Microsurgical Management of Craniopharyngiomas

— Outcomes in 56 Patients —

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Summary: Fifty-six patients with craniopharyngiomas were operated by microsurgical techniques during a 20 - year period. Of the 56 patients, 28 were males and 28 were females, with a mean age of 33 years (range 1 to 78 years). Twenty patients (35.7%) were less than 15 years of age. Remission was defined as clinical improvement with stable ophthalmological and neurological status, CT and/or MRI evidence of a decrease in tumor size or tumor disappearance when a patient was discharged from hospital. Ineffectiveness was defined as lack of change in the preoperative clinical status. The mean follow-up period in this study was 6.9 years, with 11 patients monitored. The most common presentation was visual dysfunction (69.6%). 33.9% of the patients presented with headache. The most frequent preoperative finding was a visual field defect, with 55.4% of the patients so affected; 39.5% of patients had preoperative hypothyroidism and 40% had hypoadrenalism. Diabetes insipidus was found preoperatively in 7.1%. Three female patients had amenorrhea. Hydrocephalus was uncommon, being present in only 10.7%. Unruptured aneurysms were found incidentally in 2 cases. A pterional approach was used in 29 patients (52%), a transcallosal approach in 13 (23%), a transcortical approach and a transsphenoidal approaches in 3 (5%), and a lamina terminalis approach in 1 patient. Multiple procedures were required in 8 patients in order to provide significant relief of compressive symptomatology. Overall, 12.5% of the tumors were completely resected. 92.9% of our patients were in remission, 2 had ineffectiveness result, and 2 died of postoperative complications. Except for the completely resected cases, all the other patients underwent radiotherapy postoperatively. The results of this series show that microsurgical management of craniopharyngiomas yields good operative results.

Key words craniopharyngioma, microsurgical technique, surgical approach, endocrinopathy outcome

INTRODUCTION

Craniopharyngiomas are the most common intracranial congenital tumors. They are histologically benign and relatively rare tumors, constituting between 2.5 and 4% of all intracranial tumors [1,2]. Total microsurgical resection of the tumor can cure the patient of this disease. However, the tumor usually has intimate adherence to the infundibular stalk and hypothalamus. Radical excision of the tumor is full of difficulties and great potential hazards. In order to summarize the results of microsurgical management

on patients with craniopharyngioma, we carried out a retrospective analysis on these patients who were evaluated and operated for this tumor at Kurume University Hospital in Kurume City over the past 20 years.

MATERIALS AND METHODS

From 1978 to 1997, 56 patients were operated for craniopharyngioma at Kurume University Hospital, Kurume City. Excluded from this study were patients who just received medication therapy, or patients in

whom the histological diagnosis was uncertain. An attempt was made in all cases to resect the tumor as completely as possible and with the least impairment of important surrounding structures. All patients with

TABLE 1.
Preoperative symptoms in 56 patients with craniopharyngioma

Symptom	Cases	
	No.	Percent
Visual symptoms	39	69.6
Headaches	19	33.9
Vomiting	6	10.7
Growth failure	3	5.4
General fatigue	2	3.6

TABLE 2.
Preoperative findings in 56 patients with craniopharyngioma

Findings	Cases	
	No.	Percent
Visual function		
Normal	10	17.9
Visual acuity decreased in one eye	6	10.7
Visual acuity decreased in both eyes	25	44.6
Visual field defect	31	55.4
Blindness of one eye	8	14.3
Retroactive amnesia	7	12.5
Anosmia	2	3.6
Acalculation	2	3.6
Dementia	2	3.6
Cranial nerve palsies	2	3.6
Somnolence	3	5.4
Gait disturbance	2	3.6

TABLE 3.
Pre and postoperative changes of endocrinopathies among 56 patients with craniopharyngioma

Deficit	Preoperative		Postoperative	
	No.*	Percent	No.*	Percent
Diabetes insipidus				
Permanent	4/56	7.1	33/52	63.5**
Temporary	0	0	4/52	7.7
Hypothyroidism	15/38	39.5	23/38	60.5***
Hypoadrenalism	10/25	40	19/32	59.4***
Amenorrhea	3/10	30	0	0
Panhypopituitarism	0	0	8/56	14.3**
Hypothalamic dysfunction	0	0	1/56	1.8

*: No. with that abnormality/no. of possible cases
**: Compared with preoperative data, p<0.01
***: Compared with preoperative data, p<0.05

TABLE 4.
Surgical approaches and results in 56 cases

Surgical approach	No. of cases		Total removal		Subtotal removal		Partial removal		Reservoir setting (Ommaya)	Reservoir setting (Rickham)
	Children	Adults	Children	Adults	Children	Adults	Children	Adults		
Pterional-transsylvian-parachiasmatic	12	17	4	1	6	10	2	6	1	
Pterional and subfrontal	1	2			1	2				
Transcallosal	4	9			3	5	1	4		
Transcallosal and pterional		1				1				
Translamina terminalis	1				1					
Transcortical		3		1		1		1		
Transsphenoid	1	2		1	1	1				
Transventricular and /or frontal										
Basal aspiration	1								2	1
No. of cases	20	36	4	3	12	20	3	11	3	1

subtotal or partial removal of the tumor subsequently received radiation therapy. In those patients in whom resection was thought to be radical, no irradiation was administered.

A remission was defined as clinical improvement with a stable ophthalmological and neurological status, CT or MRI evidence of a decrease in tumor size or tumor disappearance when a patient was discharged from the hospital. Ineffectiveness was defined as lack of change in the preoperative clinical status.

In this group of 56 patients, 28 were male and 28 were female. The mean age of the patients was 33 years (range 1 to 78 years). Twenty patients (35.7%) were less than 15 years of age. All patients were diagnosed as craniopharyngioma pathologically. Statistical analytic method: chi-square test.

TABLE 5.
Outcomes in 56 patients with craniopharyngioma

Results	Cases	
	No.	Percent
Remission	52	92.9
Ineffectiveness	2	3.6
Death from postoperative complications	2	3.6

RESULTS

Preoperative symptoms are summarized in (Table 1). The most frequent presentation was visual dysfunction (69.6%). 33.9% of the patients presented with headache. The most common preoperative finding was a visual field defect, with 55.4% of our patients so affected (Table 2). Retroactive amnesia was found in 12.5% of the patients preoperatively. Hydrocephalus was present in 10.7% of our patients. Unruptured aneurysms were incidentally found in 2 cases. Many patients presented preoperatively with endocrine dysfunction: 40% of the patients were hypoadrenal, and 39.5% were hypothyroid. Diabetes insipidus was present in 7.1%. Amenorrhea was found in 3 female patients (Table 3).

A pterional approach was the most commonly utilized (approximately 52% of the cases). Twenty-three percent of the patients were operated via a transcallosal approach. A transcortical approach and a transsphenoidal approach were each used in 5%, and a lamina terminalis approach in only 1 patient. Multiple procedures were required in 8 cases to produce significant relief of compressive symptomatology. Radical resection was thought to have been performed in only 7 patients. Subtotal removal in 32, and partial removal in 14 (Table 4).

The results of microsurgical management (Table 5) show that 92.9% of our patients were in

TABLE 6.
Individual follow-up and outcome data (n=11)

Year of operation	Case No.	Final follow-up time	Status at the final follow-up	Sequelae	Therapy
1978	1	1997	alive	panhypopituitarism	hormone replacement
1978	2	1997	alive	panhypopituitarism	hormone replacement
1979	3	1981	death due to postoperative complications		
1981	4	1983	alive	diabetes insipidus anemia, hypopituitarism	hormone replacement
1982	5	1984	death due to post-transfusion hepatitis, hepatocirrhosis and esophagus venectasia		
1982	6	1988	alive	panhypopituitarism	hormone replacement
1982	7	1986	alive	panhypopituitarism	hormone replacement
1985	8	1986	alive	panhypopituitarism	hormone replacement
1986	9	1991	death due to pneumonia and endotoxic shock		
1987	10	1988	death due to meningitis related with CSF rhinorrhea		
1988	11	1992	alive	panhypopituitarism	hormone replacement
Total	11	Mean follow-up time 6.9 years (range 2-20 years)			

remission and 2 had an ineffectiveness result. Two patients died of postoperative complications. Diabetes insipidus (DI) was one of the most principal sequelae (Table 3). Nearly 71% of the 52 patients without DI before surgery developed DI postoperatively. Eight percent of the newly developed DI patients recovered without hormone replacement therapy within a week. Fourteen percent of the 56 patients presented with panhypopituitarism after surgery. Postoperative hypothyroidism was present in 60.5%, and hypoadrenalism in 59.4% of the cases. Compared with preoperative endocrinopathies, the postoperative development of DI and panhypopituitarism was significantly higher (both $p < 0.01$). However, the higher morbidities of postoperative hypoadrenalism and hypothyroidism had no significance (both $p > 0.05$).

In this patient group, follow-up was carried out in 11 cases, with mean period of 6.9 years. Four discharged patients died within 2 to 6 years postoperatively (Table 6). By the end of the follow-up period, all 7 patients who still survived presented with panhypopituitarism requiring hormone replacement therapy.

DISCUSSION

At present radical microsurgical resection of the tumor is the best choice to cure the patients of craniopharyngiomas. However, because of the location and expansion of the tumor, its total removal is sometimes difficult and with high risk. The sellar region is adjacent to major blood vessels supplying the brain, to the optic and oculomotor nerves and to major endocrine structures. The above structures are frequently affected by the tumor, making the surgery a serious challenge [3]. But with modern microneurosurgical techniques and with optimal selection of the surgical approach the majority of the tumors can be excised successfully. Depending on site and location, multistaged operation with or without postoperative radiation therapy is also one of the treatment options.

Yasargil [4,5] observed postoperative diabetes insipidus in 128 of 159 microsurgically treated patients (80.5%). Among the 159 cases, 112 patients (70.4%) needed permanent hormone-replacement therapy. Surgery-induced mortality is 3/162 (1.8%). It would seem that it is impossible not to affect endocrine function in a significant number of patients postoperatively. With advances in modern endocrine replacement therapy, this is not a problem,

as patients are able to function quite well with hormonal supplementation [6].

Although great changes have taken place in the field of neuroradiological science, this lesion is still commonly diagnosed in the late phase of the disease when a big tumor occurs at the base of the skull. In this series, before operation 69.6% of our patients had deficits of visual acuity, and 55.4% presented with visual field deficit which suggested that the lesion was big enough to affect optic nerves and/or chiasm. About 40% of our patients presented with endocrine dysfunction.

The most common postoperative complication was DI (71.2%). Four patients had temporary DI which recovered spontaneously within a week. The others had permanent DI which required substitution therapy. Fourteen percent of the 56 patients without panhypopituitarism before surgery developed panhypopituitarism postoperatively. 60.5 and 59.4% of the 56 patients presented with hypothyroidism and hypoadrenalism, respectively. Compared with preoperative endocrinopathies, the postoperative morbidities of DI and hypopituitarism were significantly higher (both $p < 0.01$). Nevertheless, there wasn't significance (both $p > 0.05$) between the morbidities of pre and postoperative hypoadrenalism & hypothyroidism.

We carried out follow-up in 11 cases. Four of the patients died postoperatively within 2-6 years. By the end of the follow-up period, all 7 patients survived with panhypopituitarism which required hormone replacement therapy.

With modern microsurgical technique, 92.9% of our patients were in remission. Radiotherapy has been proved to play an important role in the treatment of all incompletely resected tumors [7]. In this series, postoperative local radiation therapy was administered to all patients except those with total removal of the tumor. However, more attention should be paid to the occurrence of endocrinopathies.

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