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Case Report

Pediatric third ventricular glioependymal cyst: A case report

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ABSTRACT

Background: Glioependymal cysts are rare lesions, especially in children.

Case Description: The authors treated a well-developed 12-year-old male who presented with 1 week of acute-onset headache. Magnetic resonance imaging revealed a thin-walled third ventricular cyst obstructing the foramen of Monro. Endoscopic fenestration of the cyst with biopsy and endoscopic third ventriculostomy was performed. Histopathologic examination revealed a glioependymal cyst. At 13-month follow-up, the patient is asymptomatic without evidence of recurrence.

Conclusion: This case represents the first glioependymal cyst reported in an adolescent.

Keywords: Endoscopic third ventriculostomy, Glioependymal cyst, Intracranial cyst, Neuroepithelial cyst, Third ventricle

INTRODUCTION

Glioependymal cysts are exceedingly rare lesions, also referred to as neuroglial or neuroepithelial cysts. Glioependymal cysts represent only 1% of all non-neoplastic intracranial cysts yet are known to grow and expand rapidly, resembling a neoplastic process. Glioependymal cysts most frequently arise in the subarachnoid space of the frontal lobes in the 4th to 5th decade of life with a female predominance but have been infrequently reported in young children with a male predominance.[10,16,25] They are lined by an innermost single layer of secretory cuboidal epithelium with surrounding glial and connective tissue and may expand, becoming rapidly symptomatic.[10] Depending on their location, they can cause headaches, macrocephalus, seizures, motor deficits, or altered sensorium.[25] Neurosurgical management consists of neuroendoscopic cyst fenestration, complete resection through craniotomy, or shunting.[1] To the author's knowledge, this is the first report of a symptomatic glioependymal cyst in an adolescent.

ILLUSTRATIVE CASE

The authors treated a well-developed 12-year-old Hispanic male who presented to our institution with an acute-onset severe worsening headache for 1 week with associated lethargy, nausea, and vomiting. Initial workup with computed tomography (CT) head demonstrated

marked ventriculomegaly of the lateral and third ventricles. Further evaluation with magnetic resonance imaging (MRI) demonstrated a thin-walled cyst within the third ventricle obstructing the foramen of Monro [Figure 1]. The cyst was isointense to the cerebrospinal fluid (CSF) on all sequences. In discussion with the patient's family, the decision to perform neuro endoscopic cyst fenestration and endoscopic third ventriculostomy (ETV) was made. An ETV was desired in this case for several reasons. First, the MRI demonstrated favorable anatomy (enlarged ventricles and generous third ventricular floor) for the performance of an ETV. The calculated ETV success score was 90%.[15] In addition, due to the symptomatic obstructive hydrocephalus, long-term management of CSF diversion was desired, which could be performed in the same endoscopic procedure. A successful ETV obviated the need for craniotomy, reoperation, or placement of potentially obstructable external hardware, namely, cysto-peritoneal or cysto-subarachnoid shunt.

Neuroendoscopic fenestration was performed through a frontal burr hole into the right anterior horn of the lateral ventricle. On entering the ventricular space with the endoscope, a smooth cyst was immediately visible through the foramen of Monro [Figure 2]. The cyst wall was fenestrated several times using a Synergy® Axiem™ neuronavigation stylet (Medtronic Navigation, Inc, Louisville, CO). The cyst was filled with clear, colorless fluid. Several samples of the cyst wall were taken for histopathologic diagnosis. Following cyst fenestration, the third ventricle was entered through the dilated foramen of Monro. The ETV was performed without complication or external ventricular drain placement. Pathologic examination of the cyst wall by light microscopy suggested a glioependymal cyst [Figure 3]. Immunohistochemical staining performed at two institutions revealed that the samples stained positive for glial fibrillary acidic protein.

Following neuroendoscopic cyst fenestration and ETV, the patient's headache resolved completely, and postoperative MRI showed cyst resolution. The patient has been symptom-free since the most recent follow-up 13 months after the procedure. Monitoring with MRI is planned for 24 months after the procedure to monitor for recurrence.

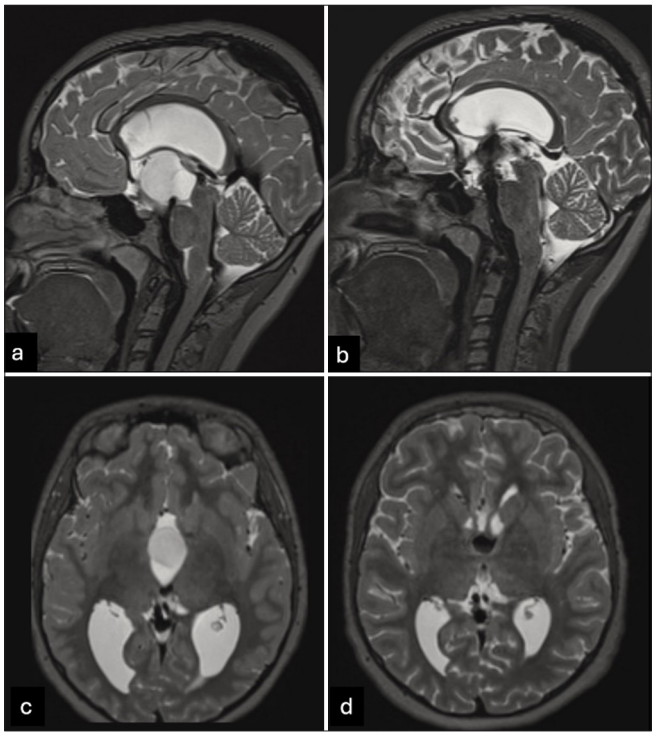

Figure 1: (a) Preoperative sagittal T2-weighted magnetic resonance (MR) imaging obtained in the patient demonstrating a cyst that is isointense to cerebrospinal fluid within the third ventricle, causing dilation of the lateral ventricles. (b) Postoperative sagittal T2-weighted MR imaging demonstrating cyst resolution. (c) Axial preoperative view. (d) Axial postoperative view.

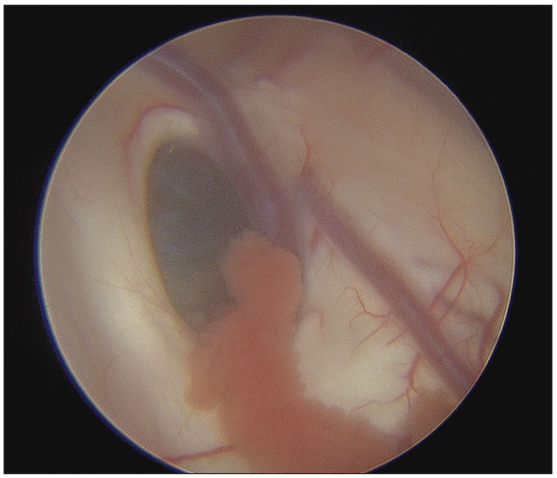

Figure 2: Intraventricular endoscopic view of the glioependymal cyst membrane through foramen of Monro.

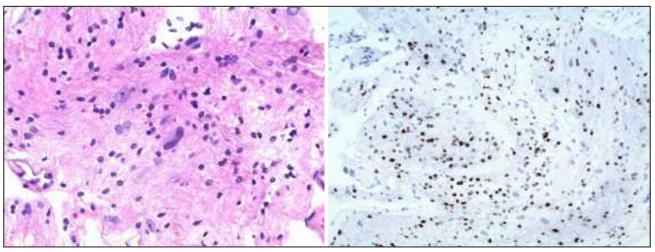

Figure 3: (Left) Light microscopy of cyst wall by hematoxylin and eosin staining demonstrating minute fragments of neuroepithelial tissue without significant presence of intact innermost cuboidal cell lining. (Right) Immunohistochemical staining for glial fibrillary acidic protein highlighting the diffuse distribution of glial cells. Samples did not stain positive for neuronal markers (NeuN and Synaptophysin).

DISCUSSION

This case is the first report of a glioependymal cyst in an adolescent, as previously reported pediatric glioependymal cysts involved neonates to children 7 years of age [Table 1].

A systematic review by Robles *et al.* suggested an updated histopathologic classification of intracranial cysts.[23] According to their classification, glioependymal cysts are considered a subtype of neuroepithelial cysts, including ependymal cysts and choroid plexus cysts, also derived from neuroepithelium. Before this update, glioependymal cysts have interchangeably been diagnosed as neuroglial and neuroepithelial cysts. However, the authors agree with using the term glioependymal cyst to describe these lesions to avoid confusion. The major histologic difference between glioependymal cysts and the more common ependymal cysts is intervening glial tissue in the former.

Case reports of glioependymal cysts in children describe a rapidly growing cyst from active ependymal secretion, ependymal proliferation, and symptoms related to mass effect.[28] The cyst expansion may be pronounced. Morigaki *et al.* reported a giant pediatric glioependymal cyst involving both the anterior and posterior fossa.[19] Children with glioependymal cysts most frequently present with macrocephaly, seizures, and motor deficits.[25] In addition, most pediatric glioependymal cysts are located interhemispheric, while intraventricular glioependymal cysts are extremely rare.[12] Our patient did not have motor deficits or seizures but presented with signs of increased intracranial pressure. Tange *et al.* observed that glioependymal cysts in infants were associated with other central nervous system abnormalities, including complete or partial agenesis of the corpus callosum or microgyria.[25] Non-enhancing thin-walled cysts, in combination with agenesis of the corpus callosum, have been suggested as sufficient for the diagnosis without necessitating histopathologic confirmation.[19] Yet, no sensitivity or specificity of these two radiographic findings has been reported. Our patient presented with an intraventricular glioependymal cyst within the third ventricle and a normally developed corpus callosum. Due to the location and likely slow growth of the glioependymal cyst in our patient, he presented acutely with symptoms of obstructive hydrocephalus in adolescence. We believe that our patient became symptomatic much later than previously reported pediatric glioependymal cysts due to the scarcity of secretory ependymal-like cells seen by light microscopy [Figure 2].

Friede and Yasargil were the first to describe adult glioependymal cysts in detail using electron microscopy.[10] In their model, a short segment of the wall becomes displaced during embryonic development into the cerebral parenchyma or the subarachnoid space from the ectopic rests of the neural tube near the site of the developing tela choroidea. They may arise anywhere in the neuraxis, accounting for both intracerebral and subarachnoid glioependymal cysts, those located in the

Table 1: Previously reported pediatric glioependymal cysts in the literature.

Case number	Author (year)	Age (sex)	Location	Operation	Outcome (complications)
1	Brihaye (1956)[4]	4 mo. (F)	Inter-hemispheric	None	Dead
2	Heiskanen and Haltia (2008)[12]	2 y.o. (F) 6 y.o. (F)	Third ventricle Third ventricle	Complete resection Complete Resection	Good Good
3	Takeshita *et al.* (1982)[24]	1 mo. (M)	Para-ventricular	Complete resection	Good
4	Barth *et al.* (2008)[3]	Neonate (M)	Inter-hemispheric	None	Dead
5	Morimoto *et al.* (1986)[18]	11 mo. (M)	Inter-hemispheric	Cyst opening	Good (subdural effusion)
6	Utsunomiya *et al.* (1987)[26]	Neonate (M)	Inter-hemispheric	Cyst opening, removal	Good (recurrence)
7	Niwa *et al.* (1991)[20]	5 y.o. (M)	Lateral ventricle	Cyst opening	Good
8	Tange *et al.* (2000)[25]	Neonate (M)	Inter-hemispheric	Complete resection with CP shunt	Good
9	Obaldo *et al.* (2007)[21]	1 day	Temporal	Fenestration to SA space	Good
10	Morigaki *et al.* (2011)[17]	2 y.o.	Supra-infratentorial	Fenestration to ventricle	Good
11	Zheng *et al.* (2013)[28]	3 mo.	Temporal peri-ventricular	Fenestration to ventricle	Good (recurrence)
12	Cho *et al.* (2014)[6]	4 mo.	Midline anterior	Partial resection	Good
13	Cavalheiro *et al.* (2019)[5]	2 y.o. (F)	Midbrain	Complete resection	Good
14	Irie *et al.* (2022)[13]	Neonate (M)	Para-ventricular	CP shunt	Good
15	Kadri *et al.* (2024)[14]	7 y.o.	Third ventricle	Fenestration to ventricle	Good
16	Bui and Tran (current)	12 y.o.	Third ventricle	Fenestration to ventricle	Good

M: Male, F: Female, y.o.: Years old, mo: Month old, CP: Cystoperitoneal, SA: Subarachnoid

posterior fossa or ambient cistern [9,10], and within nerves and the spinal cord.[2,21] Postoperative magnetic resonance (MR) ventriculocisternography has been utilized in one case of a pediatric glioependymal cyst, providing further evidence that the tela choroidea is the most likely site of origin.[17] The secretory highly ciliated ependyma lining glioependymal cysts does not resemble the ventricular ependyma, providing evidence against the theory that these lesions arise as outpouchings of the ventricular wall.[10] The secretory cellular structure explains the phenomenon of unexpected and rapid expansion. Our patient's case and other cases of third ventricular glioependymal cysts support the tela choroidea as the origin for glioependymal cysts due to proximity.[17] Furthermore, if the cysts are causing mass effects or deficits and patients need surgery for cyst fenestration, we recommend that intracranial cyst walls be biopsied and sent to pathology due to the risk of secretory ependymal-like cells increasing the risk of cyst recurrence.

These cysts have been known to recur in children and adults. Zheng *et al.* reported a glioependymal cyst in a 3-month-old treated with partial resection due to cyst wall attachment to the thalamus and internal capsule that subsequently recurred.[28] Frazier *et al.* also reported a glioependymal cyst in a woman requiring reoperation and advocated for craniotomy as the primary management of these lesions to reduce the risk of recurrence.[9] However, Alvarado *et al.* performed neuroendoscopic fenestration in three patients with glioependymal cysts and did not note a recurrence of the lesions with a median 16-month follow-up.[1] The factors influencing intra-cystic ependymal secretion, the difference between the speed of cystic expansion in children and adults, and the predominant sex discordance between adults and children remain to be elucidated. Due to the low but potential risk of recurrence, we plan to monitor our patient with periodic MRIs.

Imaging

CT imaging is insufficient to differentiate glioependymal cysts from infectious or neoplastic cysts.[11] On MRI, the wall of a glioependymal cyst does not enhance, and the fluid is isointense to CSF on T1-weighted imaging (T1WI) and T2-weighted imaging (T2WI). However, the cyst wall has been noted to enhance in a pediatric case.[25] On multiplanar imaging, glioependymal cysts can be differentiated from other cysts in communication with the subarachnoid or ventricular system.[11] The internal fluid may grossly appear identical to CSF, xanthochromic, or turbid milky due to the increased protein content.

Important to distinguish from glioependymal cysts on MRI are arachnoid cysts and epidermoid cysts, which are all isointense to CSF on T1WI and T2WI and do not enhance.[22] Arachnoid cysts may become symptomatic later in life but are extra-axial and confined to the subarachnoid space, whereas glioependymal cysts are intra-axial or in the subarachnoid space but do not communicate.[11] Epidermoid cysts can be distinguished on MR as diffusion-restricting on diffusion-weighted imaging and are heterogeneous on fluid-attenuated inversion recovery imaging. Colloid cysts are characteristically located at the foramen of Monro, which is possible for glioependymal cysts, as seen in our patient. However, colloid cysts attenuate on CT, unlike glioependymal cysts, which follow the CSF signal on CT. Choroid plexus cysts (xanthogranulomas) are enhanced on post-contrast imaging and are typically present bilaterally in the lateral ventricles with calcifications. Choroid plexus cysts do not cause symptoms even when very large, unlike glioependymal cysts. Virchow-Robin perivascular spaces may also be mistaken for glioependymal cysts but are most commonly multiple and located near the basal ganglia. Porencephalic cysts, which result from encephaloclastic processes, are also not to be mistaken for glioependymal cysts, as they communicate with the ventricle and never act as expanding lesions.[10,11,22]

Treatments

The surgical treatment for symptomatic glioependymal cysts is nearly identical to that of ependymal cysts or arachnoid cysts. Neurosurgical interventions have been described for glioependymal cysts, including neuro endoscopic fenestration to an adjacent CSF space: Cysto-ventricular, -subarachnoid, or -peritoneal shunting, and partial or complete excision.[1,9,17] A large series investigated neuroendoscopy for pediatric arachnoid cysts and described synchronous ETV performance in three patients presenting with concomitant hydrocephalus.[8] They concluded that this minimally invasive procedure was superior to open craniotomy due to low morbidity and mortality and less operative and recovery time. Open craniotomy with complete resection was once suggested as the most effective option where anatomically feasible due to the risk of recurrence.[9,16] However, reports from neuroendoscopic fenestration of glioependymal cysts with long-term follow-up have not demonstrated cyst recurrence or shunt requirement. Neuroendoscopic surgery has been reported successful for ependymal-lined cysts in children located intraventricularly or in eloquent areas such as the mesencephalon.[1,27] El Damaty *et al.* concluded that neuroendoscopic fenestration should be the treatment of choice for ependymal cysts to avoid craniotomy and shunt dependence.[7] The presence of acute hydrocephalus was noted to increase the facilitation of the procedure in our case. Neuroendoscopic fenestration with ETV utilized in our case proved successful without complication and avoided the placement of a shunt.

CONCLUSION

Glioependymal cysts are rare lesions in children. We report the first adolescent case of an intracranial glioependymal cyst. Neuroendoscopic cyst fenestration and endoscopic third ventriculostomy successfully avoided shunt placement and the patient recovered fully.

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