

Congenital fourth ventricular midline outlet obstruction

Report of two cases

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Obstruction of the foramen of Magendie unrelated to illness or trauma is rare in adults. Two cases of congenital membranous occlusion of the foramen of Magendie in adults are presented. Analysis of the reported cases of fourth ventricular outlet obstruction disclosed only three similar cases.

KEY WORDS • fourth ventricle • foramen of Magendie • obstruction • hydrocephalus

Fourth ventricular outlet obstruction may be congenital or acquired. Congenital occlusive lesions occur primarily in children and are the subject of many reports on Dandy-Walker cysts. Acquired ventricular outlet obstructions are reported in adults as well as children. The present paper describes two adult patients with congenital occlusion of the foramen of Magendie causing massive dilatation of the fourth ventricle and hydrocephalus.

Case Reports

Case 1

This 42-year-old South American man presented with a 1-year history of right-sided dysesthesias and hemiparesis which worsened when he stood up. He was evaluated for a right homonymous hemianopsia. A lumbar puncture performed to study the cerebrospinal fluid (CSF) was complicated by headache, vomiting, and blurred vision. He was then admitted to the Mount Sinai Medical Center in New York City.

The patient's history was significant for giardiasis and trichuriasis in childhood. General physical examination was normal, but neurological evaluation revealed bilateral papilledema. His mental status was intact. The patient's gait was ataxic, and there was mild right hemiparesis. Work-up for inflammatory or neoplastic disease was negative. Skull x-ray films were normal. Computerized tomography (CT) revealed an enlarged fourth ventricle with no associated masses. Ventriculography with metrizamide enhancement (4 cc, 190 mg/cc iodine) was performed via direct puncture of the right frontal horn utilizing the technique described by Rifkinson, et al.; this showed a disproportionately dilated fourth ventricle and hydrocephalus (Fig. 1). With the ventricular needle in place, a spinal tap was carried out with the patient in the sitting position, and 5 to 7 cc of air was injected. The inferior border of the obstruction to the foramen of Magendie was outlined. The CT scans after administration of metrizamide for ventriculography confirmed the massive size of the fourth ventricle and the presence of hydrocephalus.

With the patient again in the sitting position, midline suboccipital craniectomy with C-1 laminectomy was performed (by V.P.S.). Under the operating microscope, normal-appearing arachnoid mater of the cisterna magna was incised. Cerebrospinal fluid was present in the cisterna magna, and the cerebellum was normal. The tonsils were separated easily, revealing a transparent membrane without evidence of inflammation in the area of the foramen of Magendie. No CSF outflow was seen. The membrane, apparently an extension of the inferior medullary velum, was excised, resulting in a gush of clear, colorless CSF from the ventricle. There was no intraventricular pathology. The arachnoid of the cisterna magna was closed with 10-0 Neurolon.

Membrane histology was consistent with neural tissue and gliosis, without inflammation. Studies of the CSF were normal. The patient's headache and vomiting disappeared, and his papilledema resolved. Strength and ambulation returned to normal. Yearly follow-up CT
FIG. 1. Case 1. Metrizamide-enhanced ventriculogram, lateral view, showing a trapped fourth ventricle. The procedure was performed with the patient seated. Ballooning of the fourth ventricle (4V, arrowheads), with dilated lateral recesses and bulging of the anterior medullary velum (AM), is evident. The inferior portion of the dilated fourth ventricle (crossed arrow) is below the level of the foramen magnum (F). The aqueduct (Aq) is enlarged and the third ventricle (3V) is dilated, bulging into the interpeduncular and chiasmatic cisterns. The optic recess and chiasm (white arrow), the infundibular recess (I), and the mamillary eminence (M) outline the anteroinferior portion of its floor. The tuber cinereum (T) is seen bulging downward. The indentations within the lamina terminalis are caused by the anterior communicating vein (V) and anterior communicating artery (A), both of which are anteroinferiorly displaced. Due to leakage of contrast medium from the suprapineal recess, the ambient cistern (AC) is opacified. Some metrizamide appears to have seeped through the membranous portion of the foramen of Magendie to collect within the cisterna magna. The pineal gland (Pi) is outlined.

FIG. 2. Case 2. Transaxial computerized tomography scan following metrizamide ventriculography. This section is through the enlarged fourth ventricle (4V) 25 mm above the foramen magnum. Since the cut is high, the true size of the dilated lateral recesses (LR) is not appreciated. Radiopaque material is seen in both cerebellopontine cisterns (CP).

scans have been normal, and the patient remains neurologically intact.

Case 2

This 52-year-old man presented with a 10-month history of gait disturbance with a tendency to veer to the right. He developed severe morning headaches associated with decreasing vision and vomiting. His medical history was noncontributory. General physical examination was normal. Neurological examination revealed the patient to be alert and oriented but with slow speech and formulation of ideas. Fundi were normal. There was right upper-extremity dystaxia. The patient's gait was wide-based and he tended to fall to the right. Sensory examination and motor strength were normal. Workup for inflammatory or neoplastic disease was negative. The CT scans showed a nonenhancing, hypodense cystic lesion that was suspected of being an enlarged fourth ventricle. Moderate hydrocephalus of the lateral and third ventricles was seen. Angiography suggested a large avascular posterior fossa mass, but it was unclear if this represented a cyst or a dilated fourth ventricle. Metrizamide ventriculography via direct puncture of the right frontal horn showed a huge fourth ventricle, with small amounts of metrizamide in the upper cervical region, seen also on CT (Figs. 2 and 3).

Midline suboccipital craniectomy was performed (by V.P.S.). Under the operating microscope, the tonsils and vermis appeared normal. The cisterna magna was well formed with no sign of inflammation. The tonsils were loosely apposed and separated easily to expose the vallecula, which was empty of CSF. The midline area was covered by a semidiaphanous sheet, an extension of the inferior medullary velum. Through it, normal-appearing choroid plexus and CSF were seen. The membrane was excised and clear CSF flowed from the ventricle. There was no intraventricular pathology. The arachnoid of the cisterna magna was closed with 10-0 Neurolon.

Histology of the choroid and ependyma was normal. The membrane showed areas of hyalinized connective tissue and calcific deposits without inflammation. Studies of the CSF were normal. The patient's postoperative recovery was uneventful and he remains neurologically intact.
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Fig. 3. Case 2. Left: Sagittal computerized tomography (CT) reconstruction of the upper cervical canal and lower posterior fossa following metrizamide ventriculography. The fourth ventricle (4V) is dilated and the tonsils (T) are displaced almost to C-1. Metrizamide is seen in the cisterna magna, in the upper cervical subarachnoid space (S), and in the medullary cistern (MC) anterior to the medulla (M). The pons (P) is flattened and displaced anteriorly, almost obliterating the pontine cistern (PC). Right: Coronal CT reconstruction through the dilated fourth ventricle (4V) and lateral recesses (LR) following metrizamide ventriculography. The opacified subarachnoid space (S) is anterior to the cervical cord (C), and the region of the obex is shown by the arrow. The cerebellopontine cisterns (CP) are faintly outlined by radiopaque material.

intact. Yearly follow-up CT scans have been normal to date.

Discussion

There are many reports of inflammatory fourth ventricular obstruction in children and adults. The obstruction in those cases was attributed to meningoencephalitides; prenatal infection; shunting procedures; birth and head trauma; ear, ocular, and nasopharyngeal infection; venereal disease; influenza; radio- and chemotherapy; postoperative changes after cranial and lumbar procedures; granulomatosis; intraventricular hemorrhage; subarachnoid hemorrhage; and tumor. Fourth ventricular outlet obstruction has been observed with Monilia, Torula, and Toxoplasma infections. Hydrocephalus due to cystercerosis has been reported, but no reports of central nervous system involvement by Giardia lamblia or Trichuris trichuria have been found. There is one case in the literature of membranous obstruction of the foramen of Magendie in a patient with multiple sclerosis, which was believed to be coincidental.

Congenital fourth ventricular outlet occlusions are seen in children in association with such syndromes as Arnold-Chiari malformation, Dandy-Walker cysts, tureus sclerosis, and spina bifida, as well as with platybasia, achondroplasia, basilar impression, and atlanto-occipital fusion. Blocked ventricular outlet has been found in patients with cafer-au-lait spots, but no direct relationship to neurofibromatosis has been found. Other congenital lesions, such as enterogenous, arachnoid, neuroepithelial, and mesencephalic cysts, have been seen in adults.

There are reports suggesting congenital lesions in young patients similar to the lesions we found in our cases. Coleman and Troland evaluated a 17-year-old boy with chronic headaches in whom ventriculography showed a dilated fourth ventricle. At surgery, the vermis was noted to be small, but was otherwise normal with no inflammation. The fourth ventricle was said to be "hermetically sealed" by a translucent membrane, but the histology was not presented. Amacher and Page described a 21-year-old woman with recent onset of headaches and vomiting, who suffered an episode of loss of consciousness with meningismus. Angiography disclosed no vascular anomaly. Ventriculography suggested a lesion obstructing the outlet of the fourth ventricle. At surgery, a translucent membrane was found occluding the foramen of Magendie. When it was opened, free flow of CSF was seen. The vermis and cistern were normal. The pathology of the membrane was not discussed.

Review of the English-language literature revealed three cases of membranous obstruction of the foramen of Magendie in adults (all of whom were older than 30 years of age) in which the obstruction was not associated with systemic illness or trauma. In 1877, Hilton reported the case of a 34-year-old man with chronic headaches since childhood. One year prior to his death, he developed unsteady gait and head tilt. Shortly before his death, he suddenly vomited and collapsed with stertorous breathing. At autopsy, minor congenital hand deformities were found. Examination of the brain revealed hydrocephalus and a huge fourth ventricle. The midline "cerebro-spinal" opening was closed by a membrane. Because of the chronicity of the patient's symptoms, the anomaly of the hands, and the lack of inflammation, Hilton concluded that the foraminal obstruction was congenital. Levin and Gross presented the case of a 67-year-old patient with chronic depression who developed headache and cerebellar signs. The patient's medical history was noncontributory. Ventriculography disclosed a large fourth ventri-
cle. At surgery, the tonsils were seen to be adherent, but the vermis and arachnoid were normal. No CSF outflow was seen, but no mention was made of any membranous obstruction. David, et al., described the case of a 36-year-old woman with papilledema who suffered sudden onset of headache and emesis. Ventriculography revealed hydrocephalus with a huge fourth ventricle. At surgery, the cerebellum was normal with no inflammation. The fourth ventricle was dilated and the foramen of Magendie was said to be "imperforate."

Many reports detail possible developmental anomalies in cases of fourth ventricular outlet obstruction. Although in man the fourth ventricular foramina are normally patent at birth, it has been noted that 20% of otherwise normal humans have congenital imperforate foramina of Luschka, almost always bilaterally symmetrical. In 4% of cases a midline foramen fails to form, frequently accompanied by vermian agenesis, as in cases of Dandy-Walker syndrome. Two percent to 3% of Barr's pathological specimens with normal vermis had shelf-like midline membranous projections, some partially fenestrated. Another 3% of cases studied by pneumoencephalography presented with a foramen of Magendie so small that gas bubbles passed through with great difficulty. Thus, there may be partial or complete obstruction of the foramen of Magendie in up to 6% of cases.

Stasis of metrizamide in the cisterna magna, as seen in our cases, is evidence of incomplete communication between the fourth ventricle and the subarachnoid space. The presence of semipermeable membranous perforations, or of hinged or irregularly dissected membranes, could explain the delayed onset of symptomatic hydrocephalus in our patients. Semiperforate foramina of Luschka may have been sufficient to allow some CSF flow for prolonged periods of time. The fourth ventricle may have dilated slowly while CSF continued to flow at subcritical rates until decompensation occurred, leading to symptomatic intracranial hypertension.

The signs and symptoms of fourth ventricular outlet occlusion have been described in several reports. The diagnosis is suggested in patients with nonfocal signs of increased intracranial pressure without a mass lesion and hydrocephalus but with a disproportionately large fourth ventricle. The presence of a membrane occluding the foramen of Magendie can be established with metrizamide-enhanced CT and ventriculography, in conjunction with pneumoencephalography to visualize its inferior border. The radiographic appearance of the fourth ventricular outlet provides sufficient evidence to warrant posterior fossa exploration.

References

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