영아에서 후소뇌 위치의 지주막 낭종 치료에 있어 다른 뇌척수액 역동학에 따른 고려점

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Considerations in Treating Neonatal and Infantile Patients with a Retrocerebellar Arachnoid Cyst: in the Perspective of Different CSF Dynamics

Arachnoid cysts found under the age of 1 year are more likely to grow in size, relatively short term follow-up is required. Retrocerebellar location predicts a high risk of hydrocephalus, and the time window until irreversible neuronal damage is often narrow if the arachnoid cyst increases in size and becomes symptomatic. However, when and how to treat a neonatal or infantile patient with a retrocerebellar arachnoid cyst is still a controversial subject. We recently experienced 3 differently treated very young pediatric patients with retrocerebellar arachnoid cysts. One patient was treated two weeks after birth by a cystoperitoneal shunt. This patient showed normal development after the surgery. The other patient was treated by a ventriculoperitoneal shunt and subsequent cystoventriculostomy at the age of 4 months because of his mother's refusal on 14th day after birth. This one showed developmental delay despite of decreasing size of ventricles after the surgery. The last patient was treated with microscopic fenestration, which failed in its initial attempt. A revision operation by cyst excision succeeded and had no problem after the surgery. Therefore, we suggest that early surgical intervention for retrocerebellar arachnoid cyst can be considered. Although there are concerns of long term complications related to shunts, a cystoperitoneal shunt would be a feasible treatment if we consider the minor cerebrospinal fluid pathway which is the dominant cerebrospinal fluid dynamic at this age.

Key Words: Posterior cranial fossa, Arachnoid cysts, Surgery, Pediatrics, Treatment

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Introduction

Arachnoid cysts are benign, extra-axial lesions filled with cerebrospinal fluid (CSF). These developmental anomalies are known to be caused by abnormal duplication of arachnoid membranes during brain development. They are most common in the middle fossa, followed by the posterior fossa^{1,2)}. Mostly, they are asymptomatic and do not require any treatment. However, they may increase in size and thus cause neurologic symptoms by compressing adjacent neural structures. Arachnoid cysts in the posterior fossa pose another problem in that they may cause obstruction in the CSF outflow pathway and develop obstructive

hydrocephalus^{3,4)}.

Shunting and fenestration were two major surgical treatments of intracranial arachnoid cysts. Fenestration using endoscope had gained popularity with good surgical outcomes in most pediatric patients^{5,6)}. With concerns of lifelong complications regarding shunts and recent trend preferring minimal invasive surgery, it became the most popular method for the treatment of arachnoid cysts. However, the different CSF dynamics and behaviors of arachnoid cysts in neonatal and infantile age groups have been recently illuminated⁷⁾, and we believe that these characteristics should be reflected in their treatment strategies, and specifically adjust our surgical strategies in these very young age groups. In this report, we present 3 very young pediatric patients with a posterior fossa arachnoid cyst who were in similar situations but showed different outcomes. Treatments were different for each case. However, they were all popular procedures widely performed nowadays in the field of neurosurgery without any points need for attention. Focusing on the age less than one year, we are to discuss the most appropriate treatment strategy for posterior fossa arachnoid cysts with a relevant literature review.

Case reports

Case 1

A 1-day-old boy visited our hospital due to a cyst at the posterior fossa on antenatal ultrasound (Fig. 1A). The boy was born at a gestational age of 37 weeks by Cesarean section. At birth, his weight was 2.87 kg (50–75 percentile) and head circumference was 36 cm (> 90 percentile). His mother had uncomplicated pregnancy. On physical and neurologic examination, he showed normal including flat and soft fontanel. His brain magnetic resonance imaging (MRI) revealed a large retrocerebellar arachnoid cyst occupying most of the occipital and posterior fossa (Fig. 1B). His parents were greatly concerned with the underdevelopment of the brain, and requested surgery. After 2 weeks, the patient received a cystoperitoneal shunt (PS medical, ultrasmall, low pressure type; Medtronic; USA) (Fig. 1C). Postoperative care was uneventful. On follow-up brain MRI at the age of 1 month, size of the arachnoid cyst was decreased and the occipital lobe and cerebellum showed nearly normal development (Fig. 1D). At the age of 3 years, he grew normally without showing any neurological symptoms or signs.

Case 2

A 1-day-old boy visited our hospital due to an infratentorial

cystic lesion incidentally found by brain ultrasonography (Fig. 2A). The boy was born at gestational age of 38 weeks by normal delivery. His birth weight was 3.75 kg, and his head circumference was 34 cm (50–75 percentile). His mother had a diabetes, but he did not showed any symptom including hypoglycemia, large for gestational age and hypocalcemia. His physical and neurologic examination showed normal. His brain MRI revealed a large retrocerebellar arachnoid cyst compressing adjacent neural structures (Fig. 2B). He had better get a surgery but his mother refused it. He did not report any more to the out-patient clinic follow-ups. Four months later, he revisited our clinic for macrocrania. His head circumference was 45 cm (over 97 percentile). On neurologic examination, he showed a fontanel bulging and the sunset eye sign. He could not sit and could not control even his head. His follow-up brain MRI demonstrated markedly enlarged ventricles and the retrocerebellar arachnoid cyst was enlarged compared to previous MRI (Fig. 2C).

Initially, ventriculoperitoneal shunt (PS Medical, ultrasmall, medium pressure; Medtronic; USA) was performed to treat

Fig. 1. Case 1. (A) A large retrocerebellar cyst was diagnosed on antenatal ultrasound. (B) T1 weighted sagittal MRI, taken one day after birth. A large retrocerebellar arachnoid cyst previously recognized by antenatal ultrasound was noted. (C) Immediate postoperative CT scan after the cystoperitoneal shunt operation. The proximal catheter tip is located in the retrocerebellar arachnoid cyst. (D) T1 weighted sagittal MRI taken one month after the surgery. The size of the retrocerebellar arachnoid cyst remarkably decreased, and the occipital lobes and cerebellum were well developed compared to the previous month.

hydrocephalus. Two months later, endoscopic cystoventriculostomy was performed to connect two spaces, the arachnoid cyst and the lateral ventricle, where the proximal shunt catheter was placed (Fig. 2D). Computed tomography (CT) was performed 6 months later, and it showed decreased size of the arachnoid cyst and ventricles (Fig. 2E). However, the change was limited, so we decided to exchange the medium pressure valve to low pressure. At 13 months of age, the child showed the gross motor delay with a difficulty in head and trunk control but achieved normal cognitive, language and social developmental milestones.

Case 3

A 7-week-old girl visited our clinic due to the increasing size of a retrocerebellar cystic mass on routine brain ultrasonography. She was born at a gestational age of 28+3 weeks by emergent cesarean section because of breech presentation and preterm labor with premature premature rupture of membranes. Her birth weight was 1.28 kg, and her head circumference was

37 cm (25-50 percentile). She showed normal physical and neurologic examination. Brain MRI showed a retrocerebellar arachnoid cyst, located posterior to the left cerebellar hemisphere (Fig. 3A). Because no neurologic symptoms presented, we put her under close observation. She showed normal motor development until 6 months of corrected age. However, she showed developmental regression one month later. Follow-up brain MRI showed a markedly increased size of the retrocerebellar arachnoid cyst severely compressing the adjacent cerebellum anteriorly (Fig. 3B). We performed microscopic fenestration connecting the arachnoid cyst and cisterna magna instead of a cystoperitoneal shunt because her parents did not want any foreign device implanted.

Her neurologic symptoms improved after surgery. However, follow-up brain MRI performed 6 months after the initial operation revealed enlargement of the retrocerebellar arachnoid cyst and ventriculomegaly which was not observed in the previous MRI (Fig. 3C). Under the impression of recurrence, we performed revision surgery, widening the craniotomy and

Fig. 2. Case 2. (A) Brain ultrasonography image after birth. A large retrocerebellar cyst was initially recognized. (B) T1 weighted sagittal MRI, taken immediately after brain ultrasonography. A large retrocerebellar cyst compressing the adjacent neural structures was shown. Occipital lobes were not shown, and severe cerebellar atrophy was observed. (C) T1 weighted sagittal MRI taken 4 months later. The size of the retrocerebellar arachnoid cyst increased. Enlarged lateral and third ventricles, periventricular lucency, and edema at the cerebral hemisphere indicated the presence of hydrocephalus. (D) Illustrative drawing of endoscopic cystoventricular fenestration performed 2 months after the initial ventriculoperitoneal shunt. We planned to drain cystic fluids through the ventriculoperitoneal shunt by connecting the cyst and the ventricle. (E) Brain CT axial image taken 6 months after the fenestration surgery. The size of the cyst and ventricle decreased.

excising as much of the arachnoid cystic wall as possible. Follow-up MRI performed 6 months after the second surgery showed a decreased size of the retrocerebellar arachnoid cyst and the ventricles. For 4 years of follow-up, she showed normal development without any neurologic deficit (Fig. 3D).

Discussion

1. Surgical indication and timing

Most arachnoid cysts are asymptomatic, and show a benign natural history that does not require surgical treatment^{1,2}. Surgical treatment is usually considered in cases of symptomatic or radiographically expanding arachnoid cysts. The location of arachnoid cysts is known to be correlated with symptom presentation. However, those reports demonstrated that the posterior fossa location did not significantly correlate with symptom presentation^{1,2}. Therefore, the mere presence of retrocerebellar arachnoid cysts should not be taken as an

absolute surgical indication.

Enlargement of retrocerebellar arachnoid cysts should be taken seriously. Hydrocephalus might ensue, and neuronal damage by increased intracranial pressure could be devastating if not treated properly in time. As shown in our second case, the therapeutic time window could be narrow although the neuronal plasticity in these periods is known to be great. In addition, arachnoid cysts are more like to grow in size in neonatal and infantile periods^{1,3,8}. Therefore, close clinical and brain image follow-up is mandatory. Follow-up intervals should be reduced to a few weeks, not months, if the size of the cyst is large or growing. Since the anterior fontanel is opened in this age, brain sonography could be a useful tool for imaging follow-up. Although the risk is great, one should be careful in selecting surgical candidates since even enlarging retrocerebellar arachnoid cysts do not always become symptomatic⁸. For this reason, the preemptive surgical treatment of the retrocerebellar arachnoid cyst by mere presence should be avoided unless it proves to be truly symptomatic lesion.

2. Surgical methods

The optimal treatment for symptomatic arachnoid cysts is still a subject of debate. In general, there are two treatment strategies: fenestration and shunting. Both methods had been widely used and showed similar effectiveness⁵. Each approach has its unique advantages and disadvantages. When successful, fenestration offers the patient a shunt-independent state, which makes this option attractive. Risks associated with craniotomy and fenestration, such as aseptic meningitis, increased neurological deficits, and delayed intracranial hemorrhage, are well described in the literature^{9,10}. After the introduction of the neuro-endoscope, the risk of surgical morbidity had been greatly reduced while still maintaining a good outcome^{5,6}. This minimally invasive approach enabled a shorter hospital stay and less pain, and fenestration using endoscope has become one of the most popular surgical options in treating pediatric arachnoid cysts¹¹.

Shunting, mostly cystoperitoneal shunting, is also a widely adopted procedure. By directly draining the cystic fluids, this approach enables immediate decompression of the arachnoid cyst and may correct impaired CSF dynamics with lower initial risks than fenestration^{10,12,13}. However, shunt-related complications, such as early shunt failure, infection, overdrainage, and lifelong shunt dependence, are the major disadvantages of this procedure^{10,12,14}. Fearing these shortcomings, the endoscopic fenestration now seems to be the first line treatment option in most neurosurgical clinics.

Fig. 3. Case 3. (A) T1 weighted sagittal MRI taken at 3 months of age. A retrocerebellar arachnoid cyst was noted. (B) Follow-up MRI taken 9 months later. It showed the enlarged retrocerebellar cyst compressing the cerebellum. Size of the arachnoid cyst was 5.1 x 3.3 x 4.3 cm. The first surgery, craniotomy and microscopic fenestration was performed. (C) Six-month follow-up MRI after the first surgery. The size of the cyst increased (6.2 x 4.2 x 5.2 cm), and enlargements of lateral and third ventricle were noted. The presence of hydrocephalus was suspected and revision surgery was performed. (D) Follow-up MRI at 4 years old, 2 years and 6 months after the second surgery. The size of the lateral and third ventricles were normalized and the mass effect compressing the cerebellum was relieved.

Despite the concerns over lifelong shunt related complications, we think the efficacy and feasibility of a cystoperitoneal shunt should be re-evaluated in neonatal and infantile patients. As shown in Case 3, the fenestration might fail. We do not think it happened by chance. Zada et al.¹³⁾ reported that infants with intracranial arachnoid cysts presenting with nonspecific macrocephaly or ventriculomegaly showed a significantly higher rate of shunt dependency. Several authors proposed that the development of arachnoid cysts is directly related to abnormal CSF dynamics, and there was a high level of suspicion that arachnoid cysts and hydrocephalus are pathogenetically related^{9,13,20)}. Most arachnoid cysts are likely to enlarge in neonatal and infantile periods⁸⁾. In addition, Mattei et al.¹⁸⁾ reported that benign extracerebral fluid collection in infancy was a significant risk factor for the development of de novo arachnoid cysts and suggested abnormal CSF dynamics by the impairment of CSF fluid absorption in the infantile period as one of the leading causes of arachnoid cyst development and expansion. These findings are now explained by the different CSF dynamics present in the neonatal and infantile periods. It is called as the 'minor CSF pathway' in the recent literature⁷⁾. The main idea is that the major CSF pathway is not yet developed in the fetal, neonatal and infantile periods, and the minor CSF pathway works as the alternative until its full development. This is based upon the observation that the neonates and infants are lack of the arachnoid villi or granulations (Pacchionian body)⁷⁾. These major CSF absorption sites begin to appear in the infantile period, and CSF is mainly absorbed in extra-arachnoid villus sites, such as the ventricular ependyma, leptomeninges, pia-arachnoid capillaries, choroid plexus and perineural spaces until sufficient development of the arachnoid villi or granulations⁷⁾. Therefore, the CSF absorption capacity is smaller than the major CSF pathway. Initially, this notion originated from collected evidence of the high failure rates of endoscopic third ventriculostomy in treating infantile patients with hydrocephalus^{15,16)}. However, later reports suggesting the presence of the minor CSF pathway in other pathologic conditions followed^{8,17-19)}.

Considering the decreased CSF absorption capacity in infancy, a fenestration only has higher chance of failure in the neonatal and infantile patients compared to the older patients. Posterior fossa arachnoid cysts often associated with hydrocephalus^{13,20)}, and we should be kept in mind that a failure of the initial treatment might cause serious consequences. As shown in Case 1, a cystoperitoneal shunt could be performed safely with minimal initial risk. In addition, the direct diversion of CSF by shunt has advantages over the fenestration in circumstances of the minor CSF dynamics predominance. For those reasons we

described above, we suggest that the cystoperitoneal shunt would be more feasible surgical treatment in neonatal or infantile patients with a retrocerebellar arachnoid cyst.

In summary, patients with a retrocerebellar arachnoid cyst found under the age of 1 year should be closely observed, and relatively short-term follow-up is required. It is true that neuronal plasticity and the potential to regenerate is high in very young patients. However, if the arachnoid cysts increase in size and become symptomatic, early surgical intervention should be considered since the time window until irreversible neuronal damage is often narrow. We would like to suggest a cystoperitoneal shunt as an initial surgical treatment for a neonatal or infantile patient with a retrocerebellar arachnoid cyst.

요약

1세 미만에서 발견되는 지주막하 낭종의 경우, 그 크기가 커지는 경우가 많아 면밀한 추적 관찰이 필요하다. 그리고 후두개에 위치한 낭종의 경우 수두증을 일으킬 위험성이 높고, 그로 인해 종종 돌이킬 수 없는 신경학적 손상을 입는 경우가 많아 상당한 주의가 필요하다. 그러나 영아에서 후소뇌에 위치한 지주막하 낭종을 어떻게 치료할 것인지에 대해서는 아직 논란이 많다. 최근 저자들은 3명의 1세 미만 후소뇌에 위치한 지주막하 낭종 환자를 경험하였으며, 한명은 일찍 낭종-복강내 단락술을 시행, 다른 한명은 보호자의 치료거부로 상당한 시일이 경과 후 수두증이 발생한 다음 뇌실-복강내 단락술 및 개창술을, 또 다른 한명은 처음 개창술의 치료 실패로 6개월 뒤 채수술로 치료하였다. 후소뇌에 위치한 지주막하 낭종의 경우, 크기가 커져 증상이 나타날 시, 빠른 수술적 치료가 추천된다. 그동안 축적된 수많은 연구에서 단락술과 관련된 많은 장기적인 합병증들이 알려져 있어 단락술을 기피하는 경향이 있으나, 최근 보고가 이루어지고 있는 1세 미만의 소아에서 뇌척수액 역동학이 성인과 다르다는 사실을 고려하면, 이 연령대의 환자는 낭종-복강내 단락술이 개창술보다 좀 더 적합한 치료 방법으로 생각된다.

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