

## Case Report

### Infant with prenatally diagnosed arachnoid cyst

Zoe Zhang, MD; David Nascene, MD\*; Stephen Haines, MD

Departments of Neurosurgery and Radiology\*, University of Minnesota

#### Case

This patient's central nervous system abnormality was first suspected during a routine prenatal ultrasound at 29 weeks gestation. It was discovered that the patient had ventriculomegaly and a pineal region cyst (Figure 1a). A fetal MRI was performed at 31 weeks gestation (Figure 2). This showed a 3x3x3 cm unilocular cyst in the quadrigeminal plate cistern causing obstructive hydrocephalus. The fourth ventricle was normal in size. The corpus callosum was fully formed. No other anomalies were found. A repeat fetal ultrasound was performed prior to birth (Figure 1b).

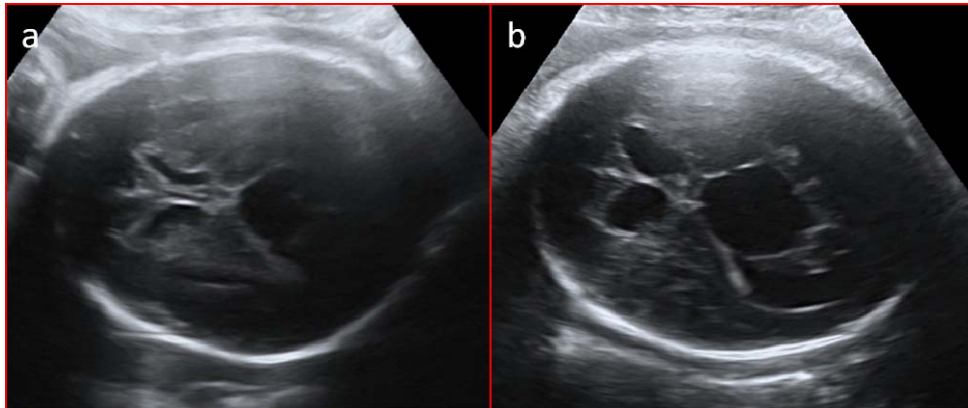


Figure 1: a) fetal ultrasound of the head at 29 weeks gestation, showing ventriculomegaly of the lateral and 3<sup>rd</sup> ventricles. b) fetal ultrasound of the head at 34 weeks gestation showing further increased ventriculomegaly. There was no color flow by Doppler imaging (not shown).

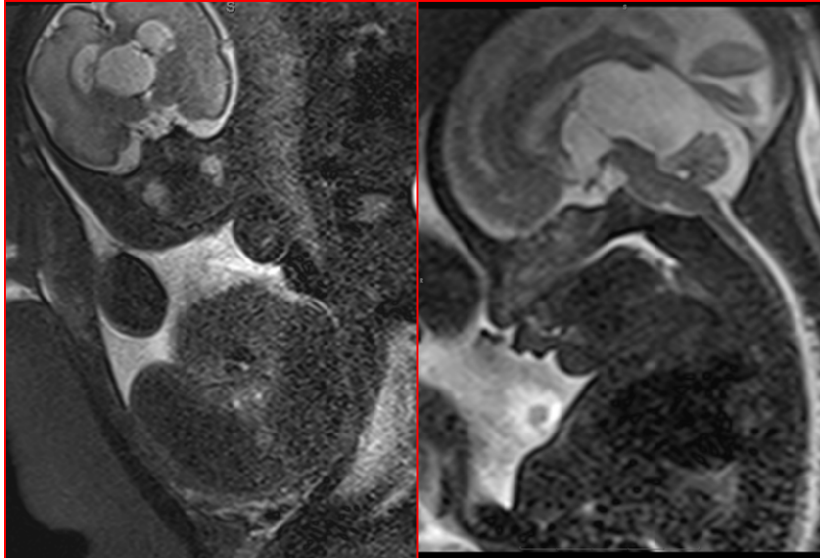


Figure 2: Prenatal MRI of the fetus with coronal and sagittal T2 weighted images showing a cyst in the quadrigeminal plate cistern, causing mass effect on the third ventricle, the cerebral aqueduct, and the cerebellar vermis.

The patient's mother is a 26 year old G3P2 female. Her previous two pregnancies were uneventful and her other children are healthy. There had been no complications with this pregnancy. At 39 weeks gestation labor was induced and a c-section performed because of macrocephaly. Apgar scores were 8 and 9 at 1 and 5 minutes respectively. Initial physical exam yielded occipito-frontal circumference of 39.5 cm, which was at the 99<sup>th</sup> percentile. Anterior and posterior fontanelles were full, but not bulging. The remainder of his exam was normal.

MRI was obtained after birth (Figure 3). This showed the cyst to be 5x4x3.5 cm in size located in the quadrigeminal plate cistern, increased in size compared to the prior study. It displaced the internal cerebral veins and the vein of Galen superiorly, and the tectum inferiorly. The pineal gland was displaced anteriorly and did not appear to be the origin of the cyst. It obstructed the cerebral aqueduct and caused lateral and third ventriculomegaly. The fourth ventricle was normal in size. After contrast was administered, there was no abnormal enhancement. Because of the increase in cyst size and ventriculomegaly, it was decided to take the patient to the operating room for endoscopic fenestration of the arachnoid cyst into both lateral ventricles and the third ventricle, followed by third ventriculostomy.

Patient returned at 2 months for a post-operative MRI. This showed CSF flow artifacts on T2 weighted imaging and significantly decreased arachnoid cyst with decreased mass effect and improved ventriculomegaly (Figure 4).

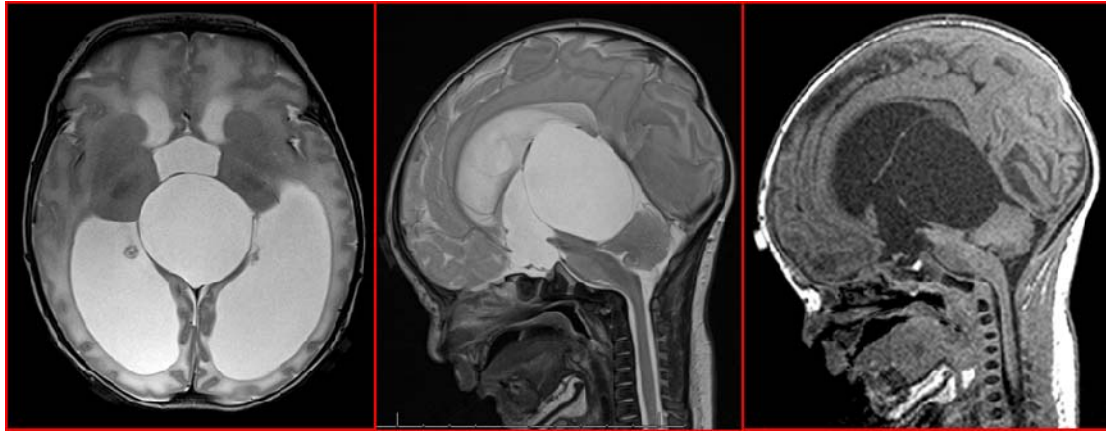


Figure 3: MRI of the brain after birth, axial and sagittal T2 weighted images, sagittal T1 weighted image, that show enlarged lateral and third ventricles with a 5x4x3.5 cm cyst in the quadrigeminal plate cistern. The pineal gland is normal in size, but displaced anteriorly. There is obstruction of the cerebral aqueduct. There is deformation of the cerebellar vermis.

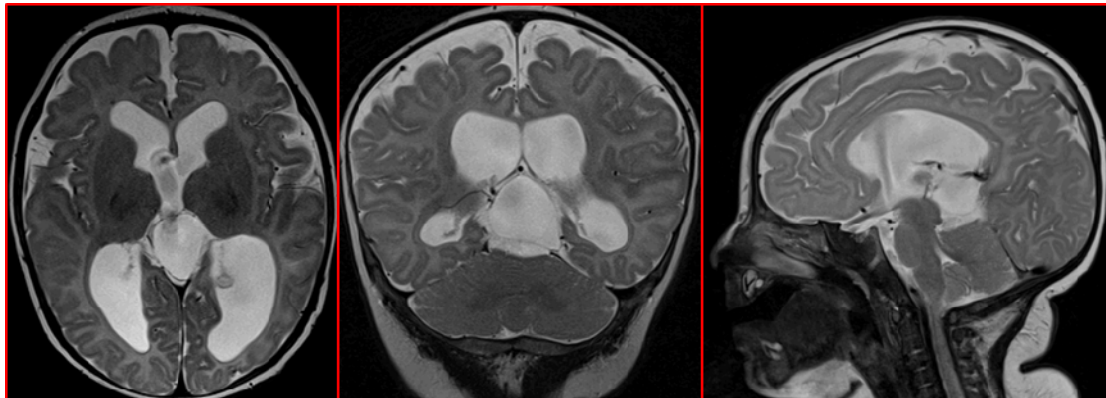


Figure 4: MRI of the brain 2 months post-operatively with axial, coronal, and sagittal T2 weighted images. There is decrease in size of the cyst, now measuring 2.4 x 2.5 x 2.5 cm. There is decreased mass on the quadrigeminal plate. The cerebral aqueduct is now patent. The cyst no longer deforms the cerebellar vermis, however it still remains distorted. There is improvement of ventriculomegaly.

## **Discussion**

Fetal MRI is an important adjunct to ultrasound in the diagnosis of central nervous system lesions. In cases such as this, the benefit to a confident, early diagnosis lies in optimizing peripartum care with cesarean section for macrocephaly, and decreasing the rates of traumatic hemorrhage and cyst rupture with birth.

Our differential diagnosis included arachnoid cyst, pineal cyst, and pineal neoplasm. A non-enhancing cyst with fluid of similar signal intensity as cerebrospinal fluid located near the third ventricle is almost certainly an arachnoid cyst. Accurate localization of the lesion plays a critical role in differential diagnosis. Pineal cyst was considered, however, with improved images post-natally, it was seen that the cyst did not originate from the pineal gland. Contrast enhancement of the lesion also narrows the differential diagnosis; based on the location, colloid cyst was a consideration, however these are generally hyper-intense on T1 imaging and iso- or hypo-intense on T2 weighted MRI imaging.

Arachnoid cysts are cysts filled with cerebrospinal fluid. They are divided into two categories, simple, in which they are wrapped in arachnoid cells only, and complex, in which they are wrapped in other tissues such as neuroglia and ependyma. These complex cysts are thought to be a result of misplaced neuroectodermal tissue and are associated with other developmental dysplasia such as agenesis of the corpus callosum and heterotopias.

These cysts can develop anywhere along the arachnoid membrane.<sup>1</sup> They are most common along the sylvian fissure and are classified into three types: type 1 is small, biconvex, and located in the temporal tip; there is communication with the subarachnoid space. Type 2 is larger and involves the proximal and intermediate segments of the fissure with partial communication with the subarachnoid space. Type 3 involves the entire fissure with midline shift, bony expansion, and no communication with the subarachnoid space (Figure 5). Much more rarely, they can be found at a cerebral convexity (Figure 6).

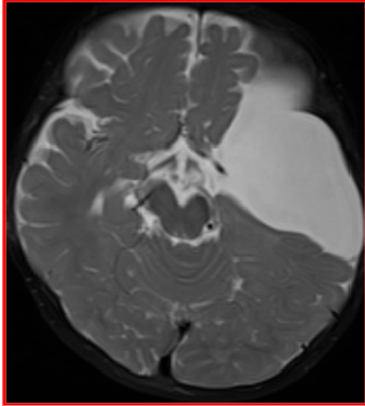


Figure 5: T2 weighted axial MRI of the brain in a different patient, depicting an arachnoid cyst that extends along the entire left Sylvian fissure.

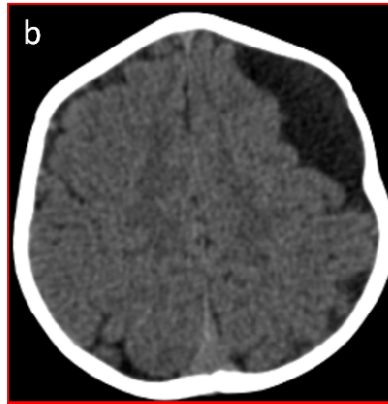
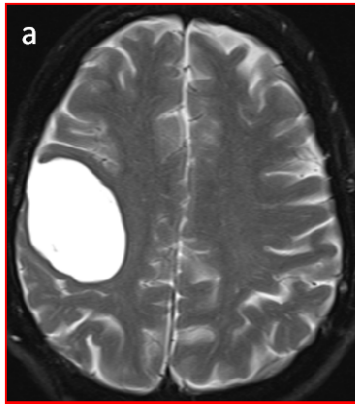


Figure 6: in two different patients, a) axial T2 weighted MRI image of the brain showing an arachnoid cyst in the right superior prefrontal sulcus. b) a CT without contrast showing focal expansion of the skull around the left frontal convexity arachnoid cyst.

Primary arachnoid cysts are believed to be a congenital disorder with developmental malformation of splitting of the arachnoid membrane. It makes up 1% of all intracranial masses and its incidence is 5/1000.<sup>2, 3</sup> It is twice as common in males as in females. These cysts can also be acquired, resulting from head injury, meningitis, tumors, or surgery, referred to as secondary arachnoid cysts.

On imaging, they have density similar to CSF with no contrast enhancement. They are generally diagnosed via imaging such as CT and MRI. MRI is a better diagnostic choice as it gives better visualization of the fluid within the cyst and the surrounding walls. CSF studies such as cisternograms and ventriculograms are rarely needed.

These cysts are usually delayed in diagnosis as they are most commonly asymptomatic. When present, symptoms vary with the location, but typically are those of hydrocephalus: nausea, vomiting, lethargy, and seizures. With sudden deterioration, one must consider the possibility of hemorrhage into the cyst, hemorrhage into a subdural compartment, or cyst rupture.

Treatment is varied and highly controversial.<sup>4</sup> Management strategies include watchful waiting, shunting of the cyst and either endoscopic or open fenestration of the cyst to a normal CSF space. Some clinicians recommend treating only those cysts which are causing symptoms, whereas others advocate that even asymptomatic cysts should be decompressed to avoid future complications.

One treatment modality involves needle aspiration of the cyst; however, if the cause was due to a lack of CSF outflow, the cyst will easily reaccumulate necessitating further aspiration. Craniotomy with cyst wall excision and fenestration of the fluid into the basal cisterns is another option. Its less invasive sister procedure is endoscopic fenestration via a burr hole (the procedure used in our case). In 2009, Spacca retrospectively studied 40 patients who had undergone endoscopic fenestration for middle fossa arachnoid cysts.<sup>5</sup> There was no death or significant morbidity associated with the endoscopic procedure. In 4 patients, further surgical treatment was required. In 4 other patients, post-traumatic intracystic bleeding occurred remotely from the date of the procedure. These rates were comparable to craniotomy and cyst wall excision. From their experience, the authors concluded that endoscopic fenestration was as effective and safe as microsurgical fenestration with cyst shunting while being less invasive.

The final modality of treatment is shunting of the cyst. This is a definitive treatment; however, the patient becomes shunt dependent with all the accompanying risks of having a shunt including infection and malfunction.

### **Teaching points**

1. Fetal MRI can be a useful modality in diagnosing arachnoid cysts prior to birth, potentially decreasing the rates of traumatic hemorrhage and cyst rupture with birth.

2. Arachnoid cyst should be near the top of the differential diagnosis of a CSF signal intensity cyst near the third ventricle.
3. Endoscopic fenestration is a less invasive modality of treating arachnoid cysts with rates of success similar to that of microsurgical fenestration with shunting.

### **References**

1. Greenberg MS. Handbook of Neurosurgery. 2006
2. Flaherty AW. The Massachusetts General Hospital Handbook of Neurology. 2000
3. Kanev PM. Arachnoid cysts. Youmans Neurological Surgery. 5<sup>th</sup> Edition
4. NIH: National Institute of Neurological Disorders and Stroke: Arachnoid Cysts Information Page. <[http://www.ninds.nih.gov/disorders/arachnoid\\_cysts/arachnoid\\_cysts.htm](http://www.ninds.nih.gov/disorders/arachnoid_cysts/arachnoid_cysts.htm)> 2010 May 5
5. Spacca B, Kandasamy J, Mallucci CL, Genitori L. Endoscopic treatment of middle fossa arachnoid cysts: a series of 40 patients treated endoscopically in two centres. Childs Nerv Syst. Jul 24 2009

### **Corresponding Author:**

Zoe Zhang, MD

Mayo Memorial Building, 4th Floor, D-429

420 Delaware Street SE

Minneapolis, MN 55455

Tel: 612-624-0196

Email: zhan1024@umn.edu