Primary arachnoid cysts of the middle cranial fossa are encountered clinically with relative frequency. Numerous reports have focused on the pathophysiology of this condition. However, many questions relating to its etiology and natural history are still unresolved. Regarding therapeutic approaches for this lesion, some investigators consider that surgery is needed in all cases, including asymptomatic ones, to prevent bleeding from the cysts, whereas others argue that surgical treatment is not necessary. Thus, there is no widely accepted therapeutic principle for this condition.

We recently encountered a 7-year-old boy in whom a very small arachnoid cyst developed immediately after birth. When examined using magnetic resonance (MR) imaging 7 years later, the cyst had increased in size and was associated with temporal lobe hypoplasia. We report this case because it may be of help in clarifying the pathophysiology of arachnoid cysts of the middle cranial fossa and establishing therapeutic criteria for such cases.

Case Report

This 7-year-old boy was hospitalized for acute lymphocytic leukemia in May 1993. During radiochemotherapy, MR imaging was performed to explore the cause of his dull headache and enlarged head circumference. The images revealed abnormalities, and the boy was referred to our department in December 1993.

Medical History. The boy was delivered by Caesarean section at the gestational age of 40 weeks and 2 days. At birth, he weighed 3680 g, was 51.3 cm long, and had a head circumference of 34 cm. The Apgar score was 8 at 1 minute and 9 at 5 minutes after birth. On the 2nd day after birth, he developed generalized convulsions. Computerized tomography (CT), performed that day, revealed a small low-density area at the tip of the temporal lobe in the middle cranial fossa on both sides (Fig. 1). This area was more evident on the left side. At that time, however, no other abnormalities were noted, and the temporal lobe was formed normally. Thereafter, the boy developed normally, without sustaining any head injury or suffering additional seizure episodes. His school performance was slightly better than average.

Examination. Neurological examination on admission showed no focal deficits. His head circumference was 57 cm. On plain skull films, the right temporal squama was less thick and protruded more laterally than the left counterpart. Elevation of the lesser wing of the sphenoid bone and dilation of the right superior orbital fissure was also disclosed on plain radiography. Head MR imaging revealed an extensive area of low signal intensity on T1-weighted images, and a signal intensity comparable to that of cerebrospinal fluid (CSF) on T2-weighted images. This area extended from the right middle cranial fossa to the frontal and parietal regions. Compression of the right lateral ventricle and a slight midline shift were noted. A small area with signal intensities similar to the area mentioned above was also noted in the left middle cranial fossa. The right temporal lobe was atrophic and hypoplastic, but the inferior horn of the lateral ventricle was pres-
ent, and no defects were seen in the cortex or gyrus (Fig. 2). On CT cisternography, performed 24 hours after the intrathecal administration of Isovist contrast material, poor communication was revealed between the subarachnoid space and the cyst. The contrast material did not fill the cyst clearly. The right middle cranial fossa was found to be markedly dilated. Cerebral angiography in the arterial phase showed the horizontal segment of the right middle cerebral artery to be markedly elevated and to run straight. The temporal branches of this artery were hypoplastic and also showed a posterosuperior shift, but no defects were noted in these branches. In the venous phase, an anteriorly shifted superficial temporal vein was revealed, and this vein was also hypoplastic.

Operation. Based on the neonatal CT data, we believed that the boy had a small arachnoid cyst on both sides immediately after birth, and that the right cyst increased in size as the body grew, inhibiting development of the right temporal lobe and causing enlargement of the head. To avoid a possible adverse influence of the cyst on future brain development, a cyst-peritoneal shunt was created from the right frontal area. During surgery, a hypertrophied cyst wall was noted below the dura mater. The fluid that filled the cyst resembled CSF and the intracystic pressure was high.

Postoperative Course. The boy no longer complained of headache after surgery. He was discharged 2 weeks postoperatively without any neurological deficits. The CT taken immediately before discharge revealed absence of mass effect and a slight reduction in the size of the cyst.

Discussion

Two different theories have been proposed regarding the pathogenesis of arachnoid cyst. According to the view propounded by Robinson and other investigators, the formation of the temporal operculum, which normally begins at a gestational age of 6 months, is disturbed by certain factors, causing primary temporal lobe hypoplasia, so-called “temporal lobe agenesis.” The resultant dilation of the subarachnoid space leads to the onset of “subarachnoid cysts.” The other view (the “intra-arachnoid cysts” theory) was primarily proposed by Starkman, et al., who speculated that very small arachnoid cysts, which are formed due to abnormal development of the arachnoid membrane in the early intrauterine period, later expand and cause secondary temporal lobe hypoplasia.

Since the increased use of CT and MR imaging, a number of cases have been reported in which temporal lobe hypoplasia improved after surgery. It has also been reported that trauma and other factors sometimes trigger spontaneous reduction of such cysts. Because of these facts, the second theory has been considered more likely. However, because the arachnoid membrane begins to develop at a gestational age of 3 months, this view can also be deemed to suggest that an arachnoid cyst, formed before the development of the temporal operculum, causes hypoplasia of the temporal operculum at a gestational age of 6 months. Thus, irrespective of the difference in the initial cause, both hypotheses strongly suggest that arachnoid cysts associated with temporal lobe hypoplasia are congenital disorders that develop during the intrauterine period. Little attention has been paid to the possibility that this kind of cyst also develops during infancy.

The features of the lesion in the present case, as assessed by diagnostic imaging techniques upon admission (at age 7 years), were those of frequently encountered arachnoid cysts of the middle cranial fossa. Considering the medical history of this boy, it is unlikely that his cyst represents a secondary arachnoid cyst caused by trauma or infection. The CT scan taken 2 days after birth to check the cause of convulsions provided valuable data in clari-
fying the natural history of this cyst. The boy showed no hypoplasia of the temporal lobe at birth. Seven years later, however, he had a large arachnoid cyst accompanied by temporal lobe hypoplasia. The protrusion of the temporal squama and dilation of the middle cranial fossa suggest that the large arachnoid cyst was gradually formed over an extended period during the period of infancy when the skull develops. It may therefore be said that the base for the arachnoid cyst of this case was formed during the intrauterine period and that secondary brain hypoplasia, which is primarily responsible for the growth of the cyst, occurred during infancy.

Although arachnoid cysts of the middle cranial fossa are often said to be congenital, very few reports of arachnoid cysts diagnosed at birth have been published. It is therefore highly possible that a relatively large percentage of middle cranial fossa arachnoid cysts follow a course of development similar to that in the present case. Additional cases need to be studied to determine whether all other cases of primary arachnoid cysts of the middle cranial fossa follow the same course. The mechanism for cyst growth should also be examined in more depth. In any event, the arachnoid cyst development in the present case is quite valuable in establishing therapeutic criteria for this disease.

If patients with this cyst have concomitant acute intracranial hypertension, bleeding within the cyst, or subdural hemorrhage, surgical treatment is indicated. No one would disagree with this. However, in cases where intracranial hypertension is absent, no marked focal symptoms are present, or the cyst has been detected incidentally, the need for surgery is controversial. Our experience with the present case would help surgeons determine the indications for surgery in such cases. Although no definitive conclusions can be drawn based on this case alone, considering the possibility that arachnoid cysts are formed in infancy, we hold the following views. Early surgery should be performed for arachnoid cysts diagnosed in infancy to promote subsequent brain development and function, at least in cases where mass effect is present. In cases without mass effect careful follow-up monitoring is mandatory, and if the cyst shows any growth at all surgical intervention should be undertaken. The accumulation of more such cases will further clarify the pathophysiology of this condition.

References


Manuscript received August 1, 1994. Accepted in final form October 17, 1994. Address reprint requests to: Yoshinari Okumura, M.D., Department of Neurosurgery, Nara Medical University, 840, Shijo-cho, Kashihara City, 634, Nara, Japan.