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Case Report

Surgical aspects of arachnoid cysts: report of 3 cases and brief review

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Since the use of prenatal ultrasonography and the widespread availability of computed tomography (CT) and magnetic resonance imaging (MRI), arachnoid cysts are able to be detected at an earlier age. If the cyst is very large or associated with hydrocephalus, neurologic deficits, or intracranial bleeding, surgical intervention is considered to be the treatment of choice for most neurosugeons.

Children with arachnoid cysts may experience symptoms such as headache, seizure, learning deficits, or behavioral disturbances. In infants increasing head circumference may be the only sign. Surgery should generally be performed as early as possible to avoid a skull-brain mismatch and prevent increasing pressure from the cyst on the underlying developing brain. Craniotomy and fenestration, cystoperitoneal shunting, and endoscopic marsupialization have been used to treat arachnoid cysts.

This paper reports two cases with surgical intervention and another case with non-surgical treatment.

Case report

Case 1

A one month-old baby was brought to Neurosurgery Clinic at Siloam Hospitals Lippo Cikarang. His mother said that when she was pregnant at about 33-34 weeks of gestation, prenatal ultrasonography had found hydrocephalus and dilatation of the right and left lateral ventricles (**Figure 1**). The large fontanelle of the baby was about 4-6 cm long, and head circumference was 40 centimeters.

Intracranial ultrasonography was performed when the baby was seven days old. Multiple cystic

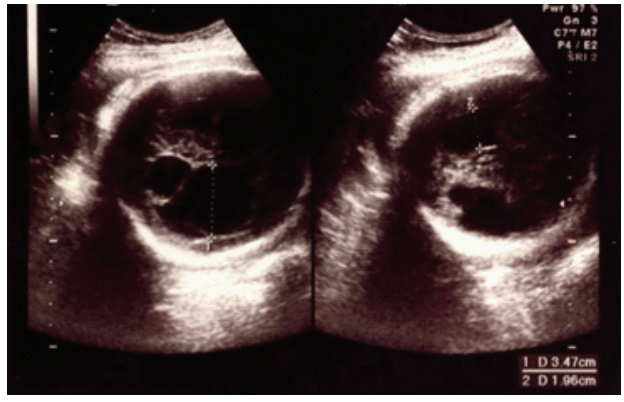

Figure 1. Prenatal ultrasonography from 33-34 weeks gestation showing dilatation of lateral ventricle.

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masses were seen in the left hemisphere, in parietal, temporal and occipital lobes, with moderate hydrocephalus on the right side and severe hydrocephalus on the left (**Figure 2**).

Cranial MRI confirms a cystic masses replacing the left parietal lobe and the posterior frontal lobe, continuous with the subarachnoid space in the left vertex region. The thin wall cyst contains CSF-like fluid suspected of arachnoid cyst. That cystic mass displaces the left lateral ventricle, with dilatation of the left lateral and third ventricles (**Figure 3**).

When the baby was 38 days old, endoscopic cysto-ventriculocystostomy was performed. Fenestration of the arachnoid cyst into the ventricular compartments was done after thorough inspection of the cystic surface. Continuous irrigation is needed during endoscopy to maintain a clear medium for image transmission and for hemostasis. It also provides real-time feedback about the translucency and consistency of the membranes onto which the fluid current is directed, allowing for the safe selection of fenestration target sites.

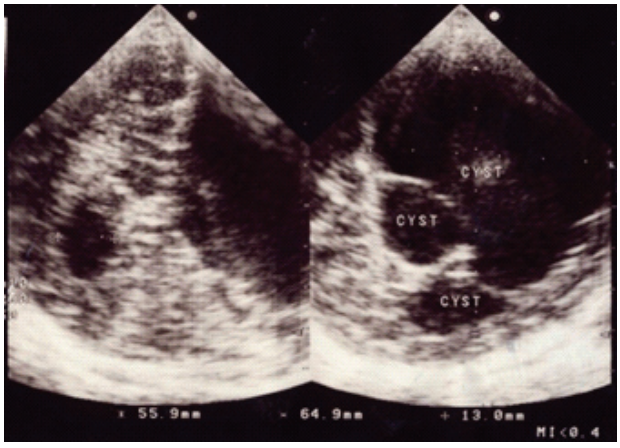

Figure 2. Intracranial ultrasonography of 7 days-old baby showed multiple cystic mass and dilatation of lateral ventricles.

Once the target site has been selected, a bipolar diathermy instrument is used both to attenuate the tissue and to provide a blunt instrument with which to perforate the cyst wall. After a small communication between the cyst and the ventricle has been made, progressive enlargement of the fenestration is created using a combination of balloon dilation, blunt perforation, and sharp scissors to connect a series of smaller fenestrations and create a large conduit for CSF egress. At the completion of the procedure irrigation is briefly withheld, and the adequacy of the cyst fenestration can be judged by the to-and-fro motion of the free edges of the fenestrations, which should move freely with the CSF pulsations of the cardiac cycle if they are in adequate communication with a CSF-containing compartment.

Two weeks after surgery, the large fontanelle of the baby was relaxed and he was discharged from the hospital with recommendation to do serial evaluation at our neurosurgery clinic. Follow up CT-scan one month later showed the ventricles were smaller and the two month-old baby was in good condition, growing up normally for his age. Fifteen months later, the 16 month-old baby was in very good condition and was learning to walk and speak properly. An MRI illustrated reduction in cyst volume. (**Figure 4**).

Case 2

This eleven year old boy came to our clinic with hyperactivity and aggressive behavior, delayed

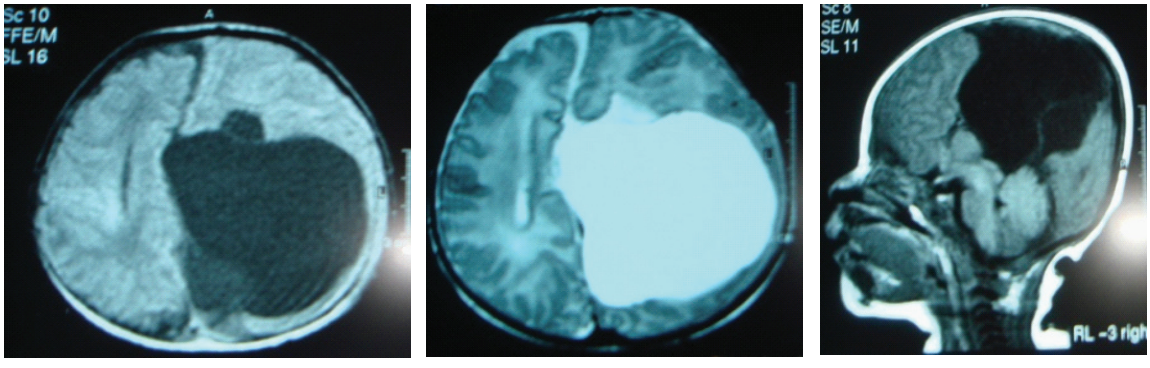

Figure 3. The preoperative MRI showed a cyst and dilatation of the ventricles

psychomotor development, and poor interpersonal relationship. Neurological examination showed no remarkable findings. MRI confirmed a giant arachnoid cyst in the right temporobasal area (**Figure 5**). EEG showed irritative dysfunction in the right temporo-parietal region.

Surgical intervention was done with craniotomy and marsupialization to reduce cyst volume and make a connection between cyst and subarachnoid space. Postoperative condition was good with improved behavior. However, serial evaluation of his condition is still needed, and he has been referred to a pediatric psychiatrist for continuing cognitive evaluation. Post-operative MRI showed reduction in cyst volume (**Figure 6**).

Case 3

A six year old boy with a history of arachnoid cyst. When he was six months old, he fell out of bed, but the neurological examination then showed unremarkable findings. MRI was performed to check for traumatic injury, but showed a large cystic mass in the left fronto-temporal region without compression of the brain. The boy was in very good condition, and has been given conservative treatment with serial observation.

The boy is growing up normally for his age and is achieving well at school. For evaluation of his anomaly periodic MRIs are performed. They show a large cystic mass in the left fronto-temporal region, compressing the left cerebral hemisphere and lateral ventricle (**Figure 7**).

Discussion

Arachnoid cysts were initially described by Bright in his *Reports of Medical Cases* published in 1831 as "serous cyst within the arachnoid".[1,2] Arachnoid cysts are fluid-filled sacs lined by arachnoid membrane and form in surface regions of the brain usually around the cranial base. The cysts are filled with clear, colorless fluid virtually identical to cerebrospinal fluid (CSF).[3-5] Its precise location is either between pia and subarachnoid, within layers of the arachnoid, or between arachnoid and dura. [6]

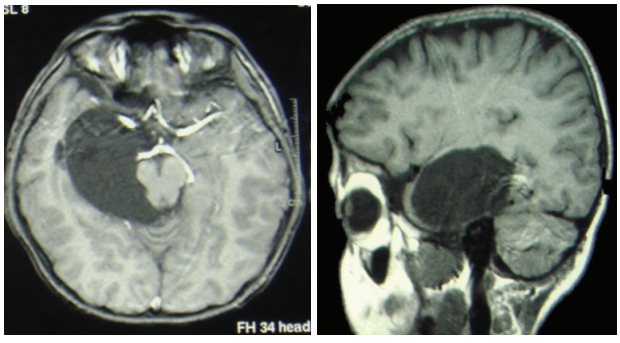

Figure 5. Preoperative MRI of a 11-year-old boy with right temporobasal cyst.

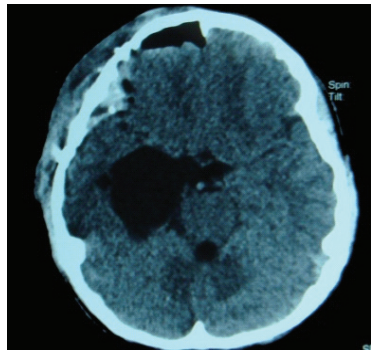

Figure 6. Post-operative MRI

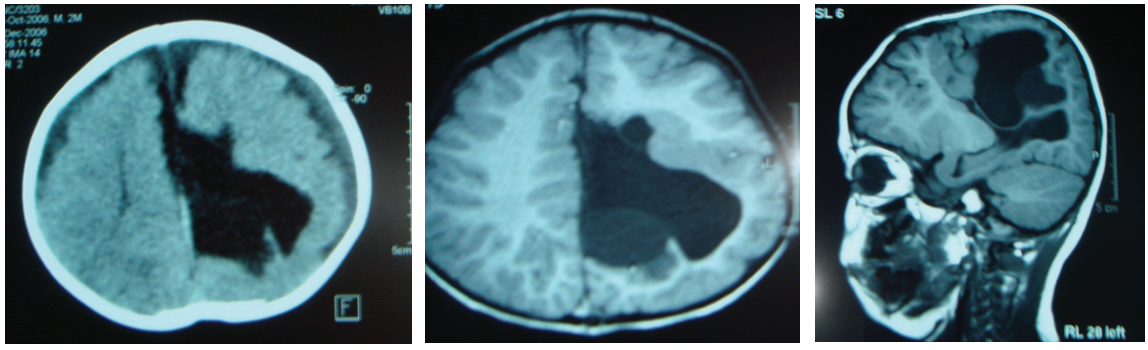

Figure 4. Serial postoperative evaluation illustrated significant reduction in cyst volume.

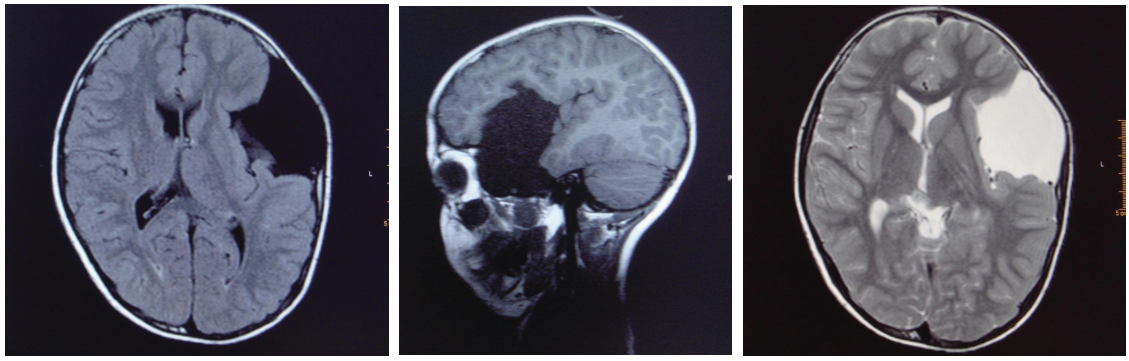

Figure 7. MRI showed a large cystic mass compress the left cerebral hemisphere and the left lateral ventricle.

In the United States, arachnoid cysts constitute approximately 1% of intracranial masses, with 50-60% occurring in the middle cranial fossa.[5-8] The male-to-female ratio is 4:1. The exact incidence of mortality and morbidity from arachnoid cysts is not known, as many people with arachnoid cyst never develop symptoms and the cyst remains undiagnosed.[3,8]

Intracranial arachnoid cysts may be primary (congenital) or secondary (acquired). Infants are most susceptible to developing arachnoid cysts, although cyst formation can occur up through adolescence.[3,6] Morbidity and mortality depend on the location of the cyst and whether complications occur, as intracystic hemorrhage can cause acute mass effect or subdural hygroma/hematoma, or obstruction of CSF outflow pathways.[1,8]

Pathologically, microscopic examination of arachnoid cyst shows that the walls are formed from a splitting of the arachnoid membrane, with an inner and an outer leaflet surrounding the cyst cavity; the cyst wall consists of fibrous connective tissue slightly denser than normal arachnoid tissue.[8,9]

Arachnoid cysts arise mainly because of an abnormality occurring in development, sometime as a result of a neonatal infection. Other cysts are congenital and presumably result from abnormal formation of the subarachnoid space during prenatal development.[3] Acquired arachnoid cysts may develop following surgery, trauma or subarachnoid hemorrhage, and can occasionally occur in association with an extra-axial neoplasm.[3,8] These acquired lesions are also referred to as leptomeningeal cysts, and are similarly thought to arise from injury to one of the arachnoid membranes and a ball valve-like mechanism of CSF pulsation, which favors gradual expansion of the cyst.[7]

Arachnoid cysts can be classified according to the location along the neural axis or by the histologic composition of the cyst wall, which is either arachnoid connective tissue or glioependymal tissue. Cysts in the cerebral hemispheres or the spinal cord are mostly arachnoid, while those in the supracollicular or retrocerebellar regions of the brain tend to be composed of either arachnoid connective tissue or glioependymal tissue.[3,8]

The potential expansion of arachnoid cysts during subsequent development probably has multifactorial causes. One theory supports growth via underlying CSF pulsation concordant with the cardiac cycle, fluid becoming entrapped between the leaflets of the membrane. It has also been reported that arachnoid cysts can expand due to an oncotic gradient resulting from increased protein concentration within the cyst.[1,3,7]

Arachnoid cysts occur in multiple locations throughout the intracranial compartment.[7] Three common location are the floor of the middle cranial fossa, the septal region, and the prepontine cistern. They can also occur in the posterior fossa, the cerebellopontine angle, interhemispherically, along the quadrigeminal plate, and anywhere along the cortical surface.[7,8] Most cysts are unilateral, smoothly rounded, and adhere loosely to the dura. Arachnoid cysts can indent deeply into the hemisphere or invaginate into major fissures, displacing and flattening the underlying cortex.[8]

Patients with arachnoid cysts usually are asymptomatic even if the cyst is quite large.[8] The cysts are often discovered as incidental findings on cranial imaging during an unrelated examination.[3,5,10] However, they may occasionally become symptomatic because of their pressure effects on surrounding structures. Since

the increasing use of prenatal ultrasonography and the widespread availability of either computed tomography (CT) or Magnetic Resonance Imaging (MRI), cysts are being detected at an earlier age. Many arachnoid cysts are recognized during the first decades of life, and some become symptomatic in children. In the European Cooperative Study, the mean age at presentation was 6 years.[11] Arachnoid cysts manifest variable clinical signs depending on their size and location within the subarachnoid space.[5] Symptoms include headache, seizures, mental retardation, cognitive impairment, calvarial bulging, intracranial hypertension, craniomegaly, precocious puberty, with focal neurological signs occurring less frequently.[7,10] Association with both intracystic and subdural hemorrhages, due to rupture of bridging veins or vessels in the cyst wall, are also well known.[10]

Headaches may localize to the site of the cyst, suggesting pain due to stretching of neurovascular structures, or they may be generalized and secondary to intracranial hypertension. In other case, the cyst may be large and associated with mass effect or is associated with signs of raised intracranial pressure.[11]

The second most common presentation of arachnoid cysts is partial or generalized epilepsy. Patients with middle fossa arachnoid cysts presented with seizures (34% of 77 patients) and 55% demonstrated abnormalities on electroencephalography. 80% of children manifest behavioral abnormalities, attention deficit disorder, or problems in school.[11]

Patients with suprasellar arachnoid cyst most commonly present with headache, visual disturbances, and neuroendocrine symptoms. Symptoms of ventricular obstruction occur relatively late. Hypopituarism is seen rarely and commonly affects growth hormone and corticotrophin.[11]

The most common clinical presentation of the arachnoid cyst in the midline posterior fossa is symptoms of ventricular obstruction such as morning headaches, nausea, vomiting, and gait ataxia. Arachnoid cysts of the cerebellopontine angle may present as facial weakness, tinnitus, diminished hearing, or gait ataxia. In the young infant, axial cerebellar signs are commonly misdiagnosed as gross motor delay because the child fails to learn to walk on schedule.[11]

MRI is the diagnostic procedure of choice because of its ability to demonstrate the exact location, extent, and relationship of the arachnoid cyst to adjacent brain or spinal cord. MRI facilitates diagnosis of these lesions with multiplanar visualization and has greater sensitivity to associated malformations than does contrast-enhanced CT. On MRI, arachnoid cysts appear as well-defined nonenhancing intracranial masses that are isotense to CSF. The extra-axial location of cysts and their relationship to adjacent neural and vascular structures are best visualized by T1-weighted sequences. Cyst fluid has low attenuation on T1 images and a high signal on T2 sequences and is nearly identical to CSF.[5,8]

On CT scan, arachnoid cysts are characterized by sharp nonenhancing borders and are isodense to CSF. They typically show compression of the subarachnoid space by a cystic structure that may be unilocular or septate and of variable size, although the septa may not always be visible.[8]

Cranial ultrasonography is an important diagnostic tool during the first year of life and is limited by the closure of the anterior fontanelle, which normally occurs in full term infants aged 9-18 months. Although infants with arachnoid cysts rarely are symptomatic, ultrasound provides a noninvasive imaging technique with a high yield in the detection and characterization of cystic masses. Intracranial cysts and ventriculomegaly also can be detected and characterized by transcranial ultrasound through a burr hole.[8]

Arachnoid cysts may be diagnosed in fetuses during routine examination and have been visualized as early as 26 weeks' gestation during prenatal ultrasonography. Ultrasound examination of arachnoid cysts demonstrates a well-defined anechoic lesion with adjacent mass effect; ultrasound may also be used to follow the growth of the cysts as well as the size of the ventricles.[5,6,11]

A large number of asymptomatic arachnoid cysts are being routinely discovered with advances in neuroimaging.[7] For asymptomatic patients, conservative treatment with observation is recommended, but it is still controversial especially in the pediatric population.[7,12] Cysts that have caused macrocephaly or hydrocephalus in infancy require surgical treatment to control head growth and to prevent development of more extreme craniocerebral disproportion.[13]

Some authors have advocated surgical intervention for most of these lesions because they have the potential of hindering the function of adjacent brain, cyst rupture, intracystic hemorrhage or subdural hemorrhage leading to sudden severe neurological deterio-

ration. Nevertheless, many other researchers favor a more conservative approach. In several studies it has been documented that even cysts causing only mild and nonspecific symptoms may affect the function of neighboring cerebral tissue, causing impaired cognition, and that such cognitive deficits tend to recover after surgery. However, a policy of treating a relatively benign condition with surgery is justified only when a clear clinical benefit and low risk of complications can be demonstrated.[14,15]

Options for surgery of symptomatic arachnoid cysts are stereotactic aspiration, excision, fenestration, cystocisternostomy, cystoventriculostomy, and the placement of cystoperitoneal shunts.[16] Reports of endoscopic cyst fenestration were rare just 7 years ago, but the field has since a large increase in the number of surgeons who are comfortably and routinely using the endoscope to approach these lesions. In addition, combined procedures, especially in the middle cranial fossa, such as small craniotomies followed by endoscopic cyst wall fenestration, are becoming the standard of care.[1,7]

In case 1, the arachnoid cyst in the baby was found in prenatal ultrasonography, and postnatal ultrasonography and MRI showed multiple cystic masses and hydrocephalus. The surgical indication for this baby was to prevent rising pressure from the cyst damaging the underlying, developing brain. In case 2, and eleven year old boy with hyperactivity and delayed psychomotoric development had a temporobasal arachnoid cyst, treated by craniotomy and marsupialization, and post-operatively showed less hypereractivity. Serial evaluation for post-operative condition and cognitive development of these children is still needed. In case 3, the six year old boy came to our clinic for serial evaluation, with a quite large arachnoid cyst in the left cerebral hemisphere. The boy is in very good condition and that cyst was found incidentally.

For asymptomatic patients even with quite large cysts but no complaints, no developmental delay, and no hydrocephalus or macrocephaly, conservative treatment is recommended. Surgical option should be done as soon as possible to avoid increasing pressure from the cyst and neurologic deficits, or delayed cognitive development. Surgical treatment is recommended in patients with focal neurological signs or symptoms of elevated intracranial pressure, and should be considered in patients with seizures linked to the presence of the cyst. The goal of intervention is reduction of the pressure exerted by the arachnoid cyst on adjacent brain structures. Surgical decompression of arachnoid cysts in children yields good long-term outcomes in patients, with a low risk of complications or additional impairment. Endoscopic assisted fenestration or marsupialization through a craniotomy seems the treatment of choice, because of better outcome and a lower rate of treatment failure.

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