Acute triventricular hydrocephalus caused by choroid plexus cysts: a diagnostic and neurosurgical challenge

Pietro Spennato, MD,¹ Carmela Chiaramonte, MD,² Domenico Cicala, MD,³ Vittoria Donofrio, MD,⁴ Manlio Barbarisi, MD,⁵ Anna Nastro, MD,³ Giuseppe Mirone, MD,¹ Vincenzo Trischitta, MD,¹ and Giuseppe Cinalli, MD¹

Departments of ¹Paediatric Neurosurgery and ³Neuroradiology, and ⁴Pathology Unit, Santobono-Pausilipon Children's Hospital; ²Division of Neurosurgery, Department of Neurosciences and Reproductive and Odontostomatological Sciences, School of Medicine and Surgery "Federico II," Naples; and ⁵Department of Neurosurgery, Second University of Naples, Italy

OBJECTIVE Intraventricular choroid plexus cysts are unusual causes of acute hydrocephalus in children. Radiological diagnosis of intraventricular choroid plexus cysts is difficult because they have very thin walls and fluid contents similar to CSF and can go undetected on routine CT studies.

METHODS This study reports the authors' experience with 5 patients affected by intraventricular cysts originating from the choroid plexus. All patients experienced acute presentation with rapid neurological deterioration, sometimes associated with hypothalamic dysfunction, and required urgent surgery. In 2 cases the symptoms were intermittent, with spontaneous remission and sudden clinical deteriorations, reflecting an intermittent obstruction of the CSF pathway.

RESULTS Radiological diagnosis was difficult in these cases because a nonenhanced CT scan revealed only triventricular hydrocephalus, with slight lateral ventricle asymmetry in all cases. MRI with driven-equilibrium sequences and CT ventriculography (in 1 case) allowed the authors to accurately diagnose the intraventricular cysts that typically occupied the posterior part of the third ventricle, occluding the aqueduct and at least 1 foramen of Monro. The patients were managed by urgent implantation of an external ventricular drain in 1 case (followed by endoscopic surgery, after completing a diagnostic workup) and by urgent endoscopic surgery in 4 cases. Endoscopic surgery allowed the shrinkage and near-complete removal of the cysts in all cases. Use of neuronavigation and a laser were indispensable. All procedures were uneventful, resulting in restoration of normal neurological conditions. Long-term follow-up (> 2 years) was available for 2 patients, and no complications or recurrences occurred.

CONCLUSIONS This case series emphasizes the necessity of an accurate and precise identification of the possible causes of triventricular hydrocephalus. Endoscopic surgery can be considered the ideal treatment of choroid plexus cysts in children.

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KEY WORDS triventricular hydrocephalus; neuroepithelial cyst; neuroendoscopy; ventriculostomy

INTRAVENTRICULAR choroid plexus cysts are unusual causes of acute hydrocephalus in children. Choroid plexus cysts are rare, benign lesions representing less than 1% of intracranial tumors.³ Several theories have been formulated about the histogenesis of these lesions, and most authors suggest an embryogenetic origin. The majority of these cysts are asymptomatic and are diagnosed incidentally; however, when they become symp-

tomatic, their clinical onset may be rapid and catastrophic. Choroid plexus cysts can be located anywhere within the neuraxis, but they are more frequently discovered adjacent to the supratentorial ventricular system or subarachnoid space. Rarely, they are located in the posterior fossa or brainstem, or adjacent to the fourth ventricle.¹⁶ Depending on their size and location, these lesions can cause hypothalamic dysfunction and obstructive hydrocephalus by

ABBREVIATIONS b-FFE = balanced-fast field echo; DRIVE = driven-equilibrium; EMA = epithelial membrane antigen; ETV = endoscopic third ventriculostomy; EVD = external ventricular drain; GFAP = glial fibrillary acidic protein; SSFP = steady-state free precession; TSE = turbo spin echo; WBC = white blood cell; WISC-R = Wechsler Intelligence Scale for Children-revised.

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compromising CSF flow in the ventricular system. For asymptomatic cysts, conservative follow-up should be recommended. However, the treatment of symptomatic lesions is strictly dependent on their locations and on the surgical treatments available. The wide range of surgical options has included stereotactic aspiration, positioning of a cystosubarachnoid or cystoventricular shunt, external drainage via a reservoir system, microsurgical cyst resection and/or fenestration, and eventually endoscopic cyst fenestration into an adjacent subarachnoid or ventricular space.¹⁶ Considering the variety of symptoms that these cysts can display at their clinical onset, the difficulty of obtaining an early and exact radiological diagnosis, and the wide spectrum of surgical alternatives proposed, choroid plexus cysts effectively constitute a significant clinical, radiological, and surgical challenge.

In this study we present our experience in 5 children with choroid plexus cysts in the region of the third ventricle. All lesions were treated by endoscopic shrinkage and removal of the cyst, in association with an endoscopic third ventriculostomy (ETV).

Methods

Patient Selection

Since 2007, 5 patients affected by intraventricular cysts originating from the choroid plexus have been operated on using an endoscopic technique at Santobono-Pausilipon Children's Hospital in Naples, Italy. During this same time frame, an average of 45 new cases of nontumoral hydrocephalus were managed per year. The ages of the patients in the study ranged from 4 months to 8 years. Clinical details of these 5 cases are shown in Table 1. All patients presented with acute neurological deterioration. In 1 case (see Illustrative Case 1 below), hypothalamic dysfunction was also present. The patients were all studied using an urgent CT scan and then MRI (Figs. 1–5, also see Fig. 7). An urgent CT scan showed triventricular hydrocephalus in all cases. The diagnosis of an intraventricular cyst was overlooked in 4 cases on the initial CT scan and was recognized only on the subsequent MRI, which in our department is systematically performed as an emergency procedure in every new case of hydrocephalus. The MRI driven-equilibrium (DRIVE; Philips) 3D T2-weighted radiofrequency reset pulse sequence in the sagittal midplane is included in our standard MRI protocol examination for hydrocephalus, mainly to detect aqueductal stenosis and/or obstruction. DRIVE sequences are 3D turbo spin echo (TSE) sequences applying a set of recovery 90° flip-back pulses that push the residual transverse magnetization back to the longitudinal axis, available on the Philips Achieva 1.5-T MRI machine. When the DRIVE pulse is added to the T2-weighted 3D TSE sequence, imaging time becomes shorter with better image quality and low CSF flow artifacts; high-resolution MR images can be obtained with high-contrast images of fluid/solid interface, allowing extremely careful identification of thin membranes and septations that could be missed on standard TSE or spin echo MRI sequences. In 1 case, the cyst was also not detected by MRI (see Illustrative Case 2; Fig. 4). In 2 cases the symptoms were intermittent, with spontaneous

remission and sudden clinical deteriorations, reflecting an intermittent obstruction of the CSF pathway. The patients were all managed by urgent surgery. In the first case of our series an external ventricular drain (EVD) was implanted (see Illustrative Case 1); in the remaining cases, urgent endoscopic surgery was performed. In 2 cases, surgery was performed at recurrence of symptoms.

Anatomoradiological Findings

MRI with DRIVE sequences and CT ventriculography (in 1 case) allowed us to accurately diagnose the intraventricular cysts that typically occupied the superior part of the third ventricle, occluding the aqueduct and at least 1 foramen of Monro. The endoscopic intraoperative aspects of the cysts were similar in all cases. The cysts, which present with very thin and transparent walls, are attached to the choroid plexus. The surface is crossed by several small vessels (Figs. 6–9, Video 1).

VIDEO 1. Clip showing preoperative MR images and endoscopic procedure in Case 5. Copyright Pietro Spennato. Published with permission. [Click here to view.](#)

The cysts are visible in 1 lateral ventricle at the level of the foramen of Monro and continue into the third ventricle, most often in the posterior part.

Endoscopic Procedure

Preoperative neuronavigational surgical planning (StealthStation AxIEM, Medtronic) was performed in all cases but 1 (Case 1, in which a preoperative EVD was implanted). In all cases a 30°, 6-mm, rigid-rod lens neuroendoscope was used (Decq endoscope, Karl Storz GmbH). A standard precoronal bur hole was drilled at the site of the larger lateral ventricle. The cysts were visualized at the foramen of Monro, in continuity with the choroid plexus. The cyst walls were thin but very elastic, and therefore difficult to perforate. A Thulium laser (RevoLix Jr, LISA Laser) or monopolar coagulation was used to coagulate the vessels on the surface of the cyst and shrink the cyst. The cyst walls were then cut by scissors and removed with forceps. In every case, a standard ETV was also performed. In 2 cases in which the size of the lateral ventricle was asymmetrical, fenestration of the septum pellucidum was also performed. The posterior part of the third ventricle was explored by rotating the endoscope (Fig. 9). Small fragments of the cyst walls strictly adherent to thalamostriate or septal veins were not removed (Video 1).

Results

Cyst shrinkage and near-total removal of the walls, as well as an ETV, were performed in all cases. Septostomy was performed in 2 cases. Even if some small fragments of the cyst walls were left behind during endoscopic surgery, no radiological evidence of cyst remnants was noted. Choroid plexus cysts were histopathologically diagnosed in all 5 cases (Fig. 2). All patients showed a marked improvement of their clinical conditions after the procedure, experiencing a complete recovery from preoperative symptoms such as seizure, headache, nausea, and vomiting. There were no intraoperative deaths and/or serious

TABLE 1. Clinical features and outcome

Case No.	Age (mos), Sex	Symptoms	CT	MRI	3rd Ventricle Lesion Site	Surgery	FU (mos)
1	12, M	Seizures, vomiting, hyponatremia, hypokalemia	Triventricular hydrocephalus	Cyst occupying the 3rd ventricle	Anterior	EVD followed by cyst shrinkage and removal + ETV	120
2	60, M	Headache, vomiting, hydrocephalus, drowsiness (intermittent symptoms)	Triventricular hydrocephalus & balloon-shaped 3rd ventricle*	Cyst in the 3rd ventricle w/ a CSF-like fluid content	Anterior	Cyst shrinkage & removal + ETV	96
3	4, M	Vomiting, drowsiness	Triventricular hydrocephalus	Cyst occupying the 3rd ventricle	Posterior	Cyst shrinkage + removal + septostomy + ETV	12
4	96, F	Headache, drowsiness (intermittent symptoms)	Triventricular hydrocephalus*	Ventricle size asymmetry, cyst occupying the 3rd ventricle†	Posterior	Cyst shrinkage + removal + septostomy + ETV	8
5	10, M	Fever, coma, lt cranial nerve VI palsy	Triventricular hydrocephalus	Cyst occupying 3rd third ventricle	Posterior	Cyst shrinkage + removal + ETV	6

FU = follow-up.

All patients experienced good outcomes after surgery.

* Ventricular size varied at different controls (first CT scan, first MR image, and second MR image).

† At initial MRI the diagnosis was missed (see Illustrative Case 2).

perioperative complications. The postoperative follow-up periods ranged from 6 months to 10 years (mean 4.03 years). All patients were followed-up at 3-month intervals in the first year and then every 12 months afterward. The follow-up evaluation consisted of a survey of clinical presentations and radiological examinations using MRI. All patients achieved a complete resolution of symptoms, with relatively reduced ventricles, in the absence of recurrence. Psychomotor testing for evaluation of neurodevelopmental outcome was available in 3 patients who exhibited normal intelligence on the Wechsler Intelligence Scale for Children–revised (WISC-R).

Illustrative Cases

Case 1

History and Examination

A 12-month old boy (Case 1, Table 1) with a 24-hour

history of repeated vomiting was admitted to another hospital for the sudden onset of generalized seizures. Body temperature was normal, blood examination on admission showed increased white blood cell (WBC) count (12,810 per mm³), mild hyponatremia (132 mmol/L), and normokalemia (4.2 mmol/L). The patient was transferred to the emergency department of our hospital, where, upon arrival, he suffered a second, prolonged generalized seizure. Blood values confirmed hyponatremia (128 mEq/L) with decreased plasmatic (255.5 mOsm/L) and urinary (255.0 mOsm/L) osmolality, and a WBC count of 20,400 per mm³ (85.6% neutrophils). An emergency CT scan showed mild triventricular hydrocephalus. Because of the progressive neurological deterioration and suspicion of a possible underlying infection, a right frontal EVD was urgently implanted. A new CT scan after the surgical placement of the EVD demonstrated a persisting enlargement of the third and left lateral ventricles (Fig. 1A). The analysis of the

FIG. 1. Case 1. **A:** Axial CT scan following EVD placement showing asymmetrical ventricular dilation and the third-ventricle dilation still present. **B:** Preoperative axial CT ventriculography showing a nonenhancing lesion arising in the third ventricle. The left frontal horn of the contralateral ventricle is prevented by the cyst of receiving contrast due to stoppage of flow through the foramen of Monro. **C:** On a postoperative axial T1-weighted sequence, no cystic remnant could be visualized.

FIG. 2. Case 1. H & E staining (A) showing single-layer nonciliated, columnar, pseudostratified, ependyma-like cells. Immunohistochemical staining shows mild positivity for EMA (B), and strong positivity for vimentin (C), S100 (D), and CD34 (E). Original magnification $\times 200$ (A, C, and E) and $\times 100$ (B and D).

collected CSF was negative for meningitis. MRI showed images compatible with a cystic lesion with a very thin membrane filled with CSF-like fluid within the third ventricle, obstructing both foramina of Monro. Due to the unclear radiological picture, we decided to further investigate the CSF dynamic using CT ventriculography (Fig. 1B), introducing a hydrophilic contrast through the EVD. This resulted in a clear absence of contrast filling in the third ventricle forming a roundish shape, but a persistent injection of contrast through the posterior half of the third ventricle, indicating the presence of a cystic formation filled by a liquid isodense to CSF. We performed a neuroendoscopic procedure with the fenestration of the cyst and an ETV to achieve a secure CSF drainage. The endoscope was introduced into the right frontal horn of the lateral ventricle through the previous EVD opening. The foramen of Monro was obstructed by a translucent cystic formation with a thin membrane rich with small vessels on its surface. A large fenestration of the membrane was obtained by monopolar coagulation, and several specimens were collected for histological examination (Fig. 2). The cyst was partially removed, leaving only a small fragment firmly attached to the choroid plexus. An ETV was eventually performed.

Pathological Findings

The examination of the cyst wall revealed a single layer of nonciliated columnar, pseudostratified, ependyma-like cells. The immunohistochemical staining was positive for vimentin and S100, focally positive for epithelial membrane antigen (EMA), and negative for cytokeratin, glial fibrillary acidic protein (GFAP), and synaptophysin (Fig. 2). This immunohistochemical result ultimately led to the diagnosis of a cyst of neuroepithelial origin, but we were

not able to precisely discriminate between a choroid plexus cyst and an ependymal cyst of the third ventricle.

Postoperative Course

The patient experienced an uncomplicated postoperative course, with rapid recovery of electrolyte balance, no further seizures, and full recovery of his neurological functions. A CT scan obtained 24 hours after the procedure revealed reduction of the ventricular volume. The patient was discharged home on postoperative Day 6. An MRI study performed 3 weeks after surgery failed to reveal a residual cyst wall within the third ventricle (Fig. 1C). Flow artifact was visible through the third-ventricle floor. There were no recurrences of cysts or symptoms at the 10-year follow up. At 7 years of age the patient underwent psychomotor testing for evaluation of neurodevelopmental outcome using the WISC-R, which revealed normal total IQ (91), normal performance IQ (92), and normal verbal IQ (85).

Case 2

History and Examination

An 8-year-old girl (Case 4, Table 1) was admitted to the emergency room of our hospital complaining of a long history of intermittent headache. Initial neuroradiological examinations with CT showed mild enlargement of the supratentorial ventricular system and asymmetrical lateral ventricles, with prevalence of the left ventricle. There were no radiological signs of intracranial hypertension (Fig. 3A). The headache resolved and the patient was discharged home. After a week, MRI performed in an outpatient clinic showed reduction in the size of the ventricles and no radiological signs of intraventricular lesions (Figs.

FIG. 3. Case 2. **A:** Axial CT scan at presentation, showing mild enlargement of the supratentorial ventricular system. **B:** Axial MR image obtained 1 week later (while the patient was asymptomatic), showing resolution of hydrocephalus. **C:** Urgent axial CT scan at second presentation, showing acute hydrocephalus with periventricular lucency.

3B and 4A). She was discharged home, and referred to neurologists for headache medications and follow-up. Two weeks later, after a severe migraine attack associated with vomiting and drowsiness, she was again admitted to the emergency room of our hospital. A CT scan showed an increasing dilation of the ventricular system with periventricular hypodensity (Fig. 3C), and an MR image finally revealed the presence of a posterior third-ventricular cystic lesion (maximum anteroposterior diameter 17 mm) in continuity with the left choroid plexus, thus explaining the dilation of the third and lateral ventricles (Fig. 4B). The optic chiasm and optic nerves showed signal hyperintensity on T2-weighted sequences. The patient underwent an emergency surgical operation.

Operation

An electromagnetic-guided neuroendoscopic procedure was performed. The cyst was widely marsupialized and the cyst walls partially removed. The remnants of the cyst walls were shrunk by Thulium laser coagulation. A septostomy and ETV were also performed.

Pathological Findings

The examination of the cyst wall revealed fibrovascular

cells with numerous ectatic vascular structures (CD31⁺), harvested by cubic-columnar cells that were positive for cytokeratin and GFAP, suggesting the diagnosis of a cyst of neuroepithelial origin.

Postoperative Course

The patient's postoperative course was uneventful, with rapid recovery from headache and drowsiness. A postoperative MR image revealed reduction of the ventricular volume and the presence of flow through the third-ventricle floor (Fig. 4D). The patient was discharged home in good clinical condition on postoperative Day 3. The explanation of our patient's clinical symptoms was intermittent hydrocephalus because of the presence of a third-ventricle cyst causing an intermittent ball-valve type of obstruction.⁶

Discussion

Cystic intracranial lesions are common findings during routine cerebral imaging examination in children.^{9,17} They can be found incidentally during antenatal ultrasonography,¹⁹ usually in the lateral ventricles. In contrast, third-ventricle cysts are uncommon. They may originate from endodermal epithelial cells or from neuroepithelial cells:

FIG. 4. Case 2. **A:** Midsagittal T2-weighted DRIVE image, showing no radiological signs of a cyst inside the third ventricle. **B:** Midsagittal T2-weighted DRIVE image 2 weeks later, showing dilation of the third ventricle, deformation of the floor, and presence of a cyst obstructing the aqueduct. **C:** Postoperative midsagittal T2-weighted DRIVE image, showing no remnants of the cyst and resolution of hydrocephalus.

FIG. 5. Case 3. Images from a 4-month-old infant presenting with vomiting and drowsiness. **Upper:** Preoperative midsagittal T2-weighted DRIVE image showing a large cyst occupying the posterior part of the third ventricle (arrow) and obstructing the aqueduct (arrowhead). **Lower:** Postoperative midsagittal MR image showing the disappearance of the cyst, normalization of the contour of the third ventricle, and flow artifact through the third-ventricle floor.

colloid cysts and Rathke cysts belong to the first group, choroid plexus and ependymal cysts to the other. In adults, the most frequent cystic formation of the third ventricle is colloid cysts. These cysts are usually filled with a mucinous and proteinaceous material that can be distinguished on CT and MRI. In children, colloid cysts of the third ventricle are less frequent than cysts originating from neuroepithelial cells.² Choroid plexus cysts have been hypothesized to originate from the folding of the neuroepithelium of the choroidal matrix during cyclic phases of growth and regression of the choroid plexus, thus leading to the formation of a cyst. The typical location of a choroid plexus cyst is the lateral ventricle, and they can move to some degree if attached to a pedunculated stalk. Their growth and movements can lead to the mechanical obstruction of CSF pathways. Choroid plexus cysts are formed in utero or early in infancy, and they are often found incidentally during antenatal ultrasonography between 9 and 24 weeks of ges-

FIG. 6. Intraoperative endoscopic images of Case 3. **Upper:** The cyst was apparent at the level of the left foramen of Monro (FM), in continuity with the choroid plexus (ChPI). **Lower:** The cyst was shrunk by laser coagulation and the walls partially removed. An ETV and septum pellucidum (SP) fenestration were also performed.

tation.⁶ The presence of such pathological anomalies detected on antenatal ultrasonography denotes the possibility of aneuploidy risk, specifically a small risk of trisomy 18. The current recommendation for antenatal detection of a choroid plexus cyst includes further testing for associated anomalies, and genetic counseling.⁸ Although most of the cysts that are less than 1 cm in diameter remain asymptomatic, those that are larger than 2 cm are more often associated with headaches and obstructive hydrocephalus.¹⁰ These symptoms may be acute (such as in every case of our series) and dramatic. Symptoms may also be intermittent, caused by an intermittent ball-valve type of obstruction, as in 2 cases of our series. Usually the symptoms are so severe that patients need immediate medical attention.

FIG. 7. Case 5. Preoperative MR images of a 10-month-old infant presenting with strabismus, drowsiness, and fever. **A:** Midsagittal T2-weighted DRIVE image reveals a noncommunicating right choroid plexus cyst that extends inferiorly into the third ventricle through the foramen of Monro. An enlarged third ventricle with prominence of the floor in the suprasellar cistern is observed. The thin membranous wall of the cyst (arrow) is detected in the lumen and along the floor of the third ventricle, resulting in obstruction of the aqueduct. No CSF flow void sign is observed through the normally patent aqueduct. Also note ischemic edema and swelling in the splenium of the corpus callosum. **B:** Phase-contrast cine MRI flow analysis reveals no forward flow within the aqueduct and fourth ventricle. **C:** Quantitative CSF flow analysis graphic performed in the transverse plane of the aqueduct shows low and inconclusive rates of flow velocity (0.7–2.1 cm/sec).

However, radiological diagnosis is difficult, because choroid plexus cysts have translucent fluid contents similar to CSF and can be missed on routine CT studies.¹⁵ In our first case, the initial CT scan, even in a setting of clear triventricular dilation, was unable to demonstrate the presence of the lesion, misleading the neurosurgeon to suspect an inflammatory cause of the acute hydrocephalus. Hyponatremia and hypokalemia, probably responsible for lethargy and seizure onset, were also misleading conditions in the acute phase. An EVD was then implanted in the acute phase. The asymmetrical reduction of the ventricular system on CT performed after EVD implantation and the

MRI DRIVE images prompted us to perform CT ventriculography, which finally delineated the intraventricular cystic lesion. Therefore, without a clear cause explaining the hydrocephalus (such as aqueductal stenosis, infection, or hemorrhage), further diagnostic investigations are mandatory to institute the appropriate therapeutic interventions.

Several heavily T2-weighted MRI sequences with thin slices are currently available, useful for demonstrating thin-walled cystic structures. Sequences with DRIVE techniques and the steady-state free precession [SSFP] method (known as balanced-fast field echo [b-FFE], balanced-turbo field echo, fast imaging employing steady-

FIG. 8. Postoperative MR images of Case 5. **A:** Midsagittal T2-weighted DRIVE image performed 1 day after surgery shows turbulent flow void (arrowhead) following ETV; reduced ventricular size and flattening of the third-ventricle floor are also observed. The wall of the cyst is not detectable, and a CSF flow void sign is revealed through the aqueduct. **B:** Phase-contrast flow analysis shows restoration of the CSF forward flow into the aqueduct and fourth ventricle, represented by the bright signal band (arrow). **C:** Quantitative CSF flow analysis graphic reveals increased rates of flow velocity (1.4–7.7 cm/sec) with a more regular waveform pattern.

FIG. 9. Intraoperative endoscopic images of Case 5. **A:** The cyst was apparent at the level of the right foramen of Monro (FM), in continuity with the choroid plexus (ChPI). The cyst wall was crossed by several small vessels and by 1 bigger vessel. **B:** The cyst was shrunk by laser coagulation, and the vessels on the surface were coagulated using alternating laser and monopolar coagulation. The wall of the cyst was partially removed, leaving a small fragment at the angle between the thalamostriate striate vein (ThSV) and anterior septal vein (ASV). **C:** Rotating a 30° endoscope posteriorly, the posterior portion of the cyst obstructing the aqueduct (Aq) became visible. **D:** The posterior portion of the cyst was also coagulated and shrunk. SP = septum pellucidum.

state acquisition, and true fast imaging with steady-state precession, according to the specific manufacturer of the scanner) are preferred, usually performed during MR cisternography. These sequences have the advantages of including excellent spatial resolution and providing an increase in contrast between fluid-containing regions and adjacent tissues (such as cranial nerves outlined by CSF, and delineation of cyst walls).¹¹ In choroid plexus cysts, these sequences are particularly useful for precise delineation of the lesion anatomy. Sometimes we prefer to use the b-FFE sequence, generally performed for obtaining MR cisternography images for visualization of the posterior fossa cranial nerves. This is a gradient echo sequence obtained with the SSFP method that requires shimming prior to acquisition: balanced gradients maintain the transverse and longitudinal magnetization, which results in representation of both T1-weighted and T2-weighted contrast in the image; it produces images with increased signal from tissues with large T2/T1 ratios, such as flu-

id, blood, and fat, obtaining excellent CSF contrast and high spatial resolution. As DRIVE sequences, b-FFE 3D sequences are superior to fast spin echo T2-weighted sequences due to high-resolution and high-quality images, and a short time acquisition with low CSF pulsation and magnetic susceptibility artifacts; in our experience, b-FFE sequences sometimes provide better high-definition images than DRIVE sequences, with fewer CSF flow artifacts (Video 1).

The radiological appearance of these cysts makes them ideal candidates for the endoscopic approach. They protrude from 1 foramen of Monro, so they can be approached with a rigid endoscope through a standard pre-coronal bur hole. Electromagnetic neuronavigation is very important to reduce morbidity when approaching the ventricular system, which usually is not very enlarged. The cyst walls are thin, but very elastic, and therefore difficult to perforate. Instead of perforating the cysts, as in cases of arachnoid cysts, we preferred to coagulate the

cyst walls utilizing a Thulium laser or monopolar coagulation, progressively shrinking the cyst until the foramen of Monro and the aqueduct were completely freed. The cyst walls were then removed using scissors and forceps. Some small fragments in the roof of the third ventricle or attached to major veins of the foramen of Monro were not removed. No cysts recurred during follow-up. ETV was also performed in every case. Nahed et al.¹⁰ suggested that ETV is not indispensable when there is good visualization of the aqueduct during the endoscopic procedure. We maintain that ETV during the same procedure is strongly advocated when removal of the cyst wall is not complete or in cases in which the cyst extended into the posterior third ventricle.¹ In fact, ETV can reduce the risk of acute triventricular hydrocephalus due to sudden obstruction of the aqueduct in cases of cyst regrowth or due to the movement of the residual cyst wall.

Few cases of third-ventricle neuroepithelial cysts have been described in the literature.^{1,4,6,7,10,12–14,18,19} In 2006, van Baalen and Stephani¹⁹ described the case of a 4-month-old child with a mobile choroid plexus cyst diagnosed at ultrasonography. They noticed that while the child was crying, an ultrasonographic study demonstrated that the cyst was prolapsing from the third ventricle into the dilated left lateral ventricle. When the child was calm, the cyst was found to be nonobstructive. Two months later, the child developed acute hydrocephalus because of a bulging third-ventricle cyst and underwent emergency endoscopic coagulation of the choroid plexus cyst wall. During endoscopic surgery, Azab et al.¹ intraoperatively documented intermittent obstruction of the foramen of Monro by a choroid plexus cyst that intermittently herniated through the foramen of Monro. In 2007, Nahed and colleagues¹⁰ described the case of a 2-year-old child with acute hydrocephalus and declining mental status. A CT scan revealed hydrocephalus, and the patient underwent an emergency ventriculostomy. The postoperative CT scan showed decompression of the right lateral ventricle but persistent enlargement of the left lateral ventricle. MRI revealed multiple choroid plexus cysts. The patient subsequently underwent endoscopic fenestration of the obstructing cyst without complications. In 2007, Tamburrini et al. described a series of 26 patients with intraventricular or paraventricular cysts.¹⁸ Two of these patients presented with choroid plexus cysts and were successfully managed with endoscopic techniques. Their results confirmed that endoscopic management of choroid plexus cysts, as well as other types of intra- and paraventricular cysts, is a valid alternative to open surgery and shunting procedures. Experience with endoscopic fenestration was also described by Gangemi et al., who reported a high success rate in a series of 6 patients, in which 5 of 6 patients were cured without a shunt.⁷ However, all patients in this series were affected by lateral ventricle cysts.

To the best of our knowledge, our series is unique because it focused on a homogeneous group of patients (all affected by third-ventricle choroid plexus cysts), treated with the same endoscopic technique (i.e., coagulation, shrinkage, and removal of the cyst walls). Until now, a good experience using cauterization and removal of choroid plexus cysts was reported only in case reports.^{1,4–6}

Conclusions

Acute triventricular hydrocephalus in children with an apparent absence of a clear cause should be accurately investigated for third-ventricle cyst formation. A radiological diagnosis of choroid plexus cysts can usually be obtained by dedicated MRI sequences. CT ventriculography remains a valuable tool for the diagnosis of a blockage in CSF circulation in cases of inconclusive MR images. The neuroendoscopic management of this type of lesion represents a safe, direct, and minimally invasive solution and offers the possibility to definitely remove the cause of hydrocephalus.

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Disclosures

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Author Contributions

Conception and design: Spennato, Cinalli. Acquisition of data: Spennato, Chiaramonte, Cicala, Donofrio, Barbarisi, Nastro, Mirone, Trischitta. Analysis and interpretation of data: Spennato, Chiaramonte, Trischitta. Drafting the article: Spennato, Chiaramonte, Barbarisi. Critically revising the article: Spennato. Reviewed submitted version of manuscript: all authors. Approved the final version of the manuscript on behalf of all authors: Spennato. Administrative/technical/material support: Cicala, Donofrio, Nastro, Trischitta. Study supervision: Donofrio, Cinalli.

Supplemental Information

Videos

Video 1. <https://vimeo.com/180181444>.

Correspondence

Pietro Spennato, Department of Pediatric Neurosurgery, Santobono-Pausilipon Children's Hospital, Via Mario Fiore n. 6, Naples 80129, Italy. email: pierospenn@gmail.com.