Acute hydrocephalus secondary to obstruction of the foramen of Monro and cerebral aqueduct caused by a choroid plexus cyst in the lateral ventricle

Case report

BRIAN V. NAHED, M.D.,1 ANEELA DARBAR, M.D.,1 ROBERT DOIRON, B.A.,1 ALI SAAD, M.D.,2 CAROLINE D. ROBSON, MB.CH.B.,3 AND EDWARD R. SMITH, M.D.1

Departments of 1Neurosurgery, 1Neuropathology, and 1Radiology, Children’s Hospital Boston, Massachusetts

Choroid plexus cysts are common and typically asymptomatic abnormal folds of the epithelial lining of the choroid plexus. Rarely, these cysts may gradually enlarge and cause outflow obstruction of cerebrospinal fluid. The authors present a case of a large choroid plexus cyst causing acute hydrocephalus in a previously healthy 2-year-old boy. The patient presented with markedly declining mental status, vomiting, and bradycardia over the course of several hours. Computed tomography scans demonstrated enlarged lateral and third ventricles with sulcal effacement, but no obvious mass lesions or hemorrhage. There was no antecedent illness or trauma. A right frontal external ventricular drain was placed in the patient, resulting in decompression of only the right lateral ventricle. Magnetic resonance (MR) imaging demonstrated a lobulated cyst arising from the choroid plexus of the left lateral ventricle and herniating through the foramen of Monro into the third ventricle, occluding both the foramen of Monro and the cerebral aqueduct. The patient underwent an endoscopic fenestration of the cyst, and histological results confirmed that it was a choroid plexus cyst. Postoperative MR imaging showed a marked reduction in the cyst size. The cyst was no longer in the third ventricle, the foramen of Monro and the aqueduct were patent, and the ventricles were decompressed. The patient was discharged home with no deficits. To the authors’ knowledge, there are no previous reports of a choroid plexus cyst causing acute hydrocephalus due to herniation into the third ventricle. This case is illustrative because it describes this entity for the first time, and more importantly highlights the need to obtain a diagnosis when a patient presents with acute hydrocephalus without a clear cause. (DOI: 10.3171/PED-07/09/236)

KEY WORDS • acute hydrocephalus • choroid plexus cyst • endoscopy • obstruction • pediatric neurosurgery

Abbreviations used in this paper: CSF = cerebrospinal fluid; CT = computed tomography; EVD = external ventricular drain; FIESTA = fast imaging employing steady-state acquisition; MR = magnetic resonance.
Acute hydrocephalus caused by a choroid plexus cyst

The patient underwent an endoscopic fenestration of the right lateral ventricle, with persistent enlargement of the left lateral ventricle. Because of the unclear nature of the incomplete decompression of the ventricles, an MR imaging examination was performed using a 1.5-Tesla system (General Electric Medical Systems). Although standard T2-weighted MR imaging was not particularly revealing, T1-weighted MR imaging with contrast enhancement and multiplanar 3D FIESTA imaging were subsequently performed during the same scan revealing multiple choroid plexus cysts, including a large lobulated cyst arising from the choroid plexus of the left lateral ventricle. This cyst had herniated through the foramen of Monro into the third ventricle, occluding both the foramen of Monro and the cerebral aqueduct with resultant hydrocephalus (Figs. 2 and 3).

Operation. The patient underwent an endoscopic fenestration of the right lateral ventricle with persistent enlargement of the left lateral ventricle. Because of the unclear nature of the incomplete decompression of the ventricles, an MR imaging examination was performed using a 1.5-Tesla system (General Electric Medical Systems). Although standard T2-weighted MR imaging was not particularly revealing, T1-weighted MR imaging with contrast enhancement and multiplanar 3D FIESTA imaging were subsequently performed during the same scan revealing multiple choroid plexus cysts, including a large lobulated cyst arising from the choroid plexus of the left lateral ventricle. This cyst had herniated through the foramen of Monro into the third ventricle, occluding both the foramen of Monro and the cerebral aqueduct with resultant hydrocephalus (Figs. 2 and 3).

Figs. 1. Preoperative CT scans. Left: Moderate dilation of the third ventricle and asymmetric dilation of the lateral ventricles (left > right) is demonstrated. The left foramen of Monro (arrow) appears widened. The fourth ventricle appeared normal (not shown). The cerebral sulci are effaced suggesting increased intracranial pressure. Right: The septum pellucidum is shifted to the right (arrow).

Figs. 2. Preoperative axial MR images. A: Fast spin echo T2-weighted image after placement of a ventriculostomy catheter demonstrates partial decompression of the right lateral ventricle with a small amount of gas within the right frontal horn. Persistent dilation of the left lateral ventricle with increased shift of the septum pellucidum to the right (arrow) is also observed. B: Fast spin echo contrast-enhanced T1-weighted image demonstrates a very thin-walled cyst (arrow) that is not identifiable on the T2-weighted image. The lesion is isointense with CSF. C: The FIESTA image at a more cephalad level demonstrates a thin-walled cyst (superior arrowhead) originating from the choroid plexus within the dilated left lateral ventricle. Another cyst is seen within the right lateral ventricle (inferior arrowhead).

Figs. 3. Sagittal MR FIESTA images. Left: A preoperative midline image reveals a 3 × 1.7 × 1.6-cm enlarged lobulated cyst (superior arrowhead) arising from the frontal horn of the left lateral ventricle with a component that herniates through the foramen of Monro into the third ventricle in an hourglass configuration. Also note the cyst wall in the middle of the third ventricle (inferior arrowhead) obstructing CSF flow via the aqueduct. Right: Slightly off-midline image obtained postoperatively reveals the absence of the portion of the choroid plexus cyst that was herniating into the third ventricle. After postoperative resolution of midline shift, the cyst (arrowhead) is now better visualized slightly to the left of midline. As a result, the aqueduct is no longer easily seen and there is volume averaging of the thalamus posterior and inferior to the cyst. Note the postoperative decreased ventricular size and decreased sulcal effacement.
decision was made to close. An EVD was left as an additional means of ensuring that both lateral ventricles would be decompressed in the postoperative period.

**Histology.** Microscopic examination of the cyst demonstrated cuboidal epithelium consistent with a choroid plexus cyst (Fig. 4). Immunostaining was performed to differentiate between an arachnoid and colloid cyst. Immunostaining was positive for transthyretin (prealbumin), negative for epithelial membrane antigen, and negative for mucin, which reaffirmed the diagnosis of a choroid plexus cyst.

**Postoperative Course.** The patient’s postoperative course was uneventful. The patient was awake, alert, interactive, following commands, and speaking at his baseline level. The patient was rapidly weaned from the EVDs, which were subsequently removed. Postoperative MR imaging confirmed a marked reduction in the cyst size (Fig. 3 right). The cyst was no longer in the third ventricle, the foramen of Monro and the aqueduct were patent, and the ventricles were decompressed. The patient was discharged home on postoperative Day 5, functioning at his baseline level and without deficits.

**Discussion**

Choroid plexus cysts are formed in utero or early in infancy, and are often found incidentally during antenatal ultrasonography, CT, or MR imaging. Although most of these cysts that are less than 1 cm in diameter remain asymptomatic, choroid plexus cysts that are larger than 2 cm have been reported in association with headaches and obstructive hydrocephalus. These lesions are often easily identified on CT or MR imaging as encapsulated, thin-walled structures in the ventricular system. As this case illustrates, however, choroid plexus cysts are not always readily visualized on initial imaging, even in the setting of enlarged ventricles associated with acute hydrocephalus. Without a clear cause for the hydrocephalus (such as aqueductal stenosis, infection, or hemorrhage), more detailed investigations are required so that appropriate therapeutic interventions can be instituted. It is important to note that significant persistence on the part of the treating physicians (both neurosurgeons and neuroradiologists) were necessary to make the diagnosis in this case, including the need for multiplanar high-resolution images. Ultimately, a herniated choroid plexus cyst was identified as the cause of the hydrocephalus.

The development of choroid plexus cysts has been postulated to result from the folding of the neuroepithelium of the choroidal matrix during periodic cycles of growth and regression of the choroid plexus, leading to the eventual formation of a cyst. These cysts are commonly found in the lateral ventricle, and have the capability to move to some degree if attached to a pedunculated stalk. These cysts can also progressively enlarge, and both enlargement and movement can result in mechanical obstruction of CSF pathways. The patient in this report had an unusual case of acute hydrocephalus arising from the choroid plexus cyst that herniated into the third ventricle and caused outflow obstruction of CSF.

Although the patient’s clinical symptoms were consistent with acute hydrocephalus, a clear cause was not identified initially. Placement of a right-sided EVD provided decompression of the right ventricle, without complete improvement of the patient’s clinical symptoms. Results of CSF studies were normal and neuroimaging identified a large left ventricle, suggesting a lack of communication between the decompressed right ventricle and the prominent left ventricle, eventually leading to our hypothesis of obstruction. Because routine neuroimaging did not reveal a cyst in the ventricles, an MR image was obtained that identified a lobulated cyst arising from the choroid plexus and occluding both the cerebral aqueduct and the foramen of Monro.

There are a range of MR imaging pulse sequences that are currently available in addition to sequences that are routinely used for brain imaging. For example, heavily T2-weighted sequences with thin slices, such as 2D and 3D fast imaging sequences, are extremely useful for demonstrating thin-walled cystic structures. One such example is the FIESTA pulse sequence. This imaging modality is a gradient echo technique with fully refocused transverse magnetization, and is also known as true fast imaging with steady-state precession (true FISP) and balanced fast-field echo (FFE). This sequence has widespread clinical applications such as cardiac, fetal, inner ear, and cranial nerve imaging. Advantages of this sequence include excellent spatial resolution, an increase in contrast between fluid-containing regions and adjacent tissues (such as cranial nerves outlined by CSF, and delineation of cyst walls), as well as a relative insensitivity to flow. A significant drawback of the FIESTA sequence is its sensitivity to off-resonance effects due to susceptibility gradients or ferromagnetic objects close to the area undergoing imaging. This sensitivity results in areas, lines, or circles of signal void surrounding the ferromagnetic object (such as a ventriculostomy catheter) or anatomical area (such as paranasal sinuses). In this case, FIESTA was particularly useful in this imaging sequence for precise delineation of the choroid plexus cyst anatomy.

After fenestration of the cyst and placement of a contralateral EVD, the patient’s symptoms returned to baseline very quickly. In this instance, cyst fenestration proved to be the easiest and safest procedure, providing immediate relief of the obstruction and improvement of the patient’s symptoms. Other techniques such as open surgery, placement of a shunt, or stereotactic puncture of the cyst were considered.
but were believed to be inferior methods of treatment. Open surgery carried a greater operative risk for bleeding and a longer time in a state of anesthesia. Placement of a shunt might not be effective if the third ventricle was not decompressed and carried the attendant risks of a permanently implanted device, including infection as well as occlusion of the catheter. Stereotactic puncture of the cyst would not allow the surgeon the benefit of direct visualization to confirm fenestration and would also not allow the additional ability to perform multiple openings in the cyst (presumably increasing the effectiveness of the procedure). We feel that in this case, endoscopic fenestration of the cyst provided the easiest, safest, and most effective method of directly treating the cause of the hydrocephalus. Although long-term follow-up is needed, thus far this treatment has provided durable relief of the patient’s symptoms and postoperative imaging has confirmed its efficacy (Fig. 3 right).

Conclusions

A choroid plexus cyst causing acute hydrocephalus from migration and herniation into the third ventricle appears to be a rare event, and has been unreported to date. If initial imaging of a patient presenting with symptoms of acute hydrocephalus fails to identify a lesion, particularly if the initial imaging studies performed are CT or ultrasonography without the added enhancements of MR imaging, further investigation into the cause of the hydrocephalus must be performed. Asymmetric ventricular enlargement (before or after EVD insertion), a normal CSF profile, and a patient history unremarkable for risk factors such as trauma or infection increase the likelihood of the involvement of a choroid plexus cyst. In such cases, using fast multiplanar MR imaging may be helpful in the identification of cysts, as exhibited in this patient. This case illustrates a novel occurrence of acute hydrocephalus arising from an obstructive choroid plexus cyst and underscores the need for persistence in identifying a diagnosis when a patient presents with acute hydrocephalus without a clear cause.

References


Address reprint requests to: Edward R. Smith, M.D., Department of Neurosurgery, Children’s Hospital Boston/Harvard Medical School, 300 Longwood Avenue, Boston, Massachusetts 02115. email: edward.smith@childrens.harvard.edu.