Dandy-Walker Syndrome Forming a Giant Occipital Meningocele
—Case Report—

Tomoki TODO, Masaaki USUI and Fumio ARAKI*

Departments of Neurosurgery and *Pediatric Surgery,
Aizu Central Hospital, Aizuwakamatsu, Fukushima

Abstract

A boy was born with Dandy-Walker syndrome associated with a giant occipital meningocele, cleft lip, and cleft palate. The meningocele was actually a component of the giant posterior fossa cyst which communicated with the fourth ventricle. A cyst-peritoneal shunt achieved a considerable decrease in the size of the meningocele, but decubital ulcers developed due to restricted head movement caused by the occipital lesion. Cranioplasty removed a wide area of the inferior occipital bone, and the boundary between the superior occipital and parietal bones was thinned to allow free bending of the bone flap. The meningocele was removed totally in the third operation, but infection of the wound and pneumonia developed, causing death. The coexistence of Dandy-Walker syndrome and occipital meningocele, together with midline facial anomalies, may suggest a later pathogenesis of Dandy-Walker syndrome than previously believed.

Key words: Dandy-Walker syndrome, meningocele, encephalocele, cranial dysraphism

Introduction

The Dandy-Walker syndrome is a congenital anomaly characterized by hydrocephalus, a posterior fossa cyst, and absence of the cerebellar vermis.2,8,9,12,13,28) This syndrome is well known, but only occurs in 4–8% of neonates with congenital hydrocephalus.20,24) The prognosis of this syndrome has been improved in the past 20 years by the establishment of therapeutic methods using shunt operations and suitable antibiotics, but the mortality is still high, ranging from 26 to 50%.6,9,20,23,28) One major cause of therapeutic problems and the poor prognosis is the frequent association with other central nervous system (CNS) anomalies, in 1/3 to 2/3 of the cases.6,12,13,23,28)

We describe an infant with Dandy-Walker syndrome associated with an unusual giant occipital meningocele which caused great therapeutic difficulty.

Case Report

This boy was delivered by caesarean section in another hospital on December 14, 1986 after 39 weeks and 1 day gestation. At birth, he had a giant occipital meningocele together with cleft lip and cleft palate. His weight at birth was 6850 g and Apgar score 10. The 25-year-old father and the 36-year-old mother were both healthy, with unremarkable family histories. Hydramnion had been diagnosed during the pregnancy, but no other prenatal information was available. He was transferred to our hospital 2 days after birth for further evaluation and treatment.

On admission, his weight was 6720 g and head circumference 69.7 cm. A giant occipital meningocele was observed with prominent superficial veins (Fig. 1). The vertical circumference of the meningocele measured 59.3 cm. Neurological examination revealed no apparent deficits. Physical examination found

Received December 8, 1992; Accepted May 14, 1993

Author's present address: T. Todo, M.D., Department of Neurosurgery, International Medical Center of Japan, Tokyo, Japan.
severe cleft lip and cleft palate, but no anomaly in the extremities. No cardiac complication was suggested from the cardiac sound. The complete blood count and biochemical analyses were within normal limits, and the chromosome analysis revealed a normal 46XY karyotype.

Plain skull x-ray films revealed cranium bifidum with the occipital bone widely divided in the center with the edges protruding laterally and inferiorly (Fig. 2). Precontrast computed tomographic (CT) scans revealed wide cleavage of the occipital bone in the center with the edges projecting laterally, and a giant low-density cyst occupying almost all the posterior fossa, communicating with the fourth ventricle ventrally and bulging out to form the meningocele through the bone defect dorsally. Furthermore, the cerebellar hemispheres were extremely atrophic and separated by a complete absence of the vermis, the tentorial edges were linear and highly positioned, and mild hydrocephalus was observed in the supratentorial ventricles with agenesis of the corpus callosum (Fig. 3). Based on these radiological findings, Dandy-Walker syndrome was diagnosed.

A two-stage operation was planned: 1) to perform a cyst-peritoneal shunt to relieve the hydrocephalus and decrease the size of the giant meningocele, and 2) a cranioplasty and total removal of the meningocele. The first stage performed on December 24 resulted in
a remarkable decrease in the size of the occipital meningocele. However, at the end of January, 1987, bilateral decubital ulcers developed on the head where the edges of the occipital bone protruded, since he could not move his head freely because of the projecting occipital lesion. On February 4, cranioplasty was performed to prevent further formation of decubital ulcers. A wide area of the inferior part of the occipital bone was removed, the boundary between the superior part of the occipital bone and the parietal bone was thinned by drilling away 95% of the thickness to allow the occipital bone flap to bend freely from outside to inside.

Fig. 4  Schematic drawings, illustrating the method of cranioplasty in the second operation. upper: Positions of the decubital ulcer and the skin incisions are shown. The opposite side was the same. lower: Schematic drawing of the skull of the patient. Part II was totally removed by rongeur and drill, and part I was thinned by drilling away 95% of the thickness to allow the occipital bone flap to bend freely from outside to inside.

On February 25, total removal of the meningocele, i.e. removal of the excess occipital scalp and dura mater with dural plasty, and revision of the cyst-peritoneal shunt were performed. Prior to the operation, cerebrospinal fluid (CSF) was aspirated from the meningocele. With him in a prone position, a linear skin incision was made in the center of the meningocele to expose the inside. The dura mater was extremely hypertrophic with abundant vessels running radially from a single point. The cyst was separated by irregular arachnoid-like membranes with a fine network of abundant vascularization existed inside the cyst. right: Inside view of the posterior fossa after excision of the occipital meningocele. The cerebellar vermis is totally defective, and the pons and the medulla oblongata can directly be observed. The cerebellar hemispheres are remarkably atrophic and separated. The cerebellar tentorium is located in a very high position.

Shortly after the third operation, CSF leaked from the wound. Additional sutures were repeatedly applied to stop the leakage, but the wound was eventually infected, causing an abscess in the posterior fossa cyst. Staphylococcus epidermidis grew in the

Neurol Med Chir (Tokyo) 33, December, 1993
bacterial culture of the CSF. On March 3, a fourth operation was performed to drain the cyst and remove the shunt system. After the operation, the cyst was continuously irrigated with saline containing antibiotics, although without much effect. His general condition gradually deteriorated complicated by pneumonia, and he died on March 18, 1987. Autopsy was not permitted.

Discussion

Agenesis of corpus callosum is the most common CNS anomaly associated with Dandy-Walker syndrome.6,13,20,23,28) Other anomalies include microcephaly, cerebral gyral abnormalities, heterotopias, syringomyelia, malformations of the brainstem, aqueductal stenosis, Klippel-Feil deformity, six lumbar vertebrae, subdural hygroma, and cranium bifidum.6,12,13) Only 24 cases of Dandy-Walker syndrome associated with occipital meningocele or encephalocele have been reported.4,14,21,22,26,28) Kojima et al.16) reviewed 11 cases of Dandy-Walker syndrome associated with occipital encephalocele, finding the encephalocele communicated with the posterior fossa cyst in eight cases. Recently, Bindal et al.10) found eight cases of associated occipital meningocele in their series of 50 cases of Dandy-Walker syndrome, suggesting that this anomaly is not so rare. However, such a large occipital meningocele as in our case has never been described. In all previous cases, the associated occipital meningocele was not the main issue of therapy. Bindal et al. even reported that the occipital meningocele disappeared after shunting without surgical repair in two patients. In contrast, the occipital meningocele in our case was formed by the huge Dandy-Walker cyst and was the major threat to the patient's survival.

The present case could also be classified as "tectocerebellar dysraphia with occipital encephalocele," which Friede10) proposed as a new clinicopathological entity distinguished from Dandy-Walker syndrome in the classification of aplasias of the cerebellar vermis. The nine reported cases of this syndrome were characterized by occipital encephalocele, aplasia of the vermis, and deformity of the tectum.7,10,23)

Several theories have been proposed for the pathogenesis of Dandy-Walker syndrome. The original theory was based on atresia of the foramina of Luschka and Magendie,8) supposedly during the 8th week of embryonic life.27) This theory was later rejected on the basis that the cerebellar anlagen are fused in the midline before the foramina become patent5) and because some patients demonstrated patent foramina.25) Benda3) and others2,13) consider that this syndrome is a complex developmental anomaly, possibly of the fourth ventricle with membrane alterations and cleaving of the cerebellum. Brodal and Hauglie-Hanssen26) believed that early bulging of embryonic membranes of the rhombencephalon prior to formation of foramina is the crucial abnormality. Carmel et al.6) postulated that Dandy-Walker syndrome is a primary developmental anomaly that affects the closure of the neural tube at the level of the cerebellum before the 8th week of embryonic life, based on the complete absence of both the vermis and the choroid plexus in two of their 18 cases, structures which initially appear in the 7th to 8th embryonic week. In contrast, Padget19) explained the syndrome as the result of reopening of the neural tube rather than failure to close. The syndrome may also be a manifestation of a spectrum of neural tube defects resulting from overdistention of the tube during development.11) These earlier hypotheses generally postulated the origin of this syndrome to occur quite early in embryonic life.

In the present case, however, the coexistence of the occipital meningocele and the midline facial anomalies suggests that the origin is much later than neural tube closure. A cleft palate is considered to originate during the 8th to 12th embryonic week.13) Oi et al.10) recently showed that exencephaly induced in the chick embryo, which is comparable to human anencephaly, occurs by reopening of the rostral end of the neural tube. Furthermore, histological and immunohistochemical analyses of human neonate autopsy and surgical specimens demonstrated that, in contrast to anencephaly which is associated with interruption of the neuronal developmental process, cranium bifidum cysticum presents with completed cortical formation of the telencephalon with an intact central neural canal, implying that cranium bifidum cysticum originates later than anencephaly.17) Based on this evidence, the associated occipital meningocele in our case suggests that Dandy-Walker syndrome may not be an early embryonal maldevelopment, but an event occurring much later than the closure of the neural tube. The exact pathogenesis of Dandy-Walker syndrome awaits further studies.

Four surgical methods have been used to treat Dandy-Walker syndrome: 1) excision of the posterior fossa cyst membrane, 2) shunt of the lateral ventricle, 3) shunt of the cyst, and 4) simultaneous Y-shunt of the lateral ventricle and the cyst.1,3,6,9,12,18,20,23,28) Excision of the cyst membrane

Neurol Med Chir (Tokyo) 33, December, 1993
has been ineffective in most cases, requiring reoperation for shunting.\textsuperscript{9,23} No type of shunt is clearly superior,\textsuperscript{12,28} but Sawaya and McLaurin\textsuperscript{23} claim shunt of the posterior fossa cyst to be the first choice as stenosis is not usually present between the supratentorial and infratentorial compartments, and the absence of cyst wall collapse around the shunt tubing minimizes the risks of shunt malfunction. Several reported cases required replacement or additional shunting of the cyst after the initial ventricular shunt proved ineffective.\textsuperscript{12,23,28} Carmel et al.\textsuperscript{6} pointed out that the cyst shunt has the theoretical advantage of maintaining the CSF flow through the aqueduct in the physiological direction, although they found an especially high complication rate (5/6). Simultaneous shunting of the lateral ventricle and the cyst was originally introduced to prevent upward herniation following decompression of only the lateral ventricles.\textsuperscript{20} A lateral ventricular shunt frequently results in acquired aqueductal stenosis with isolated fourth ventricle because of a disturbance in the physiological direction of CSF flow.\textsuperscript{1,18} Several workers prefer combined shunting of the lateral ventricle and the cyst to the other methods.\textsuperscript{3,18} Bindal et al.\textsuperscript{3} suggested that patients needing combined shunting may manifest different CSF composition or flow mechanics in the posterior fossa, based on a higher incidence of malfunction in the cyst component than the ventricular component.

When cranium bifidum is present, the hydrocephalus may be concealed by the pressure-buffering effect of the encephalocele.\textsuperscript{15} Based on this hypothesis, Oi\textsuperscript{15} recommended a two-step "ventricular volume reduction technique" for repair of cranium bifidum: 1) reduction in the buffering capacity of the cranial bifidum, resulting in manifestation of hydrocephalus; and 2) removal of CSF from the ventricles allowing the transposition of the protruded encephalocele into the intracranial space. No special method has been proposed for treating occipital meningocele associated with Dandy-Walker syndrome, but no other case has occurred with such a giant occipital meningocele.

Complications after a shunt operation for Dandy-Walker syndrome occur frequently; shunt infection is the most common (18-32\%), followed by CSF leakage, peritonitis, and subdural effusion.\textsuperscript{5,23} Acute exacerbation by abscess formation in the posterior fossa cyst has been reported in one case after an asymptomatic period of 13 years.\textsuperscript{20} Once infection occurs within the posterior fossa cyst as in our case, an extremely poor outcome can be expected due to consequent occlusion of the aqueduct, isolation of the fourth ventricle, increased pressure in the posterior fossa, shunt malfunction, and increased difficulty in controlling the hydrocephalus, as well as the difficulty in treating the abscess.

Dandy-Walker syndrome is still considered to have a poor prognosis with a mortality of 26-50\%.\textsuperscript{9,20,23,28} Tal et al.\textsuperscript{28} reported a mortality in their series before 1965 of 9/13 but after 1966 of 2/8, showing that, although the mortality remains quite high, it has been improved by the use of shunting and the prevention of complications by more potent antibiotics. Most studies also reported that less than 50\% of the cases have normal intelligence,\textsuperscript{5,9} or even less than 30\%.\textsuperscript{23,28} Golden et al.\textsuperscript{12} found that residual focal neurological deficits may indicate poor intellectual development, while CT evidence for reestablishment of posterior fossa architecture after shunting indicates the opposite. Moreover, the prognosis depends greatly on the presence of associated anomalies. Other CNS anomalies definitely result in a poorer outcome, accounting for 50-80\% of deaths.\textsuperscript{6,23,28} The presence of occipital meningocele may not be as important as other associated CNS anomalies in determining poor intellectual development,\textsuperscript{4} but whether this applies to giant meningocele is unknown. Improvement in the prognosis of Dandy-Walker syndrome requires appropriate treatment for associated anomalies, especially those of the CNS, as well as the Dandy-Walker syndrome.

References

2) Benda CE: The Dandy-Walker syndrome or the so-called atresia of the foramen Magendie. \textit{J Neuropath Exp Neurol} 13: 14-29, 1954

Address reprint requests to: T. Todo, M.D., Department of Neurosurgery, International Medical Center of Japan, 1-21-1 Toyama, Shinjuku-ku, Tokyo 162, Japan.