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(CASE REPORT)

A rare case of arachnoid cyst of the fourth ventricle resulting in an Isolated fourth ventricle: Case report and review of the literature

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Abstract

Introduction: Post-hemorrhagic or post-infectious isolated or trapped fourth ventricles are rare occurrences associated with cystic lesions of the V4. However, the even less frequent arachnoid cyst of the fourth ventricle poses a differential diagnosis challenge and may be easily overlooked during initial medical assessments of V4 cystic lesions.

Case Report: An 11-year-old child, previously healthy, presented with symptoms indicative of intracranial hypertension syndrome, including headaches, vomiting, and bilateral Grade II papilledema. Neuroradiological examination revealed a significantly enlarged fourth ventricle (V4) alongside small lateral and third ventricles. Initially suspected as a V4 arachnoid cyst, the patient underwent surgical intervention with a sub-occipital approach for cyst marsupialization, showing positive progress in the first postoperative month. However, one month later, intracranial hypertension symptoms recurred, and imaging indicated cyst recurrence within V4. A second procedure involved a V4-peritoneal shunt, resulting in a favorable postoperative outcome for one month. One month later, headaches and vomiting without papilledema reappeared, and imaging revealed moderate ventricle dilation. The therapeutic approach included ventriculocisternostomy alongside the V4 peritoneal shunt, leading to a favorable outcome with no signs of intracranial hypertension during follow-up consultations and a normal clinical examination.

Conclusion: Trapped fourth ventricle is a rare clinico-radiologic entity, with limited cases reported. Treatment options encompass direct microsurgical approaches, fourth ventricle outflow fenestration, and alternative treatments like fourth ventriculoperitoneal shunts. Additionally, the combination of ventriculoperitoneal shunt and endoscopic treatment (VCS) may be considered for comprehensive management.

Keywords: Isolated; Fourth ventricle; Cyst arachnoid; Rare; Surgery

1. Introduction

The isolated or entrapped fourth ventricle is a relatively uncommon neurosurgical condition recognized as a consequence of various pathological conditions. This occurs when there is an obstruction of both the Sylvian aqueduct and the exit foramina, namely Magendie and Luschka or the basal cisterns of the fourth ventricle (10). This leads to a progressive accumulation of cerebrospinal fluid (CSF), initially causing a mass effect on the cerebellum resembling a space-occupying lesion in the posterior fossa, and subsequently affecting the brainstem anteriorly. Although the exact mechanism by which the fourth ventricle becomes trapped is not fully understood, it occurs more frequently in the context of post-hemorrhagic and post-infectious hydrocephalus, as well as in patients who have undergone multiple shunt procedures (Youman's Isolated Fourth Ventricle Syndrome) (2).

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The treatment of symptomatic isolated fourth ventricle varies depending on resource availability. In advanced centers, endoscopic approaches are the primary means of treatment (1). In other settings, the insertion of a fourth ventriculo-peritoneal shunt may be considered (9), although this procedure is generally challenging.

2. Case report

This concerns an 11-year-old child, with no prior medical history, admitted for the management of intracranial hypertension syndrome presenting with headaches, vomiting, and bilateral Grade II papilledema. Neuroradiological examination revealed a significantly dilated fourth ventricle, associated with small lateral and third ventricles. The rounded shape of the fourth ventricle resulted in a ventral displacement of the brainstem (Figures 1, 2). The initial diagnostic hypothesis was a fourth ventricle arachnoid cyst, leading to urgent surgical intervention. Marsupialization of the arachnoid cyst was performed using a classic sub-occipital approach (Figure 3). Postoperative recovery was uneventful, without complications, and the patient exhibited significant clinical improvement with complete regression of intracranial hypertension symptoms.

The postoperative cerebral CT scan was favorable, demonstrating complete regression of the fourth ventricle dilation (Figure 4).

However, one month after the surgery, the child was readmitted due to the recurrence of intracranial hypertension symptoms (headaches, vomiting, bilateral Grade II papilledema). A subsequent neuroradiological investigation revealed cyst formation in the fourth ventricle (Figure 5). The decision was made to perform a aqueduct-peritoneal shunt, resulting in a highly favorable postoperative course characterized by marked improvement in intracranial hypertension symptoms and complete regression of papilledema. And the cerebral computed tomography (CT) scan is highly satisfactory (Figure 6).

After a month and a half of positive progress, the child presented again with valve dysfunction clinically manifesting as headaches and vomiting without papilledema. Imaging revealed a subtle ventricular dilation (Figure 9). Following appropriate laboratory assessments and a sterile lumbar puncture for cerebrospinal fluid analysis, the decision was made to perform a ventriculo-cisternostomy (Figure 7) in conjunction with the existing aqueduct-peritoneal shunt. The patient underwent surgery, receiving a right-sided ventriculo-cisternostomy, and experienced a good outcome without therapeutic relapse with a good follow-up cerebral CT scan (Figure 8). The child continues to be monitored in follow-up appointments, exhibiting satisfactory progress and an absence of clinical symptoms.

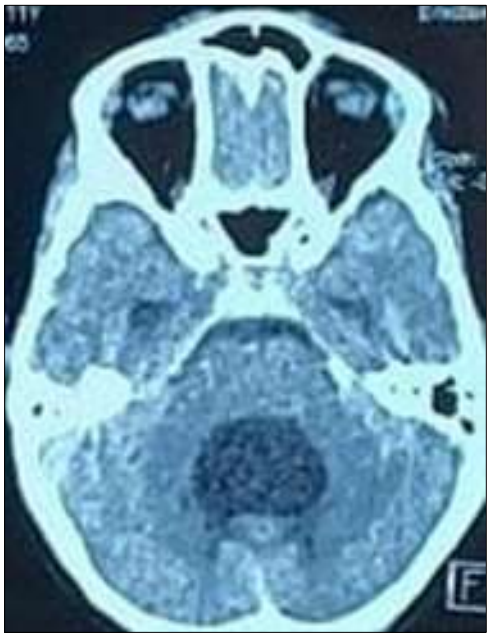

Figure 1 Initial preoperative brain CT Scan showing very large fourth ventricle

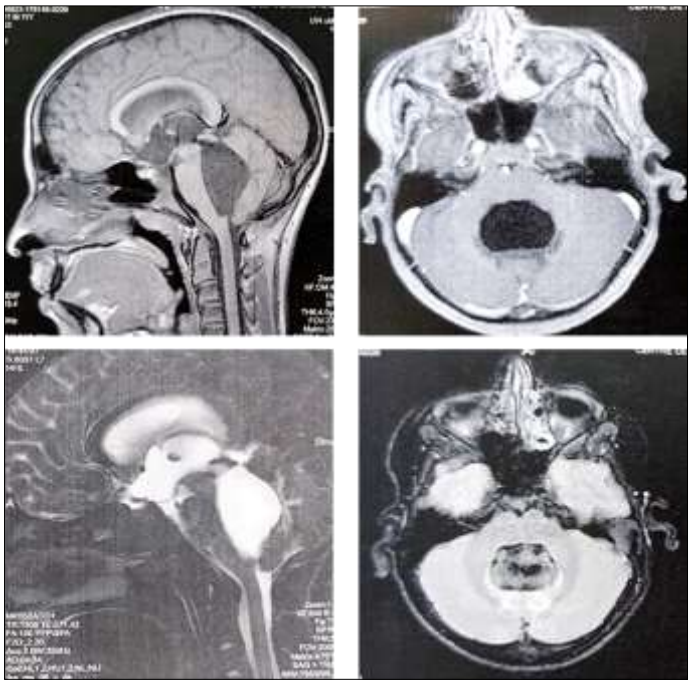

Figure 2 The preoperative MRI reveals a fourth ventricle arachnoid cyst, displaying a rounded configuration and inducing ventral displacement of the brainstem, along with altered cerebrospinal fluid (CSF) signal intensity. Sagittal T1-weighted MRI image demonstrates a hypointense arachnoid cyst compressing the fourth ventricle and cerebellum with the same signal intensity as CSF(A). Hyperintense(C) on T2-weighted images with FLAIR suppression (D)

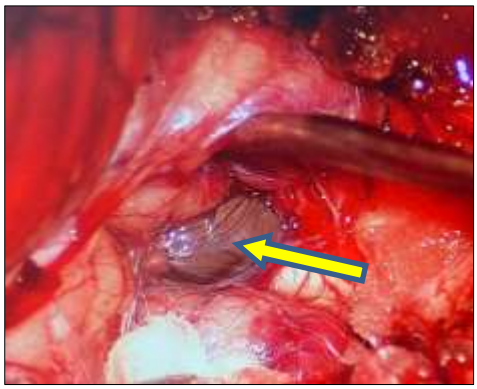

Figure 3 The intraoperative appearance of the V4 arachnoid cyst membrane (yellow arrow)

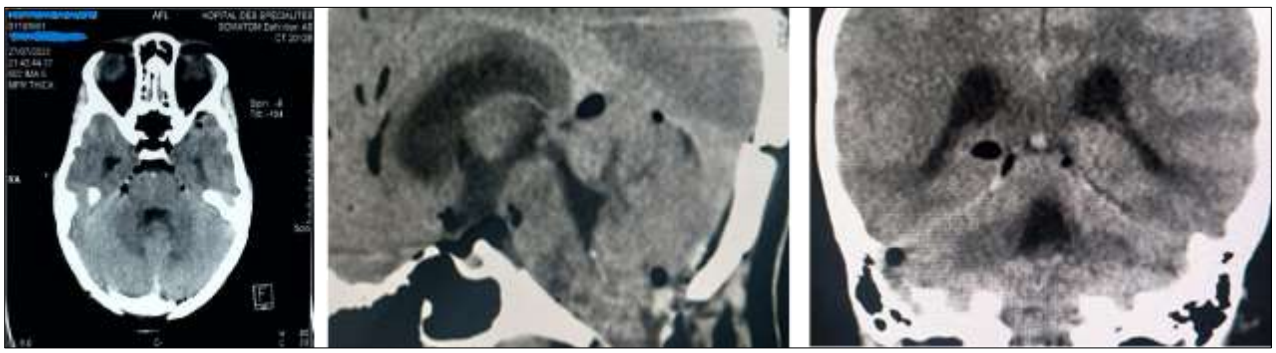

Figure 4 Post-operative cerebral CT scan without contrast, depicting a normal appearance of the fourth ventricle. Axial (A), sagittal (B), and coronal (D) sections

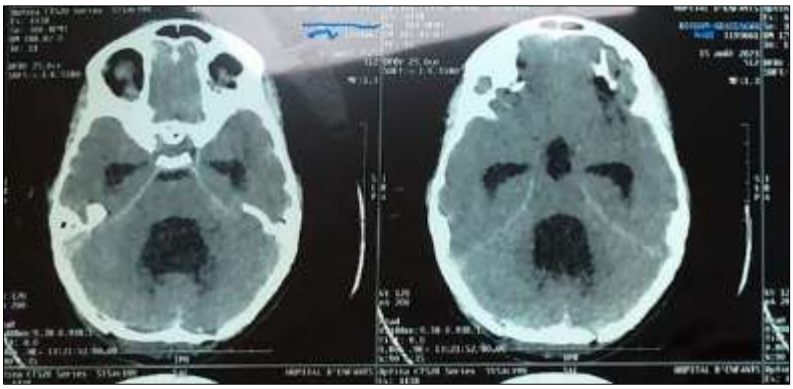

Figure 5 Brain scan without injection, showing an encystment of the 4th ventricle

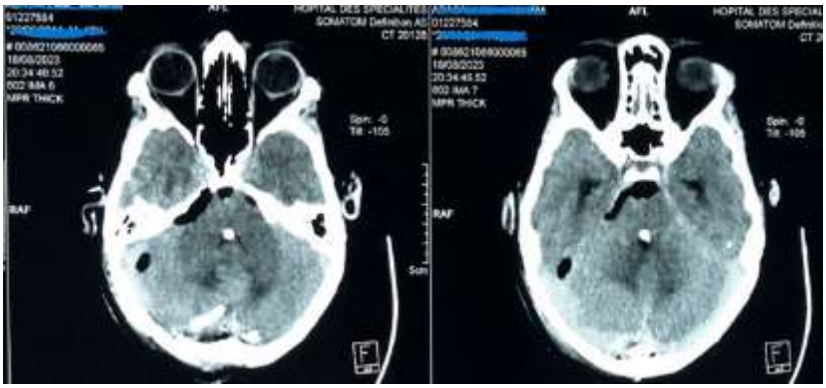

Figure 6 Control scan after surgery, which shows a clear regression of the dilation of the 4th ventricle, with a ventricular catheter in place

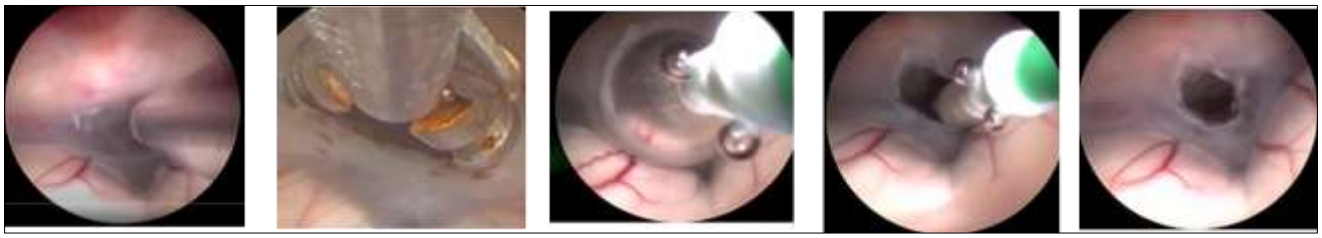

Figure 7 Perforation of the floor of the third ventricle (a,b). Balloon-dilated Stoma. (c)inflated balloon. (d) deflated balloon. (e)End-of-procedure view, stoma located on the midline midway between the infundibulum and the mammillary bodies

3. Discussion

A trapped fourth ventricle is diagnosed when it no longer communicates with the third ventricle and the basal cisterns, leading to persistent and progressive dilation. This condition, also known as isolated fourth ventricle (IFV), is rare with few cases reported (2).

It often results from occlusion due to ependymal reactions following intraventricular hemorrhage or infection, causing a buildup of cerebrospinal fluid (CSF) that exerts pressure on the cerebellum and brainstem. The exact mechanism is unclear but is frequently associated with post-hemorrhagic or post-infectious hydrocephalus and multiple shunt procedures. Other potential causes include cystic tumors and arachnoid cysts, which can be differentiated through CT scans and medical history (6).

The development of obstructive hydrocephalus due to fourth ventricle outlet occlusion can be categorized into several subcategories (Among the causes, there is the arachnoid cyst of the fourth ventricle) (7);

Arachnoid cysts are congenital, benign cystic cavities filled with cerebrospinal fluid (CSF) and are typically found between the arachnoid membranes. They occur in about 0.5-1% of the population (4), Arachnoid cysts in the posterior fossa in adults are rare and are more frequently observed in the pediatric population than in adults**,** these cysts are usually asymptomatic but can cause symptoms like headaches, nausea, and seizures if they exert pressure on surrounding brain structures. Imaging techniques such as CT and MRI are crucial for diagnosis, revealing cysts as smooth, non-calcified lesions with CSF density. Differentiating arachnoid cysts from conditions like Blake's pouch cysts and other causes of fourth ventricle obstruction is essential.

Treatment options include microsurgical excision, ventriculoperitoneal shunting, and endoscopic approaches, with the choice of method depending on the specific case and symptoms. In our case, the suboccipital approach was selected as the initial therapeutic option, followed by an aqueducto-peritoneal shunt for the fourth ventricle due to recurrence (1, 3, 5).

The placement of a fourth ventricle shunt can be done through a medial or lateral approach, which can then be connected to the peritoneal cavity (1), as is the case for our patient. Some authors recommend carrying it out using a direct stereotactic technique or under ultrasound or endoscopic guidance to enhance the safety and efficiency of the shunt, prevent potential direct traumas to the brainstem, and ensure precise placement (3,8).

In our case, it is important to emphasize that the implementation of a new shunting system in the fourth ventricle in our case significantly alleviated symptoms associated with increased intracranial pressure, without leading to complications related to the fourth ventricle shunt. The postoperative period proceeded without incident, and no complications were observed during the one-month follow-up period, and after 1 month, the recurrence of symptoms of intracranial hypertension.

The implementation of a ventriculocisternostomy in addition to the aqueduct-peritoneal shunt has significantly improved the symptoms in our case. The combination of endoscopic treatment with concomitant cerebrospinal fluid shunting can be a crucial strategy to prevent therapeutic failure (5).

4. Conclusion

Arachnoid cysts of the fourth ventricle are rare, and when they occur, they can be confused with other diagnoses, highlighting the importance of medical imaging and the patient's medical history. Restoring normal CSF circulation in the cyst through various surgical techniques enables the patient to recover.

Compliance with ethical standards

Disclosure of conflict of interest

No conflict of interest to be disclosed.

Statement of informed consent

Informed consent was obtained from all individual participants included in the study.

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