CASE REPORTE

Spontaneous ventriculostomy of the third ventricle mimicking an arachnoid cyst of the quadrigeminal cistern: a case report

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ABSTRACT

Background: Spontaneous third ventriculostomy (STV) occurs due to spontaneous rupture of the thinned walls of the third ventricle due to hypertensive hydrocephalus, establishing direct communication from the ventricular system to the subarachnoid space. STV may resolve or diminish the clinical picture of hydrocephalus.
Case description: We present the case of a 16-year-old female patient with persistent headache, dizziness, and vomiting. Initial neuroradiological studies showed hydrocephalus. Subsequent studies showed increased hydrocephalus with the appearance of a collection in the quadrigeminal cistern, for which an endoscopic third ventriculostomy (ETV) was indicated as surgical treatment. Symptoms improved immediately after surgery and in subsequent evaluations the patient evolved asymptomatically.
Conclusion: This case shows that ETV represents a suitable surgical approach for patients with chronic hydrocephalus and stenosis of the aqueduct of Sylvius with symptoms of progressive endocranial hypertension, with neuroradiological evolution of STV. This approach decreases the complications of the ventricular cerebrospinal fluid shunt system.

Keywords: Arachnoid Cyst; Hydrocephalus; Endoscopic Third Ventriculostomy (Source: MeSH)

Ventriculostomía espontánea del tercer ventrículo simulando un quiste aracnoideo de la cisterna cuadrigeminal: reporte de caso

RESUMEN

Antecedentes: La ventriculostomía espontánea del tercer ventrículo (VET) ocurre por la ruptura espontánea de las paredes adelgazadas del tercer ventrículo debido a la hidrocefalia hipertensiva, estableciéndose una comunicación directa entre el sistema ventricular y el espacio subaracnoideo. La VET puede resolver o disminuir el cuadro clínico de la hidrocefalia.
Descripción del caso: Se presenta el caso de una paciente de 16 años con cefalea, mareo y vómitos persistentes. Los estudios neurorradiológicos iniciales mostraron hidrocefalia. Los estudios posteriores evidenciaron un incremento de la hidrocefalia con la aparición de una colección en la cisterna cuadrigeminal, por lo cual se indicó como tratamiento quirúrgico una ventriculostomía endoscópica del tercer ventrículo. Los síntomas mejoraron inmediatamente después de la cirugía y, en las evaluaciones posteriores, la paciente evolucionó de forma asintomática.
Conclusión: Este caso muestra que la ventriculostomía endoscópica representa un abordaje quirúrgico adecuado para pacientes con hidrocefalia crónica y estenosis del acueducto de Silvio con síntomas de hipertensión endocraneana progresiva, con evolución neurorradiológica de VET. Este abordaje disminuye las complicaciones asociadas al sistema de derivación ventricular de líquido cefalorraquídeo.

Palabras clave: Quistes Aracnoideos; Hidrocefalia; Ventriculostomía Endoscópica del Tercer Ventrículo (Fuente: DeCS)

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INTRODUCTION

Obstructive hydrocephalus, also known as non-communicating hydrocephalus, is characterized by an anatomical or functional obstruction of cerebrospinal fluid circulation within the ventricular system. Obstructive hydrocephalus is the most common form of hydrocephalus in children, with a prevalence of congenital forms of 0.5 to 1 case per 1,000 live births and acquired forms of 3 to 5 cases per 1,000 live births (1,2).

The accumulation of cerebrospinal fluid in the cerebral ventricles due to obstruction leads to ventricular dilatation and intracranial hypertension. The obstruction may be due to congenital stenosis of the aqueduct of Sylvian, congenital infections, synechiae secondary to previous hemorrhages, Blake's pouch cyst, Chiari type II malformation, intraventricular lesions (e.g., neoplasms of the posterior region of the third ventricle), or external compression by lesions adjacent to the ventricular system (e.g., posterior fossa tumors, brain stem tumors, and pineal region tumors) (1,3,4).

Spontaneous ventriculostomy occurs due to spontaneous rupture of the walls separating the ventricular system from the subarachnoid space in patients with chronic obstructive hydrocephalus (3,5). The rupture causes the spontaneous resolution of symptoms and a reduction in head circumference in newborns (1). De Lange first described the entity of spontaneous third ventriculostomy (STV) (6). Spontaneous ventriculostomy can occur at the level of the lamina terminalis, at the floor of the third ventricle, in the pineal recess of the third ventricle, and in the ventricular atrium (4,5,7).

STV appears as signal voids at the site of rupture on conventional sagittal magnetic resonance imaging (MRI) projections. At the same time, phase-contrast MRI (PC-MRI) can detect abnormal flow through the site of spontaneous rupture in the third ventricle (8).

There are few reports of STV, with approximately 40 cases in the literature, making it a rare condition. Even less frequent is the occurrence of rupture at the level of the posterior wall of the third ventricle within the pineal recess, which mimics an arachnoid cyst of the quadrigeminal cistern.

We present the case of a 16-year-old female patient with obstructive hydrocephalus and Sylvian aqueduct stenosis who developed a rare form of STV and was treated with endoscopic third ventriculostomy, which is the ideal treatment with good results, reducing the complications associated with the implantation of cerebrospinal fluid shunt systems.

CLINICAL CASE

We present the case of a 16-year-old female patient who began her illness with headache, dizziness, and vomiting, with episodes recurring once a week and increasing in intensity, duration, and frequency. For this reason, she was evaluated by a neurosurgeon at a private medical institution in Peru, who requested a brain CT scan, which was performed two months later. This study revealed the presence of hydrocephalus, characterized by an Evans index of 0.38 (reference value for her age: 0.237 ± 0.006), a third ventricle diameter of 1.41 cm (reference value for her age: 4.04 ± 0.16 mm), and stenosis of the aqueduct of Sylvius (Figure 1).

Two weeks after the initial CT scan, the diagnosis was confirmed by MRI, which showed the presence of non-communicating hydrocephalus, an Evans index of 0.42, and aqueductal stenosis (Figure 2).

The patient was then evaluated by the ophthalmology service, which requested an optical coherence tomography study that revealed bilateral papilledema.

One year after the onset of symptoms, after being evaluated by neurosurgeons with different opinions during that time, the patient was referred to the neurosurgery department of our national pediatric referral center for a second opinion, as she was again experiencing more severe headaches lasting up to 2 hours, nausea, and occasional vomiting.

A new brain CT scan showed non-communicating hydrocephalus, transependymal edema, and an Evans index of 0.46. A new MRI study then revealed a collection at the level of the quadrigeminal cistern, resulting in a descent of up to 5 mm of the cerebellar tonsils, stenosis of the Sylvian aqueduct, hydrocephalus affecting the lateral ventricles, transependymal edema, and an Evans index of 0.44 (Figure 3).

Upon evaluating the initial CT and MRI studies and comparing them with the latest ones, a collection of cerebrospinal fluid was observed at the level of the quadrigeminal cistern, mimicking an arachnoid cyst, evidencing a rupture of the posterior wall of the third ventricle at the level of the pineal recess.

The patient was re-evaluated by the ophthalmology service, which suggested a new optical coherence tomography and optic nerve tomography study. With these studies, the ophthalmologist concluded that the patient had papilledema and probable intracranial hypertension.

After confirming the diagnosis of hydrocephalus and Sylvian aqueduct stenosis, the endoscopic third ventriculostomy success score was assessed, and the shape of the third ventricle was evaluated in the sagittal view of the MRI to decide whether the patient was a candidate for this procedure. Immediately after confirming her eligibility, she was prepared for surgical treatment.

A non-contrast CT scan was performed prior to the endoscopic third ventriculostomy, which showed increased hydrocephalus, stenosis of the aqueduct of Sylvius, and a collection of cerebrospinal fluid at the level of the quadrigeminal cistern (7.5 cc) that displaced the cerebellum toward the foramen magnum and caused compression of the brainstem against the clivus (Figure 4).

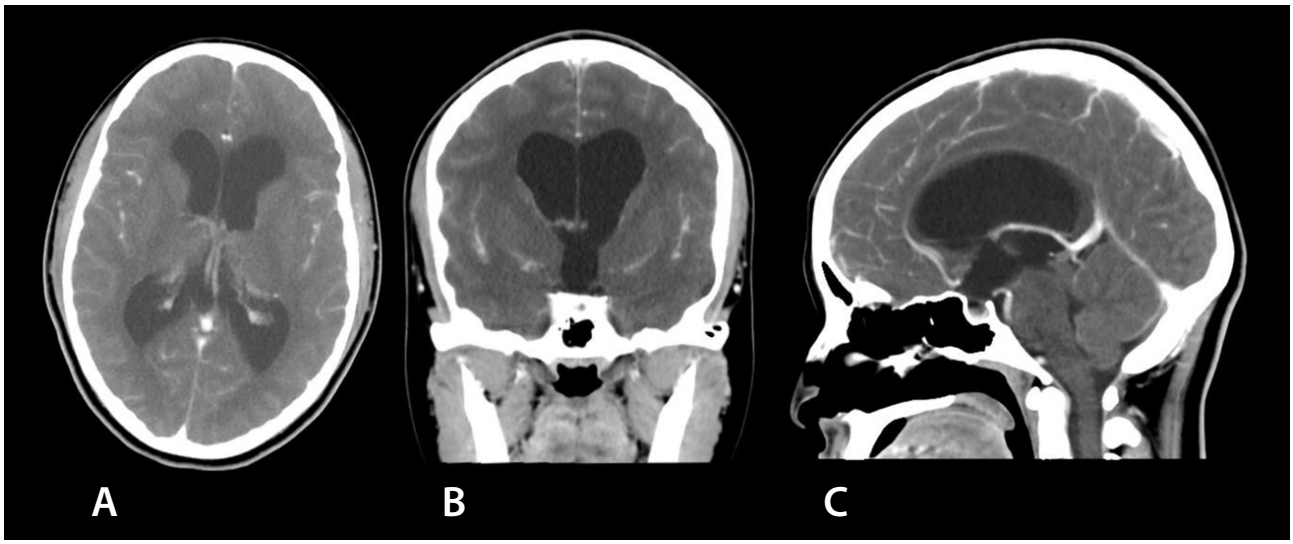

Figure 1. Contrast-enhanced brain CT scan. **A**. Non-communicating supratentorial hydrocephalus with periventricular edema. **B**. Dilation of the third ventricle. **C**. Thinning of the interthalamic adhesion; bulging of the infundibular and suprachiasmatic recesses; presence of an empty sella turcica; and stenosis of the aqueduct of Sylvius.

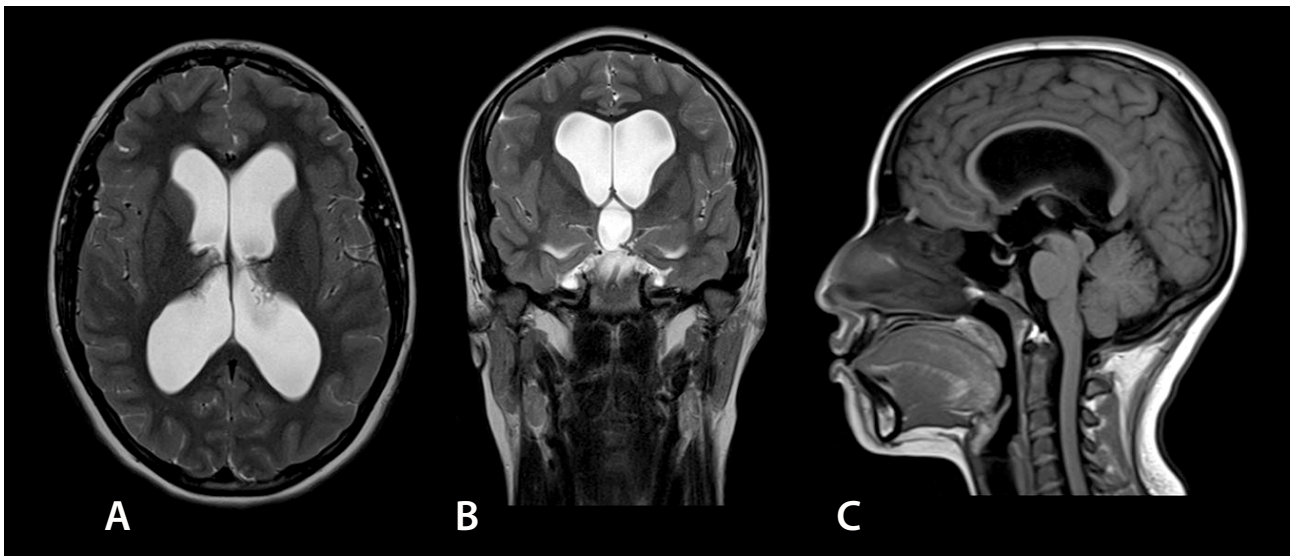

Figure 2. Brain MRI showing hydrocephalus. **A**. Non-communicating supratentorial hydrocephalus with periventricular edema predominantly affecting the frontal horns. **B**. Dilation of the third ventricle. **C**. Hydrocephalus resulting in compression of the cingulate gyrus and paracentral lobule; thinning of the corpus callosum and interthalamic adhesion; bulging of the infundibular and suprachiasmatic recesses; depression of the third ventricle floor; empty sella turcica; and aqueductal stenosis.

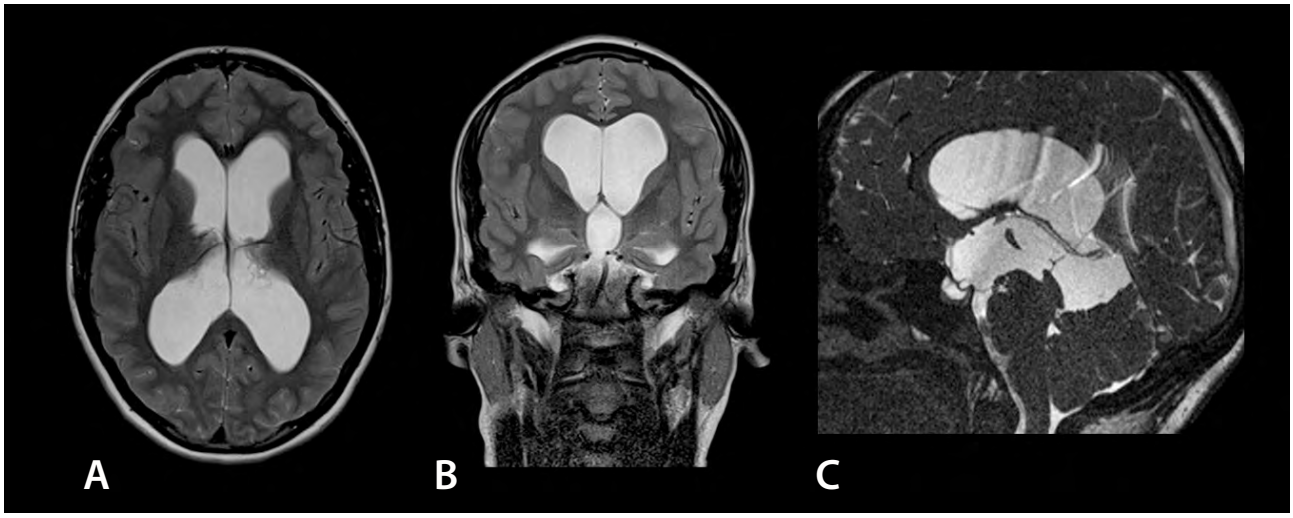

Figure 3. Brain MRI with increased hydrocephalus severity. **A**. Non-communicating supratentorial hydrocephalus with notable periventricular edema. **B**. Greater dilation of the third ventricle and temporal horns. **C**. Compression of the cingulate gyrus and paracentral lobule; thinning of the corpus callosum and interthalamic adhesion; increased bulging of the infundibular and suprachiasmatic recesses; depression of the third ventricle floor; appearance of an empty sella turcica; aqueductal stenosis; and cerebrospinal fluid collection in the quadrigeminal cistern compressing the brainstem and causing descent of the cerebellar tonsils.

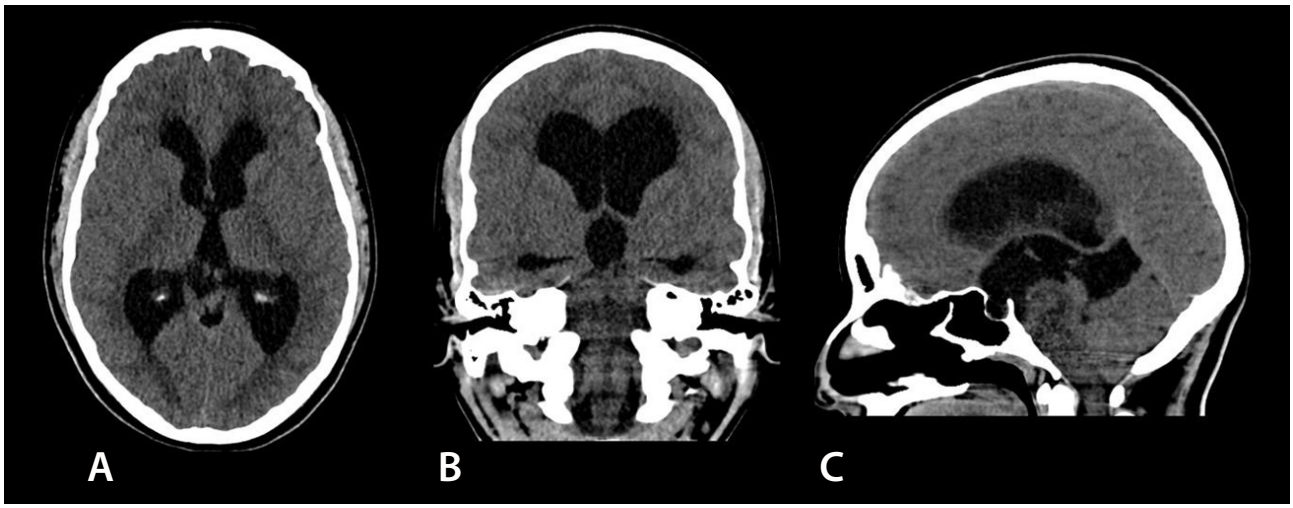

Figure 4. Non-contrast brain CT scan obtained one day before surgery. **A**. Non-communicating supratentorial hydrocephalus with periventricular edema. **B**. Dilation of the third ventricle. **C**. Bulging of the infundibular and suprachiasmatic recesses; a 7.5 cc collection of cerebrospinal fluid in the quadrigeminal cistern compressing the cerebellum and brainstem; thinning of the interthalamic adhesion; empty sella turcica; and stenosis of the aqueduct of Sylvius.

A third endoscopic ventriculostomy was performed using a rigid neuroendoscope with 30° optics, with an approach through a trephine hole, using the right pupillary line and 1 cm in front of the coronal suture as anatomical landmarks.

On the third postoperative day, the patient's favorable evolution was confirmed by a non-contrast CT scan (Figure 5), which confirmed the decrease in hydrocephalus and cerebrospinal fluid collection in the quadrigeminal cistern (2.3 cc). The patient was discharged on the fourth postoperative day with a marked decrease in headache and nausea.

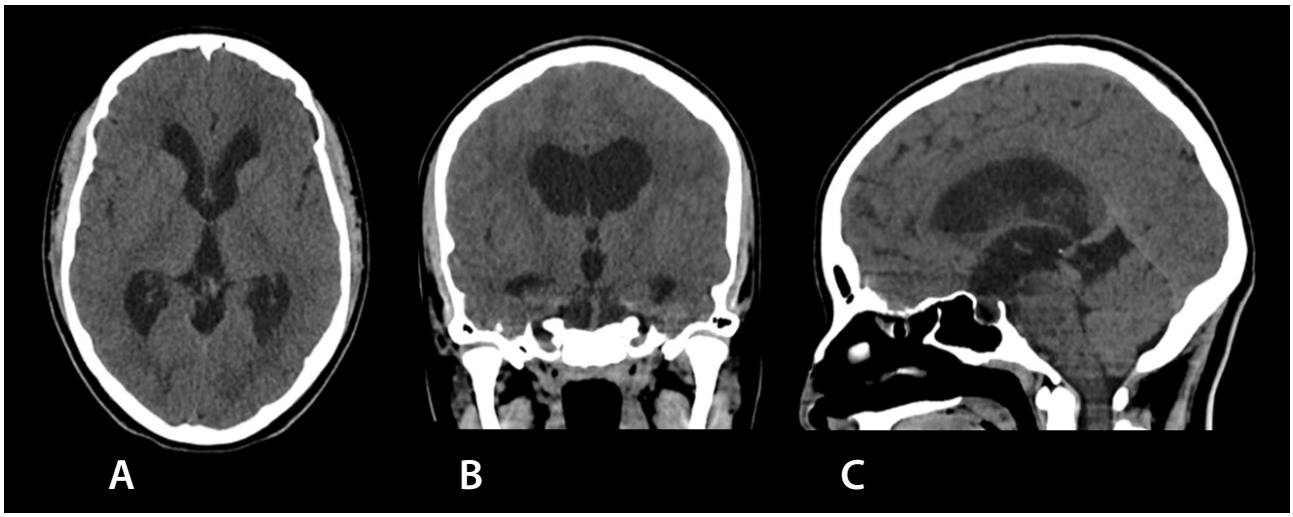

Figure 5. Non-contrast brain CT scan obtained three days post-endoscopic third ventriculostomy. **A**. Non-communicating supratentorial hydrocephalus with reduced ventricular size compared to preoperative study; cerebral sulci now visible. **B**. Decreased size of the third ventricle. **C**. A 2.3 cc decrease in the cerebrospinal fluid collection in the quadrigeminal cistern, resulting in less compression of the cerebellum and brainstem.

A follow-up MRI was performed one month after surgery (Figure 6), which showed a decrease in ventricular size and a reduction in cerebrospinal fluid collection in the quadrigeminal cistern. Additionally, a flow artifact was observed in the sagittal view, indicating the effectiveness of endoscopic third ventriculostomy.

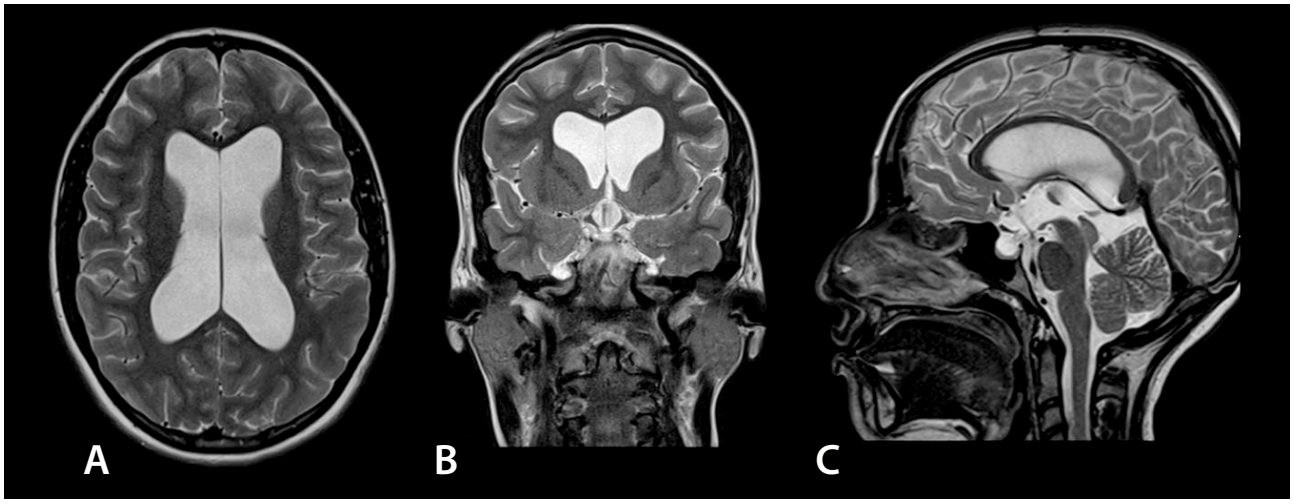

Figura 6. Brain MRI obtained one month after surgery. **A**. Smaller lateral ventricles compared to previous studies, with visible cerebral sulci and absence of periventricular edema. **B**. Reduced size of the third ventricle. **C**. Flow artifact noted through the floor of the third ventricle, with minimal cerebrospinal fluid collection in the quadrigeminal cistern, and brainstem and cerebellum positioned normally.

The patient continued with outpatient follow-up and was asymptomatic. Six months after surgery, a new MRI scan was performed, which showed no hypertensive hydrocephalus or collection of cerebrospinal fluid at the level of the quadrigeminal cistern (Figure 7).

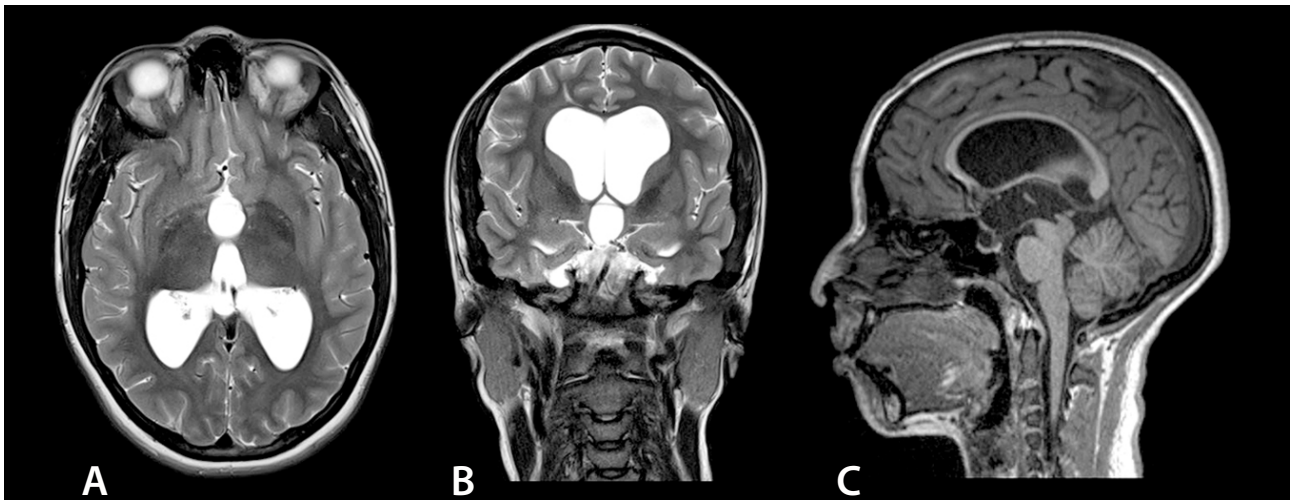

Figura 7. Brain MRI obtained six months after surgery. **A**. Markedly decreased ventricular size; visible cerebral sulci and no signs of periventricular edema. **B**. Further reduction in the size of the third ventricle compared to preoperative studies. **C**. Absence of cerebrospinal fluid collection in the quadrigeminal cistern; brainstem and cerebellum positioned normally.

CT and MRI studies performed during the diagnosis and treatment of the disease showed a progressive increase in the Evans index, reaching a peak of 0.47 one day before the endoscopic third ventriculostomy. After surgery, a progressive decrease was observed, reaching 0.38 almost 8 months later (Table 1, Figure 8).

Table 1. Variations in the Evans index pre- and post-endoscopic third ventriculostomy

Date	Evans index	Imaging modality
10/14/22	0.38	Brain CT scan
10/28/22	0.42	Brain MRI
08/11/23	0.46	Brain CT scan
08/24/23	0.44	Brain MRI
09/20/23	0.47	Brain CT scan
09/24/23	0.43	Brain CT scan
11/02/23	0.42	Brain MRI
03/24/24	0.39	Brain MRI
05/12/24	0.38	Brain MRI

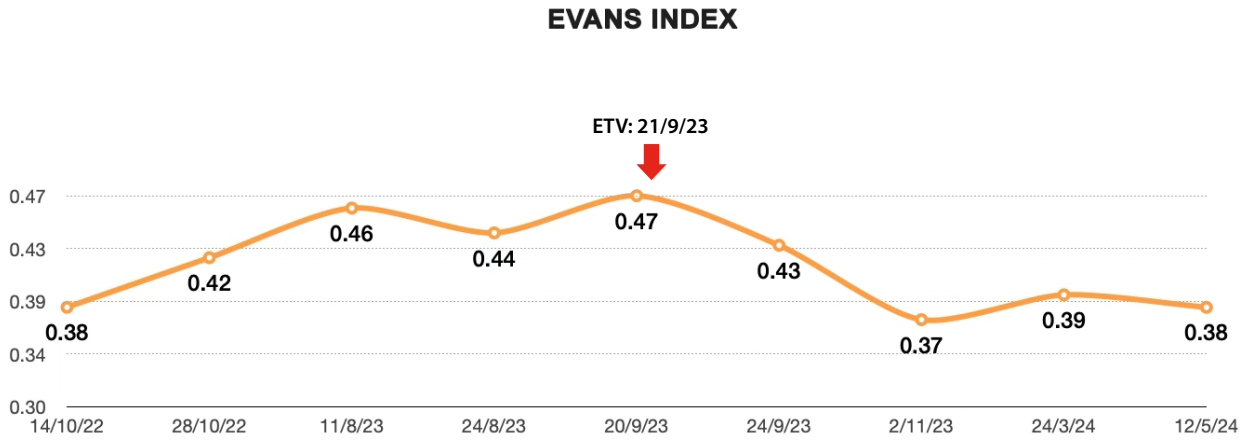

Figure 8. Temporal changes in Evans index before and after endoscopic third ventriculostomy (ETV). This figure illustrates a progressive increase in the Evans index leading up to the surgical intervention (ETV). Post-surgery, a gradual decrease in the index is observed, ultimately stabilizing at a value of 0.38.

Similarly, the evolution of the third ventricle diameter was evaluated, showing an increase to 1.8 cm one day before the endoscopic third ventriculostomy. After surgery, a decrease in the diameter of the third ventricle to 1.3 cm was observed (Table 2, Figure 9).

Table 2. Third ventricle diameter as assessed by CT and MRI imaging

Date	Third ventricle diameter (cm)	Imaging modality
14/10/22	1.41	Brain CT Scan
09/20/23	1.80	Brain CT Scan
09/24/23	1.20	Brain CT Scan
11/02/23	1.20	Brain MRI
03/24/24	1.30	Brain MRI
05/12/24	1.30	Brain MRI

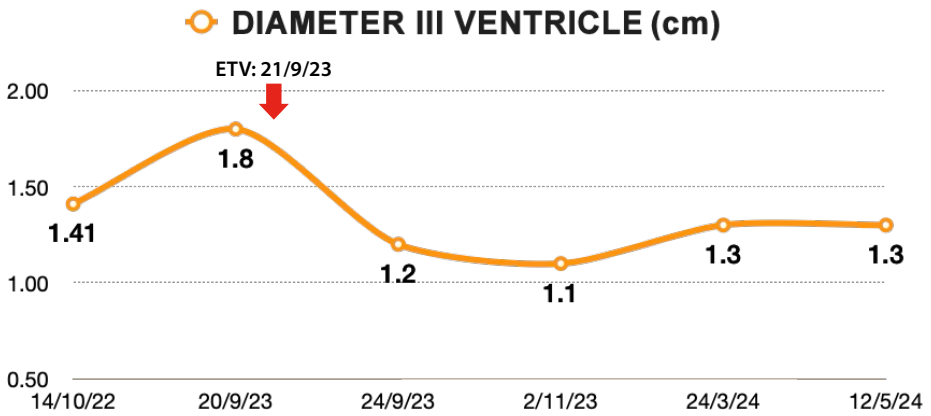

Figure 9. Temporal changes in third ventricle diameter before and after endoscopic third ventriculostomy (ETV)**, as measured by CT and MRI**. This figure demonstrates an increase in third ventricle diameter, peaking at 1.8 cm. Following the ETV procedure, a steady decrease in diameter is noted, reaching a final measurement of 1.3 cm.

DISCUSSION

Spontaneous rupture of the third ventricle due to chronic hydrocephalus remains a rare condition (approximately 1 in every 5,000 patients with chronic hydrocephalus). A limited number of cases have been reported in the literature (8). In the retrospective study by Aleem Ragab et al. (5), conducted between 2015 and 2022 and comprising the most extensive series of ETVs (14 patients), 8 presented with spontaneous rupture of the floor of the third ventricle, 5 at the level of the lamina terminalis, and 1 with two rupture sites: one at the floor of the third ventricle and another at the lamina terminalis (5).

The patient's clinical presentation was consistent with intracranial hypertension secondary to obstructive hydrocephalus, as also reported by Aleem Ragab et al. (5), whose cases predominantly presented with headache, dizziness, increased head circumference, and gait disturbances, and by Calvaheiro et al. (1), whose patients presented with tense fontanelle and increased head circumference.

The diagnosis was based on CT and MRI studies, through which the diameter of the third ventricle and the Evans' index were assessed. These data were used for both preoperative and postoperative monitoring, taking into account the reference values for age (9).

MRI is the imaging modality of choice, allowing for the evaluation of various sequences useful for ETV assessment, such as phase-contrast MRI (PC-MRI), the FIESTA protocol (Fast Imaging Employing Steady-state Acquisition), or cine-MRI, all of which can assess cerebrospinal fluid (CSF) flow, including through the ETV site (5,8,10,11). T2-weighted high-resolution sequences, in particular, enable visualization of the defect in the floor of the third ventricle and the CSF flow artifact (5,11).

In the present case, CT and MRI were performed at different centers. The MRI studies included only basic protocols with fine T2-weighted sequences, omitting specific sequences to evaluate CSF circulation. Nonetheless, phase-contrast imaging remains a valuable diagnostic tool, capable of measuring and accurately visualizing CSF flow and allowing for assessment of the ETV site. Furthermore, the flow artifact permits the evaluation of ETV functionality or patency (1,5,11).

Before surgery, the patient developed a CSF collection behind the posterior wall (pineal recess) of the third ventricle, mimicking a type II arachnoid cyst of the quadrigeminal cistern (12). Similar cases involving rupture of the posterior wall of the third ventricle with communication to the quadrigeminal cistern have been reported by Calvaheiro et al. (1) and Cinalli et al. (3).

Surgical treatment options include ETV and ventriculoperitoneal shunting (1, 4, 5). In this case, clinical evaluation was supported by CT and MRI findings. The Endoscopic Third Ventriculostomy Success Score (ETVSS) was applied, and the morphology of the third ventricle was assessed (13–16). Both tools are used to predict the success of ETV and can enhance the likelihood of a favorable surgical outcome. It was determined that the patient would benefit from ETV.

Following the procedure, the patient showed a favorable clinical course; the Evans' index and ventricular diameter decreased, although they did not reach normal age-adjusted values (9). In addition, the CSF collection in the quadrigeminal cistern, secondary to spontaneous rupture of the pineal recess caused by chronic hydrocephalus, progressively diminished after surgery, ultimately leading to complete resolution. Symptoms improved immediately postoperatively, and in subsequent evaluations, the patient remained asymptomatic.

LIMITATIONS

This case of STV complicated by a cerebrospinal fluid collection in the quadrigeminal cistern, resembling an arachnoid cyst, highlights several limitations due to the variability in neuroradiological studies conducted at different institutions and across distinct time points. Given the rarity of this case, caution is warranted when extrapolating our findings. Additionally, no data were available regarding the timing of symptom resolution relative to the occurrence of STV.

CONCLUSIONS

We report a 16-year-old female patient with chronic hydrocephalus secondary to the aqueduct of Sylvius stenosis, which progressed to STV and a cerebrospinal fluid collection at the quadrigeminal cistern, simulating a quadrigeminal cisternal arachnoid cyst.

Although STV is rare and may be overlooked as a potential progression of hydrocephalus due to aqueduct stenosis, it is crucial to consider STV or the emergence of cerebrospinal fluid collections following an improvement in symptoms associated with hypertensive hydrocephalus.

STV should not be regarded as a definitive treatment for hydrocephalus, as patients may subsequently develop hypertensive hydrocephalus and require surgical intervention, including emergency procedures.

Hydrocephalus with aqueduct stenosis may evolve with STV. In cases of hypertensive hydrocephalus, endoscopic third ventriculostomy is the preferred treatment, as it minimizes reliance on medical implants like cerebrospinal fluid shunt systems, which are associated with higher complication rates compared to endoscopic approaches.

Author contributions

The author confirms responsibility for the conceptualization and design of the study, data collection, analysis, interpretation, and preparation of the final manuscript.

Conflicts of interest

The author declares that he has no conflicts of interest associated with the material presented in the manuscript.

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This study was self-funded.

Ethical aspects

The study was conducted in accordance with the fundamental ethical principles outlined in the Declaration of Helsinki, including non-maleficence and confidentiality. All collected information was treated as strictly confidential and used solely for the purposes of this study. Authorization was obtained to review medical records and informed consent documentation. In addition, the study was approved by the Institutional Research Ethics Committee of the Instituto Nacional de Salud del Niño San Borja (CIEI INSNSB).

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