



Case Report

Intracranial arachnoid cysts in an infant: A technical note on the innovative use of navigation and flexible endoscopy for cyst fenestration

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ABSTRACT

Background: Intracranial arachnoid cysts (ACs) are a cerebral spinal fluid (CSF) collection within the meninges. They typically arise during embryologic development. Some are stable overtime with little consequence, but large or growing cysts may require surgical intervention. The optimal surgical technique is debated and may be more technically challenging in the infant age group.

Case Description: Our unique case report details a 10-month-old (6 months corrected age) infant who presented with a drastic increase in head circumference and was found to have midline shift and three cysts – one large and two smaller ones. He was treated with an innovative surgical approach combining stereotactic introduction of a catheter to facilitate subsequent flexible endoscopy allowing three separate cysts to be treated through one small surgical incision with no complications and a stable examination on 2-year follow-up.

Conclusion: Symptomatic ACs in the infant population that require treatment can be addressed with open surgery to fenestrate the cyst, endoscopic cyst fenestration, or cystoperitoneal shunting. Typically, surgeons must choose between a rigid endoscope which allows stereotactic navigation or a flexible endoscope which allows multiple trajectories but precludes navigation. Our case demonstrates that combining stereotactic ventricular placement before flexible endoscopy provides the benefit of both approaches and allows for successful endoscopic treatment in a young patient with durable results.

Keywords: Endoscopy, Infant, Intracranial arachnoid cysts, Minimally invasive

INTRODUCTION

Intracranial arachnoid cysts (ACs) are collections of cerebrospinal fluid (CSF) within the meninges.^[3,6] They are classically thought to arise from nonphysiologic arachnoid membrane splitting or duplication, typically during embryologic development. Interestingly, there are rare exemplary case descriptions in the literature illustrating *de novo* cyst formation from head trauma or intraventricular hemorrhage of prematurity.^[8,9,14] The prevalence of these lesions has incidentally increased with rising utilization of imaging modalities, namely, computed tomography and magnetic resonant imaging (MRI). The majority of these cysts are asymptomatic, and their long-term behavior is often unknown. Lesions typically become symptomatic due to cyst growth, a process that has several postulated mechanisms. While

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surgical management is indicated in symptomatic patients, the optimal approach to use remains under debate. Cysts that require treatment can be addressed with open surgery to fenestrate the cyst, endoscopic cyst peritoneal shunting. Surgical management in infants, with the approach carefully tailored to the individual patient with respect to age and cyst location, is an effective and safe way to manage this unique group of patients.

ILLUSTRATIVE CASE

History

An ex-25.2-week premature infant with extensive complications of prematurity and a history of a $2.2 \times 1.2 \times 1.4$ cm parieto-occipital cystic mass [Figure 1] was noted to have a drastic 3-month increase in head circumference at his 10-month (6 months corrected age) neurology appointment. The patient was twin B of a twin pregnancy, and twin A did not survive. The patient was sent for head ultrasound and mother was instructed to take the patient to the emergency department after head ultrasound obtained demonstrated large increase in cyst size with midline shift.

The mother denied any episodes of seizure activity, vomiting, or change in behavior. His head circumference had increased over 1 month to 47.5 cm (>99th percentile) from 46 cm (97th percentile). MRI demonstrated increased mass effect on the adjacent brain and approximately 1.3 cm leftward midline shift. The patient was noted to have three cysts – one large cyst and two smaller ones [Figure 2].

Due to the developing midline shift and growing cysts, the patient required surgical treatment. We utilized a combined approach of introducing a ventricular catheter stereotactically to fenestrate the site of the fenestration followed by flexible endoscopy to widen the fenestration and fenestrate the two additional cysts into the larger cyst and the system into the temporal horn [Figure 3].

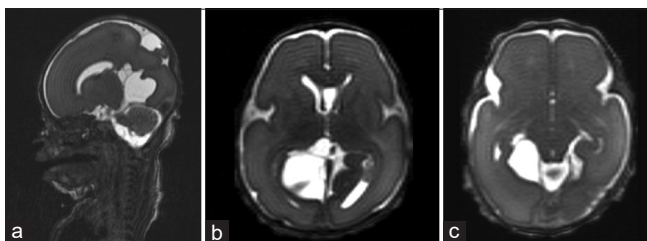


Figure 1: MRI performed on day of life 4. T2-weighted MRI of sagittal (a) and axial (a&c) cuts demonstrating a multiloculated cystic mass measuring approximately $1.7 \times 1.3 \times 3.2$ cm. There is an additional CSF equivalent cystic mass located superiorly and posteriorly, immediately adjacent to the right side of the superior sagittal sinus.

Operative details

The patient was placed supine with the head turned left exposing the right parieto-occipital region. The patient was registered to the stealth axiom navigation system. We planned a high right parieto-occipital trajectory from the largest cyst into the right temporal horn. We made a small mini-craniotomy in the standard fashion. A ventricular catheter was placed through the largest of the cysts and into the right temporal horn with a direct trajectory utilizing stereotactic navigation of the catheter tip. We then tied off this catheter and cut it, leaving it in place, and subsequently introduced the flexible endoscope into the cyst next to this catheter. The ventricular catheter was visualized with tip puncturing the cyst wall. A Bugbee wire was introduced adjacent to the ventricular catheter which marked the entrance into the temporal horn. Manipulating the wire enlarged the hole, and then after removal of the wire, a #3 embolectomy catheter was placed through the hole and inflated to widen this hole further. We then removed the ventricular catheter and advanced the endoscope into the temporal horn through this fenestration. We were able to visualize ependyma and choroid plexus confirming our presence in the ventricular system. At this point, the large cyst was significantly decompressed.

Next, we withdrew the flexible endoscope back into the larger cyst and redirected it posteriorly to focus on the inferior of the two medial cysts. We also utilized ultrasound adjacent to the endoscope to assess the configuration of the remaining two smaller cysts. We introduced the Bugbee wire through the endoscope to make a hole between the larger cyst we were within and the inferior smaller cyst. This was widened with the #3 embolectomy catheter and then the endoscope was passed into the cyst for visual inspection. We then similarly fenestrated from the largest cyst to the superior small cyst through the same sequence and then repeated this process to fenestrate the superior small cyst into the inferior small cyst [Figure 4].

In total, we had created four fenestrations – a communication from the large cyst into the right temporal horn, a

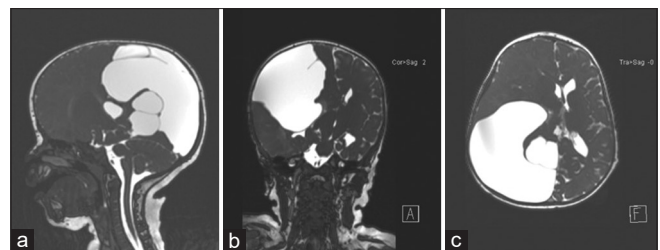


Figure 2: MRI performed at 10 months old uncorrected (6 months corrected age). T2-weighted MRI of sagittal (a), coronal (b), and axial (c) cuts demonstrating three cysts with significant enlargement of the cysts in the right parieto-occipital temporal lobe causing significant mass effect on the adjacent brain and approximately 1.3 cm leftward midline shift.

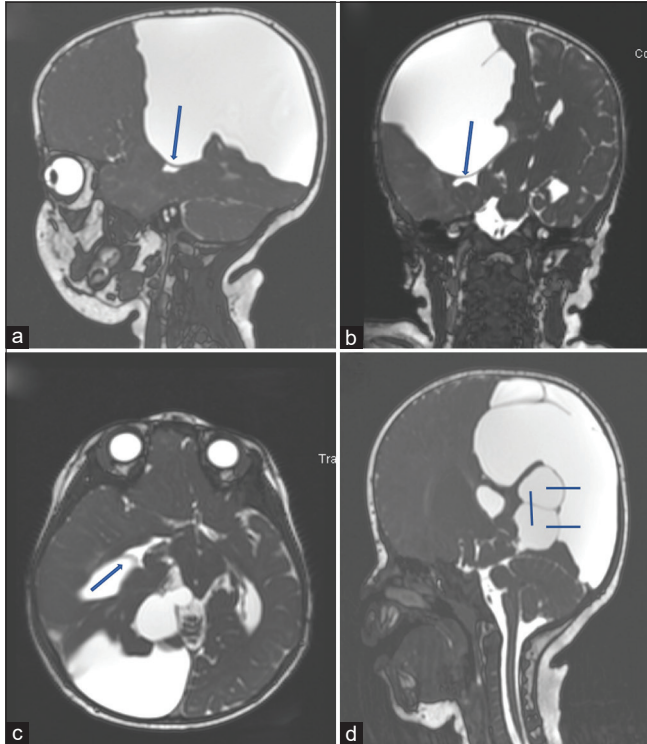


Figure 3: Preoperative T2-weighted MRI of sagittal (a), coronal (b), and axial (c) cuts with location of planned fenestrations between the large cyst into the right temporal horn (arrow). T2-weighted sagittal MRI (d) demonstrating planned fenestrations from the larger cyst into the two smaller cysts and fenestration between the two smaller cysts (lines).

communication from the large cyst into each of the smaller cysts, and a communication between the two smaller cysts, thus allowing for full communication of the cystic structures and decompression of the mass effect they caused. There were no complications.

Postoperative course

The patient was followed at regular intervals postoperatively. At his 2-year postoperative follow-up, he had a stable head circumference at the 85th percentile for corrected age on CDC growth curve, imaging was stable, and patient demonstrated appropriate developmental progress with significant increase in language maturity [Figure 5].

DISCUSSION

We report a rare case of multiple, symptomatic ACs treated with an innovative surgical approach combining stereotactic introduction of a catheter to facilitate subsequent flexible endoscopy allowing three separate cysts to be treated through one small surgical incision.

Natural history

ACs are classically thought to arise from either erroneous arachnoid membrane splitting or duplication, typically during embryologic development. Other etiologies in the literature illustrate *de novo* cyst formation from head

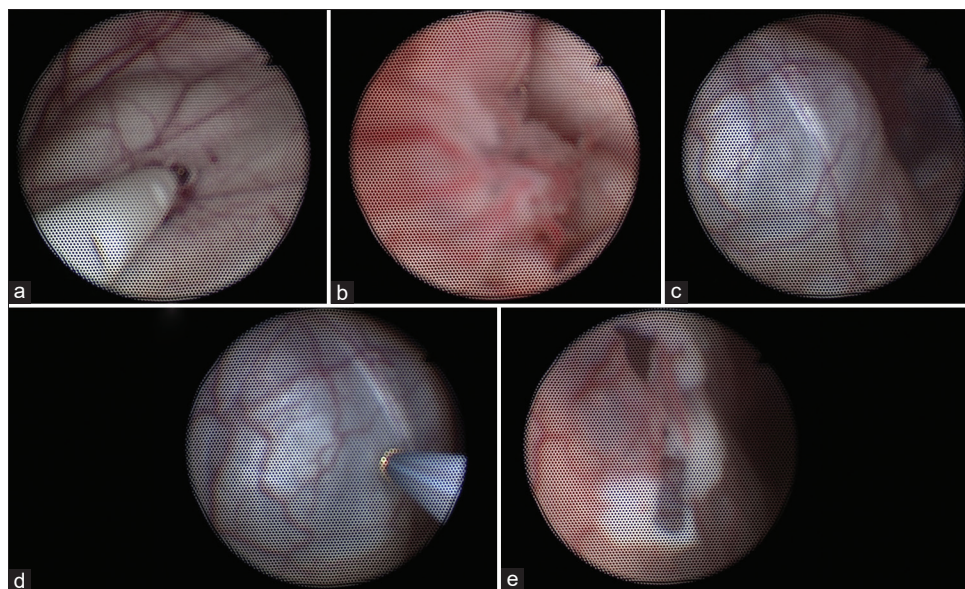


Figure 4: Intraoperative endoscopic view of a ventricular catheter (a) placed in the largest parieto-occipital cyst with trajectory into the right temporal horn. View through this fenestration into the temporal horn with visualization of choroid plexus to confirm intraventricular location (b). Medial fluid-filled arachnoid cysts (c and d). Fenestrations made from largest cyst into medial arachnoid cysts (e).

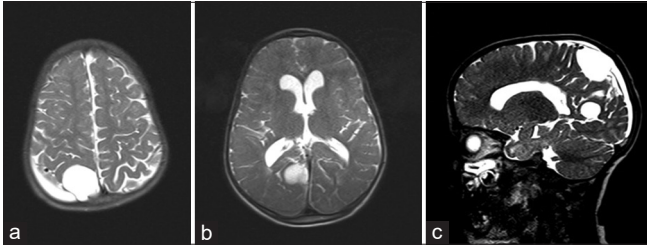


Figure 5: Two-year postoperative T2-weighted MRI of axial (a/b) and sagittal (c) cuts demonstrating grossly stable size and appearance of arachnoid cysts in the right parietal and occipital regions and right parieto-occipital subdural fluid collection. There is stable mild dilation of the lateral and third ventricles.

trauma or intraventricular hemorrhage of prematurity.^[8,9,14] Enlargement, which occurs in an unpredictable manner, is thought to occur by either active fluid secretion from cyst walls, expansion from osmotic gradients, or unidirectional valve mechanisms.^[3,6,10]

A recent single-institution retrospective review of 11,738 patients found a pediatric prevalence rate of 2.6%, an increase from prior studies which reported AC prevalence between 0.2 and 1.7%.^[2] The prevalence within the infant population (<24 months) is often underreported, due to this cohort's lack of predictable manifestation of symptoms associated with cyst enlargement. Most surgically treated infants described in the literature presented with insidious symptoms secondary to increased intracranial pressure, such as macrocephaly and developmental delay.

Treatment

Cysts can be treated through cystoperitoneal shunt, open craniotomy for fenestration, or endoscopic fenestration. CP shunting is relatively straightforward and provides a gradual, predictable change in cyst volume and pressure dynamics.^[4] An outcome-based, prospective single-institution experience demonstrated that this technique was largely successful in reducing cyst size by 95% but argued that surgeons must be attuned to the surgery's related complications. Thus, the practicality of this option must be weighed against risk of early shunt failure, infection, and lifelong shunt failure among other morbidities.^[15]

Open craniotomy for cyst fenestration allows for the avoidance of a permanent shunt; however, some express concern about its true efficacy due to cyst recurrence, especially in the younger population.^[1] Craniotomy and marsupialization can also cause rapid cyst decompression, with risk for the development of subdural hygromas due to the wide surgical aperture and predisposition to CSF accumulation within the subdural space, where absorption is insufficient.^[12] In addition, this strategy is more challenging in younger patients, and some surgeons argue against this

strategy given the invasiveness of the technique and risk of closure of the fenestrations with cyst recurrence.

Neuroendoscopy, a rapidly advancing technique within the field of neurosurgery during the past two decades, has gained momentum for treatment of ACs as a sound compromise which minimizes the extent of surgery while avoiding a permanent shunt.^[7] This newer approach sacrifices a larger working space for a less invasive procedure, thereby avoiding both the invasiveness of an open fenestration as well as known complications associated with CP shunting.^[11] As with most techniques, endoscopy is not without pitfalls. Particularly as relates to complicated cysts, the literature describes a steep learning curve along with the need for a surgeon with ample endoscopic experience to lessen the potential for adverse outcomes. With large cysts, identifying the site for fenestration with an endoscope once inside can be complicated as the cyst wall looks similar throughout. Fenestration for deeper cysts poses a unique technical challenge, typically accomplished with the use of a rigid endoscope, where trajectory must be steadfast to surrounding neural and vascular structures.^[5,7,13] When endoscopy is used, surgeons must traditionally choose either a rigid approach which allows for the use of stereotactic navigation or a flexible endoscope which allows for more flexibility in trajectory but typically precludes the use of stereotaxy. In addition, many of the favored rigid scopes for stereotactic endoscopic approaches are significantly larger in diameter than the flexible endoscope, making them less ideal for treating an infant.

Technical pearls from illustrative case

Our patient, with an extensive medical history and multiple cysts causing mass effect, required careful surgical planning. Given the presence of three cysts and need to make four fenestrations, multiple trajectories for fenestration were required. This need for multiple trajectories favored the use of the flexible endoscope. However, flexible endoscopy typically precludes the use of stereotactic navigation, which was critical in this case to identify the site to fenestrate from the larger cyst into the ventricular system. Identifying this site once inside the cyst without navigation would have been challenging and posed a significant risk if the site could not be appropriately identified. By the use of a novel combined approach of introducing a ventricular catheter stereotactically to fenestrate the site of this fenestration followed by flexible endoscopy to widen this fenestration, fenestrate the two additional cysts into the larger cyst, and fenestrate the two smaller cysts into each other, the patient was able to safely be treated endoscopically through one small incision. This innovative surgical approach allowed safe, minimally invasive treatment with lasting clinical improvement.

CONCLUSION

The management of symptomatic ACs in infants is mostly based on small, single-center series and there are no current guidelines for management. While craniotomies are performed in older patients, there is thought that this technique is less durable and efficacious in infants compared to their older counterparts. CP shunting is commonly utilized in the infant population as it is less technically challenging but leads to lifelong shunt dependence. Combining stereotactic ventricular placement before flexible endoscopy may allow for a greater number of these cysts to be treated endoscopically in younger patients with durable results.

Declaration of patient consent

Patient's consent not required as patients identity is not disclosed or compromised.

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Conflicts of interest

There are no conflicts of interest.

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