Different Modalities of Management of Craniopharyngiomas

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Abstract: Objective: To evaluate the different surgical approaches (transcranial & transnasal transphenoidal) and adjuvant modalities (Gamma Knife, Interferon, Bleomycin, Ommaya Reservoir,…..) for treatment of craniopharyngiomas. **Patients and Methods:** This study was conducted in neurosurgery department in Al-Azhar University (Al-Hussein & Bab El-Sha'ariya Hospitals) during the period from June 2012 to March 2017. This work was done on twenty patients with craniopharyngioma. The patients were evaluated according to the modality of treatment used, and follow-up (by clinical, endocrinological, ophthalmological and radiological assessment) were done six months after the intervention for assessment of the results of each treatment modality. **Results**: The outcome of management in this work is represented as Causes of poor outcome were absence of functional vision in the four cases (case 6, 10, 12, 17) plus loss of employability in case 6 and poor school performance in case 17, but it is worth noting that poor vision and cognitive impairment were pretreatment presentations **Conclusion**: Beyond treatment success ratings of craniopharyngioma related to diagnostic and treatment strategies is the experience level of the neurosurgeon.
[Yousef Abd- Elgaliel Barakat, Hamdy Mohammad Behairy, Ibrahim Hassan Mohammad, Mokhtar Ragab Ramadan, Abdullah Fathy Abdullah Abd El-Wahed. **Different Modalities of Management of Craniopharyngiomas.** *Rep Opinion* 2017;9(6):27-35]. ISSN 1553-9873 (print); ISSN 2375-7205 (online). http://www.sciencepub.net/report. 4. doi:10.7537/marsroj090617.04.

Keywords: Different; Modality; Management; Craniopharyngiomas

1. Introduction:

Craniopharyngiomas are benign extra-axial tumor which follows progressive clinical course leading to significant morbidity. (13). A craniopharyngioma is a slow-growing, extra-axial, epithelial-squamous, calcified, and may cystic or mixed tumor arising from remnants of the craniopharyngeal duct and Rathke cleft and occupying the suprasellar region (5).

There is a bimodal age distribution, with one peak in children between 5 and 14 years, and a second peak in adults between 50 and 75 years of age. (14). Adamantinomatous craniopharyngiomas are more common in the pediatric age group, while papillary craniopharyngiomas are seen predominantly in adults (21). Surgical management of craniopharyngiomasis still remain a big challenge. The difficulties of surgical treatment are caused by complicated relationship of the tumor with surrounding nervous and vascular structures of important functions. Due to benign histological nature and malignant behavior, there are two different attitudes to the management of craniopharyngiomas:

a) Radical surgical excision,

b) Planned incomplete excision and radiotherapy.. (14). Computed tomography have high sensitivity to detect calcifications which are present in about 85% of childrenand in 40% of adult patients with craniopharyngiomas. MRI is essential for defining the anatomy of the tumor and surrounding.

2. Patients and methods

This study was conducted in neurosurgery department in Al-Azhar university (Al-Hussein & Bab El-Sha'ariya Hospitals) during the period from June 2012 to March 2017. This work was done on twenty patients with craniopharyngioma.

The patients were evaluated according to the modality of treatment used, and follow-up (by clinical, endocrinological, ophthalmological and radiological assessment) were done six months after the intervention for assessment of the results of each treatment modality.

All patients signed an informed consent. The outcome of the management was graded according to the criteria adopted by (27).

Our inclusion criteria were patients aged from 6 to 56 years with both. The following approaches were used in this study: subfrontal, lateral supraorbital, pterional, transsphenoial, and combined approaches.

-The reported surgical findings included; tumor consistency (cystic and/or solid), presence of

calcifications, and adhesions (to the hypothalamus, optic apparatus, arteries, or other brain structures).

-The surgical procedures included tumor removal (total, subtotal, cyst evacuation and biopsy or biopsy only), CSF diversion (shunting), or Ommaya reservoir placement. the second most common approach & adjuvant therapy (External Beam Radiation Therapy (**EBRT**) or Gamma Knife Radiosurgery (**GKS**) were done for selected patients. **Intracystic treatment: Bliomycin** Intracystic **interferon** was used for cystic lesions with Ommaya reservoir. The outcome of the management was graded according to the criteria adopted by (27).

I- Good:

1- Alive at follow-up examination.

2- No major motor deficit related to operation or tumor progression.

3- Functional vision.

4- Katz Grade A (able to perform basic activities of daily living "ADLs").

5- Karnofsky Performance Scale score ≥80.

6- No more than 1 year behind in expected school grade.

7- Employability for adults of working age.

8- Absence of debilitating psychological or emotional problems.

II-Poor:

- All patients not meeting the “good” criteria.

Results

This section presents the results obtained in twenty cases treated for craniopharyngioma. The age ranged in this study from 6 years to 55 years. 9 cases were children (≤ 16 years) and 11 were adults (>16 years). The median age was 16.5 years and the mean age was 25.45 years. The peak incidence in this work was below 10 years. The study was conducted on 20 cases, 11cases were of primary disease presentation, and 9 recurrent cases. All cases presented with gradual onset and progressive course, the duration of symptoms ranged from 12 months to 36 months. The mean duration was 18.85 months; the median duration was 9.0 months. Visual impairment was the most common presenting complaint (all the twenty cases). Headache was the second most common complaint (16 cases). Endocrinological symptoms come third (9 cases) Routine laboratory investigations were within normal range in most cases in this work, while hormone assay revealed GH deficiency in 3 cases (15%), Hypothyroidism in 2 cases (10%), Hyperprolactinemia in 8 cases (40%), Hypogonadism in 7 cases (35%), adrenal failure in one case (5%).. All cases in this work were studied with MRI, 19 cases with CT brain, and 3 cases with plain x-ray. Treatment modalities used in this work included surgical excision utilized in 16 cases (10 primary cases and 6 recurrent), Gamma knife radiosurgery was used as adjuvant therapy in 4 primary cases for postoperative residual, 1 recurrent case, and as sole treatment in 1 primary case, external beam radiation therapy was used in 1 recurrent case after surgical debulking, Ommaya reservoir was inserted in the cystic part of 3 primary and 4 recurrent cases, intracystic interferon was tried in 3 recurrent cases, and ventriculoperitoneal shunt was used to treat hydrocephalus in 3 primary and 4 recurrent cases. Surgical approaches used in this work included; transsphenoidal in 2 cases, transciliary lateral supraorbital approach "via eye brow skin incision" in 3 cases, subfrontal approach in 6 cases, pterional approach in 4 cases, and combined transcallosal and subfrontal approach in 1 case Three cases died in the early postoperative period (2 recurrent and 1 primary); case 3 and case 20 showed nearly similar history of disturbed conscious level, polyuria, and electrolyte disorders, moreover case 20 developed uncontrolled fever. Case 16 showed postoperative left hemiplegia with unremarkable CT brain changes, patient died on the 5th postoperative day after having respiratory irregularity, polyuria, electrolyte disorders, and disturbed conscious level.

The follow-up period in this work ranged from 2 months to 24 months, the mean follow up period was 9.7 months.

Visual function evaluation during follow up period revealed improvement in 3 cases; case 5, 7, and 19. Other cases showed no change in the pretreatment visual function (acuity and field), and no case developed deterioration of vision.

No change in the pretreatment endocrinological status (whether improvement or deterioration) apart from 3 surgically treated cases requiring long-term ADH treatment. Patients with preoperative hormone dysfunction continued their follow-up with the endocrinologist.

Complications were infrequent in these cases; only 1 case developed transient CSF rhinorrhea after transsephenoidal approach (case 12) and resolved with conservative measures, another case developed acute subdural hematoma after attempted excision of recurrent tumor (case 5) but was not sizable and was managed expectantly. No other complications reported with surgery, radiation therapy, or intracystic interferon.

The outcome of management in this work is represented as Causes of poor outcome were absence of functional vision in the four cases (case 6, 10, 12, 17) plus loss of employability in case 6 and poor school performance in case 17, but it is worth noting that poor vision and cognitive impairment were pretreatment presentations.

Table (1) Age incidence in 20 cases of craniopharyngioma.

Age group in years	Number of cases	Percentage from total
0-10	6	30%
11-20	4	25%
21-30	1	5%
31-40	3	15%
41-50	5	20%
>50	1	5%

Table (2) Sex distribution relative to age.

Sex / Age group	Male	Female	Total
Children	3 (15%)	7 (35%)	10 (50%)
Adults	5 (25%)	5 (25%)	10 (50%)
Total	8 (40%)	12 (60%)	20 (100%)

Table (3) Clinical presentation relative to tumor type.

Patient group / Clinical presentation	Primary tumor (11 case)	Recurrent tumor (9 cases)	Total
Visual impairment	11 (100%)	9 (100%)	20 (100%)
Headache	9 (83%)	8 (87.5%)	17 (85%)
Endocrinological symptoms	2 (16.6%)	6 (75%)	8 (40%)
Hypothalamic regulatory symptoms	2 (16.6%)	2 (25%)	4 (25%)
Neurobehavioral disorders	2 (16.6%)	1 (12.5%)	3 (15%)
Seizures	1 (8.3%)	0 (0%)	1 (5%)
Vomiting	1 (8.3%)	2 (25%)	3 (15%)
Autonomic disturbance	0 (0%)	1 (12.5%)	1 (5%)

Table (4) Clinical presentation relative to age group.

Patient group / Clinical presentation	Children (10 cases)	Adults (10 cases)
Visual impairment	10	10
Headache	9	8
Endocrinological symptoms	4	4
Hypothalamic regulatory symptoms	0	4
Neurobehavioral disorders	0	3
Seizures	0	1
Vomiting	2	1
Autonomic disturbance	0	1

Table (5) Results of hormone assay.

Hormone	Number of cases		
	Children	Adults	Total
GH deficiency	3	0	3 (15%)
Hypothyroidism	0	2	2 (10%)
Hyperprolactinemia	3	5	8 (40%)
Hypogonadism	2	5	7 (35%)
Adrenal failure	0	1	1 (5%)

Table (7) Treatment modalities relative to tumor type.

Patient group / Treatment modalities	Primary tumor (12 case)	Recurrent tumor (8 cases)	Total (20 cases)
Surgical excision	10	6	15
Gamma knife radiosurgery	5	1	6
External beam radiation therapy	0	1	1
Ommaya reservoir	3	4	7
Intracystic interferon	0	3	3
Ventriculoperitoneal shunt	3	4	7

Table (6) Imaging features relative to tumor type

Imaging features \ Patient group		Number of cases		
		Primary tumor	Recurrent tumor	Total
Shape	Oval	6	3	9
	Lobular	5	5	10
	Bilobed	1	0	1
Calcifications	-	9	1	10
	+	2	6	8
	++	1	0	1
	+++	0	1	1
Cysts	-	1	0	1
	+	11	6	17
	++	0	2	2
Solid parts	-	2	0	2
	+	10	8	18
Enhancement	-	1	0	1
	+	11	8	19
Surrounding edema	-	11	5	16
	+	1	3	4
Hydrocephalus	-	9	4	13
	+	3	4	7

Table (8) Surgical approach relative to tumor type.

Surgical approach \ Patient group	Primary tumor (12 case)	Recurrent tumor (8 cases)	Total (20 cases)
Transsphenoidal	1	1	2
Lat. Supraorbital	2	1	3
Subfrontal	3	2	6
Combined Transcallosal & subfrontal	0	1	1
Pterional	3	1	4

Table (9) Mean ẋ± SD of age distribution according to outcome.

Outcome \ Age	ẋ ± SD	t	P
Died (3 cases)	30.3±20.5 Range (9-50)	0.45	>0.05
Alive (17 cases)	24.6±17.3 Range (5-50)		

Table (10) Mean ẋ± SD of sex distribution according to outcome.

Outcome \ Sex	Male 8 cases		Female 12 cases		Total 20 cases		Z
	No.	%	No.	%	No.	%	
Died (3cases)	1	12.5	2	16.6	3	15	0.24
Alive (17cases)	7	87.5	10	83.4	17	85	0.1

Table (11) Mean ẋ± SD of duration according to outcome.

Outcome \ Duration	ẋ± SD	t	P
Died (3 cases)	15.3±17.9 (Range 4-36)	0.39	>0.05
Alive (17 cases)	11.2±6.5 (Range 2-24)		

As shown in tables 9, 10, 11 no statistically significant correlation were found between age, sex, or pretreatment duration of symptoms and outcome.

Table (12) Correlation between presenting symptoms and outcome.

Symptom \ Outcome	Died		Alive		Z	P
	No.	%	No.	%		
Headache	3	100	14	82.3	0.31	>0.05
Vomiting	0	0.0	3	17.6	0.79	>0.05
Diminution of vision	3	100	17	100	_	_
Field defect	3	100	15	88.2	0.2	>0.05
Growth retardation	0	0.0	3	17.6	0.79	>0.05
Galactorrhea	1	33.3	1	5.9	1.39	>0.05
Obesity	2	66.7	2	11.8	1.96	<0.05
Fatigability & decreased activity	2	66.7	2	11.8	1.96	<0.05
Loss of libido	1	33.3	2	11.8	0.89	>0.05
Amenorrhea	1	33.3	2	11.8	0.89	>0.05
Sleep disturbance	2	66.7	1	5.9	1.39	>0.05
Behavioral disorders	2	66.7	1	5.9	1.39	>0.05
Polyuria & polydepsia	3	100	1	5.9	3.36	<0.05
Seizures	0	0.0	1	5.9	0.43	>0.05
Autonomic disturbance	1	33.3	0	0	2.44	<0.05
Previous surgery	2	66.7	6	30%	3.55	<0.05

Correlation between the presenting symptoms and outcome were statistically significant regarding; the presence of obesity, fatigability & decreased activity, polyuria & polydepsia, autonomic disturbance, and previous surgery to poor outcome.

Table (13) Correlation between imaging features and outcome.

Imaging features \ Outcome		Died		Alive		Z	P
		No.	%	No.	%		
Shape	Oval	1	33.3	8	47.1	0.33	>0.05
	Lobular	2	66.7	8	47.1	0.44	>0.05
	Bilobed	0	0	1	5.9	0.43	>0.05
Calcifications	-	0	0	10	58.8	1.88	<0.05
	+	1	33.3	7	41.2	0.2	
	++	1	33.3	0	0	2.44	<0.01
	+++	1	33.3	0	0	2.44	<0.01
Cysts	-	0	0	1	5.9	0.43	
	+	2	66.7	15	88.2	0.37	
	++	1	33.3	1	5.9	1.39	
Solid parts	-	0	0	2	11.8	0.63	
	+	3	100	15	88.2	0.2	
Enhancement	-	0	0	1	5.9	0.43	
	+	3	100	16	94.1	0.1	
Surrounding edema	-	1	33.3	15	88.2	0.98	
	+	2	66.7	2	11.8	1.96	<0.05
Hydrocephalus	-	0	0	13	76.5	2.56	<0.01
	+	3	100	4	23.5	2.06	<0.05

Analysis of imaging features of the tumor revealed no significant relationship between outcome and shape, cystic component, solid parts, or enhancement. There were statistically significant relationships between outcome and presence of hydrocephalus, peritumoral edema, and calcification. The relationship was highly significant between outcome and presence of heavy calcification.

Table (14) Correlation between tumor size and outcome.

Size / Outcome	Range	ẋ ±SD	T	P
Died	30-165 cc	101.7±47.9	2.31	<0.05
Alive	6-123.8 cc	35.3±31.7		

Also, there was statistically significant relationship between outcome and tumor size.

4. Discussion

Craniopharyngiomas are the most common non glial brain tumor of childhood, comprising 6–8% of pediatric brain tumors. They have a bimodal distribution in age at diagnosis. Despite their benign histology, craniopharyngiomas have malignant clinical course, especially in children. This work was done on twenty patients with craniopharyngioma over a period of two years, and subjected to one of the modalities of treatment.

The age ranged from 5 years to 55 years. 10 cases were children (≤ 16 years) and 10 were adults (>16 years). The median age was 16.5 years and the mean age was 25.45 years. The peak incidence in this work was below 10 years, and about 55% of cases were younger than 20 years. The peak incidence in this work conforms well to the first peak of the bimodal age distribution in the literature (7). No second peak was faced, but having 55% of cases younger than 20 years conforms with the results of (Bunin et al., 1998).

Females represented 65% of cases in this work, and males were 35 %; that is quite similar to the results of (30) in which females were 52.78% and males 47.22%, but is different from most case series that reported equal sex distribution (7), and the large English series of (20) in which males were affected 30% more often than females.

The duration of symptoms ranged from12 months to 36 months. This duration is within the range described in most series (27). The mean duration was 11.85 months; the median duration was 9.0 months, which is very close to the median duration in the literature (10 months) (4). Visual disturbances were the most common presenting symptom in this work; the same with (2).

The first half of this case's scenario in which complete recovery of vision was achieved after surgery is similar to the case report of (22) which is considered exceptional. Headache was the second most common complaint faced with 85%, and was associated with vomiting in15% of cases; this was close to the results of (27) in which headache occurred in 79% of cases and vomiting in 22%. Approximately 40% of adults present with symptoms of endocrine disturbances. Gonadal insufficiency was the most common endocrine abnormality at presentation (in 50% of cases). This consists of loss of libido and impotence in men (30% of cases), galactorrhea and secondary amenorrhea in women (20% of cases). Also about 40% of children presented with complaints attributable to an endocrinologic deficit; growth retardation represented 15% of them and delayed puberty 35%. Diabetes insipidus presented in 20% of cases, hypothyroidism and hypocortisolemia were manifest in 10% and 5%, respectively; this was more or less similar to the findings of (23 & 3).

Obesity was seen in 20% of cases, sleep disturbance was present in 15%, behavior disorders 15%, cognitive functions were impaired in 20% of cases. Case number 20 showed characteristic clinical course alternating between cachexia and obesity before each recurrence; her overall clinical condition resulted in social failure and she died shortly after the last surgery. Presence of cachexia and emaciation could be explained by the rare occurrence of diencephalic syndrome in adults according to (24). The relation between weight abnormalities and impairment of cognitive functions was statistically significant that conform with the description of (7) about the typical child with a craniopharyngioma who is short and obese with poor eyesight and learning difficulties. Also, this association allows the classification of these manifestations as hypothalamic dysfunction manifestations according to (10) who adopted hypothalamic dysfunction. the tumor consistency of the recurrent lesion did not differ significantly from the primary lesion, compatible finding with (2). Also, there was no significant difference in the tumor consistency between children and adults, compatible finding with (3).

Calcifications were evident in 55% of cases, within the range of 45–57% reported in the works of (27 & 23). Hydrocephalus was evident in 33% of cases, within the range 20–38% reported by (27 & 23).

Perifocal edema was demonstrated in 4 cases, its presence in axial CT and MRI scans was described by (17) as "moustache sign" which is apparent in case number 5 of the illustrative cases.

All pathologically proven craniopharyngioma (19 cases) were of the adamantinomatous subtype apart from case number 12 that was of the papillary variety representing 5.26% of all cases, and 11% of adult

cases in comparison to the work of (31) in which papillary variety was reported in 14–50% in adult patients, and in less than 2% of pediatrics.

The goal of treatment was to relieve the raised ICP and optic nerve compression, preserve existing hypothalamic function and vision, provide long-term tumor control, avoid repeat surgery and minimize neurotoxic effects from surgery and radiotherapy. Treatment modalities used in this work included; surgical excision utilized in 16 cases (10 primary cases and 6 recurrent), Gamma knife radiosurgery was used as adjuvant therapy in 4 primary cases for postoperative residual, 1 recurrent case, and as sole treatment in 1 primary case, external beam radiation therapy was used in 1 recurrent case after surgical debulking, Ommaya reservoir was inserted in the cystic part of 3 primary and 4 recurrent cases, intracystic interferon was tried in 2 recurrent cases, and ventriculoperitoneal shunt was used to treat hydrocephalus in 3 primary and 4 recurrent cases.

The degree of resection was graded according to the parameters of (9) which depend on both the surgeon's assessment at surgery and the results of postoperative follow-up MR imaging.

If the postoperative MR imaging indicated no residual tumor but residual tumor was known to have been left behind at surgery, then the surgery was not considered a GTR (gross-total resection). Radical STR (radical subtotal resection) was defined as 95% tumor removal, STR as at least 50% tumor removal, and partial resection as < 50% removed. Accordingly, in this work GTR was achieved in 6.25%, radical STR in 56.25%, STR in 31.25%, partial resection in 6.25% of surgically treated cases.

Complete surgical eradication of the tumor tissue was not reachable in many cases because of:

(a) complex relationship of the tumor with the structures of the chiasmatic-hypothalamic-pituitary region (difficult accessibility),

(b) its firm adherence to surrounding structures and its large size in some cases (problematic resectability), and

(c) intentionally in some cases following the concept of (12) who recommended radical removal only for tumors not involving, or just compressing the hypothalamus and intentional subtotal/ partial excision and radiotherapy for tumors involving hypothalamus.

Surgical approaches used in this work included;

- Transsphenoidal in 2 cases,

-Supraorbital approach "via eye brow skin incision" in 3 cases,

-Subfrontal approach in 6 cases, (4 primary and 2 recurrent cases).

-Pterional approach in 4 cases, and

-Combined transcallosal and subfrontal approach in 1 case.

-Transsphenoidal approach was used in case1.

Choice of the surgical approach was based on the surgeon's preference and experience, as well as, the location, the consistency, the degree of calcification, the shape and size, and whether the tumor is primary or recurrent.

Craniopharyngioma has potentially greater radiosensitivity not only when fractionated therapy is undertaken but also with radiosurgery. (19 & 8).

Gamma Knife radiosurgery "GKS" was used as adjuvant therapy in 4 primary cases for postoperative residual, 1 recurrent case,

GKS introduced as secondary treatment for:

(a) residual tumor after redo transsphenoidal debulking of recurrent craniopharyngioma in case1,

(b) residual tumor in case 9 when radical excision was difficult during subfrontal approach due to marked adhesion to the basilar artery and brain stem, Ommaya reservoir was inserted and patient was referred to GKS,

(c) residual and right thalamic extension in case 13,

(d) residual growth detected on follow up MRI performed 3 months after sub-frontal approach in case 14.

Intracystic interferon was tried in 3 recurrent predominantly cystic tumors and resulted in reduction of cyst size and satisfactory tumor control over follow up period of about 12 months with no reported complications.

Interferon is easy to handle and available at a low cost with a low morbidity rate (11).

The follow-up period in this work ranged from 2 months to 24 months, the mean follow-up period was 9.7 months. However, due to financial considerations, poor compliance with follow-up appointments, or logistical reasons, not all patients completed both pre- and postoperative testing.

Three cases died in the early postoperative period, representing about 15.7% of surgically treated cases.

Visual function evaluation during follow-up period revealed modest improvement in 3 cases; case 5, 7, and 19. It can be accepted that visual function only occasionally improves after surgery, according to (15).

Diabetes insipidus was faced with 35% of cases in the post-operative period, compatible with (20,18 & 3) who believe that restoration of preexisting hormone deficits after treatment is absent or uncommon, and reported diabetes insipidus within the range of 25–86%.

The outcome of the management was graded according to the criteria adopted by (27). That classify

patient outcomes as either good or poor; patients not meeting all criteria were considered to have had poor outcomes.

Correlation between the presenting symptoms and outcome were statistically significant regarding the presence of hypothalamic dysfunction manifestations and poor outcome, consistent with (26).

Correlation between recurrence and poor outcome was statistically significant. Analysis of imaging features of the tumor revealed no significant relationship between outcome and shape, consistency, or enhancement, compatible with (3).

There were statistically significant relationships between outcome and presence of hydrocephalus, consistent with (26 & 28), on contrary to (3 & 6) who denied the impact of hydrocephalus on outcome. Also, there was statistically significant relationship between outcome and tumor size, consistent with (26), and on contrary to (Wen et al., 1989).

The relationships between outcome and presence of peritumoral edema, and calcifications were statistically significant. (16) reported high recurrence rate and poor outcome with calcified tumors.

Conclusion

The role of aggressive surgical removal of these tumors is still somewhat controversial. Some authors contend that Gross total resection, when safe, is the treatment of choice to offers the best chance for tumor-free survival. Other authors support a more conservative approach consisting of subtotal resection combined with post operative radiation. Such management has been shown to be associated with equivalent cognitive outcomes and less neurological impairment and deterioration of quality of life. For acraniophryngioma with large cystic components, stereotactic, or open implantation of an intracystic catheter with subcutaneous reservoir is a proven treatment both for the relief of pressure and, in some cases, for the instillation of sclerosing substances (Bleomycin).

Radiation therapy has long been used as in adjuvant treatment of craniopharyngioma after subtotal resection an das primary treatment of recurrence. Conformal radiotherapy, intensity – modulated radiotherapy, stereotactic radiotherapy, steriotasctic radiosurgery (SRS), and multisession SRS have all been used to maximize treatment effect and minimize damage to surrounding tissue.

Beyond treatment success ratings of craniopharyngioma related to diagnostic and treatment strategies is the experience level of the neurosurgeon.

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6/11/2017