



Neuroendoscopic Fenestration of Quadrigeminal Cistern Arachnoid Cyst Presenting with Developmental Regression

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OPEN ACCESS

Received: August 9, 2022

Revised: August 25, 2022

Accepted: September 12, 2022

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Quadrigeminal cistern arachnoid cyst is rare and not usually located in the tectal plate region. Such a cyst may extend to the surrounding area and compress adjacent structures due to cystic expansion. Arachnoid cysts occasionally are associated with hydrocephalus and produce related symptoms. In our case, a 20-month-old boy presented with developmental regression and macrocephaly. Image study of the brain demonstrated a large cyst on the superior portion of the posterior fossa with extension to the supratentorial space and a compressed cerebellum inferiorly and brain stem anteriorly. Lateral and third ventricles were enlarged severely. We performed lateral ventriculo-cystostomy by neuroendoscope via a precoronal approach. Additional third ventriculostomy was performed on the floor of the third ventricle. At 9 months after surgery, the patient showed restored development, and postoperative image showed a reduction of arachnoid cysts and lateral and third ventricles.

Keywords: Arachnoid cysts; Cystostomy; Endoscopy; Ventriculostomy

INTRODUCTION

Arachnoid cyst is a congenital cerebrospinal fluid (CSF) collection between arachnoid membranes. The fluid is not communicated with adjacent spaces, such as ventricles and subarachnoid space. The prevalence of cysts is 1.1% to 2.6% in the general population^{1,3}. The most common location of an arachnoid cyst is a Sylvian fissure, followed by hemisphere convexity, cerebellopontine angle, and suprasellar cistern^{1,6}. Quadrigeminal cistern arachnoid cyst originates in the tectal or pineal region and may extend to surrounding areas such as the trigone, supracerebellar area, third ventri-

cle, and ambient cistern⁷. Quadrigeminal cistern arachnoid cyst is rare and accounts for 5% to 10% of all intracranial arachnoid cyst¹². Though arachnoid cysts are generally asymptomatic, they can produce symptoms with growth and lead to hydrocephalus and compressed adjacent brain structures due to fluid secretion from the cyst wall or entrapment of CSF through a valve-like mechanism¹¹. Children with quadrigeminal cistern arachnoid cysts present with hydrocephalus symptoms, macrocephaly, ocular symptoms, and rare developmental delay^{2,16}. Treatments of quadrigeminal cistern arachnoid cysts are numerous and include open craniotomy with cyst excision and shunting between

cyst and cistern. Recently, endoscopic fenestration of cysts and cistern has been used for the treatment of symptomatic quadrigeminal arachnoid cyst. We present an interesting case of quadrigeminal arachnoid cyst-producing developmental delay that is resolved by endoscopic treatment.

CASE REPORT

A 20-month-old child was admitted to our institute with developmental regression, bilateral leg rigidity, and macrocephaly. The child showed normal development until 12 months of age. He could stand up and walk by himself and was able to babble and say small words such as 'mama and papa.' However, he had not shown developmental or verbal progress since 15 months of age and had lost the ability to walk. The patient's leg revealed hypertonia and rigidity. The head circumference was 99th percentile by age. There was no medical history such as infectious disease and head injury and was no cranial nerve palsy or sensory change on neurological examination. There was no increased intracranial hypertension. Computed tomography (CT) scan and magnetic resonance image of the brain demonstrated a

large cyst on the superior portion of the posterior fossa (Fig. 1). The cyst compressed the cerebellum inferiorly and the brain stem anteriorly, and the lateral and third ventricles were enlarged severely.

Surgery of endoscopic fenestration between the lateral ventricle and cyst and third ventriculostomy was planned. A paramedian and semicircular incision was created, and burr hole trephination was performed at the right Kocher's point. After the dura mater was opened, a 0° rigid endoscope (Karl Storz, Tuttlingen, Germany) was inserted and preceded to the lateral ventricle by neuronavigation (Fig. 2A, B). First, lateral ventriculo-cystostomy (right lateral ventricle to quadrigeminal cistern arachnoid cyst) was performed at a size of 10 mm at the trigone and medial to choroid plexus in an avascular region. The ventricle and cyst wall were fenestrated using a unipolar coagulator and widened by forceps and 3 Fr Fogarty balloon catheter (Fig. 2C). The endoscope was inserted into the third ventricle through the foramen of Monro (Fig. 2D). Third ventriculostomy (third ventricle to interpeduncular cistern) was performed at a size of 5 mm at the floor of the third ventricle (tuber cinereum) using the same armamentarium as above (Fig. 2E).

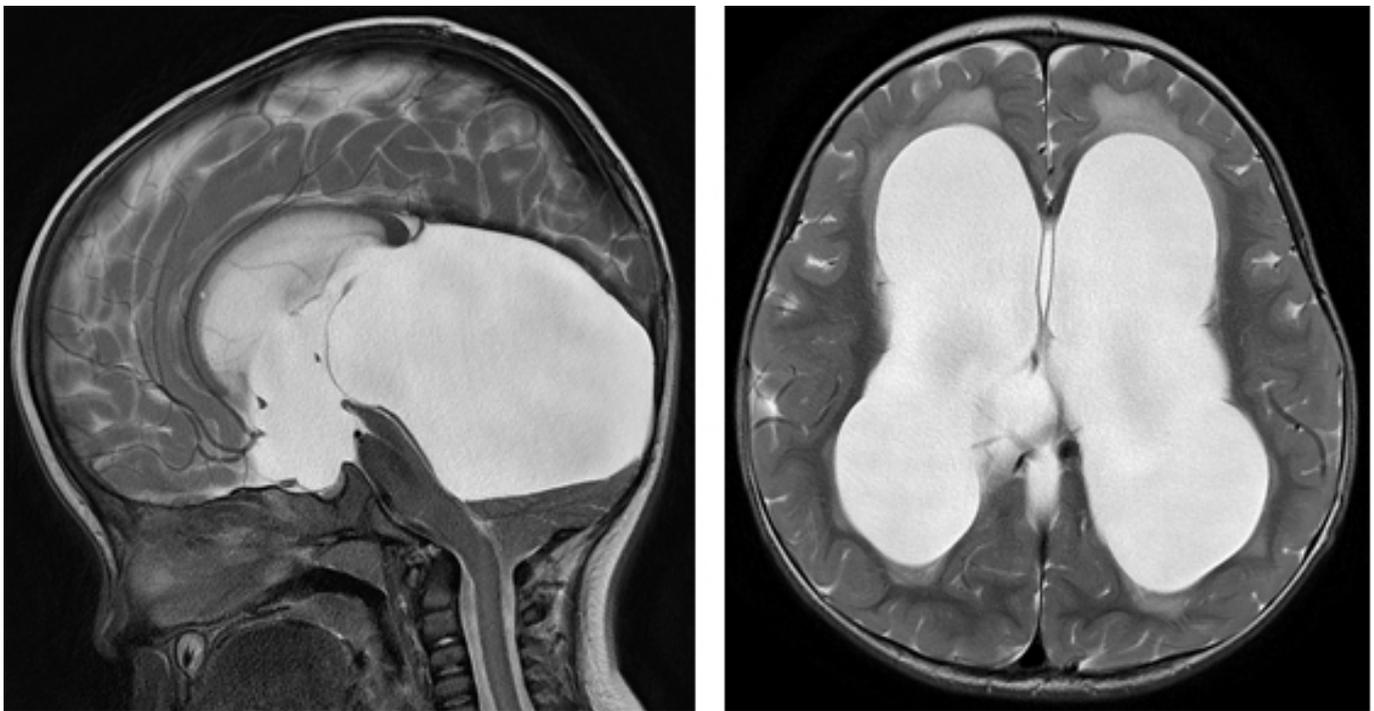


Fig. 1. Magnetic resonance imaging demonstrated a large cyst on the superior portion of the posterior fossa with enlarged the third and lateral ventricles. The cerebellum and brain stem were compressed severely.

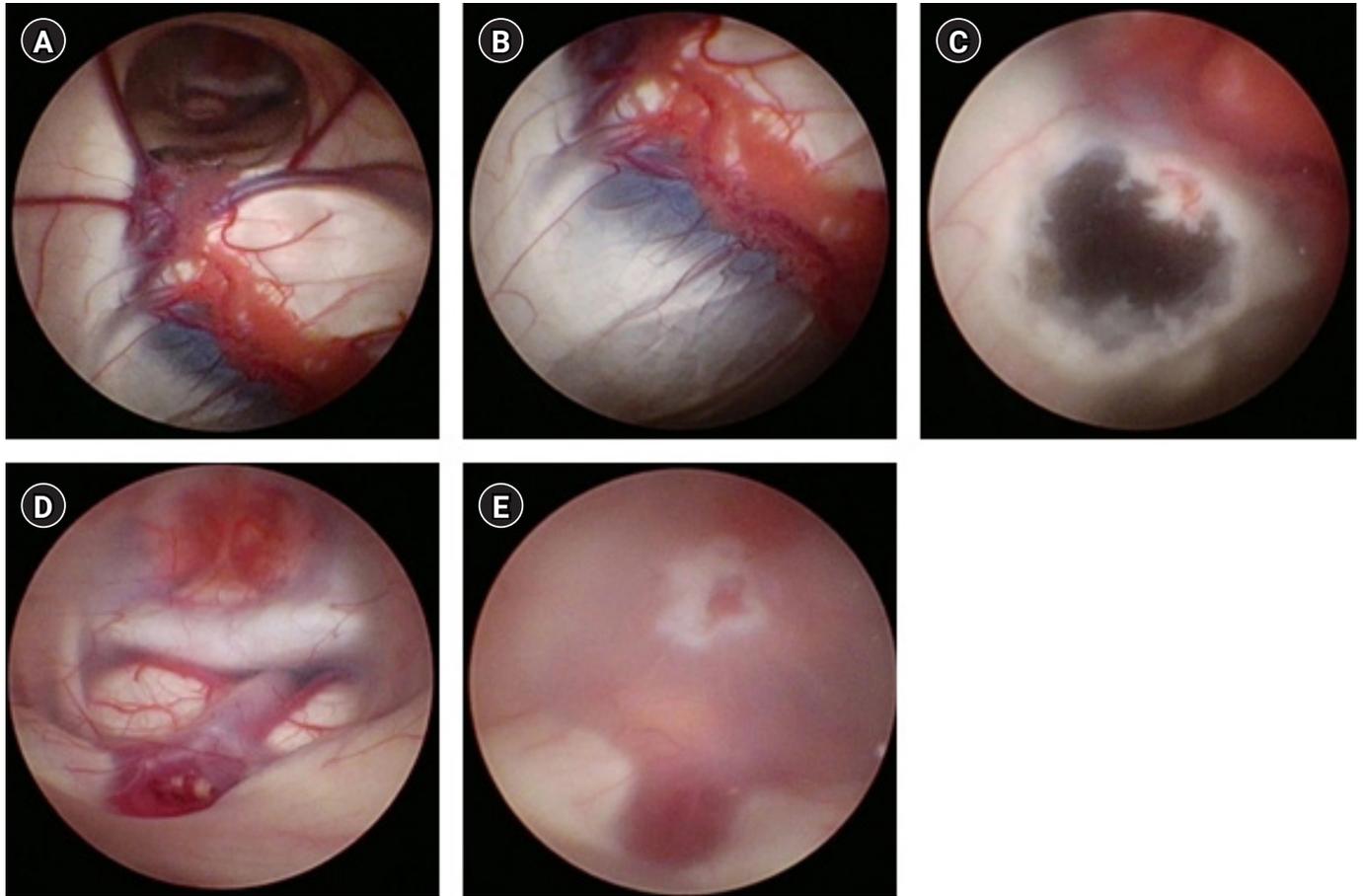


Fig. 2. Endoscopic view of the right lateral ventricle (A, B). The fenestration was made on trigone of the lateral ventricle and arachnoid cyst wall (C). Third ventriculostomy was performed at the floor of the third ventricle (tuber cinereum) (D, E).

The rigidity of both legs was relieved one day after surgery. One-week postoperative CT showed a reduction in the size of the ventricles and cysts accompanied by bilateral subdural hygroma. The patient was able to stand unassisted and could walk with two-hand assistance. At nine months after surgery at 29 months of age, the patient was able to run and speak in sentences. A CT image showed further reduction of the arachnoid cyst, lateral and third ventricles, and hygroma (Fig. 3).

DISCUSSION

Quadrigeminal cistern arachnoid cyst develops in the tectal plate region along the midline, supracerebellar, and infratentorial cisternal space⁹. Because of the arachnoid cyst increase caused by CSF formation from the cyst wall or entrapment of CSF, there are many possible extensions to ad-

jacent brain structures. An arachnoid cyst may extend to the trigone cranially, supracerebellar cistern caudally, the third ventricle anteriorly, and ambient cistern laterally⁷. Cinalli et al.⁷ classified quadrigeminal cistern arachnoid cysts into three types: type I, cyst with supratentorial and infratentorial extension; type II, infratentorial extension; type III, lateral extension. In anterior extension, the Sylvius aqueduct and fourth ventricle can be compressed and displaced downward, and the third ventricle can be distorted and displaced anteriorly³. A quadrigeminal cistern arachnoid cyst is usually associated with obstructive hydrocephalus.

Because an arachnoid cyst is located in a deep location and near important neural structures, many different and critical symptoms can be created by compression. Usually, an arachnoid cyst compresses the tectal plate of the mid-brain and leads to obstructive hydrocephalus by compression of the Sylvius aqueduct. The associated symptoms

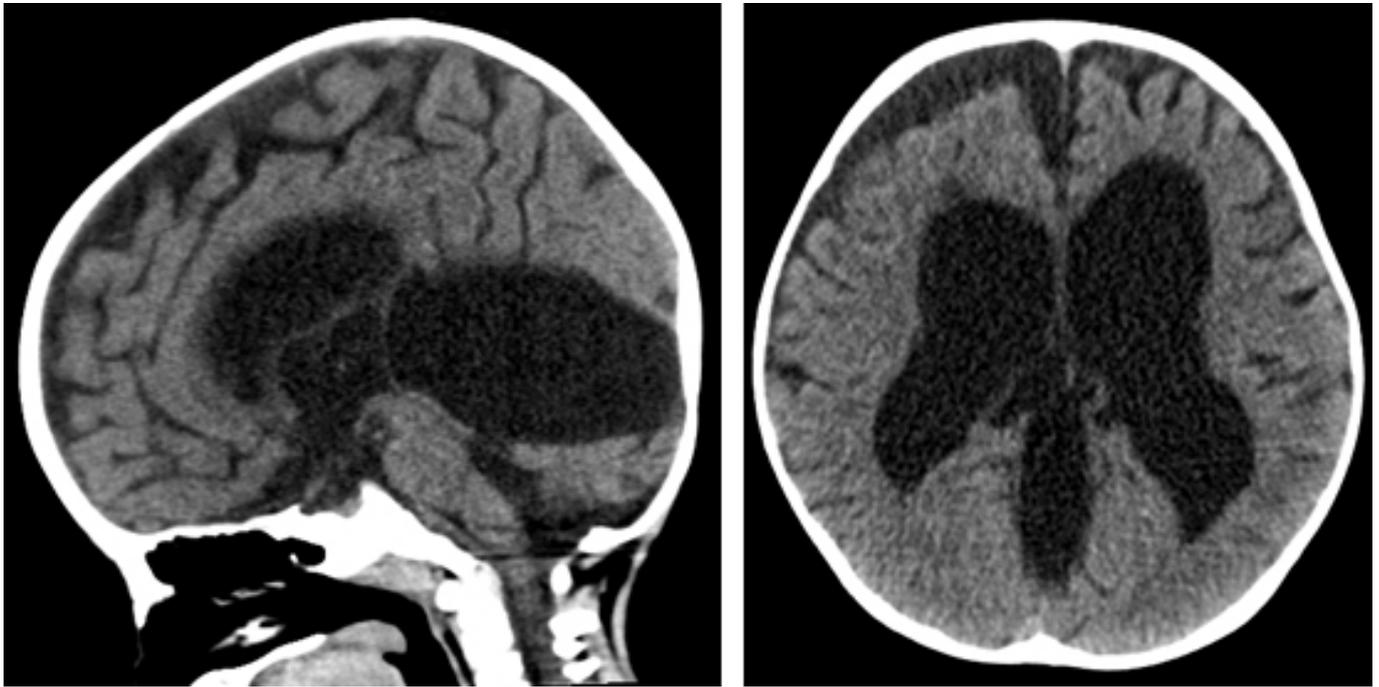


Fig. 3. Computed tomography scan showed a reduction of the arachnoid cyst, lateral and third ventricles.

are mainly related to hydrocephalus, such as headache, nausea, vomiting, lethargy, and ataxic gait. Due to direct compression of the tectal plate, visual symptoms may occur. In children, macrocephaly can result from obstructive hydrocephalus and developmental regression can develop but is rare²⁾. In our case, though the patient had rare symptoms, such as bilateral leg rigidity and developmental regression, the symptoms resolved after treatment.

Optimal treatment of quadrigeminal cistern arachnoid cyst is not obvious. Traditional treatment is open craniotomy with cyst wall removal or fenestration and cysto-peritoneal shunting. However, open craniotomy can produce many complications in children, such as meningitis, oculomotor palsy, subdural hematoma, and seizure^{4,8)}. Arachnoid cysts can recur occasionally, and additional shunt replacement is needed for the treatment of hydrocephalus after surgery. Shunt procedure of cysto-peritoneal shunting or ventriculo-peritoneal shunting for hydrocephalus has many complications, such as infection, CSF leakage, disconnection, shunt malfunction, over-drainage, and skin breakdown over the shunt device⁵⁾. Recently, neuroendoscopic fenestration has been performed to treat intracranial arachnoid cysts. It is minimally invasive and effective, with a high success rate of 71% to 81%^{10,14)}. There are two approaches to neuroen-

doscopic surgery to treat quadrigeminal cistern arachnoid cysts. One is a direct approach through the infratentorial and supracerebellar route¹⁵⁾. This approach involves paramedian burr hole drilling and neuroendoscope insertion into the cyst after removal of the posterior wall. The anterior wall of the arachnoid cyst is fenestrated carefully, avoiding the vein of Galen and other important vascular structures. The other is precoronal approach of ventriculo-cystostomy. Because quadrigeminal cistern arachnoid cyst is usually associated with hydrocephalus and an enlarged lateral ventricle, an endoscopic can be used in the large working space. Because the trigone of the lateral ventricle is attached to the enlarged cyst wall, ventriculo-cystostomy can be performed through the lateral ventricle.

The limitation of neuroendoscopic fenestration is an insufficient opening lead that results in a recurrence of cyst re-expansion. Many authors have reported additional third ventriculostomy to prevent a recurrence. In cases of simple fenestration between arachnoid cyst and lateral ventricle, the CSF circulation remains complex, and turbulent flow at the cyst level can lead to fenestration closure. With the third ventriculostomy, direct flow from the third ventricle to the prebular cistern decreases the turbulent flow, maintaining the fenestration of cysto-ventriculostomy. Ventriculo-cys-

tostomy and third ventriculostomy could be performed with one trajectory in the precoronal approach. Also, in cases of anterior extension of the arachnoid cyst to the third ventricle, ventriculo-cystostomy can be performed through the third ventricle via the foramen of Monro. In our case, ventriculo-cystostomy and third ventriculostomy were performed by neuroendoscope, and there was no complication or recurrence over nine months.

CONCLUSION

Quadrigeminal cistern arachnoid cyst is rare and is usually associated with hydrocephalus. Neuroendoscopic fenestration of arachnoid cysts with the third ventriculostomy is a safe and effective treatment for quadrigeminal cistern arachnoid cysts associated with hydrocephalus.

CONFLICTS OF INTEREST

No potential conflict of interest relevant to this article was reported.

REFERENCES

1. Abbott R: The endoscopic management of arachnoidal cysts. *Neurosurg Clin N Am* 15:9–17, 2004
2. Akutagawa K, Tamura G, Tsurubuchi T, Ishikawa E, Matsumura A, Inagaki T: Quadrigeminal arachnoid cyst with perinatal encephalocele. *Childs Nerv Syst* 36:1393–1397, 2020
3. Almeida JP, Quinino S, Faquini IV, Silva DO, Albuquerque LA, Mendes JC, et al.: Neuroendoscopic treatment of quadrigeminal arachnoid cyst in a two-year-old child. *Arq Neuropsiquiatr* 66:758–760, 2008
4. Choi JU, Kim DS, Huh R: Endoscopic approach to arachnoid cyst. *Childs Nerv Syst* 15:285–291, 1999
5. Cinalli G: Alternatives to shunting. *Childs Nerv Syst* 15:718–731, 1999
6. Cinalli G, Cappabianca P, de Falco R, Spennato P, Cianciulli E, Cavallo LM, et al.: Current state and future development of intracranial neuroendoscopic surgery. *Expert Rev Med Devices* 2:351–373, 2005
7. Cinalli G, Spennato P, Columbano L, Ruggiero C, Aliberti F, Trischitta V, et al.: Neuroendoscopic treatment of arachnoid cysts of the quadrigeminal cistern: a series of 14 cases. *J Neurosurg Pediatr* 6:489–497, 2010
8. Ciricillo SE, Cogen PH, Harsh GR, Edwards MS: Intracranial arachnoid cysts in children. A comparison of the effects of fenestration and shunting. *J Neurosurg* 74:230–235, 1991
9. di Rocco C, Caldarelli M, di Trapani G: Infratentorial arachnoid cysts in children. *Childs Brain* 8:119–133, 1981
10. Hopf NJ, Perneczky A: Endoscopic neurosurgery and endoscope-assisted microneurosurgery for the treatment of intracranial cysts. *Neurosurgery* 43:1330–1336, 1998
11. Hornig GW, Zervas NT: Slit defect of the diaphragma sellae with valve effect: observation of a "slit valve". *Neurosurgery* 30:265–267, 1992
12. Inamasu J, Ohira T, Nakamura Y, Saito R, Kuroshima Y, Mayanagi K, et al.: Endoscopic ventriculo-cystomy for non-communicating hydrocephalus secondary to quadrigeminal cistern arachnoid cyst. *Acta Neurol Scand* 107:67–71, 2003
13. Muhlestein WE, Maher CO: Incidental intracranial cysts in children. *Pediatr Clin North Am* 68:775–782, 2021
14. Pierre-Kahn A, Capelle L, Brauner R, Sainte-Rose C, Renier D, Rappaport R, et al.: Presentation and management of suprasellar arachnoid cysts. Review of 20 cases. *J Neurosurg* 73:355–359, 1990
15. Ruge JR, Johnson RF, Bauer J: Burr hole neuroendoscopic fenestration of quadrigeminal cistern arachnoid cyst: technical case report. *Neurosurgery* 38:830–837, 1996
16. Yu L, Qi S, Peng Y, Fan J: Endoscopic approach for quadrigeminal cistern arachnoid cyst. *Br J Neurosurg* 30:429–437, 2016