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Shunt-independent surgical treatment of middle cranial fossa arachnoid cysts in children

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Abstract The best operative intervention for children with arachnoid cysts remains the subject of controversy. Recent reports stress that craniotomy for cyst fenestration is associated with a low incidence of morbidity and mortality and may leave the child shunt-independent. Among a total of 66 intracranial arachnoid cysts operated on in the authors' department from 1985 to 1997, 44 cases (67%) were located in the middle cranial fossa. A higher incidence in the first decade of life (53 cases) and a marked male predominance (45 cases) were recognized. Headache, cranial deformities, symptoms of raised intracranial pressure, and seizures constituted the most frequent features of the clinical presentation. To determine which treatment provides the greatest benefit with the lowest incidence of complications, the records of the 44 patients with arachnoid cysts in the middle cranial fossa were reviewed. The mean age of these patients was 4.6 years (range 0–16 years). Different types of initial surgical procedures were performed. In 33 patients with middle cranial fossa arachnoid cysts (MCFAC) the initial surgery took the form of craniotomy with excision of the cyst walls and fenestration into the basal cisterns. Shunting procedures were performed in 9 patients: cysto-peritoneal shunts (CPS) were placed in 4 patients and ventriculo-peritoneal shunts (VPS), in 3 patients, and cyst excision was performed in addition to CPS in 2 patients. Excision of the cyst membrane alone without fenestration was performed in 2 patients. The initial treatment was successful in terms of reduced symptoms and decreased cyst size, with no additional treatment needed for the cyst, in 79% (26/33) of patients who had undergone excision of the cyst walls and fenestration into the basal cisterns, compared with 66% (6/9) of patients who had undergone shunting procedures. Cyst membrane excision was not successful in any of the patients who underwent this procedure alone. No significant difference in morbidity was noted between these different treatment options. On follow-up CT scan and MRI, cysts of types I and II (Galassi classification) exhibited a steady tendency to reduction or obliteration. These results confirm that radical excision of the outer and inner membranes of the cyst wall with fenestration into the basal cistern is a safe and effective shunt-independent procedure for MCFAC, especially for those of types I and II.

Key words Arachnoid cyst · Middle cranial fossa · Fenestration · Shunt-independent

Introduction

Arachnoid cysts are intra-arachnoid collections of cerebrospinal fluid (CSF). By compression of adjacent neural tissue or by obstruction of CSF flow, they produce symptoms such as craniomegaly, developmental delay, seizures, and headaches [1, 12, 25]. The majority of these lesions are discovered during childhood, with 60–80% occurring in patients under the age of 16 in the reported series [2, 8]. There is now little controversy about the origin, histopathology, pathophysiology, and radiological appearance of these lesions [1, 12]. In patients whose cysts are symptomatic surgical intervention is called for, but the best method of treatment is still disputed. The principal goal of surgical treatment is to eliminate the pressure effect on adjacent structures and the obstructive effect on the normal CSF pathway. Different therapeutic options, including needle aspiration, cysto-peritoneal shunting, ventriculo-cystostomy, and craniotomy for partial or complete cystectomy for marsupialization into the subarachnoid space, basal cisterns, or ventricle have been developed [22]. In contrast to other series, this study claims that good results can be obtained with removal of the cyst wall and communication of the cyst with the normal subarachnoid space [2, 5, 16]. Having gained additional insight into this problem through our work, we carried out a prospective evaluation of different methods of shunt-independent surgical treatment for symptomatic MCFAC, involving fenestration with radical opening into the basal cisterns. Special attention was paid to the initial operative approach and outcome to allow comparison of the different treatment options.

Patients and methods

This series included 44 patients with MCFAC, who made up a subgroup of the 66 patients with intracranial arachnoid cysts treated at Kangnam St. Mary's Hospital during the period 1985–1997(Table 1). The prospective study of direct surgical treatment involved all our cases of symptomatic arachnoid cyst. MRI, CT with or without metrizamide, and cerebral angiography were done before and after surgery. Surgical treatments consisted in fenestration, shunting procedures, and excision only. For the operative fenestration procedures, extensive excisions of both the outer and inner membranes with radical opening to the parasellar and basal cisterns were performed. Shunting procedures included placement of cysto-peritoneal shunts (CPS) and of ventriculo-peritoneal shunts (VPS). The diagnosis was confirmed by histological examination. The mean follow-up period was 4.2 years (8 months to 10 years).

Table 1 Locations of 66 intracranial arachnoid cysts

Location	No. of patients (%)
Middle fossa	44 (67)
Posterior fossa	12 (18)
Suprasellar	4 (6)
Interhemispheric	3 (5)
Quadrigeminal	1 (1)
Others	2 (3)
Total	66 (100)

Results

The mean age was 4.6 years and the male-to-female ratio was 29:15 (Table 2). Table 3 summarizes the clinical features of the 44 patients with MCFAC. Signs and symptoms of increased intracranial pressure and compression of the adjacent area caused by the arachnoid cysts were present in one half of these patients, but were mostly mild. In 14% of patients delayed development was observed. Hydrocephalus was noted in 16% of the patients. From MRI and X-ray CT findings, three morphological types [9] were distinguishable. Type I, the mildest form

Table 2 Age distribution in 66 patients with arachnoid cysts (mean age 4.6 years, M-to-F ratio = 29:15)

Age (years)	No. of cases (%)
0–5	19 (43)
6–10	16 (36)
11–16	9 (21)
Total	66 (100)

Table 3 Clinical features and presenting symptoms and signs in 44 patients with middle fossa arachnoid cysts (mean age 4.6 years, M-to-F ratio = 29:15)

Symptom/sign	No. of patients (%)
Headache	38 (86)
Focal neurological deficits	19 (42)
Cranial deformity	14 (32)
Seizure	15 (40)
Delayed development	6 (14)
Hydrocephalus	7 (16)
Mental retardation	1 (2)
Decreased visual acuity	4 (9)
Psychosis	3 (7)

Table 4 Surgical management of 44 children with middle cranial fossa arachnoid cyst (*CPS* cysto-peritoneal shunt, *VPS* ventriculo-peritoneal shunt)

Surgical procedures	No. of patients (%)
Fenestration with communication into basal cisterns	33 (74)
Shunting procedures	9 (21)
CPS	4 (9)
Excision + CPS	2 (5)
VPS	3 (7)
Excision only	2 (5)
Total	44 (100)

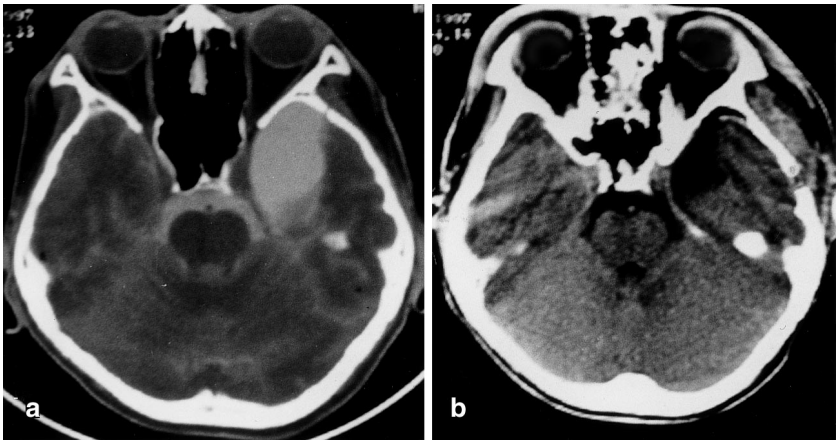

Fig. 1a, b Type II arachnoid cyst. **a** Preoperative metrizamide CT showing delayed penetration (6 h) of the dye into the middle cranial fossa arachnoid cyst. **b** Postoperative CT 2 weeks after the fenestration procedure, showing marked reduction in size of the cyst

of MCFAC, accounting for 18% (8/44), was limited to the anterior aspect of the temporal fossa. Type II, medium-sized, triangular, accounting for 55% (24/44), occupied the anterior and middle parts of the temporal fossa and the temporal lobe was foreshortened. Type III, the most severe form, is huge and oval and accounted for 27% (12/44) of those in this series; these cysts occupied the temporal fossa almost entirely, with a striking mass effect, and the temporal lobe was severely atrophic. Metrizamide CT cisternography was performed in 9 patients (2 type I, 4 type II, 3 type III). These three types displayed different patterns of metrizamide circulation around and inside the lesions. This suggested different relationships between the three kinds of cysts and the adjacent subarachnoid spaces, with distinct characteristics of CSF kinetics. Type I cysts (2 cases) were massively filled with metrizamide 1 h after intrathecal injection of metrizamide and there was evidence of free communications between the cystic cavities and the basal cisterns. Type II cysts (4 cases) became visible with some delay and there was absent or negligible metrizamide penetration at the 1-h CT cisternography. The filling of the type III cysts (3 cases) was quite different from that of types I and II. Evidence of the metrizamide stasis in the surrounding subarachnoid space was observed, with no clear filling of the cysts at any of the early and late control cisternograms. We therefore consider it can be assumed that type III cysts have no communication with the rest of the subarachnoid space. Three types of surgical procedures were performed to manage the children with MCFAC (Table 4).

Cysts initially treated by fenestration

Thirty-three cysts were treated initially by craniotomy with removal of large portions of the cyst wall and wide fenestration of the cyst into the basal cisterns. Of these 33 patients, 26 (79%) achieved successful results in terms of reduced symptoms and size of the cysts and required no additional operation (Table 5, Fig. 1). Seven patients (21%) were in failure after initial fenestration; 3 patients needed CPS beccause of re-expansion of the cyst after craniotomy, 3 patients required VPS for persistent hydrocephalus after craniotomy, and 1 required a ventriculo-cysto-peritoneal shunt.

Table 5 Outcomes of 33 children with middle cranial fossa arachnoid cyst treated initially by fenestration

Treatment needed	No. of patients (%)
No treatment	26 (79)
CPS	3 (9)
Ventriculo-cysto-peritoneal shunt	1 (2)
VPS	3 (9)
Total	33 (100)

Cysts initially treated by shunting procedures

Nine patients underwent shunting procedures as their initial treatment. Six patients (66%) showed successful results, but a higher incidence of failure, 34% (3/9), than after the fenestration procedure was noted. There were 6 patients who had CPS and 3 who had VPS as their initial operation (Table 4). Among the 6 patients with CPS, cyst excisions were combined in 2 patients. Subsequently, 2 patients were in failure and needed additional treatment for their cysts; 1 patient underwent a subsequent fenestration and 1 refused further treatment. The 3 patients initially treated by VPS showed some clinical and radiographic improvement postoperatively, but 1 of them required a subsequent fenestration due to shunt malfunction.

Table 6 Postoperative follow-up[a] evaluation of middle cranial fossa arachnoid cyst by CT scan and MRI

Type[b]	No. of patients (%)	Complete obliteration	Marked reduction	Moderate reduction	Unchanged
I	8 (18)	4	3	1	
II	24 (55)	3	15	4	2
III	12 (27)		6	5	1
Total	44 (100)	7 (16)	24 (55)	10 (23)	3 (6)

[a] Mean follow-up: 4.2 years
[b] Galassi classification [9]

Cysts initially treated by excision only

Two patients were initially treated by cyst excision only, and they both required subsequent fenestration. One patient initially treated by excision with CPS obtained clinical and radiographic improvement postoperatively.

Operative results according to type of arachnoid cyst

A significance difference is detectable in the postoperative outcome among the patients with the three types of arachnoid cyst. Radiologically, type I and II cysts exhibited a major and steady tendency to reduction in size following the operation, with infrequent complete obliteration (Table 6). Clinical and radiographic results at follow-up seemed satisfactory in all three types after surgical procedures. Prognosis was somewhat better in patients with types I and II lesions in whom fenestration was performed. In patients with type III lesions, cerebral re-expansion occurred in 11 patients examined postoperatively, but it proved less evident than in the patients with type I and II cysts and never led to obliteration and disappearance of the cyst cavity.

Complications and follow-up

There was no mortality in the entire subgroup, regardless of the type of surgery. Five of the 33 patients with fenestrated cysts suffered complications after procedure. Two patients developed subdural hematoma after fenestration and required craniotomy for evacuation of the hematoma. The other complications were hemiparesis (1 case), IIIrd nerve palsy (1 case) and infection (1 case), and they all resolved completely. Of the 6 patients with CPS, 2 required revisions. Of the 3 patients with VPS, 2 patients required shunt revisions and there was 1 shunt infection. For 44 patients with MCFAC, the mean follow-up has been 4.2 years with a range of 0–16 years. Serial CT scan and MRI were used to determine the size of the cysts, and 41 of the 44 cysts (94%) were decreased in size on follow-up scans. Seven patients showed complete obliteration of the cysts (Table 6). Among 7 patients with hydrocephalus, 6 have normal-sized or smaller-than-normal ventricles. Nine of 15 patients with seizures became seizure free with no need for anticonvulsant medication, and 6 continued to require medication to control the seizures. In 3 of 4 patients who presented with decreased visual acuity improved vision was noted after the surgical procedures.

Discussion

Intracranial arachnoid cysts are congenital lesions that are thought most likely to arise from anomalous splitting and duplication during the development of the endomeninges [25]. The congenital nature of these lesions is suggested by their prevalence in the pediatric population and by the absence of inflammatory or traumatic antecedents [2, 8, 16, 21]. Two features seem to be well documented: arachnoid cysts of the middle fossa have an intraarachnoid location [3, 8, 21, 25] and they compete for space with the brain and have potential expanding properties. Concerning the pathogenesis of such possible expansion due to the increase in intracystic fluid, three hypotheses have been put forward and variously related to all intraarachnoid collections [11, 15]: (1) secretion of fluid by the cyst walls, (2) fluid infiltration through the cyst wall due to an osmotic gradient, and (3) trapping of fluid within the cyst by a ball-valve mechanism, presumably located where a small communication exists between the cyst and the rest of the subarachnoid spaces. The key for finding the pathophysiological explanation for these malformations is therefore based on the relationship between the cystic cavity and the remaining CSF spaces, in order to find a possible communication. To this aim various authors have carried out surgical or postmortem inspection [28], radionuclide cisternography [13] and finally, metrizamide CT cisternography [6, 8, 15]. Our experience with metrizamide CT cisternography limited to arachnoid cysts of the middle cranial fossa would seem to show that the presence of a communication is not incidental and its occlusion leads to re-expansion of the fluid cavity. The cysts progressively expand and compress the adjacent brain. In our series, the sites of origin of the cysts were the middle fossa in 67%, the posterior fossa in 18%, the midline including interhemispheric in 12%, and other locations in 3%. Arachnoid cysts give rise to symptoms by compressing nearby neural structures, increasing intracranial pressure by their own mass effect and by obstruction of CSF flow. In this series, the most common signs and symptoms, headache

and seizures, were closely related to increased intracranial pressure and compression effect to adjacent neural structures, respectively. Other frequent clinical features were abnormally enlarging head circumference, developmental delay, and decreased visual acuity. Hydrocephalus of either communicating types or obstructive types were found. Again, these symptoms and signs are similar to those reported in other series [7, 23]. We think that, in children, all arachnoid cysts documented as causing significant displacement of normal brain structures warrant surgical treatment. Certainly, there is consensus that symptomatic cysts causing seizures, hydrocephalus, raised intracranial pressure, or focal deficit should be treated [4, 16]. In addition, asymptomatic cysts can exert pressure on the brain with inhibition of development and function [12]. Children with untreated arachnoid cysts are also at risk of intracystic or subdural hemorrhage occurring spontaneously or after minor head injury [1]. We are concerned about other authors' proposal that all sylvian cysts exerting detectable mass effects call for surgical treatment even if clinically silent [8, 11, 12, 20]. The choice of the optimal surgical method still remains a controversial topic. Recent papers [3, 18, 27] have advocated the superiority of primary cysto-peritoneal shunting alone, or combined with VPS when hydrocephalus is associated. Although these series seem mostly heterogeneous, including intracranial arachnoid cysts at different locations, the CSF diversions proved safe and frequently effective. Nevertheless some failures caused by shunt malfunction or by infection and defective cyst collapse have also been recorded [11, 27]. Notably, in the case of sylvian cysts, unlike those in midline and infratentorial locations [10], hydrocephalus is usually absent, since changes in the CSF kinetics are mainly limited to the cyst cavity and its periphery [9]. Therefore, we assume that extensive resection of the outer and inner membranes and establishment of a wide communication with the subarachnoid pathways can normalize the fluid circulation adequately [8, 9, 12] and avoid the need for CSF shunting. The advantage of fenestration is the possibility of leaving the patient shunt-independent [2, 7, 16]. In our series, 3 revisions in 9 shunted patients, 2 with CPS and 1 with VPS, were required due to shunt-related complications. Studies of other series, even those supporting shunting as the treatment of choice, have also documented a high rate of shunt-related complications in patients with CPS, i. e., 3 revisions in 12 shunted patients in the series of Harsh et al. [14], and 3 revisions and 1 death due to infection in 30 patients in the series of Locatelli et al. [19]. On the other hand, fenestration is not always successful, and some patients with cysts and concomitant hydrocephalus will need a VPS despite successful treatment of their cyst by fenestration [27]. On this point, patients should be divided into two groups depending upon the presence or absence of hydrocephalus. Patients without hydrocephalus should undergo craniotomy with resection of all easily removed cyst wall and fenestration of the cyst into all accessible subarachnoid spaces. In Raffel and McComb's series [24], 26 (76%) of 34 patients with arachnoid cysts without hydrocephalus at presentation underwent successful cyst removal and fenestration and remained without shunts. Lange and Oeckler [17] reported a series of 35 patients in which 13 (68%) of 19 patients treated by craniotomy showed significant clinical improvement. Galassi et al. reported a series of 25 temporal fossa cysts without associated hydrocephalus, who were all treated successfully by fenestration [8]. Similarly, 16 of 18 patients with temporal arachnoid cysts in the series of Sato et al. were improved by fenestration alone [26]. In our series, 26 of 33 patients (79%) without hydrocephalus underwent successful cyst removal and fenestration and remained shunt free. Thus, fenestration seems to leave a child shunt-independent and to eliminate the possibility of future shunt revisions in most cases. We were unable to identify the factors for re-expansion of the arachnoid cyst after fenestration procedures. However, this would allow us to predict that the cyst wall may reconstitute, the extent of the fenestration may have been inadequate, or adequate flow of CSF from the cyst and its surrounding cisterns may not occur [14]. We think that the fenestration approach to arachnoid cysts suggested in this paper is rational on the basis of the data presented. Fenestration procedures for the middle fossa arachnoid cysts without hydrocephalus had 79% of the patients shunt-independent in our series.

In conclusion, we assume that craniotomy with wide excision of the membranes and fenestration of the cyst into the basal cisterns is the most rational approach to a shunt-independent procedure for the treatment of middle cranial fossa arachnoid cyst. Because surgical therapy is more effective in children, it is recommended that arachnoid cysts that show poor communication between the cyst and the subarachnoid spaces should be treated as early as possible with regard to reversible brain growth in childhood. We believe that further experience with different therapeutic alternatives and long-term studies will give us the optimal guidelines for the management of this complex anomaly.

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