



Original Article

Endoscopic resection of the third ventricular epidermoid cysts: A case review and review of literature

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ABSTRACT

Background: Epidermoid cysts are benign, congenital lesions that originate from ectodermal cells, they are most commonly found in the cerebellopontine angle, but rarely in the ventricular system. There is limited literature regarding the different microsurgical techniques utilized to approach these lesions.

Methods: A 63-year-old female with a recurrent third ventricular epidermoid cyst underwent gross total resection utilizing an endoscopic technique. We also reviewed the various endoscopic/microsurgical approaches and outcomes reported in literature.

Results: We identified 15 cases, including our own. Nine of these were managed using microsurgical techniques, while six cases (including ours) were treated endoscopically; gross total resection was achieved in 10/15 cases (67%). Most commonly, surgeons utilized the interhemispheric transcallosal approach (five cases). Tumor recurrence was seen in two cases. Complications attributed to these resections included: disorders of the hypothalamic-pituitary axis (3) – diabetes insipidus, galactorrhea, and hypopituitarism; tumor recurrence (2); aseptic meningitis (1); and a transient Korsakoff syndrome (1).

Conclusion: Epidermoid cysts of the third ventricle are exceedingly rare, and surgical resection is generally well tolerated. Microsurgical transcortical, transcallosal, and endoscopic approaches each have advantages and disadvantages, and are associated with unique procedure-specific complications. Continuing technological improvement would favor endoscopic approaches for resection of tumors of the third ventricle in the future.

Keywords: Epidermoid cyst, Neuroendoscopy, Third ventricle

INTRODUCTION

Epidermoid cysts are benign, congenital lesions that originate from ectodermal cells.^[3,19] They are most commonly found in the cerebellopontine angle and are only rarely seen in the ventricular system.^[16,19,25] Symptoms associated with these lesions included: headaches, nausea, vomiting, and visual disturbances caused by increased intracranial pressure due to obstructive hydrocephalus. When symptomatic, the definitive treatment for epidermoid cysts is surgical resection. While many different microsurgical techniques have been described for resecting these lesions, neuroendoscopic approaches have not been as thoroughly evaluated. Here, we report a case of a 63-year-old female presenting with confusion and somnolence attributed to a recurrent epidermoid cyst of the third ventricle and its effective treatment utilizing an endoscopic technique, while also reviewing the appropriate literature.

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Clinical presentation

A 63-year-old female presented with several days of confusion/ altered level of consciousness. She had a Glasgow Coma Score of 14, was lethargic, and demonstrated a left homonymous hemianopsia. In 2015, she had undergone a craniotomy for subtotal resection of an epidermoid cyst. Now, the computed tomography (CT) scan revealed a large mixed-density suprasellar mass effacing the third ventricle, with a notable increase in the size of the cyst [Figure 1].

Surgical management

Through a small, elliptical right frontal craniotomy and corticectomy, the third ventricle was entered using an 11.5 mm Thoracoport with the assistance of stereotactic navigation. An endoscope was placed down the port and through the foramen of Monroe; tumor was identified as adherent to the floor of the third ventricle. An Angiocath, attached to a syringe, was utilized to drain the cyst. Pulsatile bacitracin/normal saline irrigation was then utilized to flush the ventricular system, and the cyst contents were gently suctioned and removed. Lilliequist's membrane was identified, and additional tumor was noted to be deep to the membrane. When it was opened, keratin fragments were mobilized with pulsatile irrigation. Endoscopic biopsy instruments then afforded removal of more adherent tissue. The remaining fragments that had settled in the occipital horn were also mobilized and removed. A ventricular catheter was then left in place as the Thoracoport was removed. Postoperatively, the patient exhibited a left gaze preference (without a residual field cut) that persisted. Imaging consistent with gross total resection [Figure 2]. Her GCS improved from 14 to 15. The external ventricular drain (EVD) was successfully removed on postoperative day 3, and she was discharged back to the nursing facility on day 6. Four weeks later, she developed a pseudomeningocele over the craniotomy site that was treated with a head wrap for 1 week. Her gaze preference was noted to have resolved on outpatient

follow-up. Imaging of 4 month followup demonstrated no signs of recurrence [Figure 3].

DISCUSSION

Review of literature

A review of literature, including our case, yielded 15 cases undergoing resection of the third ventricular epidermoid cysts [Table 1]. Of these, nine lesions were removed microsurgically, while six were approached endoscopically. Patients averaged 40.1 years of age and included eight males and seven females. Hydrocephalus was noted in all but one of the cases. For the one patient without hydrocephalus, the majority of the tumor was within the cavum septum pellucidum (e.g., did not obstruct normal cerebrospinal fluid [CSF] flow). Gross total resection was achieved in 10/15 cases (67%). Transcallosal approaches achieved gross total resection in 3/5 (60%) cases, while endoscopic approaches yielded gross total resection in 5 of 6 cases (83%). Gross total resection was also achieved utilizing the supracerebellar infratentorial and transventricular approaches. The three transbasal, parieto-occipital transcortical, or endoscopic transcortical approaches all resulted in subtotal resections; >95% of the tumor was removed, as residual tumor was adherent to major neurovascular structures (e.g. vein of Galen and posterior choroidal artery). Similarly, Kumar reported leaving residual tumor due to its adherence to the floor of the third ventricle.^[14] Further, Gaab and Schroeder reported a gross total resection with wide margins; however, the cyst recurred despite complete removal.^[6,7] In all but one case, hydrocephalus resolved. In cases where an EVD was used postoperatively, there were no reported cases of aseptic meningitis.

Anatomy and pathophysiology of epidermoid tumors

Epidermoid cysts are slow-growing, benign lesions that comprise 0.2%–1.8% of all intracranial tumors.^[15] A review of 432 cases of intracranial epidermoid cysts revealed that only

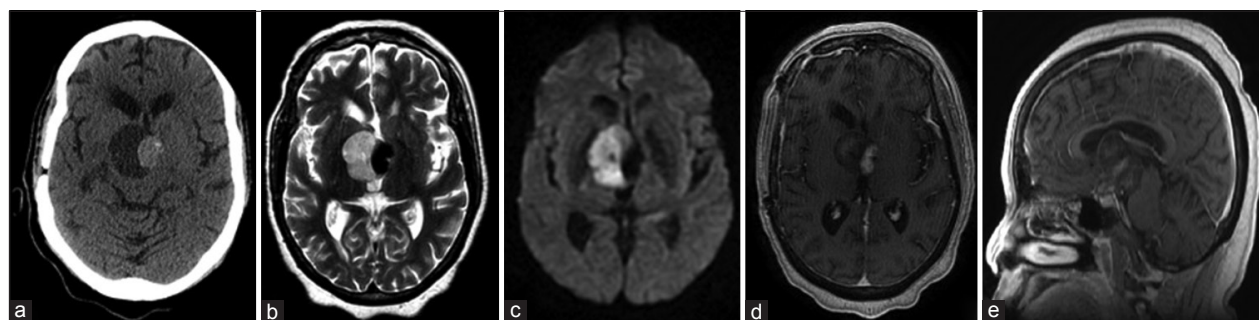


Figure 1: Preoperative – *(a) Noncontrast computed tomography head demonstrating epidermoid cysts of the third ventricle. (b) Axial T2 sequence showing the cystic and solid components of the tumor. (c) Diffusion-weighted imaging showing the characteristic diffusion restriction associated with epidermoid cysts. (d) T1 postcontrast axial sequence showing enhancement of the solid component of the cyst with minimal enhancement within the cystic component. (e) T1 postcontrast sagittal sequence showing occupation of the third ventricle by the tumor.

Table 1: Review of literature on epidermoid cysts in the third ventricle.

Authors	Age/Sex	Symptoms	Location in the third ventricle	Surgical technique	Gross total resection	Preoperative hydrocephalus	EVD preoperative	EVD/VP shunt postoperative	Complications
Smith and Chamoun, 2014 ^[27]	51, male	Headaches, confusion, disorientation, memory difficulties	Posterior third	Endoscopic transcortical	Yes (residual capsule)	Yes	No	EVDx2-day postoperative	None
Santosh <i>et al.</i> , 2001 ^[24]	23, female	Mastalgia and galactorrhea	Posterior extending to the right thalamic region	Parieto-occipital transcortical	No	N/A	No	No	Galactorrhea improved, but persisted
Paz <i>et al.</i> , 2017 ^[9]	50, female	Drowsiness, disorientation, apathy	Anterior third extending through foramen of Monroe	Endoscopic transcortical	No	Yes	Yes	No	None
Kontoangelos <i>et al.</i> , 2013 ^[2]	32, male	Severe headache, fatigue, depressed mood, insomnia, reduced ability to think/concentrate	Posterior with obstruction of cerebral aqueduct	Endoscopic transcortical+ETV	Yes	Yes	No	No	None
Shimizu <i>et al.</i> , 1985 ^[26]	15, male	Severe headache, nausea	Complete obstruction extending into preoptine cistern	Interhemispheric transcalsal	Yes	Yes	Yes	No	Mild diabetes insipidus, subdural effusion
Iaconetta <i>et al.</i> , 2001 ^[11]	25, male	Headache, visual disturbances, bilateral papilloedema	N/A	Supracerebellar infratentorial	Yes	N/A	No	No	None
Riviérez <i>et al.</i> , 1998 ^[23]	36, male	Headache, memory disturbances	Entire third ventricle developing mainly from the left side	Trans-ventricular approach	Yes	Yes	No	VP shunt	Aseptic meningitis, transient Korsakoff's syndrome, hypopituitarism
Koumtchev <i>et al.</i> , 2002 ^[13]	34, male	Headache, double vision, staggering gate, recurrent tumor	Anterior third obstructing both foramen of Monroe	Endoscopic transforaminal	Yes	Yes	No	No	None
Bikmaz <i>et al.</i> , 2007 ^[1]	29, male	None (incidental discovery due to head trauma)	Third ventricle, cavum septum	Interhemispheric transcalsal	Yes	No	No	No	None
Kumar <i>et al.</i> , 2014 ^[4]	40, male	Diabetes insipidus, headache, mild bilateral papilledema	Anterior third	Transbasal translamina terminalis	No (near total)	Yes	No	No	None

(Contd...)

Table 1: Continued

Authors	Age/Sex	Symptoms	Location in the third ventricle	Surgical technique	Gross total resection	Preoperative hydrocephalus	EVD preoperative	EVD/VP shunt postoperative	Complications
Hendricks <i>et al.</i> , 2016 ^[10]	36, male	Hemiparesis, gait disturbance, recurrent tumor	Complete, extending into lateral ventricles and suprapineal region	Interhemispheric transcalsal	No	Yes	No	No	N/A
Gelabert-Gonzalez <i>et al.</i> , 2002 ^[8]	68, female	Headaches, nausea, vomiting	Complete third ventricle	Interhemispheric transcalsal	Yes	Yes	No	No	None
Giannotta <i>et al.</i> , 1976 ^[9]	N/A, female	Headache, mental disturbances	N/A	Interhemispheric transcalsal	No	N/A	N/A	N/A	N/A
Gaab and Schroeder, 1998 ^[6]	61, female	Headache, confusion, gait disturbance	Anterior	Endoscopic transcortical	Yes	Yes	No	EVD postoperative	Recurrence of epidermoid cyst at 12 months
Our case	63, female	Lethargy, confusion, decreased left visual field	Anterior, extending into lateral ventricles, sella	Endoscopic transcortical	Yes	Yes	No	EVD×3-day postoperative	None

EVD: External ventricular drain, VP: Ventriculoperitoneal

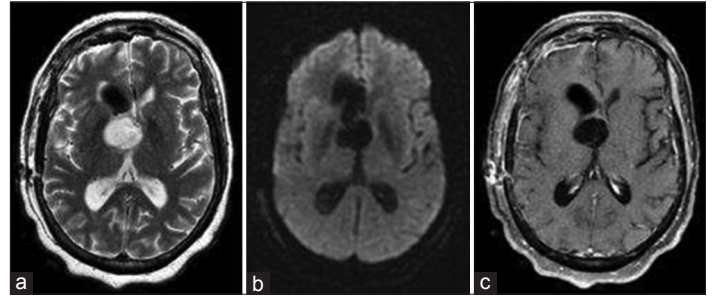


Figure 2: Postoperative day 1 – Postoperative imaging demonstrating gross total resection on (a) T2, (b) diffusion-weighted imaging, and (c) T1 postcontrast axial sequences. Expected intraventricular air is also seen.

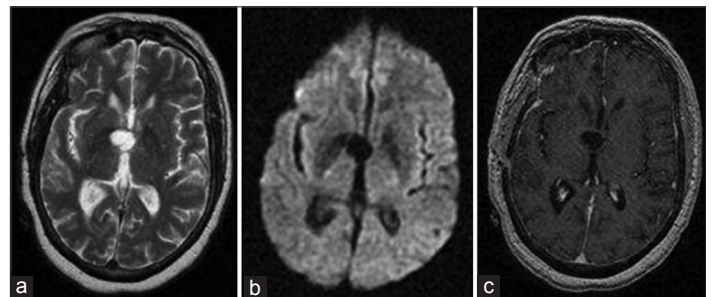


Figure 3: Postoperative (4 months later) – 4-month postoperative imaging showing gross total resection on (a) T2, (b) diffusion-weighted imaging, and (c) T1 postcontrast axial sequences without any signs of recurrence.

0.7% were located in the third ventricle.^[16] Their intracerebral or intraventricular locations result from segregation of epithelium at the 3rd week of embryogenesis, rather than the 5th week.^[22] Their bulging, irregular shape is formed by squamous epithelium, while desquamation of the cyst wall produces a liquid interior composed of keratin, water, and cholesterol – giving them a pearly white appearance intraoperatively.^[18,28] Yaşargil *et al.* described these tumors as “flowing” into any available subarachnoid spaces – defining the extra-axial nature of these lesions. Due to their slow-growing nature, the peak incidence of these tumors is in the fourth decade.^[4] Most patients are asymptomatic, with patients reporting vague symptoms such as headache, feeling “foggy,” or blurry vision.^[11,16]

Clinical, radiographic diagnosis, and treatment of the third ventricular epidermoid tumors

These lesions are typically incidental findings on CT, but magnetic resonance imaging is often the optimal confirmatory test.^[15,18,22] Both display hypointensity on T1 and hyperintensity on T2; however, epidermoid cysts will exhibit diffusion restriction on diffusion-weighted imaging while arachnoid cysts typically do not.^[17,18] Those with hydrocephalus can be effectively treated with ventriculoperitoneal shunts, but this does not resolve the underlying tumor progression. Where surgical resection is

indicated, it is the best to avoid placing a ventriculostomy, due to the risk of aseptic meningitis from entry of epidermoid crystals into the CSF. Optimally, gross total resection and cyst removal should be performed to avoid lesion and cyst recurrence.

Surgical approaches to the third ventricle

There are three essential approaches to the third ventricle: transcortical, transcallosal, and endoscopic. The transcortical approach traditionally utilizes entry to the ventricle through a right frontal trajectory. There are, however, three variations of this approach utilizing a frontal, parietal, or middle temporal gyrus trajectory.^[5] The interhemispheric transcallosal approach, first described by Dandy, in 1915, employs an entry through the midline utilizing gravity to aid in brain retraction and to minimize any physical retraction required for exposure.^[20] Notably, it provides maximal exposure to the third ventricle. Nevertheless, this route poses an increased risk of damage to the superior sagittal sinus and bridging veins, and the fornix (e.g., causing memory loss). Although Apuzzo and Litofsky reported a seizure rate of 25% with transcallosal versus just 8% with transcortical operations in their series of 127 Patients.^[23] The neuroendoscopic technique was first described in 1983 by Powell *et al.* and was modernized by Gaab and Schroeder, in 1998.^[2,6,7] The standard endoscopic approach to the third ventricle utilizes a right frontal trajectory, decreasing the amount of eloquent tissue that must be passed to reach the third ventricle. The major limitation of this technique includes the limited flexibility in the field of view and the limited number of instruments that can be passed through the endoscope. In the cases reviewed, the endoscopic approach has yielded four cases, including our own, of total resections of the cysts with no postoperative complications. We believe this approach minimizes damage to deep white matter structures and the Thoracoport-assisted flexible endoscopes, and higher definition endoscopes provide better maneuverability to visualize the surrounding structures.

CONCLUSION

There are various microsurgical and endoscopic techniques for approaching epidermoid cysts/tumors in the third ventricle. The aim is to limit disruption of normal tissue and injury to the limbic system, while still providing adequate surgical exposure. In our case and reviewing the literature, we noted gross total resection of these lesions could be achieved in 83% of endoscopic cases (5/6), compared to 3/5 utilizing the interhemispheric approach. Further, advances in endoscopy may eventually establish this technique as the new standard of care for the surgical management of the third ventricular epidermoid cysts.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given his consent for

his images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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