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Proposal of a New Grading System Based on Surgical Results of 100 Craniopharyngiomas

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Abstract

Objective: Craniopharyngiomas pose a baffling problem to neurosurgeons because of the difficulty in total removal and tendency to recur even operated by experienced hands. We propose a new classification or grading system of craniopharyngiomas based on MRI findings including not only sagittal diameter but also coronal diameter of the tumor, the usefulness of which was evaluated in 100 consecutive patients mainly operated by one neurosurgeon (T.H.).

Methods: Between 1981 and 2012, 100 patients comprising 55 males and 45 females aged from 1 year to 75 years (mean 33.1 ± 22.7) underwent surgeries for craniopharyngiomas, including endoscopic removal in two patients. Thirty-six pediatric (younger than 15 years) patients comprising 23 males and 13 females with a mean age of 8.1 ± 4.3 years were included. Surgeries were conducted by a transnasal transsphenoidal (TSR), pterional (PTR) or anterior interhemispheric (AIH) approach, orbyendoscopic removal (END).Based on MRI findings with or without gadolinium enhancement, tumor size was classified by the maximum sagittal diameter into <2 cm (score 1), 2-4 cm (score 2), and >4 cm (score 3). Tumor size was also classified by the maximum coronal diameter perpendicular to midline into<2 cm (score 1), 2-4 cm (score 2), and >4 cm (score 3).A score of 1 was added when the lower limit of the tumor was below the clinoidal line, when the tumor extended to the mammillarybody, or when the tumor reached the foramen of Monro. From MRI findings, tumor composition was classified as cystic only (score 0), multi-cystic (score 1), mixture of cystic and solid (score 2), and solid only (score 3). In each patient, the total score was calculated and graded as follows: score 2 as grade I, scores 3-5 as grade II, scores 6-8 as grade III, scores 9-11 as grade IV, and score 12 as grade V.

Results: Mean age was 35.6 ± 22.8 in grade II patients, 34.9 ± 22.8 years in grade III, 24.1 ± 20.6 in grade IV, and 1 year in grade V (1 patient).Although tumor grade was apparently higher in younger patients, there was no significant difference. Higher grades were associated with lower pre- and postoperative performance status. Preoperative grade was significantly lower in patients operated via the TSR compared to PTR or AIH approach.

Conclusion: The new grading system is useful for analyzing pre- and post-operative performance status, and for selecting operative approach. It is also useful to compare treatment outcomes reported in the literature.

Keywords: Craniopharyngioma; Lateral extension; Grading; Interhemispheric approach; Pterional; Transsphenoidal; Sagittal diameter; Coronal diameter; Foramen Monro; Mammillary body

Introduction

Craniopharyngiomas are one of the most difficult challenges for neurosurgeons, because of the difficulty in total removal and the tendency to recur even operated by experienced hands [1-11]. Craniopharyngiomas have been classified according to the relationship with the sella turcica, optic chiasm, and third ventricle [5,10,11]. Most classification systems reported until now take into account the relationship between the tumor and the third ventricle. However, lateral extension of the tumor should be also considered for selecting appropriate operative approach and comparing surgical results reported in the literature.

To classify craniopharyngiomas appropriately, it is necessary to establish a new classification or grading system based on MRI findings including not only the sagittal diameter but also the coronal diameter of the tumor, as in the widely used Knosp grading system for pituitary adenomas [12]. The grading system should be simple, easy to use by all physicians and surgeons, and consider not only the size but also MRI features of the tumor as well as the relationship of the tumor with surrounding structures such as the mammillary body, foramen of Monro and posterior clinoid process. For example, invasion of the mammillary body by the tumor will cause memory dysfunction, while obstruction of the foramen of Monro will cause hydrocephalus. If the tumor extends below the clinoidal line, it is relatively difficult for most neurosurgeons to extirpate the tumor completely by a transsphenoidal or subfrontal interhemispheric approach. The grading system should also take into consideration the MRI features of the tumor, whether it has an entirely cystic or multicystic component, mixed cystic and solid components, or solid component only [10]. The grading system should be useful not only for selecting the best approach to remove the tumor satisfactorily, but also for predicting the out come. Such system should be developed by experienced neurosurgeons who have operated on many patients with good surgical results. Perioperative management should be discussed based on the same grading system, not only by neurosurgeons but also among physicians especially endocrinologists, and by radiotherapists.

Here, the authors propose a new grading system of

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craniopharyngiomas based on their surgical experience of 100 patients with long-term (mean>10 years) follow-up.

Patients and Methods

Patients

The new grading system was evaluated by analyzing 100 surgeries for craniopharyngiomas mainly conducted by one surgeon (TH) between 1981 and 2012, except two endoscopic operations in 2000 or 2002. Nineteen of the surgeries were conducted in Tottori University from 1981 to 1997, and 81 surgeries in Tokyo Women's Medical University from 1998 to 2012. There were 55 males and 45 females with ages at operation ranging from 1 year to 75 years (mean 33.1 ± 22.7). Thirty-six pediatric (younger than 15 years of age) patients (36.0%) were included, comprising 23 males and 13 females with a mean age of 8.1 ± 4.3 years. Fifteen patients were treated by gamma knife in other institutions before undergoing operation in our institution and 13 patients were treated by gamma knife (GK)after subtotal removal in our institution, while 72 patients did not receive stereotactic radiosurgical treatment. Fourteen patients had undergone prior surgery in other institutions, while 86 patients received primary surgery in our institution.

Surgical approach

In principle, a midline approach was used in surgeries for craniopharyngiomas, with a choice of either a transnasal transsphenoidal (TSR) or an anterior interhemispheric (AIH) approach dissecting the lamina terminal is. When the tumor extended far laterally beyond the lateral limit of the internal carotid artery, a pterional approach (PTR) was used. In these three approaches, an operating microscope was mainly used with or without endoscopic assistance for removing the hidden part of the tumor such as the ventral portion of the optic chiasm and upward or laterally extended portion. The techniques of these three approaches were not substantially different from conventional methods, and the technique of AIH used in this series has been reported in 2010 [4].

Scoring and grading systems

The scoring and grading systems used in our proposed method are shown in Table 1. Based on MRI findings with or without gadolinium enhancement, tumor size was classified by the maximum sagittal diameter into smaller than 2 cm (score 1), 2 to 4 cm (score 2), and greater than 4 cm (score 3). Tumor size was also classified by the maximum coronal diameter perpendicular to midline into smaller than 2 cm (score 1), 2 to 4 cm (score 2), and greater than 4 cm (score 3).A score of 1 was added when the lower limit of the tumor was below the clinoidal line, when the tumor extended to the mammillary body, or when the tumor reached the foramen of Monro. From MRI findings, the tumor composition was easily identified as cystic only (score 0), multi-cystic (score 1), mixture of cystic and solid (score 2), and solid only (score 3). In each patient, the total score was calculated and graded as follows: a score of 2 was classified as grade I, scores from 3 to 5 as grade II, scores from 6 to 8 as grade III, scores from 9 to 11 as grade IV, and a score of 12 as grade V.

When the new scoring and grading system was applied to the 100 cases in this study, the numbers of cases classified in different grades are shown in Table 1.

Statistical analysis

Data are expressed as mean ± SD. One-way analysis of variance (ANOVA) followed by post hoc Fisher's test was used to determine the statistical significance of the differences in Karnofsky performance status score (KPS) among tumor grades and the differences in tumor grade among surgical approaches. A p value less than 0.05 was considered statistically significant.

Illustrative cases

Case 1

A 1-year-old boy was operated via an AIH approach. The tumor had been partially removed previously in another institution, but re growth of the tumor was rapid and a second operation was performed in our department. From the preoperative MRI (Figure 1a), the total score of this patient was 12(3+3+1+1+1+3) and was classified as grade V. Surgery via an AIH approach resulted in total removal of the tumor

MRI findings		**Score**		
Tumor size		*<2 cm*	*2-4 cm*	*>4 cm*
Maximum sagittal diameter		1	2	3
Maximum coronal diameter		1	2	3
Relation with surrounding structure				
Below clinoidal line		1		
Reach foramen of Monro		1		
Reach mammillary body		1		
Tumor composition				
	cystic only	*multi-cystic*	*mixed*	*solid only*
	0	1	2	3
Grade		**Score**	**N**	
Grade I		2	0	
Grade II		3–5	38	
Grade III		6–8	45	
Grade IV		9–11	16	
Grade V		12	1	

N: number of cases analyzed in the present study

Table 1: The proposed new classification system for craniopharyngiomas.

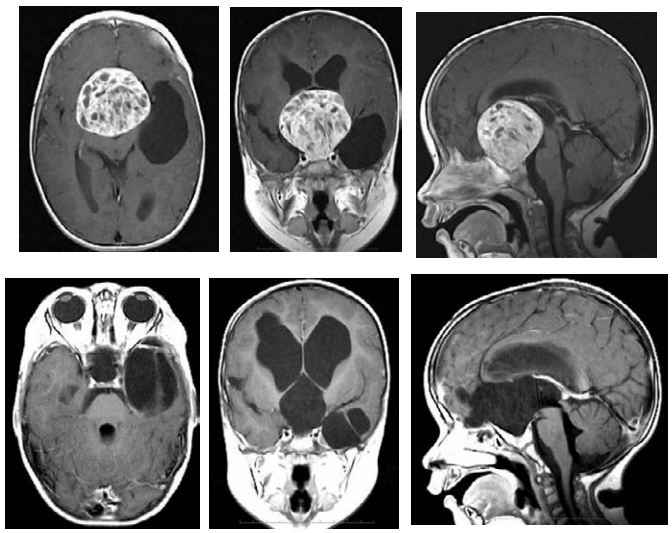

Figure 1: MRI with gadolinium enhancement of a 1-year-old boy. (a) Preoperative MRI show a huge suprasellar solid gadolinium-enhanced mass lesion (left: axial, center: coronal, right: sagittal section). According to the proposed new system, the tumor was grade V (score: 3+3+1+1+1+3=12). (b) Postoperative MRI with gadolinium enhancement at the last follow-up 5 years after surgery demonstrate total tumor removal without recurrence (left: axial, center: coronal, right: sagittal section).

as demonstrated in Figure 1b. No recurrence was detected at the last follow-up 5 years after surgery, although the patient was already blind bilaterally before the second operation and was affected by hypopituitarism.

Case 2

A 5-year-old girl was operated via a PTR approach. As illustrated in Figure 2a, a large cystic tumor extended laterally to the right lateral part of the sphenoidal ridge. This patient was classified as grade IV (score: 3+3+1+1+0+2=10) by the new grading system. Surgery via a right PTR approach resulted in subtotal removal of the tumor (Figure 2b) with mild perforator injury and no significant neurological deficits. Two years later, additional GK treatment was conducted for the residual tumor. Since then, no recurrence was detected at the last follow-up in 2012, 5 years after GK treatment.

Case 3

A 56-year-old woman was operated via a TSR approach. The patient presented with headache, hormonal insufficiency, and diplopia. The tumor was classified as grade III (Figure 3a). A TSR approach with opening the clival dura mater was used to remove the tumor. After total tumor removal, the dural defect was repaired by suturing the dura with a patch graft using the abdominal fascia [13]. At 12 years after the operation, there was no recurrence (Figure 3b) and no definite signs and symptoms of hormonal insufficiency, although occasional steroid replacement was necessary during the long-term postoperative course.

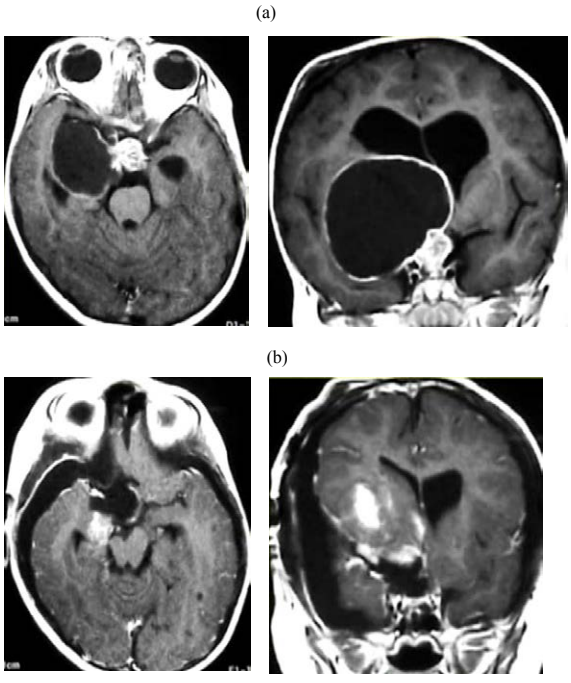

Figure 2: MRI with gadolinium enhancement of a 5-year-old girl. (a) Preoperative MRI show a huge suprasellar cystic gadolinium-enhanced mass lesion with lateral extension and hydrocephalus (left: axial, right: coronal section). According to the proposed new system, the tumor was grade IV (score: 3+3+1+1+0+2=10). (b) Postoperative MRI with gadolinium enhancement demonstrate apparently total removal of the tumor with small lacunae infarction (left: axial, right: coronal section).However, two years later, residual tumor was detected and additional gamma knife (GK) treatment was conducted. Five years after GK treatment until now, no recurrence has been detected both clinically and radiologically.

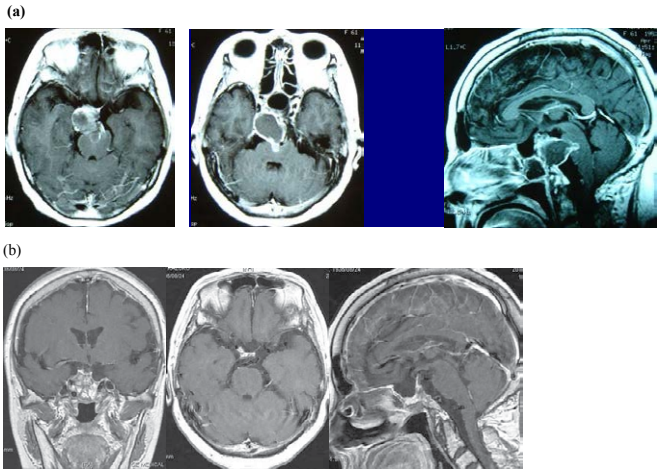

Figure 3: MRI with gadolinium enhancement of a56-year-old woman. (a) Preoperative MRI show a cystic lesion with a gadolinium-enhanced component (left and center: axial, right: sagittal section).According to the proposed new system, the tumor was grade III (score: 2+2+0+0+1+2=7). (b) Postoperative MRI with gadolinium enhancement performed 12 years after the operation demonstrate no recurrence (left: coronal, center: axial, right: sagittal section).

Results

Among 100 patients in the present study, no patient was classified as grade I, while 38 patients were grade II, 45 patients were grade III, 16 patients were grade IV, and 1 patient was grade V (Table 1).

Analysis of score-grading system in 100 patients

Age: The mean age was 35.6 ± 22.8 years in grade II, 34.9 ± 22.8 years in grade III, and 24.1 ± 20.6 years in grade IV patients, with one grade V patient aged 1 year. Although the grade was apparently higher in younger patients, there was no significant difference.

Karnofsky performance status: Preoperative KPS decreased significantly corresponding to an increase in tumor grade (Figure 4). Mean preoperative KPS was 79.5 ± 13.1 in grade II (n=38), 73.1 ± 9.7 in grade III (n=45), 64.4 ± 18.6 in grade IV, (n=16), and 40.0 in grade V (n=1) patients (Figure 4). Gamma knife treatment (preoperative 15 patients, postoperative 13 patients; total 28 patients) or surgery in other institution (reoperation 14 patients) did not influence preoperative KPS score (Figure 4). Likewise, postoperative KPS also decreased significantly with an increase in tumor grade. Mean postoperative KPS was 88.9 ± 13.3 in grade II (n=38), 87.6 ± 9.1 in grade III (n=45), 81.9 ± 19.7 in grade IV (n=16), and 50.0 in grade V (n=1) patients (Figure 5). Gamma knife treatment (preoperative 15 patients, postoperative 13 patients; total 28 patients) or surgery in other institution (reoperation 14 patients) did not influence postoperative KPS score.

Surgical approach: Preoperative tumor scores for various surgical approaches are shown in Figure 6. Mean preoperative grade was 2.48 ± 0.63 for TSR approach (n=25), 2.90 ± 0.77 for AIH (n=61), and 3.08 ± 0.67 for PTR approach (n=12).Preoperative grade was significantly different between TSR and AIH approaches, and between TSR and PTR approaches.

Total removal rate

Among 100 patients, 5 patients were lost to follow-up, and these patients had subtotal tumor removal at surgery and were analyzed as subtotal tumor removal cases. Among 95 patients, the mean follow-up period was 121.0 ± 61.9 months, ranging from 6 to 301 months.

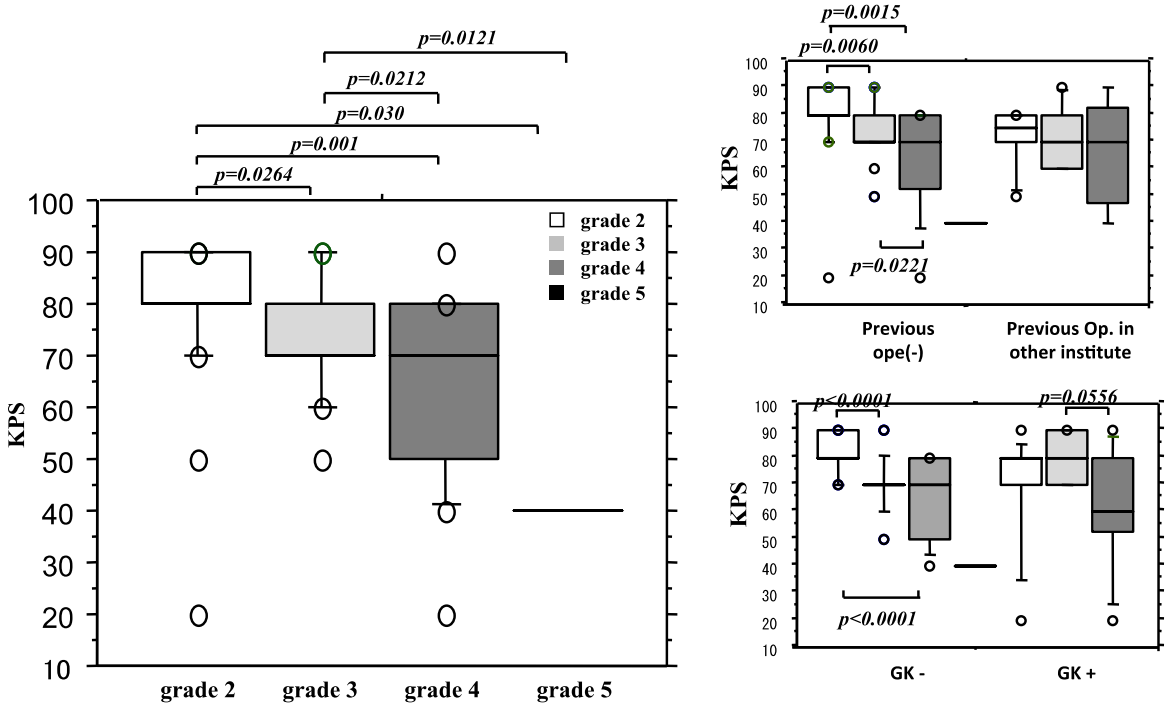

Figure 4: Relationship between preoperative Karnofsky performance status score (KPS) and tumor grade according to the proposed new classification system for craniopharyngiomas. Preoperative KPS decreases significantly corresponding to an increase in tumor grade. Mean preoperative KPS is 79.5 ± 13.1 for grade II (n=38), 73.1 ± 9.7 for grade III (n=45), 64.4 ± 18.6 for grade IV (n=16) is, and 40.0 for grade V (n=1). Gamma knife (GK) treatment (right lower panel) (preoperative; n=15, postoperative; n=13, total; n=28) or surgery in other institution (right upper panel: previous Op. in other institute; n=14) does not influence preoperative KPS score. GK+; GK treatment before or after surgery, GK-; No GK treatment, Previous Op. in other institute; 1st operation done in other institution, Previous Op. (-); primary surgery done in our institution.

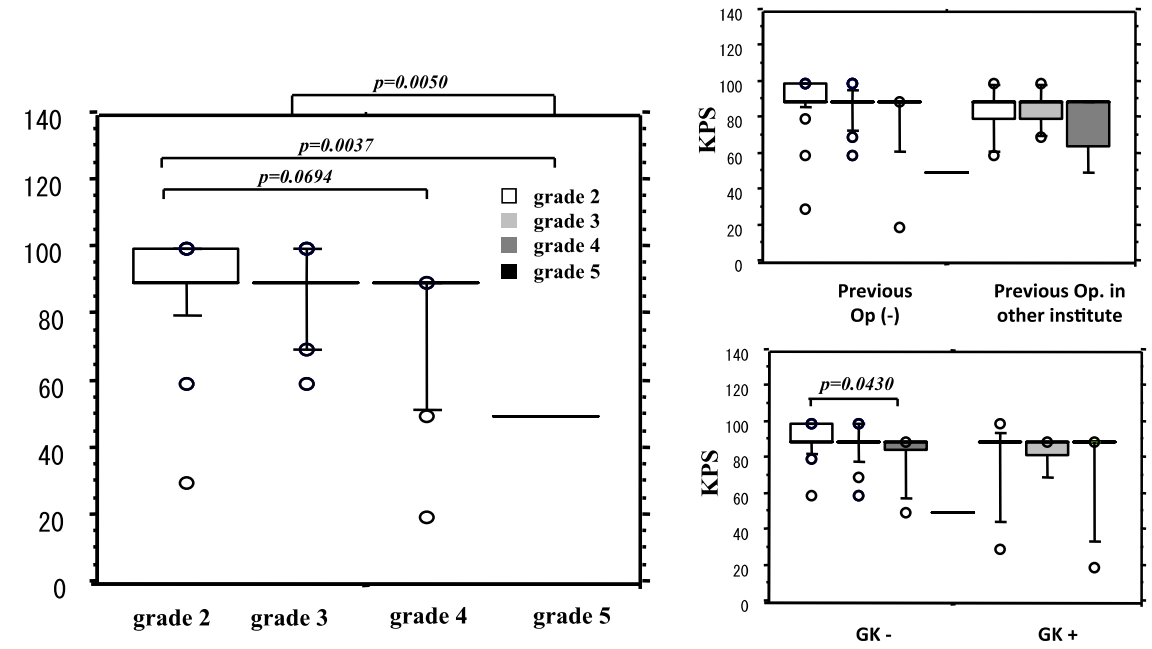

Figure 5: Relationship between postoperative Karnofsky performance status score (KPS) and tumor grade according to the proposed new classification system for craniopharyngiomas. Postoperative KPS decreases significantly with an increase in tumor grade. Mean postoperative KPS is 88.9 ± 13.3 for grade II (n=38), 87.6 ± 9.1 for grade III (n=45), 81.9 ± 19.7 for grade IV (n=16), and 50 for grade V (n=1).
Gamma knife (GK) treatment (right lower panel) (preoperative; n=15, postoperative; n=13, total; n=28) or surgery in other institution (right upper panel: previous Op. in other institute; n=14) does not influence postoperative KPS score. GK+;GK treatment before or after surgery, GK- ; No GK treatment, Previous Op. in other institute; 1st operation done in other institution, Previous Op. (-); primary surgery done in our institution.

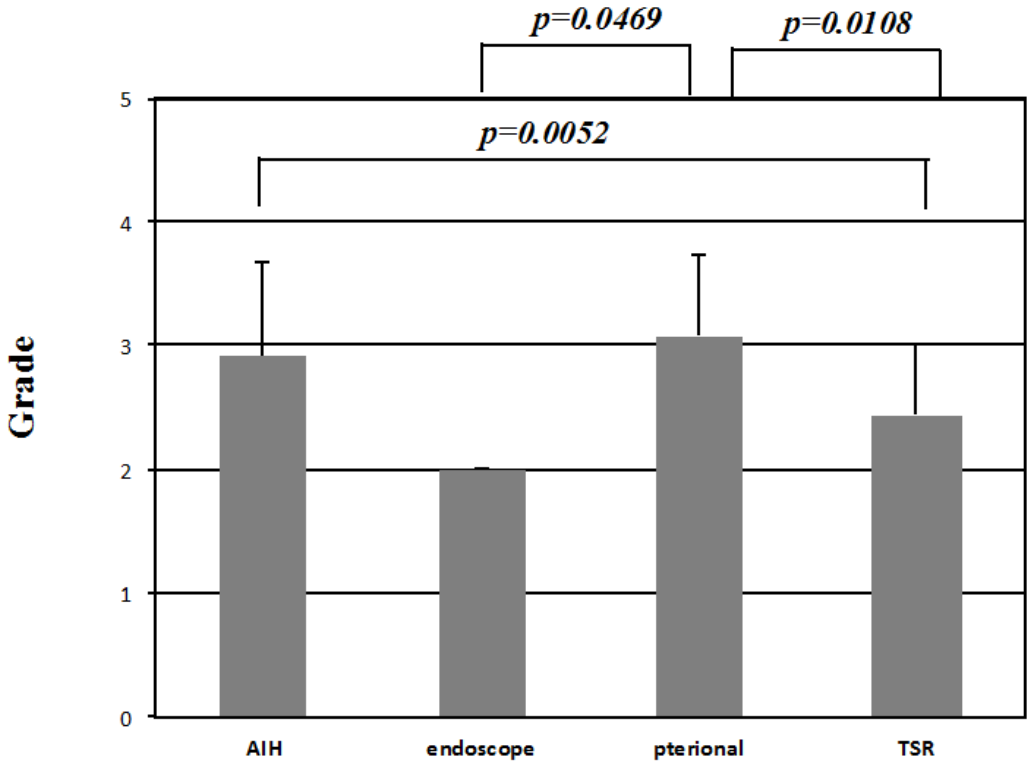

Figure 6: Relationship of operative approach and tumor grade according to the proposed new classification system for craniopharyngiomas. AIH: Anterior Interhemispheric, TSR: Transnasal Transsphenoidal. The mean preoperative grade was 2.48 ± 0.63 for TSR approach (n=25), 2.90 ± 0.77 for AIH (n=61), and 3.08 ± 0.67 for pterional approach (n=12). Preoperative grade was significantly different between TSR and AIH approaches, and between TSR and PTR approaches.

As shown in Table 2, using the AIH approach, tumor was completely removed in 52 of 61 patients (85.2%). Tumor was completely removed in 21 of 25 cases (84.0%) by the TSR approach and in 7 of 12 cases (58.3%) by the PTR approach. Total removal rate by the PTR approach was apparently lower than that by AIH or TSR approach, but there was no statistical difference among these three approaches. Of the two patients who underwent endoscopic operation, the cystic tumor was totally removed in a boy after two operations, while the tumor was only partially removed in another old patient with renal insufficiency. Totally, tumor was completely removed by surgery alone in 81 of 100 patients. However, two surgeries were necessary in 9 patients, and three surgeries were required in 3 patients to accomplish total removal without further recurrence. Therefore, multiple surgeries were necessary to obtain total removal in 12 of 81 patients (14.8%).

Discussion

We propose a new classification system for grading craniopharyngiomas. Analysis of this system using 100 cases showed a tendency of higher tumor grade in younger patients but with no significant difference. On the other hand, a significant reverse relationship was observed between preoperative tumor grade and preoperative as well as postoperative KPS. Furthermore, the mean grade of tumors operated by a TSR approach was significantly lower than those using an AIH or a PTR approach, suggesting that this grading system may also be useful for selecting operative method.

Several classification systems of craniopharyngiomas have been proposed [5,10,11]. Some systems classify the tumor according to operative findings such as the relationship between the tumor and infundibulum [5], but it is sometimes very difficult to clarify the relationship between the tumor and infundibulum preoperatively. The relationship between the tumor and pituitary stalk is established only after meticulous intraoperative observation. Therefore, these classifications have limited use for deciding a suitable operative approach. Kassam et al. [5] classified craniopharyngiomas by intraoperative findings. Yasargil et al. [11] also classified the common locations of craniopharyngiomas diagrammatically based on mainly MRI and operative findings. Yamada et al. [10] illustrated the tumor-third ventricle relationship of supradiaphragmatic craniopharyngiomas based on MRI and operative findings. These classification systems are very useful to understand the tumor and surrounding structures. However, a clinically valuable classification system should be simple to use, applicable before operation, and useful for determining operative approach and comparing treatment results not only among surgeons but also among various treatment modalities such as stereotactic radiosurgery and stereotactic radiotherapy. Yamada et al. [10] reported a simple tumor classification system based on mainly preoperative MRI findings, but their classification system was developed only for transsphenoidal approach. Our new system classifies craniopharyngiomas by preoperative MRI findings and considers the relationship between the tumor and surrounding structures as well as the extent of lateral, superior, posterior, and inferior extensions of the tumor. Our system can be used for the selection of operative approach and comparison of operative results, and correlates well with patient's pre- and postoperative performance status.

Concerning the rate of total removal, Yasargil et al. [11] and Hoffmann et al. [3] reported 90% total removal rate in their series.

Surgical approach	No. of cases with complete removal/ No. of all cases	%
Anterior inter-hemispheric approach	52/61	85.2%
Transsphenoidal approach	21/25	84.0%
Pterional approach	7/12	58.3%
Endoscope	1/2	50.0%
Total	81/100	81.0%

Table 2: Rates of total tumor removal for different surgical approaches.

Although Yasargil et al. [11] reported a high rate of total removal in their series, they also reported a high mortality rate; whereas Hoffman et al. [3] reported 90% total removal rate with only 2% mortality for pediatric craniopharyngiomas. There are large differences in total tumor removal rate among surgeons, but the demographic data of the patients also vary. Therefore, it is difficult to compare the surgery success rates among surgeons simply based on total removal rate, morbidity, and mortality. Considering these situations, grading craniopharyngiomas by the same classification system among clinicians is important. Pituitary adenomas are classified by size into microadenomas and macroadenomas, or by the Knosp grading system considering lateral extension of the tumor. For craniopharyngiomas, using a similar classification system among surgeons is mandatory to discuss the operative approach, surgical removal rate, and success rate. Our proposal of a new classification system would at least generate discussions on a wide variety of issues concerning tumor properties, surgical techniques and surgical outcome of craniopharyngiomas based on the same standard.

Conclusion

The new grading system is useful to analyze preoperative and postoperative KPS, and to select surgical approaches. It is also useful to analyze surgical results. Based on a common grading system, surgeons who treat craniopharyngiomas can discuss the best management for the tumors, which is one of the most difficult issues in infants and adults in the neurosurgical field.

References

1. Elliott RE, Jane JA Jr, Wisoff JH (2011) Surgical management of craniopharyngiomas in children: meta-analysis and comparison of transcranial and transsphenoidal approaches. Neurosurgery 69: 630-643.
2. Fahlbusch R, Honegger J, Paulus W, Huk W, Buchfelder M (1999) Surgical treatment of craniopharyngiomas: experience with 168 patients. J Neurosurg 90: 237-250.
3. Hoffman HJ, De Silva M, Humphreys RP, Drake JM, Smith ML, et al. (1992) Aggressive surgical management of craniopharyngiomas in children. J Neurosurg 76: 47-52.
4. Hori T, Kawamata T, Amano K, Aihara Y, Ono M, et al. (2010) Anterior interhemispheric approach for 100 tumors in and around the anterior third ventricle. Neurosurgery 66: 65-74.
5. Kassam AB, Gardner PA, Snyderman CH, Carrau RL, Mintz AH, et al. (2008) Expanded endonasal approach, a fully endoscopic transnasal approach for the resection of midline suprasellar craniopharyngiomas: a new classification based on the infundibulum. J Neurosurg 108: 715-728.
6. Kawamata T, Amano K, Aihara Y, Kubo O, Hori T (2010) Optimal treatment strategy for craniopharyngiomas based on long-term functional outcomes of recent and past treatment modalities. Neurosurg Rev 33: 71-81.
7. Leng LZ, Greenfield JP, Souweidane MM, Anand VK, Schwartz TH (2012) Endoscopic endonasal resection of craniopharyngiomas:Analysis of outcome including extent of resection, cerebrospinal fluid leak, returnto preoperative productivity, and body mass index. Neurosurgery 70: 110-124.
8. Mortini P, Losa M, Pozzobon G, Barzaghi R, Riva M, et al. (2011) Neurosurgical treatment of craniopharyngioma in adults and children: early and long-term results in a large case series. J Neurosurg 114: 1350-1359.
9. Van Effenterre R, Boch AL (2002) Craniopharyngioma in adults and children: a study of 122 surgical cases. J Neurosurg 97: 3-11.
10. Yamada S, Fukuhara N, Oyama K, Takeshita A, Takeuchi Y, et al. (2010) Surgical outcome in 90 patients with craniopharyngioma: an evaluation of transsphenoidal surgery. World Neurosurg 74: 320-330.
11. Yaşargil MG, Curcic M, Kis M, Siegenthaler G, Teddy PJ, et al. (1990) Total removal of craniopharyngiomas. Approaches and long-term results in 144 patients. J Neurosurg 73: 3-11.
12. Knosp E, Steiner E, Kitz K, Matula C (1993) Pituitary adenomas with invasion of the cavernous sinus space: a magnetic resonance imaging classification compared with surgical findings. Neurosurgery 33: 610-617.
13. Hori T (1986) Advancement of pituitary surgery. -Dural suture technique-. Japan Medical Journal (Japanese) 3254: 16-20.

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