Expansion Diverticulum of the Suprapineal Recess Causing Cerebellar Ataxia

A Case Report

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Key words: ventricular diverticulum, hydrocephalus, endoscopic third ventriculostomy, cerebellar ataxia

SUMMARY – *As a result of long-standing cerebrospinal fluid (CSF) pulsation against the thinnest segments of the ventricular walls, focal enlargement of the ventricular system (diverticulum) may occur, mainly at the medial wall of the trigone of the lateral ventricles (atrial diverticula) or at the posterior wall of the third ventricle (expansion of the suprapineal recess). In the latter case, ocular signs are the most common symptoms, due to the severe deformation of the periaqueductal region. We describe a case of non-communicating hydrocephalus in a 36-year-old woman who presented a three-year history of cerebellar ataxia. Preoperative brain magnetic resonance (MR) scan showed marked supratentorial hydrocephalus with an apparently patent aqueduct of Sylvius, and an enlarged suprapineal recess causing cerebellar and tentorial dislocation. The patient was successfully treated by endoscopic third ventriculostomy and monitored by MR scans with phase-contrast sequences for assessment of CSF flow. Cerebellar ataxia is a very rare symptomatic onset for a suprapineal recess expansion diverticulum, which may cause obstructive hydrocephalus that can be effectively treated by endoscopic third ventriculostomy.*

Introduction

The formation of a diverticulum of the ventricular walls may be subsequent to long-standing cerebrospinal fluid (CSF) pulsation against the thinnest segments of the ependymal lining. The third ventricle represents the point of origin of about 30% of ventricular diverticula¹. The weakest parts of the third ventricle are the lamina terminalis, the infundibular and suprapineal recesses², the latter being among the most common sites of diverticulum formation. An expansion diverticulum of the suprapineal recess usually causes a midbrain dislocation with most severe deformation at the level of the periaqueductal region and subsequent ocular signs. We describe a case of an expansion diverticulum of the suprapineal re-

cess with cerebellar ataxia as the only symptom reported.

Case Report

Three years before admission, after a spontaneous abortion, a 36-year-old woman with otherwise unremarkable medical history started to complain of fluctuating ataxia. One year before admission, the patient underwent a brain magnetic resonance (MR) study that showed supratentorial hydrocephalus with an expansion diverticulum of the suprapineal recess. At that time, the patient refused endoscopic third ventriculostomy (ETV) treatment due to a concomitant pregnancy. After spontaneous interruption of the second pregnancy, she came to

Figure 1 Preoperative (A,D: left column), 7 month-postoperative (B,E: middle column) and 30 month-postoperative (C,F: right column) T2-weighted sagittal (upper row) and axial (lower row) MR scans. A) Marked dilatation of the supratentorial ventricular system and suprapineal recess, which bulges posteriorly in the quadrigeminal and superior vermian cisterns, severely compressing the cerebellum and displacing upwards the tentorium and the midline cerebral veins. The corpus callosum is stretched and thinned; the aqueduct appears patent. B,C) Progressive size reduction of the diverticulum and re-expansion of the vermian folia. D) Symmetrical dilatation of the ventricular trigones with intact medial walls and normal appearance of the pericerebral CSF spaces. Measurement of the width of the third ventricle (1.65 cm) is reported, as part of the semiquantitative assessment of the hydrocephalus. E,F) Return to normal of third ventricle width, size reduction of the ventricular atria and decreased “splitting” of the occipital lobes after endoscopic third ventriculostomy (ETV).

our observation because of progression of the gait disturbance, unresponsive to steroids. At admission she showed gait incoordination with ataxia, but without dysmetria at the finger-nose test; the remaining neurological examination was within normal range. There was no nystagmus or diplopia, or Parinaud’s syndrome, and the optic discs were normal.

Brain MR was performed on a 1.5 T scanner using a phased-array head coil and conventional morphologic sequences. A markedly dilated supratentorial ventricular system was observed, without direct signs of aqueductal stenosis, but with a suprapineal recess expanded to such an extent that the quadrigeminal and superior vermian cisterns were obliterated, the vermis and cerebellar hemispheres were displaced downwards, and the great vein of Galen and tentorium were displaced upwards (Figure

1 A,D). No significant splitting of the mammillary bodies was observed, a finding which has been associated with thickening of the third ventricle floor³.

ETV was performed with a 6 mm rigid endoscope (Zeppelin) under general anaesthesia and antibiotic prophylaxis. The stoma was placed within the tuber cinereum by means of monopolar coagulation and then dilated using a 2 French Fogarty balloon catheter and Decq’s forceps (Figure 2). Visual control of the aqueduct showed no gliotic membranes. At the end of the ETV, as routinely performed in our endoscopic procedure⁴, a ventricular transducer (Codman Microsensor Ventricular Catheter) was left in the lateral ventricle and connected to a monitor for bedside intracranial pressure (ICP) monitoring in the first postoperative 24-36 hours (ICP Express - Codman), showing a normal ICP

Figure 2 ETV intraoperative snapshots. A) Third ventricle floor with translucent tuber cinereum. B-D) Progressive enlargement of the stoma using Decq's forceps.

range. Cerebellar ataxia greatly improved and finally disappeared in a few weeks.

The follow-up MR scans at seven and 30 months after ETV showed a progressive size reduction of the ventricles and suprapineal recess, with re-expansion of the vermis and cerebellar hemispheres (Figure 1 B,C,E,F). Patency of the third ventricle floor stoma was documented with morphologic and phase-contrast CSF flow-sensitive sequences, at that time available on the MR scanner, while no significant flow in the aqueduct was observed (Figure 3).

Discussion

Because the ventricular enlargement in many cases of aqueductal stenosis progresses slowly, with late onset of symptoms of raised

intracranial pressure, the diagnosis may be delayed and the ventricular dilatation may be massive. The ependymal lining often presents individual variations, being absent for short distances (denuded areas) or thicker than one cell layer in others, or even wrinkly, with formation of small diverticula². As a result of long-standing CSF pulsations against the thinnest segment of the ventricular walls, focal enlargement of the ventricular system may occur, leading to the formation of pulsion diverticula, consisting of cystic spaces lined by pia mater that are walled off from the subarachnoid spaces and communicate only with the ventricles⁵. The most frequent sites of ventricular rupture and diverticular formation are the medial walls of the trigone of the lateral ventricles (atrial diverticula)⁶. Atrial diverticula were reported to occur in 66% of

Figure 3 Postoperative MR follow-up at 7 (A,C, left column) and 30 months (B,D, right column) after ETV. A,B) Axial FLAIR MR scans at the level of the interpeduncular cistern showing progressive size reduction of the temporal horns of the lateral ventricles, the CSF artifact induced by ETV, and absence of periaqueductal gliosis. C) Coronal 3mm-thick T1-weighted MR scan at the level of the third ventricle floor, showing the patency of the ETV site. D) Midline sagittal cardiac-gated phase-contrast MR scan showing high signal in the interpeduncular and pre-pontine cisterns, due to CSF flow through the stoma, and no adequate CSF flow through the aqueduct.

cases, while the origin from the third ventricle is reported in about 30% of cases¹.

The suprapineal, lamina terminalis, and infundibular recesses are the weakest parts of the third ventricle. Their walls are not covered by neural or vascular structures, unlike the lateral and inferior walls of the third ventricle, which are bordered by the thalami laterally and the

midbrain inferiorly². In normal subjects, the suprapineal recess is merely a diverticulum of the ependymal roof⁷ and, in cases of aqueductal stenosis, may become so dilated to fill the quadrigeminal and superior vermician cisterns, displacing the vermis and cerebellar hemispheres downwards, and dislodging and compressing the dorsal midbrain and posterior commissure.

The rostral interstitial nucleus of the medial longitudinal fasciculus, a group of cells in the prerubric field of the mesencephalic reticular formation, is considered the location of vertical gaze⁸. The compression of this nucleus, consequent to the severe deformation of the periaqueductal region is related to the ocular signs, the typical clinical onset of a diverticulum of the suprapineal recess^{9,10}. Cerebellar ataxia is seldom reported⁶, and extremely rare as the only symptom, as was the case in our patient.

Our case confirms that aqueductal stenosis may be difficult to demonstrate on conventional morphologic MR sequences, as direct signs of flow obstruction may be lacking¹¹. The apparent patency of the aqueduct on the preoperative morphologic MR images (Figure 1) confirms the need for a functional assessment of CSF dynamics in cases of suspect aqueduc-

tal stenosis, which is best accomplished by cine phase-contrast MR sequences¹². Postoperatively, besides the size reduction of the supratentorial ventricular system, the efficacy of ETV can be monitored by assessing CSF flow at the level of the third ventricle floor stoma (Figures 1 and 3).

Conclusions

To the best of our knowledge, this is the first report of cerebellar ataxia as the only clinical presentation related to an expansion diverticulum of the suprapineal recess. Endoscopic third ventriculostomy is the treatment of choice for obstructive hydrocephalus, and MRI with cine phase-contrast sequences should be used pre- and post-operatively.

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