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Childhood craniopharyngioma: Vancouver experience

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Abstract *Objective:* To present our institution's experience in the management of childhood craniopharyngioma since 1982. *Methods:* We retrospectively reviewed the records of all children diagnosed with craniopharyngioma at our children's hospital from its opening in 1982 through to 2003. One neuroradiologist systematically reviewed the neuroimaging. Kaplan–Meier curves were used to analyze the progression-free survival and the overall survival from the time of the first definitive intervention. *Conclusions:* Most children diagnosed with craniopharyngioma are long-term survivors. Survivors suffer from multiple deficits in the long term. A conservative surgical and radiotherapeutic approach and avoiding interventions that are known to cause severe morbidity may minimize these. The use of intracystic bleomycin is a strategy that allows the delay of more aggressive therapies in select patients.

Keywords Surgery · Bleomycin · Radiotherapy · Outcome · Survival · Morbidity

Introduction

Craniopharyngioma represents between 6 and 9% of primary central nervous system (CNS) tumors in childhood [2]. Although the histology is benign, the intimate relationship of the tumor with important structures such as the pituitary, optic chiasm, circle of Willis, and hypothalamus poses a major challenge in management. The management of a newly diagnosed craniopharyngioma at our institution has involved a relatively conservative surgical approach designed to control tumor growth while attempting to limit the morbidity of the treatment and the tumor. The goal at

diagnosis was to decompress the optic apparatus and to relieve intracranial hypertension from associated hydrocephalus, if such existed. For predominantly cystic tumors, consideration was given to cyst drainage for the immediate decompression, followed by instillation of intracystic bleomycin via an indwelling cyst catheter attached to a subgaleal reservoir since 1994, with the hope of delaying or avoiding a more aggressive surgical procedure or radiation. For predominantly solid tumors, open surgery was performed with the goal of partial resection unless the tumor could be dissected away from the adjacent structures readily. In patients with a mainly solid residual tumor postsurgery, adjuvant radiation therapy (XRT) was considered, especially in the older children.

Methods

We retrospectively reviewed all children diagnosed with craniopharyngioma since the opening of the British Columbia Children's Hospital (BCCH) in 1982 through to 2003. BCCH is the only tertiary care facility in the province, serving a population of 4.2 million. Cases were identified through our neurosurgical and oncology databases. Patients who were diagnosed and commenced their treatment outside the province were excluded from the analysis. The patients' charts were retrospectively reviewed to extract information about demographics, tumor diagnosis, therapy, endocrine, ophthalmologic and neurological deficits, clinical response, and long-term outcome. The neuroradiologist systematically reviewed the neuroimaging to evaluate tumor characteristics at diagnosis, response to first definitive therapy, time to progression, and tumor status at last radiological follow-up. The location of the tumor at diagnosis was evaluated specifically in regard to whether the following could be determined on imaging: ny retrochiasmatic component on midsagittal section magnetic resonance imaging (MRI), if the tumor distorted the hypothalamus on computed tomography (CT) or MRI, if the tumor reached up to the middle of the third ventricle, or if the tumor extended into the sylvian fissure. This evaluation was limited by the fact that many patients had only CT scanning at diagnosis, and in these patients, particularly, it was difficult to judge the location of the chiasm. The tumor was measured in three dimensions: AP, transverse, and craniocaudal; the volume was calculated based on the elliptical model, i.e., the product of these measurements times 0.5. The degree of resection was based on review of imaging and was graded as follows: >95%=gross total resection (GTR); 50–95%=subtotal resection (STR); 10–49%=partial resection; and <10%=biopsy (Bx). The degree of best response to radiation (XRT) or bleomycin was defined radiographically as follows: >90% reduction in volume compared with prior to therapy = complete response (CR); >50%=partial response; and >25%=minor response. Progression was defined as at least a 40% increase in tumor size or a clinical deterioration that required intervention with less than 40% increase in tumor size. Kaplan–Meier curves were used to analyze progression-free survival (PFS) and overall survival (OS) from the time of the first definitive intervention.

Results

Twenty nine children fulfilled the study criteria. Of these, 26 had neuroimaging available for review to enable evaluation of the degree of response to therapy and PFS.

Features at diagnosis

The median age at diagnosis was 7.4 years (0.6–16.2). The sex distribution was approximately equal. Twenty eight of the 29 patients were symptomatic at the time of diagnosis, with headache, vomiting, weight loss, endocrine dysfunction, or visual disturbance or combination thereof (Table 1). One patient had imaging for a head injury, and the tumor was diagnosed incidentally. Nine out of 29 had papilledema, 21 of 29 had a visual deficit, and 2 were blind at diagnosis. Eighteen had growth hormone deficiency. Twelve had multiple endocrinopathies at the time of diagnosis, and for ease of reporting, these patients were categorized as having panhypopituitarism. Twenty patients had a mainly cystic craniopharyngioma, 13 of which had single cysts. As many patients did not undergo MRI at diagnosis, the relation of the tumor to the chiasm could not be evaluated on imaging. We assumed for this study that all tumors that extended to the mid third ventricle were retrochiasmatic and involved the hypothalamus, and that all patients who had distortion of the hypothalamus were also retrochiasmatic. Based on these assumptions, 20 had hypothalamic involvement and 23 were retrochiasmatic at the time of diagnosis. Sixteen children had hydrocephalus defined by ventriculomegaly on CT or MRI, although not all had transependymal spread. Twenty five were both intrasellar and suprasellar.

Therapy

The same neurosurgeon was involved in the primary treatment in 23 of the 29 children. Seven patients had a surgical resection as the initial surgical treatment: four GTR, two STR, and one partial resection. This was immediately followed by radiation in the three with an incomplete resection. The initial surgical intervention was acute decompression of hydrocephalus or cyst drainage only in 22, and ventriculoperitoneal shunts were performed in five. Of these 22 patients, the subsequent definitive therapy was resection in five, resection and radiation in six, radiation alone in three, and bleomycin in eight. Nineteen of 22

Table 1 Clinical and radiological features at diagnosis

Code	Diagnosis date	Age	Endocrine	Visual defect	Cognitive deficits	Tumor location (hydrocephalus)	Cystic/solid/ mixed	Cyst: single/ multiple	Tumor volume (ml)
1	1982	4.15	N	VA	N	IS, SS, 3rd V, (H)	Mainly cystic	Multiple	N/a
2	1982	7.44	N/a	VA	School work[a]	N/a (H)			
3	1983	3.79	Panhypopit	VA	N	N/a (H)	Solid		
4	1983	11.24	GH, obesity	VF	N/a	IS, SS, 3rd V	Mainly cystic	Single	49.7
5	1984	4.23	Panhypopit	VF, VA	N/a	N/a			
6	1985	12.68	Panhypopit	VA	N	IS, SS, 3rd V, (H)	Mainly cystic	Single	15.3
7	1985	4.24	GH, obesity	Blind uni	N	IS, SS, 3rd V, (H)	Mainly cystic	Single	8.5
8	1986	2.98	GH	N	N	IS, SS, 3rd V	Solid		2
9	1990	14.76	Panhypopit, obesity	VF	N/a	IS, SS, 3rd V, (H), hypothalamus	Mixed	Multiple	46.5
10	1991	11.02	N	VA	N	IS, SS, (H), SF	Cystic	Multiple	227.5
11	1992	7.87	N/a	VF	N	IS, SS, 3rd V, (H)	Mixed	Single	9
12	1994	3.48	N	N	N	IS, SS, 3rd V, hypothalamus, SF	Mainly cystic	Multiple	117.2
13	1995	7.20	GH	VF, VA	N	IS, SS, 3rd V, (H), hypothalamus	Mainly cystic	Multiple	28.7
14	1995	12.00	Panhypopit	VF	N	IS, SS, 3rd V, retrochiasmatic,	Mainly cystic	Single	13.4
15	1995	0.58	N	Blind	N	IS, SS, retrochiasmatic, 3rd V, (H), hypothalamus, SF	Mainly cystic	Multiple	7.7
16	1995	2.44	N	N	N	IS, SS, 3rd V, (H)	Cystic	Single	101.8
17	1997	7.26	N	N	N	IS, SS, 3rd V, (H), retrochiasmatic	Mainly cystic	Single	10.1
18	1997	10.02	GH	N	N	IS, SS, 3rd V, (H), SF	Mainly cystic	Multiple	35.7
19	1997	14.71	Panhypopit	VF, VA	N/a	IS, SS	Cystic	Single	5.2
20	1997	7.05	GH, obesity	Blind	N/a	IS, SS, 3rd V, (H), hypothalamus	Mainly cystic	Multiple	33.5
21	1998	14.77	Panhypopit	VA, VF	N	SS, hypothalamus, SF	Cystic	Single	58.1
22	1998	5.87	N	VA	N	IS, SS	Cystic	Single	11.9
23	1999	14.07	Panhypopit	N	Learning[a]	IS, SS, retrochiasmatic	Cystic	Single	1.5
24	2001	9.22	N	VF	N	IS, SS, retrochiasmatic	Cystic	Single	109.8
25	2002	14.77	Panhypopit	N	School work[a]	IS, SS, retrochiasmatic	Mainly cystic	Single	4.3
26	2002	7.22	Panhypopit	VF, blind uni	N	IS, SS, 3rd V, retrochiasmatic	Mixed	Single	15.7
27	2003	11.47	N	N	N	IS, SS, retrochiasmatic, 3rd V, (H) hypothalamus	Mixed	Multiple	13.6
28	2003	15	Panhypopit	VF, VA	Memory[a]	IS, SS retrochiasmatic, 3rd V	Cystic	Single	20.2
29	2003	14.46	Panhypopit	VF, VA	N/a	SS, 3rd V, (H), retrochiasmatic	Mixed	Multiple	19

Abbreviations: *N* Normal, *N/a* not available, *VA* visual acuity, *VF* visual field, *Panhypopit* panhypopituitarism, *GH* growth hormone, *IS* intrasellar, *SS* suprasellar, *3rd V* third ventricle, *H* hydrocephalus, *SF* sylvian fissure, *uni* unilateral

[a]Worse

Table 2 Treatment of each patient and long-term deficits

Code	Diagnosis	1st definitive Rx (surgical intent)	Response to Rx cyst: solid: overall	LT change 2° initial Rx	Subsequent Rx (year)	Residual deficits
1	1982	GTR (partial)+10 MV photon 2F 50 Gy	PG: PG	Vision[a]	Surgery (83)	***Pan***; ***VF***; ***obesity***; ***LA***
2	1982	Partial (GTR)−films n/a		VF[b], pan	Surgery+XRT (86)	***Pan***; ***blind***; ***obesity***
3	1983	GTR-scans n/a (GTR)		Sp quad, epilepsy	Nil	Pan; ***obesity***; ***delay***[c]
4	1983	STR (safe)+photon 2F 10 MV 52 Gy	CR: CR		Nil	***Pan***; ***epilepsy***
5	1984	GTR (try GTR) films n/a for review			Surgery (85, 95); XRT (85)	***Pan***
6	1985	GTR (safe)+3-field photon 50 Gy	MR: MR	VF[b]	Nil	Pan; ***VF***
7	1985	Partial (safe)+3-field photon 50 Gy	Partial: partial		Nil	VF; obesity, ***pan***
8	1986	GTR (GTR)		Vision[a]	XRT (89), bleo (97), surgery (03)	***VF***; ***pan***; ***obesity***; ***memory***
9	1990	GTR (GTR)		DI, memory[b]	XRT (95)	N/a
10	1991	STR (safe)+3-field photon 50 Gy	PG: stable: PG		Surgery (91)	***Pan***; ***VF***; ***LA***; ***memory***[c]
11	1992	GTR (GTR)		VF[b], pan	XRT (96)	VF; ***pan***; ***obesity***
12	1994	Bleo	CR: nil		Surgery (03) + XRT (04)	***Pan***
13	1995	Bleo	Partial: nil	Pan	Surgery+XRT (96)	***Pan***; ***obesity***
14	1995	Bleo	Partial: nil	VF[a]	Nil	Pan; VF
15	1995	GTR (urgent decompression)	CR: CR	Vision[a]	Nil	VF; ***cortisol***; ***obesity***; ***cognitive***
16	1995	Bleo	MR: CR		Bleo (96, 97); surgery (98, 03); XRT (99)	***Pan***; ***blind***; ***obese***; ***cognitive***[c]
17	1997	Proton 56 Gy	MR: CR		Surgery (03)	***Pan***; ***VF***
18	1997	STR (safe)+25 MV photon 2F 50 Gy	Partial: CR	Pan	Surgery (01)	Pan; ***blind***; ***obese***
19	1997	GTR (GTR if possible)		Pan, vision[a]	Surgery (00); XRT (04)	Pan; blind; memory
20	1997	STR (safe)+proton 58 Gy	Partial: partial		Nil	VF; obesity; ***pan***; ***LA***
21	1998	Bleo	CR: nil	DI, vision[a]	Bleo (01)	Pan; VF
22	1998	Bleo	CR: CR	Vision[a]	Nil	Pan; ***VF***; ***VA***
23	1999	GTR (safe)			Nil	Pan; LA; memory
24	2001	Bleo	CR: nil	VF[b], uni blind	Bleo (01); surgery + XRT (02)	VF; ***pan***
25	2002	Stereo 54 Gy	Partial: nil	Learning[a]	Nil	Pan; ***obesity***
26	2002	Bleo	Partial: CR	DI, obesity	Nil	Pan; VF; ***obesity***
27	2003	Stereo 54 Gy	Nil: MR	Obesity	Nil	***Obesity***
28	2003	GTR (GTR)+stereo 54 Gy	PG: PG	Vision[a], DI	Nil	Pan
29	2003	Partial (safe)+stereo 45 Gy	CR: PG : partial	Pan	Nil	Pan; VF

Entries in bold italics indicate onset since diagnosis

Abbreviations: *Pan* Panhypopituitarism, *XRT* radiation therapy, *VF* visual field, *VA* visual acuity, *DI* diabetes insipidus, *N/a* not available, *Rx* therapy, *bleo* bleomycin, *stereo* stereotactic photon XRT, *PG* progression, *safe* remove what is safe and not adherent to structures that are clinically intact preoperatively, *proton* proton beam XRT, *sp quad* spastic quadraparesis, *uni* unilateral, *photon* photon beam XRT, *2F* two-field, *LT* long-term, *2°* secondary

[a]Improved

[b]Defect

[c]Severe

went on to the definitive therapy for the tumor within 1 year. Thus, nine patients had surgery alone as the first definitive therapy, three of which have not required any further tumor intervention. In those who have needed radiation, this was delayed by a median of 4 years (range 1–7). The postoperative course following resection in one patient was complicated by status epilepticus, frontal and cerebellar hemorrhage, and shunt infection resulting in severe spastic quadraparesis and subsequent epilepsy. The details of management, including extent of surgical resection, response to therapy, and long-term deficits secondary to therapy, are outlined in Table 2. All children treated with bleomycin responded in terms of tumor shrinkage, and three had a complete response. The technique of bleomycin installation has been reported previously [5]. Three of eight children who received bleomycin have not required any surgery or radiation to date, and in those who did receive radiation after bleomycin, this was delayed by a median of 3 years (range 1–9). All patients undergoing XRT received fractionated radiotherapy; however, the modality changed over time as new techniques became available to deliver more focused radiation. Nine of 12 craniopharyngiomas treated with radiotherapy as part of the initial treatment responded, two of them subsequently progressed. Three of 12 did not respond to radiotherapy. A more conservative surgical approach evolved over the years, with 63% undergoing GTR as the first definitive therapy up to 1993 and only 22% in the last 10 years. In addition, there was a trend towards decreased use of radiation in the last decade: 45% in the first decade vs 33% in the second decade. These trends coincide with the onset of the use of bleomycin therapy and despite the availability of more focused radiation in the second decade.

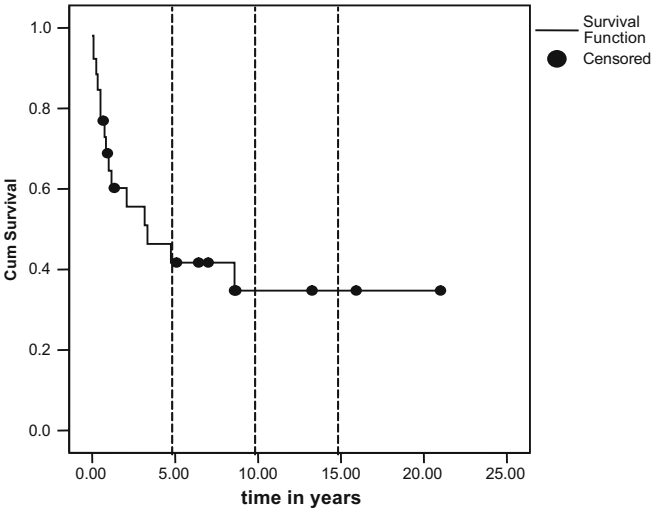

Fig. 1 Progression-free survival from first definitive therapy in children with craniopharyngioma

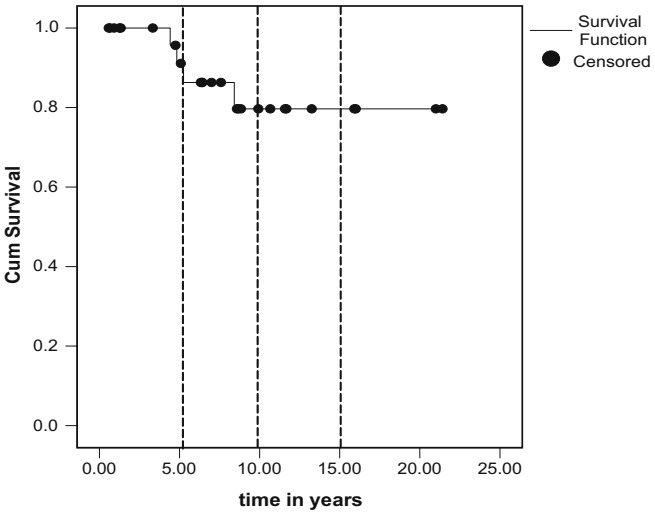

Fig. 2 Overall survival from definitive therapy in children with craniopharyngioma

Long-term deficits

All patients had resolution of raised intracranial pressure following the initial surgery. Vision improved following the first definitive therapy in 7 of 21 patients with visual impairment at diagnosis. Data regarding long-term function were available on 28 of 29 patients. Twenty six had multiple endocrine deficits at the last follow-up compared to 12 at diagnosis, and one has isolated cortisol deficiency. The single patient with no endocrine dysfunction is only 11 months post-XRT, and endocrine dysfunction will probably become apparent on longer follow-up. Fourteen of 28 are obese at follow-up compared with only four at diagnosis. Nineteen have visual impairment, and four of these are blind in both eyes. Although the number of patients with any visual impairment has decreased following therapy, eight have had permanent deterioration in their vision secondary to the tumor or its treatment. Nine have learning difficulties with or without behavioral difficulties, and in three, these deficits are considered severe. The severe cognitive dysfunction was secondary to immediate complications of GTR in two, and most likely secondary to radiation in the third.

Survival

The median follow-up was 84 months (7–257). We were able to review the scans and determine the date of progression in 26 of 29 patients. Fifteen of 26 patients had tumor progression. The survival curves are demonstrated in Figs. 1 and 2. The 5- and 10-yr PFS is 42 [95% confidence interval (CI): 22–62%] and 35% (95% CI: 14–56%), respectively. The 5- and 10-yr OS is 93 (95% CI: 79–100%) and 80% (95% CI: 61–98%), respectively.

Discussion

The management of childhood and adolescent craniopharyngioma continues to be controversial. The philosophy at BCCH has been relatively conservative in terms of both radiation and surgery in an attempt to minimize irreversible and devastating complications of surgery and the well-recognized long-term complications of radiotherapy particularly in the young child. The use of surgical decompression and, in some cases, bleomycin has permitted at least the delay of more aggressive approaches in the child with craniopharyngioma, potentially decreasing associated morbidity.

Comparison of our population with other pediatric craniopharyngioma studies shows our study group to be similar in terms of the patients' sex and age. The median age reported at presentation is 8 years (1.5–24.8) [3, 7, 11].

Reporting of deficits in children at the time of diagnosis is variable. Baskin and Wilson [1] reported 93% having growth failure. Stripp et al. [11] reported 23% having endocrine abnormalities, 58% visual impairment, and 7% obesity. Gonc et al. [4] reported that 33% had endocrine disorders, 42% optic atrophy, 34% visual field defects, 22% unilateral blindness, and 16% were obese at the time of presentation.

The incidence of hydrocephalus in other case series of childhood craniopharyngioma varies between 15 and 46% [1, 3, 4, 8]. The incidence in our series was 57% (16 of 28), which is somewhat higher than that reported in the literature. However, in three of the series reported above, several of the patients were diagnosed prior to the CT era, and their numbers regarding hydrocephalus may be an underestimate. Alternatively, it may be that our series was skewed towards larger tumors obstructing the foramina of Monro. Danoff et al. [3] report that 43% were mainly cystic, which is lower than in our series. The majority of our patients had both an intra- and suprasellar component to the tumor at diagnosis in contrast to Gonc et al. [4], who only reports 41%. Of our series, 77% had hypothalamic involvement compared with 81% in the series of Poretti et al. [8]. Hydrocephalus and hypothalamic involvement are radiological features that are thought to be associated with poor functional outcome, potentially related to a more aggressive tumor and the role that hypothalamic dysfunction plays on quality of life [6, 8]. There was no clear association in our study between these radiological features and long-term deficits in this series.

Therapy

Although OS is excellent in our current series, the morbidity related to this chronic disease of childhood remains substantial. Controversy prevails in the literature in regard to the approach that provides the least morbidity. The main split is between an aggressive surgical approach and conservative surgery with radiation given either immediately after surgery or at the time of progression. There are no recent reports wherein children were treated uniformly with limited surgery followed by immediate radiation, suggesting that even in centers where a conservative surgical approach occurs, some patients' tumors are deemed to be resectable. This also suggests consensus in delaying radiation in the young patient. There are no prospective randomized studies due to the rarity of this disease and different philosophical approaches. Current reviews include patients diagnosed as far back as the 1960s, and these might be expected to report higher morbidity and mortality related to surgery and radiation compared with patients being treated with modern techniques. We agree with Gonc et al. [4], who state that the choice of treatment should depend on various aspects: the age of the patient, the volume and localization of the mass, and quality of life considerations. We would also add that the ability to do a safe resection should guide the neurosurgeon in this non-malignant tumor. In the presence of a large cystic component to the tumor, intracystic therapies such as bleomycin should be considered, particularly in the younger child. Although recurrence rates may be higher with this approach, delaying or avoiding the potential hazards of aggressive surgery or radiation may improve the child's quality of life and functional outcome.

Stripp et al. [11] described the Philadelphia experience between 1974 and 2001, where all 76 children underwent an attempted GTR. Less than GTR was achieved in 27. The surgery was followed immediately by radiation in 18. The initial radiological features of the tumors are not described; there is no comment with respect to any change in the ability to achieve a GTR and postoperative morbidity over this large time span. Stripp et al. reported 85% 10-year OS, and 48% relapse-free survival with median follow-up of 7.6 years, similar to ours. Poretti et al [8], whose philosophy was an aggressive surgical approach to childhood craniopharyngioma, reported on 25 children treated between 1980 and 2002: GTR was achieved in 23, and STR in 2. The 10-yr OS was 92%, and the 10-yr PFS was 68%; however, schooling difficulties were reported in 50%, and impaired quality of life was reported in 75%. Thus, although the aggressive surgical approach of Poretti et al. has led to a PFS superior to ours, their data suggest worse morbidity.

In a review of 66 children diagnosed between 1969 and 1999, Gonc et al. [4] noted that 31% underwent GTR and 55% received XRT a median of 3 months after the operation. The recurrence rate was 56%, and the median time to recurrence was 12 months (2 months–18 years), with an overall mortality of 24% at a median follow-up of 5 years (0.5–24). Merchant et al. [7] retrospectively compared patients treated between 1984 and 1997 with an aggressive surgical approach (eight GTR, seven STR) versus a limited surgical approach followed by radiation (13 external beam, two intratumoral ^{32}P). However, the features of the tumor,

clinical characteristics at diagnosis, and the preoperative aim were not compared. At a median follow-up of 72 months (41–146), 11 of 30 progressed and 1 died. Twenty five percent of the patients not treated with radiation initially were less than 4 years at diagnosis, compared with only one in the radiation group. Those treated initially with radiation appeared to have a lower recurrence rate. Danoff et al. [3] described 19 children who received radiation between 1961 and 1978, of whom only three had a GTR. All patients received radiation, 14 immediately after surgery and five at relapse. At 10 years, 64% were alive, and 36% had progressive disease. These three studies [3, 4, 7] had a more aggressive radiation approach than in our series, with at least half receiving radiation early in the management. The relapse rates of 36–56% [3, 4, 7] are in keeping with ours. Our OS data is in keeping with that of recent publications [3, 4, 7, 8, 11], whether the center's thrust was an aggressive surgical approach or not.

Long-term deficits

Different authors have used different measures of long-term outcome, and there was an inconsistent reporting of baseline deficits at the time of diagnosis. However, most provide some indication of endocrine, visual and cognitive function, or disability index in the long term.

Multiple endocrinopathies are the rule, 84–97% [7–9, 11], which is in keeping with our experience. Obesity is present in 50% of our patients, compared with those with an aggressive surgical approach who report obesity ranging between 49 and 61% [8, 10, 11]. In our series, vision seems to be relatively spared compared to other series: 31% have normal visual fields, 79% normal visual acuity, compared with Merchant et al. [7] (37 and 60%, respectively), Poretti et al. [8] (42 and 46%, respectively), and Riva et al. [10] (25 and 25%, respectively).

Poretti et al. [8] reported 17% of the survivors with epilepsy, 50% with significant schooling problems, and 75% with an impaired quality of life. Riva et al. [10] reported on 12 children following radical surgery for craniopharyngioma and noted the absence of cognitive deficits and only minor attention deficits, but only 17% achieved a good academic performance. Merchant et al. [7] reported mean functional IQ in the average range, 10% had a hemiparesis, 20% had epilepsy, and one patient had devastating parenchymal necrosis following radiation. Danoff et al. [3] classified patients according to disability based on the neurological examination: 43% had a mild handicap and 21% a major handicap but were capable of self-care. In our series, 22% have mild cognitive dysfunction, 10% have moderately severe cognitive dysfunction, and 7% have epilepsy on long-term follow-up. The cognitive outcome in our series appears favorable compared to those with an aggressive surgical or radiotherapeutic approach; however, the use of a quality of life tool would be helpful in evaluating this further.

Conclusion

The Vancouver experience demonstrates that a conservative surgical and radiotherapeutic approach to the management of craniopharyngioma and the use of bleomycin results in similar OS to more aggressive approaches. Survivors suffer from multiple deficits in the long term, and avoiding interventions that are known to cause severe morbidity may minimize these. The use of intracystic bleomycin is a strategy that allows the delay of more aggressive therapies in selected patients. The management of craniopharyngioma remains a challenge, and in the absence of a multicenter clinical trial for this rare disorder, it will remain difficult to determine the best management strategy.

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