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Case Report

# Endoscopic management of a cavernous malformation on the floor of third ventricle and aqueduct of Sylvius: Technical case report and review of the literature

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## Abstract

**Background:** Intraventricular cavernous malformations are unusual intracranial vascular malformations; their deep anatomical location complicates their surgical management. Microsurgical approaches are the gold standard approaches for the resection of ventricular lesions, however, they imply considerable neurovascular risks.

**Case Description:** A 51-year-old patient presented with acute headache, diplopia, vertigo, blurred vision, and a depressed level of consciousness. A ventricular hemorrhage was treated with a ventriculostomy and the patient was discharged without hydrocephalus. After 11 days, he developed ataxia, diplopia, and a depressed level of consciousness. The patient was diagnosed with hydrocephalus secondary to the previous third ventricle hemorrhage. An endoscopic exploration using a 30° rigid ventricular endoscope was performed; after the third ventriculostomy, an intraventricular cavernous malformation located on the floor of the third ventricle and the aqueduct of Sylvius was resected.

**Conclusions:** Three days after the surgery, magnetic resonance imaging demonstrated a gross total resection and adequate third ventriculostomy flow. One year after the surgery, the patient was asymptomatic. Neuroendoscopy has evolved towards minimally invasiveness, and in selected cases is an equally effective surgical approach to ventricular lesions. It provides minimal cerebral cortex disruption and vascular manipulation.

**Key Words:** Aqueduct of Sylvius, cavernous malformation, endoscopy, intraventricular, minimally invasive, third ventricle

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## INTRODUCTION

A cavernous malformation (CM) is an unusual vascular lesion characterized by its bleeding predisposition and composed of sinusoidal vascular spaces lined by a single layer of endothelium not separated by parenchyma.[3,12,34,41] Their prevalence is estimated to be 0.4–0.6% of the population, which constitutes 8–15% among vascular malformations.[12,19,21,41,43] Even more unusual are intraventricular cavernous malformations (IVCMs), representing 2.5–10.8% of cerebral CMs.[13,24,29,44,49] The most frequent localization is in the lateral ventricles (54%), following the third (34%) and fourth (12%) ventricles.[13] The clinical findings are mostly associated with mass effect, although seizures and intraventricular hemorrhage have been reported.[13,24] The goal of treatment is gross total resection,[4] and the surgical corridor is usually selected using the two-point method.[7] Several microsurgical corridors have been successfully described for the resection of IVCMs.[10,22-24] However, only three cases[15,17,39] have reported endoscopic resection of an IVCM. This is the first case report of a purely endoscopic resection of a CM on the posterior floor of the third ventricle (TV) and aqueduct of Sylvius.

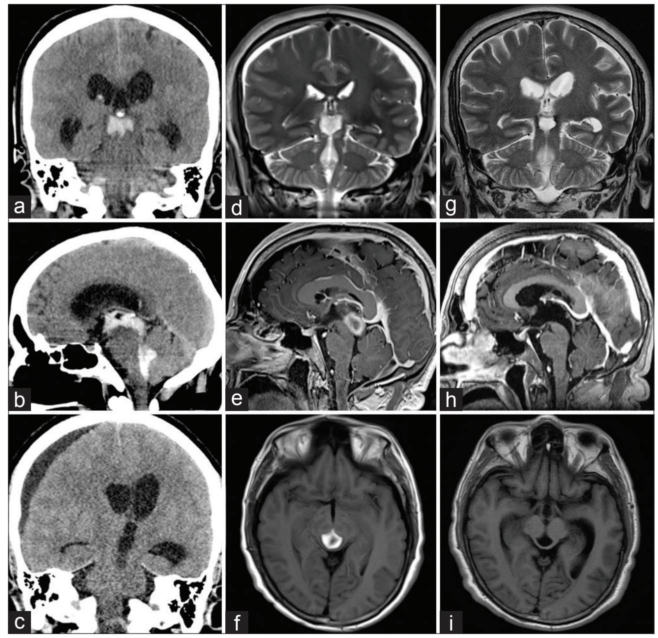

**Figure 1: (a and b) Initial coronal and sagittal CT study that demonstrated an intraventricular hemorrhage modified Graeb of 11. (c) Notice in the second preoperative CT a right subdural hygroma and obstructive hydrocephalus. (d-f) Magnetic resonance imaging (MRI) showed a late subacute hemorrhage at the posterior TV and the aqueduct of Sylvius. T2-weighted, T1-weighted simple and contrasted T1 were all hyperintense. (g-i) MRI three days after the surgery demonstrated a gross total resection**

## CASE DESCRIPTION

### Clinical findings

A 51-year-old patient presented with acute headache, diplopia, vertigo, and blurred vision. During the following hours, the level of consciousness was acutely depressed. He required ventilatory support. Computed tomography (CT) demonstrated an intraventricular hemorrhage Graeb[18] 6 and modified Graeb[35] of 11 [Figure 1a and b]. He was treated with a ventriculostomy for 9 days and was discharged 16 days after the acute hemorrhage without hydrocephalus. Magnetic resonance imaging (MRI) showed a late subacute hemorrhage at the posterior TV and the aqueduct of Sylvius, involving the pars anterior, ampulla, and pars posterior. T1-weighted, T2-weighted and gradient-echo images were all hyperintense [Figure 1]. Both T2 and gradient-echo images showed a hypointense rim owing to hemosiderin [Figure 2]. Hydrocephalus and a developmental venous anomaly (DVA) were not observed. Eleven days after the discharge, he developed ataxia and diplopia. Three days later, the level of consciousness was acutely depressed and he was admitted to our emergency department. He was unresponsive to verbal and painful stimuli. Cranial nerves revealed papilledema and Parinaud syndrome. CT revealed a right subdural hygroma [Figure 1], obstructive hydrocephalus, and an isodense lesion at the posterior TV and the aqueduct.

### Surgical technique

Neuroanesthesiology used an armored oral endotracheal tube and total intravenous anesthesia. The patient was placed supine with a 30° head elevation so that the burr hole site was at the highest point. The site of the burr hole and the trajectory was planned using preoperative MRI measurements; the trajectory was planned in between the aqueduct and tuber cinereum through the foramen of Monro. A 30° 6 mm ventricular endoscope (DECQ neuroendoscope, Karl Storz, Tuttlingen, Deutschland) was used for the procedure. An endoscopic third ventriculostomy (ETV) was performed using blunt forceps [Figure 2]. Posterior TV exploration was performed by rotating the endoscope 90° and advancing towards the massa intermedia and aqueduct. A subacute hemorrhagic lesion was observed; the classical "mulberry" appearance was not visualized. Sharp dissection and bipolar cautery was used to mobilize the lesion from the TV margins. Grasping forceps were used to dissect the lesion from the borders of the aqueduct and TV until the pars posterior and entrance into the fourth ventricle were visualized. Given the friable nature of the lesion, the clot and cavernous malformation [Figure 2] were removed in multiple pieces through the cerebral endoscopic path. Finally, endoscopic inspection revealed no residual lesion [Figure 2], and the cortex was plugged with an absorbable gelatin compressed sponge (Gelfoam, Puurs, Belgium). A ventricular catheter was not placed due to the absence of hemorrhage and the ventricular-cistern connection. The incision was closed in a multilayered fashion. Surgical time was 210 minutes and estimated blood loss was 50 ml [Video 1].

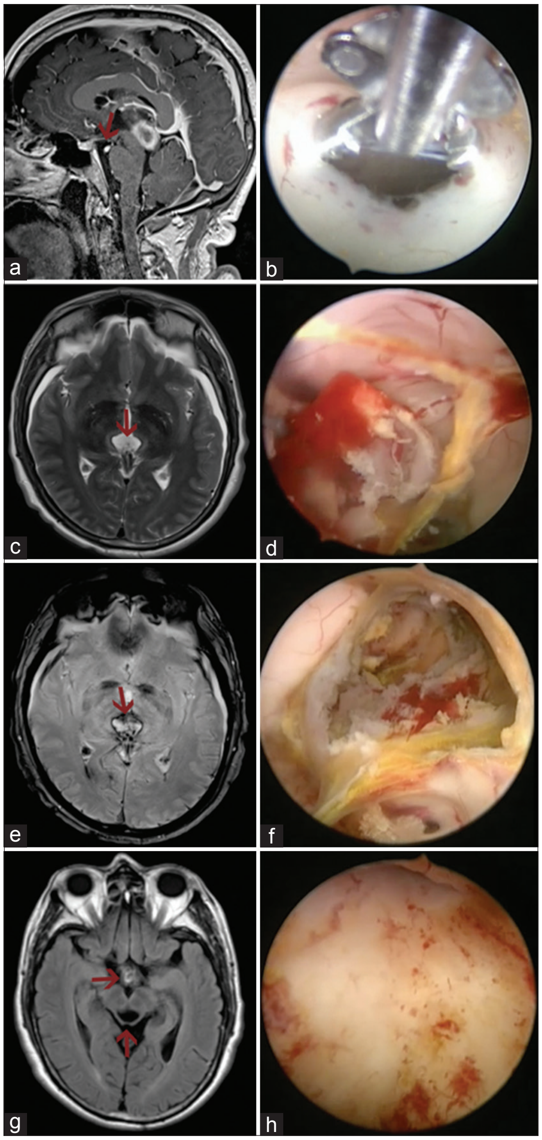


**Figure 2: (a and b) Endoscopic third ventriculostomy was performed using blunt forceps. (c-f) A subacute hemorrhagic lesion was observed. Grasping forceps and bipolar coagulator were used to dissect away the CM and clots from the borders of the aqueduct and TV. Notice that T2 and gradient-echo images showed a hypointense rim due to hemosiderin. (g and h) Endoscopic inspection revealed no residual lesion. Postoperative MRI demonstrated a gross total resection (caudal arrow) and adequate third ventriculostomy flow (rostral arrow)**

## Postoperative course

Immediately after the surgery, the patient was awake and his ocular movements improved. The patient was discharged on the fifth postoperative day. MRI obtained 3 days after the surgery demonstrated a gross total resection and adequate third ventriculostomy flow [Figure 1]. One year after the surgery, the patient was asymptomatic modified rankin scale (MRs 0). Neuropathology reported a cavernous malformation surrounded by inflammatory and granulation tissue associated with hemosiderin deposition.

# DISCUSSION

## Surgical approach

Microsurgical approaches have been considered the gold standard for managing intraventricular lesions.[11,15] However, improvements of neuroendoscopic image quality, surgical techniques, and instruments have increased effectiveness and safety during endoscopic procedures. TV approaches can be divided into three categories – anterior, lateral, and posterior routes.[10] The trajectory is selected based on tumor characteristics (location, origin, extension, vascular supply, and size), suspected diagnosis, clinical findings, hydrocephalus, and surgical experience.[10,48]

The TV can be accessed from above using either an interhemispheric anterior transcallosal approach (IATcA) or a frontal transcortical approach (FTA). Once within lateral ventricle, it is possible to reach the TV through several pathways – transforaminal, transchoroidal, subchoroidal, or interforniceal.[47,50] In all of them, complexity and depth of TV lesions put unaffected neurovascular structures (fornix, thalamostriate, and internal cerebral veins) at risk, which means potential temporal or permanent deficits involving memory, consciousness, strength, and sensibility.[47,50] Furthermore, several variants of the subfrontal approach can be used to reach the anterior TV through the lamina terminalis, which is suitable for lesions that occupy the anterior and inferior part of TV.[25,26,28]

Three main approaches have been described to reach the posterior TV through the velum interpositum of the suprapineal recess: the supracerebellar infratentorial approach (ScItA), the interhemispheric posterior transcallosal approach (IPTcA), and the occipital transtentorial approach (OTtA).[5,31] The risk of complications depends on the approach. In all the approaches, posterior deep veins (basal Rosenthal veins, internal cerebral veins, and Galen vein) are at risk, IPTcA adds memory and disconnection syndrome risk and the OTtA puts the visual cortex at risk.[5]

Although microsurgical techniques to reach intraventricular lesions with less morbidity have improved, the endoscopic transfrontal approach offers interesting benefits.[15] First, the cerebral parenchyma is minimally disrupted by the endoscope. Second, cortex retraction is negligible. Third, a keyhole bone access is used instead of a wider craniotomy. Fourth, use of natural anatomical cavities (ventricles and foramen of Monro) as working corridors without manipulation of the roof of the TV. Fifth, safe and accurate third ventriculostomy can be performed. Finally, a wide-angled view from a "fish-mouth" lens provides excellent illumination and image.[9,15,27] Endoscopic transforaminal[42] or transcavum interforniceal[45] approaches have been described to reach

TV lesions. Regarding surgical planning, it is critical to define surgical objectives, determine the best entry point based on trajectories, and to be prepared with the microscope for any potential complications. In our opinion, it is better to start with the simplest procedures, so before biopsy or tumor resection we always start performing the ETV. For posterior ventricular exploration, a 30° endoscope is preferred because the visualization angle is improved. It is important to consider that during posterior exploration, gentle traction is applied to the ipsilateral fornix, so when the endoscope is pulled back, it is important to follow the same entrance movements. Frameless stereotactic navigation can be useful for accurate anatomical orientation.[46] The risk of intraoperative intraventricular hemorrhage is the main concern during intraventricular tumor resection. The bleeding could hinder the vision, and if it is not adequately controlled, it is impossible to continue with the procedure. This aspect is particularly important with vascular lesions such as IVCMs. Active bleeding was minimal from this lesion, continuous irrigation using warm Hartman's solution is enough in the majority of cases, but bipolar cautery is required for hemostasis and dissection. When the bleeding is profuse, it is recommended to leave a ventricular drain in place at the end of the procedure.[9] Approximately 25% of CMs are associated to DVAs,[1,52] it is important to identify them preoperatively because its obliteration carries a significant risk of venous infarction.[2,6,40] Even though coagulation of the associated large transcerebral vein has been proposed to avoid recurrent or *de novo* malformations,[52] the potential morbidity around the TV outweighs the risk of recurrence and does not support DVA resection or coagulation.

## Intraventricular cavernous malformations

Faropoulos *et al*.[13] reported the most recent IVCMs review, which involved 91 cases. In the last years, ten new IVC cases have been published, giving a cumulative total of 101 cases.[8,14,20,30,32,33,36-38,51] Thirty-six percent (36/101) of the reported cases had their location in the TV. Of the 101 IVC cases, 3 were successfully treated using purely endoscopic means.[15,17,39] Gaab and Schroeder described the first trigonal CM in 1998; unfortunately, the patient had permanent memory loss attributed to forniceal injury. No serious bleeding was reported.[15] A decade later, Prat and Galeano performed the endoscopic resection of a CM located in the foramen of Monro. They reported postoperative transient recent memory loss and negligible bleeding that was controlled with irrigation and bipolar coagulation.[39] In 2013, Giannetti published a thalamic CM case that was successfully resected without surgical complications and had a good clinical outcome, however, the patient was not operated of a second midbrain

**Table 1: Surgical reports of IVCM resected using ventricular endoscopy**

| Author and year | Age, sex | IV Location | Clinical Findings | Hydrocephalus/ ETV | Endoscope | Endoscopic approach | Resection | Follow-up (months) | IVCM Status | Outcomes |
|---|---|---|---|---|---|---|---|---|---|---|
| Gaab and Schroeder, 1998[16] | 44, F | Trigone | Headache, nausea and vomiting | -/- | Rigid | Transcallosal | Total | 19 | NR | Permanent memory loss |
| Prat and Galeano, 2008[39] | 56, M | Foramen of Monro | Progressive headache and confusion | +/- | Rigid | Transcortical | Total | 1 | NR | Transient recent memory loss that resolved within 1 month after surgery |
| Giannetti, 2012[17] | 56, M | Midbrain and dorsum thalamus | Confusion, gait, and visual acuity disturbances. Disorientation, papilledema, bilateral medial rectus paresis, and dysmetria | +/+ | Rigid | Transcortical | Total (thalamic)/ not removed (midbrain) | 28 | NR (thalamic)/ enlargement (midbrain) | Improved mental status. Gaze palsy persisted. Visual acuity and gait had partial recovery. Cerebellar symptoms worsened in the last 6 months of follow-up |
| Present case, 2016 | 51, M | Floor of TV | Headache, diplopia, vertigo, and blurred vision. Papilledema and Parinaud syndrome | +/+ | Rigid | Transcortical | Total | 12 | NR | Asymptomatic |

*IVCM: Intraventricular cavernous malformation, IV: Intraventricular, ETV: Endoscopic third ventriculostomy, F: Female, M: Male, NR: No recurrence, TV: Third ventricle, +: Present, -: Absent

CM and the cerebellar symptoms worsened in the last 6 months of follow-up.[17] To our knowledge, this is the first endoscopic resection of a CM located at the posterior floor of the TV and aqueduct [Table 1]. In our case, no significant bleeding was observed and no memory dysfunction was reported. Surgery is considered the gold standard for the treatment of IVCs, and the best clinical outcome is obtained when total resection is achieved.[13] In all the previously described endoscopic cases, total resection was attained [Table 1].

## CONCLUSIONS

Ventricular endoscopy should be considered as a useful minimally invasive technique for some well-selected patients harboring intraventricular lesions.


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### Conflicts of interest

There are no conflicts of interest.

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