inordinate head growth continued. Exploration of the posterior fossa revealed cohesion of the cerebellar tonsils and adhesive obliteration of the foramen of Magendie. A ventriculo-cervical subarachnoid shunt failed to provide adequate drainage of CSF. Two weeks later a lumbar subarachnoid-ureteral shunt was done. She did well for 3½ years. Then an acute episode of vomiting produced severe dehydration. This responded well to intravenous salt replacement, but streptococcus fecalis meningitis intervened. In spite of removal of the shunts and institution of continuous ventricular drainage, the patient died in September, 1958.

Case 3

An 8-year-old boy was admitted with a 2-month history of generalized, throbbing headache. He had always been poorly coordinated, but had become worse in the previous month. School performance had deteriorated. There was bilateral papilledema, a mild left facial crural hyperreflexia with bilaterally extensor plantar responses, a positive Romberg sign, gait ataxia, and lateralized cerebellar signs to the left. Plain skull films showed enlargement of the sella turcica, craniomegaly, and absence of the posterior clinoid processes. The CSF protein (ventricular) was 8 mg%. A ventriculogram was done (Fig. 3). The fourth ventricle was large and appeared high, with no air passing beyond it. The presumptive diagnosis was fourth ventricular tumor. Posterior fossa craniectomy revealed a transparent membrane
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FIG. 3. Case 3. Air ventriculogram, lateral brow-down view. The aqueduct and fourth ventricle are huge. There is no air in the lower fourth ventricle. Note that the convex borders of the fourth ventricular shadow are everywhere directed away from the lumen (arrows).

Two weeks prior to admission, she witnessed a minor accident in which a friend was knocked down by a car. As she bent over her friend, she suddenly collapsed and lost consciousness for several seconds. Following arousal, she complained of the most severe headache she had yet experienced, and of neck stiffness. A lumbar puncture the next day produced slightly xanthochromic CSF, and skull films were normal. Hydrocephalus was evident on carotid angiography. The EEG showed general slowing, more pronounced occipitally. The neurological examination was normal on admission, as was a vertebral angiogram. A Pantopaque-air ventriculogram (Fig. 5) appeared to show a lobulated mass totally obstructing the lower fourth ventricle. Because of the presumptive evidence of subarachnoid hemorrhage the preoperative diagnosis was choroid plexus papilloma. At operation, a translucent membrane was found 7 mm above the foramen of Magendie totally obstructing the lower fourth ventricle. Saline put into the ventricular catheter above would ooze slowly out of

(Fig. 4), stretched across the lower fourth ventricle, 1 cm above the foramen of Magendie. Both the membrane and the inferior vermis were divided. Near the aqueduct was another transverse membrane containing two small holes; this membrane was also incised. Catheter exploration of the aqueduct encountered no further obstruction. His postoperative course was complicated by coma and decerebrate rigidity for 7 days, and inadequate CSF circulation. A ventriculoatrial shunt was done. Two years later he is doing well in school, his coordination is intact, and his neurological examination is normal.

Case 4

This 21-year-old married woman had had severe, periodic frontal headache for 2 years. The pain radiated occipitally, and was frequently associated with nausea and vomiting. Headaches occurred several times each week, at any time of the day. Then she enjoyed a 1-year interval of freedom from headache. Four weeks prior to admission the symptoms returned, associated with nausea, vomiting, and low-grade fever. Two weeks prior to admission, she witnessed a minor accident in which a friend was knocked down by a car. As she bent over her friend, she suddenly collapsed and lost consciousness for several seconds. Following arousal, she complained of the most severe headache she had yet experienced, and of neck stiffness. A lumbar puncture the next day produced slightly xanthochromic CSF, and skull films were normal. Hydrocephalus was evident on carotid angiography. The EEG showed general slowing, more pronounced occipitally. The neurological examination was normal on admission, as was a vertebral angiogram. A Pantopaque-air ventriculogram (Fig. 5) appeared to show a lobulated mass totally obstructing the lower fourth ventricle. Because of the presumptive evidence of subarachnoid hemorrhage the preoperative diagnosis was choroid plexus papilloma. At operation, a translucent membrane was found 7 mm above the foramen of Magendie totally obstructing the lower fourth ventricle. Saline put into the ventricular catheter above would ooze slowly out of

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the foramina of Luschka, but would not pass through the web. The membrane was torn, and CSF gushed into the wound. The cerebellar vermis, the aqueduct, and the cisterna magna were normal. Lest postoperative adhesions compromise CSF flow, a Silastic tube was left across the torn membrane and so secured to the dura that its lower end lay in the cisterna magna. The postoperative course was benign, and the patient remains perfectly normal and free from headache 1 year after surgery.

Discussion

In all cases, the preoperative diagnosis from clinical and radiological data was posterior fossa tumor. Duration of preoperative complaints was 3 years, 5 months, 2 months, and 3 years respectively. In no case could we find historical events to explain the presence of the membranes. The one microscopic section available (Fig. 4) provides no clue to etiology, although the presence of abundant mature fibroblasts might be interpreted as evidence of previous inflammation. The radiological and operative findings preclude the diagnosis of Dandy-Walker syndrome. The 18 years prior to onset of symptoms in Case 4 is difficult to reconcile with a congenital etiology unless, as suggested by Holland and Graham, a subcritical reduction in normal CSF flow such as may occur with aqueductal stenosis. This situation might be compatible with delayed symptomatology. Certainly, the ventricular sizes in Cases 1 and 3 suggest longstanding obstruction. Case 2 illustrates that the obstruction may become critical within the first few months of life. Turnbull and Drake felt that membranes within the aqueduct were postinflammatory, the attenuated remnants of gliotic aqueductal overgrowth. In the case of Holland and Graham, a congenital etiology was assumed in the absence of any evidence of neoplasm or inflammation.

Membranous obstruction of the fourth ventricle produces no definitive clinical syndrome. The ventriculogram may suggest an ependymoma arising from the floor of the lower fourth ventricle, a low midline cyst, or a tumor of the lower vermis. The possibility of membranous obstruction of the fourth ventricle should be kept in mind when the ventriculogram indicates a sharp cut-off of the lower end of the fourth ventricle, especially when the obstruction presents a convex surface away from the enlarged fourth ventricle.

Even if the correct diagnosis is suspected preoperatively, we feel strongly that posterior fossa exploration should be done to rule out a mass lesion. The membrane then should be widely opened. Our experience indicates that these patients will usually re-
quire a shunting procedure. This may be accomplished at the initial operation by fourth ventriculocisternostomy with a Silastic tube fixed at one point to the dura. With any evidence of basal cisternal obstruction, a ventriculoatrial or ventriculoperitoneal shunt will be necessary. Where operative manipulation has been minimal, and basal obstruction is absent, it might seem reasonable to defer a shunting procedure.

References


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