Neuroendoscopic fenestration of gliependymal cysts to the ventricle: report of 3 cases

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Gliependymal cysts are rare congenital lesions of the central nervous system. Reported surgical treatments of these lesions have varied and yielded mixed results, and the optimal surgical strategy is still controversial. The authors here report the clinical and surgical outcomes for three adult patients successfully treated with neuroendoscopic fenestration into the ventricular system. The patients had presented with symptomatic gliependymal cysts in the period from 2013 to 2016 at the authors' institution. All underwent minimally invasive neuroendoscopic fenestration of the gliependymal cyst into the lateral ventricle via a stereotactically guided burr hole. Presenting clinical and radiological findings, operative courses, and postintervention outcomes were evaluated.

All three patients initially presented with symptoms related to regional mass effect of the underlying gliependymal cyst, including headaches, visual disturbances, and hemiparesis. All patients were successfully treated with endoscopic fenestration of the cyst wall into the lateral ventricle, where the wall was thinnest. Postoperatively, all patients reported improvement in their presenting symptoms, and neuroimaging demonstrated decompression of the cyst. Clinical follow-up ranged from 4 months to 5 years without evidence of reexpansion of the cyst or shunt requirement.

Compared to open resection and shunting of the cyst contents, minimally invasive endoscopic fenestration of a gliependymal cyst into the ventricular system is a safe and effective surgical option. This approach is practical, is less invasive than open resection, and appears to provide a long-term solution.

<https://thejns.org/doi/abs/10.3171/2018.7.JNS172501>

KEYWORDS gliependymal cyst; ependymal cyst; neuroepithelial cyst; endoscopy; surgical technique

GLIOPENDYMAL cysts are exceedingly rare congenital lesions of the central nervous system. They are considered benign in nature and compose an estimated 1% of all intracranial cysts.^{6,7} The origin of such lesions remains a matter of debate, and it is postulated that they arise from ectopic ependymal cells during the embryonic period. The ectopic cells thus develop into an epithelial-lined cyst, either intraaxial or extraaxial.⁵ Imaging characteristically demonstrates a parenchymal cyst with smooth, rounded borders and fluid content similar to cerebrospinal fluid (CSF). The cystic fluid follows CSF characteristics on both CT and MRI. Clinically, presentation is highly variable and related to the size and location of the lesion. Most of these cysts are asymptomatic and can be simply watched; in certain cases, however, they can be clearly symptomatic. The symptoms are typically attributable to regional mass effect and include headaches, hemiparesis, hemianesthesia, or even seizure.²

The treatment of choice for a symptomatic gliependymal cyst remains unclear. To date, numerous surgical procedures have been employed, including burr hole craniotomy for evacuation of the fluid component, open craniotomy with cyst wall excision with or without fenestration into the ventricular or subarachnoid system, and shunting. Some authors recommend resection of the cyst wall given the recurrent nature of the cyst and concerns regarding additional surgical intervention in the future. For patients undergoing cystoperitoneal shunting, there is concern about shunt obstruction due to the high protein content of the cystic fluid.¹⁰

Given the lesion's clinical rarity and the lack of agreement regarding its management, the optimal surgical treatment of gliependymal cysts remains uncertain. In this paper, we propose a minimally invasive neuroendoscopic approach to treat a gliependymal cyst. We describe a series of three patients who were successfully treated with

ABBREVIATIONS CSF = cerebrospinal fluid; POD = postoperative day.

SUBMITTED October 6, 2017. **ACCEPTED** July 16, 2018.

INCLUDE WHEN CITING Published online November 30, 2018; DOI: 10.3171/2018.7.JNS172501.

endoscopic fenestration of the cyst wall into the ventricular system. We also discuss the clinical presentations and surgical outcomes of our patient cohort.

Case Reports

Case 1

History and Examination

A 41-year-old man presented with a 1-year history of worsening headaches that gradually became associated with blurred vision. His neurological examination revealed left hemianopia, but the remainder of his physical examination was unremarkable. MRI of the head demonstrated a large right occipital intraparenchymal cyst measuring 70 mm and causing significant mass effect on surrounding brain parenchyma (Fig. 1A). The cystic fluid component demonstrated imaging characteristics similar to CSF.

Surgical Procedure

The patient underwent neuroendoscopic fenestration of the right occipital gliopendymal cyst to the right lateral ventricle. With the aid of an endoscope, the cyst wall was fenestrated into the lateral ventricle at the thinnest portion. Wide communication of the cyst with the ventricle was visualized.

Postoperative Course

The patient tolerated the procedure well without complications. On postoperative day (POD) 1, the patient reported mild headache improvement, and postfenestration CT demonstrated moderate cyst decompression and improved mass effect. The patient was discharged home on POD 2 without issues. At the 1-month follow-up, the patient reported significant headache improvement and resolution of his blurry vision. His neurological exam showed resolution of the hemianopia, and brain MRI showed decompression of the cyst with nearly resolved adjacent brain compression. Brain MRI performed 16 months after surgical intervention demonstrated continued decompression of the gliopendymal cyst (Fig. 1B–D).

Case 2

History and Examination

A 60-year-old woman presented with severe headache and left-sided numbness. On examination, she was neurologically intact. One year earlier, a right parafalcine gliopendymal cyst had been found during a workup for complaints of occipital headaches. At the time of initial presentation, she was intact on examination and was therefore managed nonsurgically with serial imaging. Later brain MRI demonstrated enlargement of the existing right parafalcine cyst, measuring 79 mm, with mass effect upon the adjacent brain parenchyma (Fig. 2A and B).

Surgical Procedure

After a thorough discussion regarding the interval enlargement of the gliopendymal cyst and the adjacent brain compression noted on imaging, the patient elected to proceed with a minimally invasive surgical intervention. She underwent right-sided neuroendoscopic fenestration of the cyst to the ventricular system.

Postoperative Course

She tolerated the procedure well without issues. Her postoperative course was uneventful, and she was discharged home on POD 1. At the 1-month follow-up she reported significant improvement in her headaches and resolution of the preoperative numbness. Brain MRI demonstrated a marked decrease in the size of the cyst and significant improvement of the mass effect and midline shift (Fig. 2C and D). At the 3- and 4-year follow-ups, she remained symptom free without focal neurological deficits on examination. At the 5-year follow-up, MRI demonstrated stable decompression of the cyst.

Case 3

History and Examination

A 55-year-old woman presented with a 3-month history of subjective headaches, dizziness, and progressive

FIG. 1. A: Preoperative axial T1-weighted MR image demonstrating a right occipital gliopendymal cyst. **B and C:** Postintervention axial T1-weighted MR images obtained at different levels, demonstrating decompression of the cyst following fenestration with connection to the right lateral ventricle.

FIG. 2. Preoperative axial T2-weighted (A) and sagittal T1-weighted (B) MR images demonstrating the periventricular location of a gliependymal cyst adjacent to right lateral ventricle. Postintervention axial T2-weighted (C) and sagittal T1-weighted (D) MR images showing decompression of the right occipital gliependymal cyst with communication to the right lateral ventricle following neuroendoscopic cyst fenestration.

visual decline. She underwent formal preoperative ophthalmological visual testing, which demonstrated a right visual field cut. Neurological examination confirmed right superior quadrantanopia, but the remainder of her exam was unremarkable. Brain MRI showed a large, right intraparenchymal temporooccipital cyst of CSF-like intensity, consistent with a gliependymal cyst. It measured approximately 57 mm (Fig. 3A).

Surgical Procedure

Given the right visual field deficit demonstrated on formal visual testing and significant mass effect related to the cyst, surgical intervention was recommended. The patient elected to proceed with neuroendoscopic fenestration of the gliependymal cyst to the ventricle. The cyst wall was widely fenestrated into the right lateral ventricle at the level of the trigone.

Postoperative Course

The patient tolerated the procedure well without compli-

FIG. 3. A: Preoperative axial T2-weighted MR image demonstrating a right temporooccipital gliependymal cyst adjacent to the right lateral ventricle. B–D: Postintervention axial T2-weighted MR images obtained at different levels, demonstrating cyst decompression and fenestration to the right lateral ventricle.

cations. On POD 1, she reported mild improvement in her headaches and existing visual deficit. Postfenestration head CT demonstrated a mild decrease in cyst size. Her hospital course was uneventful, and she was ultimately discharged home 1 day after cyst fenestration. At the 1-month follow-up visit, she reported resolution of her headaches and improved vision. Physical examination demonstrated full visual fields by confrontation. Formal visual field testing performed 2 months after the intervention demonstrated improvement of the preoperative right superior quadrantanopia. At the 4-month follow-up visit, her exam remained stable, and MRI demonstrated continued decompression of the gliependymal cyst (Fig. 3B–D).

Discussion

Gliependymal cysts are rare entities, and documented findings in adults are infrequent. The nomenclature used to describe these lesions is confusing because they are known by several names including “gliependymal cysts,” “neuroglial cysts,” and “neuroepithelial cysts.” They are

TABLE 1. Clinical, radiological, and follow-up data of treated patients

Case No.	Age (yrs)/ Sex	Symptoms	Lesion Location	Cyst Size			Length of Stay (days)	FU (mos)	Outcome
				Preop (mm)	Postop (mm)	Reduction In (%)			
1	41/M	Headache, rt blurry vision	Rt occipital	70	48	31	2	16	Symptom free
2	60/F	Headache, lt hypesthesia	Rt parafalcine	79	45	43	1	61	Symptom free
3	55/F	Headaches, dizziness, visual decline	Rt temporooccipital	57	27	52	1	4	Symptom free

FU = follow-up.

considered benign lesions of congenital origin, yet their exact origin is controversial. Given that most of the literature, in the form of case reports, cites occurrences in fetuses or infants, there is a belief that they arise early during embryogenesis. Multiple hypotheses support the notion that they arise from ectopic rests of primitive neuroectoderm that become trapped during neurulation.^{5,9} It is further thought that these cysts arise from ectopic displacement of segments of the neural tube wall at the site of the formation of the tela choroidea.^{3-5,8} As a result, gliependymal cysts are usually lined by ependymal cells that have become separated from the ventricular system. Moreover, given the secretory nature of the ependymal cells, these cysts can grow to considerable size over time and progressively result in neurological symptoms.

The term “gliependymal cyst” was introduced by Friede and Yasargil in 1977 to describe the cystic lesions that are typically located intracerebrally, extracerebrally, or anywhere along the neuroaxis and do not communicate with the ventricles.^{3-5,8} However, they exhibit a propensity for a periventricular location, thus supporting the thought that the cyst arises from ectopic segments of the neural tube.^{3,4,8} In our series, all patients demonstrated a periventricular location of the gliependymal cyst. Frazier and colleagues documented an infratentorial gliependymal cyst located near the fourth ventricle.³ Qi et al. reported a giant right frontal cyst located near the lateral ventricle.⁸

Age at clinical presentation varies widely from fetuses to adults, with most cases documented in infants or children; however, the focus of our writing is the adult presentation. In our series, the average age of presentation was 51 years, and two of the three patients were female (Table 1). These findings are similar to those of Frazier et al., who reported that gliependymal cysts often present in the fourth and fifth decades of life and show a slight female predominance.³ Clinical presentations vary from seizures to symptoms related to focal mass effect such as headaches, nausea, visual symptoms, weakness, or sensory changes.⁹ In our series, patients presented with headache, visual disturbance, and hypesthesia. The progressive growth of these cysts is attributed to the secretory activity of the ependymal cells, resulting in mass effect.

Imaging modalities used to establish a diagnosis include CT scanning and MRI, though MRI is the preferred modality for the radiological diagnosis of gliependymal cyst. The parenchymal cyst characteristically has smooth, rounded borders composed of unilocular thin walls and CSF-like fluid that can be readily identified on CT or MRI. CT scans show a cystic focus with fluid of low den-

sity without enhancement. MRI displays a cystic filled lesion with fluid that is hypointense on T1-weighted images and hyperintense on T2-weighted images.^{1,3,6,7} In our series, all patients exhibited intraparenchymal periventricular lesions with smooth rounded borders and cystic fluid resembling CSF on CT or MRI.

The differential diagnosis of gliependymal cyst includes arachnoid cyst, epidermoid cyst, ependymal cyst, porencephalic cyst, and trapped ventricle. Though the diagnosis can be definitively made by histopathological analysis, imaging of such lesions is fairly characteristic. Pathological analysis was not performed to confirm gliependymal cyst as the diagnosis in our patients, but it could be inferred based on imaging characteristics and thin-walled morphology. Pathological examination would not have affected the treatment decision at the time of surgery, so the cost of doing the examination was avoided. These patients' cysts were most likely gliependymal cysts because they were smooth, thin-walled cysts (thicker than intraparenchymal arachnoid cyst), which did not appear irregular or porencephalic in nature (porencephalic cyst) or arise from the cortex. Additionally, they did not appear to communicate with or arise from the ventricle (ependymal cyst or trapped ventricle).

Because of the rarity of gliependymal cysts and the paucity of literature describing their optimal treatment, management remains controversial. Most gliependymal cysts are small and asymptomatic. In these instances, surgical treatment is typically not necessary, as the cyst is commonly found incidentally during evaluation for various other reasons. However, patients who are symptomatic tend to present with symptoms related to raised intracranial pressure and local mass effect. The intervention of choice in symptomatic patients is undetermined, as a multitude of treatment options exist. Common surgical approaches for gliependymal cyst include open craniotomy for total extirpation, marsupialization, fenestration of the cyst into the ventricle or subarachnoid space, shunting, or a combination of these approaches.^{1,3} In our experience, we successfully treated these three symptomatic adult patients harboring gliependymal cyst by using neuroendoscopic fenestration of the cyst wall to the ventricular system via burr hole access. Current literature on the treatment of such cysts in adults is highly variable. Morigaki and colleagues reported the successful treatment of a giant gliependymal cyst in a 35-month-old girl with endoscopic fenestration of the cyst wall through a frontal burr hole.⁵ At her 5-year follow-up, there was no evidence of recurrence.⁵ Frazier and colleagues described the case of

a 55-year-old woman with a 10-year history of a recurrent posterior fossa gliependymal cyst, which required two operations and eventually a third procedure with gross-total resection.³ Considering the recurrent nature of the cyst, the authors advocated complete resection of the gliependymal cyst via open craniotomy.³ Qi et al. described the treatment of a giant right frontal gliependymal cyst in a 38-year-old man through a combination of open partial resection and cystoperitoneal shunting, yet the authors advocated complete removal as the ideal treatment for gliependymal cyst.⁸ To the best of our knowledge, endoscopic fenestration of a gliependymal cyst to the ventricular system has never been reported in adults.

Each surgical approach to gliependymal cysts entails its own drawbacks. Open craniotomy for complete or partial cyst wall resection presents a more invasive approach with an increased risk of morbidity and presumed length of stay. Shunting exposes the patient to the placement of a foreign body and the risk of shunt malfunction or obstruction. The advantages of endoscopic fenestration are the minimally invasive nature of the procedure and its association with low morbidity, short hospital stay, and apparent long-term efficacy. The average length of stay in our case series was 1.6 days (Table 1). The median follow-up period was 16 months without evidence of cyst reaccumulation or symptom return.

Conclusions

In summary, the utility of endoscopy in neurosurgery has progressively expanded in recent years and is a feasible option for the treatment of gliependymal cyst. In our series, we discuss periventricular gliependymal cysts in three patients who were successfully treated with a minimally invasive approach via fenestration of the cyst to the ventricular system. All patients had resolution or improvement of their preoperative symptoms, brief hospital course, and stable decompression of the cyst on radiological follow-up. Therefore, we conclude that the neuroendoscopic technique is a favorable treatment option with the advantage of minimal invasiveness and fewer complications. The gliependymal cyst location and whether the involved hemisphere is dominant determine the ideal endoscopic trajectory. The long-term efficacy of the endoscopic technique in treating gliependymal cysts needs further evaluation.

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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: Chamoun. Acquisition of data: Alvarado. Drafting the article: Alvarado, Smith. Critically revising the article: all authors. Reviewed submitted version of manuscript: all authors. Approved the final version of the manuscript on behalf of all authors: Chamoun. Administrative/technical/material support: Chamoun. Study supervision: Chamoun, Smith.

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