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Original Article

Surgical techniques in the management of supratentorial pediatric brain tumors: 10 years' experience at a tertiary care center in the Middle East

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ABSTRACT

Background: The goal of this retrospective study is to present the first epidemiological data on pediatric supratentorial central nervous system (CNS) tumors in Lebanon and to review the various surgical management strategies used.

Methods: We conducted a retrospective case series of all pediatric patients who presented with a supratentorial CNS tumor and underwent surgery at our institution between 2006 and 2016. We collected and analyzed demographic characteristics, tumor location, clinical manifestations, histopathology, and surgical management strategies and outcome, and discussed them after dividing the tumors as per location and in view of published literature.

Results: Ninety-nine children were studied with a male-to-female ratio of 2.3:1 and a mean age of 8.5 years. The most common location was convexity (44%) and included low-grade and high-grade glial tumors, along with other miscellaneous lesions. The next location was sellar/diencephalic (34%), including craniopharyngiomas, hypothalamic/optic pathway/thalamic gliomas, hamartomas, and pituitary/Rathke's cyst, where there was notable use of endoscopic techniques (21%). Tumors in the pineal region (13%) were tectal gliomas, germ cell tumors, and pineoblastomas and were mostly treated endoscopically. The last group was lateral intraventricular tumors (8%) and was mostly choroid plexus lesions and ependymomas. Overall, the surgical objective was achieved in 95% with mild/moderate complications in 17%.

Conclusion: A variety of pathologies may affect the pediatric population in the supratentorial region. Different surgical strategies, including microsurgical and endoscopic techniques, may be employed to remove, debulk, or biopsy these tumors depending on their location, suspected diagnosis, prognosis, and the need for treatment of possible associated hydrocephalus.

Keywords: Endoscopic neurosurgery, Microsurgery, Pediatric brain tumors, Supratentorial

INTRODUCTION

Primary central nervous system (CNS) tumors of childhood arise in 5.4 cases/100,000 person-years. This incidence is rare when compared to adulthood, but, as a whole, brain tumors continue to represent a significant source of morbidity and mortality in the pediatric population. They are

the most common solid malignancy and the leading cause of death among all childhood cancers.[49] They comprise a myriad of different tumor types, of which nearly 50% arise above the tentorium.

Pediatric supratentorial tumors form a separate entity as far as incidence, histology, and surgical approach are concerned. They predominate under the age of 3 years, whereas posterior fossa tumors are more common between the ages of 3 and 11 years. Subsequently, the incidence of supra- and infratentorial tumors is identical.[9] The largest percentage of these tumors (17%) is located within the frontal, temporal, parietal, and occipital lobes of the brain.[49] Histologically, the majority of these tumors are of neuroepithelial origin, where roughly one-quarter consist of pilocytic astrocytomas followed by malignant gliomas and embryonal tumors.[49] The presentation is dependent on the type of tumor, its location, and the age of the child.

Survival has improved for pediatric brain tumors, as a result of improved surgical techniques and rational use of postoperative radiation and chemotherapy. Management of children with brain tumors requires a multidisciplinary approach, and these children are best served at facilities with the necessary resources and personnel. While gross total resection (GTR) is the key in many of these tumors,[26,45] location of some tumors can make them unsafe to remove surgically and may negatively impact survival. In such cases, practical goals of surgical therapy may include one or more of the following: (1) obtaining a tissue diagnosis, (2) debulking the tumor, and (3) restoring normal cerebrospinal fluid (CSF) flow through ventriculoperitoneal shunt (VPS) or endoscopic third ventriculostomy (ETV).[26] The surgical approach is unique to each brain tumor. For instance, primary endoscopic procedures are increasingly becoming the preferred surgical strategy for patients with small intraventricular lesions, pineal region lesions, and cystic craniopharyngiomas.[1,6,7,48] As such, as we stratify the different supratentorial brain tumors in pediatric and adolescent patients operated at the American University of Beirut Medical Center (AUBMC) primarily according to their location, we will evaluate the variety of surgical strategies used in the management of such tumors to achieve optimal surgical outcomes and reduce postoperative complications, taking into consideration the presumed pathologies. We will also review the current literature and highlight some of the controversies associated with the surgical management of certain supratentorial tumors and emphasizing the role of neuroendoscopic techniques in minimizing surgical morbidity.

MATERIALS AND METHODS

In this study, we retrospectively reviewed all cases with confirmed diagnosis of CNS tumors admitted to AUBMC, Lebanon, from November 2006 to November 2016 and operated by the senior author. Patients aged <19 years with a histopathologic diagnosis of a supratentorial CNS tumor (primary or secondary) were included in the study. Colloid cysts, vascular malformations, and infectious lesions were excluded from the study. This study was approved by the Institutional Review Board.

Demographic, clinical, pathological, and imaging findings at diagnosis as well as the treatment modalities, their complications, and outcomes were collected. Tumor-specific data included location, pathology, goal of surgery, extent of surgery, and postoperative complications. Tumors were classified according to the 2007 WHO classification. Extent of resection was based on the postoperative magnetic resonance imaging with GTR defined as no evidence of residual tumor. The surgical approach to children with supratentorial tumors has been relatively constant through the years ranging from maximal safe surgical resection with preservation of neurological function to more conservative therapy such as a biopsy and CSF diversion. As the anatomic location of a brain tumor and its presumed pathology would influence the surgical goals set and affect the surgical approach and outcome, lesions in our study were divided into four groups according to location: convexity lesions, sellar/diencephalic lesions, pineal/tectal plate region lesions, and lateral intraventricular lesions. As such, while lesions in our study were grouped according to tumor locations, tumor histopathology of each supratentorial tumor was taken into account before any surgical intervention. We did not perform a statistical analysis due to the limited nature of the data not being amenable to a quantitative analysis. After narratively synthesizing our findings for supratentorial tumors with different locations, we reviewed the different surgical strategies used, their complications, and whether the set surgical objective (set by the senior author taking into consideration tumor location and its presumed pathology) was achieved or not.

RESULTS

A total of 99 children were operated on for a supratentorial tumor by the senior author between 2006 and 2016. The median age at diagnosis was 9 years (6 months–18 years). There was a male preponderance with male-to-female ratio of 2.3:1. Surgical procedures included diagnostic biopsy, GTR, surgical debulking, CSF diversion, and reresection for disease progression. There were frequent applications for neuroendoscopy in our series, namely, for endoscopic biopsy, endoscopic debulking/evacuation of cystic lesions, and ETV. [Table 1] shows the characteristics of patients with supratentorial tumors and the commonly encountered pathologies.

Table 1: Demographic characteristics of the surgical series of 99 pediatric patients with supratentorial CNS tumors and the commonly encountered pathologies.

Characteristic	Number (%)
Gender	
Male	69 (69.7)
Female	30 (30.3)
Common tumor pathologies	
High grade glial tumors	18%
Convexity low-grade glial tumors	15%
Craniopharyngiomas	15%
Hypothalamic/thalamic/OPG	11%
Ependymomas	8%
Tectal plate gliomas	6%
Hamartomas	5%
Germ cell tumors	4%
Choroid plexus tumors	4%

OPG: Optic pathway glioma

Convexity tumors

The cerebral hemispheres were the commonest tumor site in our series (n = 44 or 44%). Various types were found including low-grade glial and neuronal-glial tumors (34%), high-grade glial and neuronal-glial gliomas (40%), and miscellaneous lesions including ependymoma, ATRT, Ewing's sarcoma, and embryonal tumors (25%). The low-grade glial tumors included oligodendrogliomas, pilocytic astrocytomas, gangliogliomas, and DNETs, and presented mostly with seizures (80%) and less so with headaches (13%) or focal deficit (7%). The high-grade gliomas included glioblastomas, anaplastic oligodendrogliomas, and anaplastic ependymomas, and in contrast to their low-grade counterparts, presented mostly with headaches and vomiting (50% and 25%, respectively), compared to seizures (13%). The miscellaneous lesions had a variable presentation.

GTR was the primary goal in all tumors except for three patients who underwent a diagnostic biopsy because of extensive or nonsurgical disease. Tumor removal was performed with the aid of neuronavigation, neurophysiological monitoring, and histopathological monitoring with frozen sections whenever applicable. The surgical objective was achieved in all the patients in this group. None of these patients experienced any morbidity including transient postoperative events (seizures, hemiparesis, or sensory deficits) or permanent deficits (hemiparesis, hemianopsia, or sensory deficits). Two patients continued to have seizures (5%).

Sellar/diencephalic tumors

The next common location in our cohort was sellar/diencephalic region (n = 34, 34%), where the lesions were distributed among craniopharyngiomas (44%), hypothalamic/optic pathway/thalamic gliomas (32%), hamartomas (15%), and pituitary/Rathke's cyst (9%). These presented mostly with visual and endocrine-related symptoms, except for the hamartomas presenting exclusively with intractable epilepsy.

Among the 15 craniopharyngiomas operated, 6 were mostly solid, whereas 9 were predominantly cystic (including some cystic recurrences) and presented with visual symptoms in 47%, pituitary insufficiency in 27%, and progression of disease on imaging in 34% of patients. These surgeries were performed on nine children, which meant that several procedures were done for recurrent disease (10/15 or 66% of the procedures). The solid tumors underwent microsurgical resection, with total/near-total removal in five patients (three of whom were recurrent) and partial removal in one. Hypopituitarism and recurrent disease complicated these difficult tumors, with one patient suffering mild hemiparesis after total removal of his recurrent tumor. There were no perioperative mortalities.

As for the cystic tumors, a variety of less invasive techniques were used: Ommaya reservoir and intracystic treatment in four patients; endoscopic biopsy in two patients; endoscopic transsphenoidal surgery in three patients with recurrent cystic disease; and microsurgical removal in one patient. These patients had minimal complications (failure of Ommaya reservoir placement in one patient, and mild hypothalamic syndrome in the microsurgical resection surgery). Of noteworthy importance, of the two patients who underwent transventricular endoscopic biopsy/fenestration, one had remission for 2 years and later had microsurgical removal, whereas the other is still in remission with collapse of the cyst and no other intervention for the past 4 years [Figure 1]. The immediate surgical objective was thus achieved in 13/15 surgeries (87%) for craniopharyngiomas, with complications mostly related to hypopituitarism, and endoscopic techniques carrying less morbidity when done for cystic tumors.

Optic pathway glioma was the second most common entity in this area (n = 11). Initial manifestations were diverse at the time of diagnosis and included weakness (45%), visual impairment (27%), and headache (27%). The role of surgery was very limited in this subset of tumors and included biopsy and/or debulking surgery (endoscopic biopsy/debulking in three patients, stereotactic biopsy in four, and debulking cranial surgeries in four). The surgical objective was achieved in all patients. No postoperative complications were encountered. Postoperative adjuvant therapy was needed for all patients with chemotherapy being the chief modality of treatment with carboplatin/vincristine used as a first-line therapy in all of our patients.

Hamartomas presented with intractable seizures, and three patients underwent microsurgical resection/disconnection,

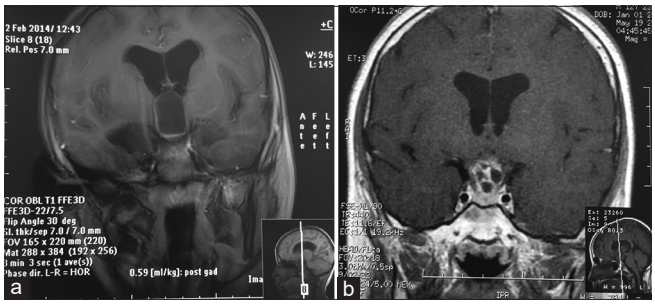

Figure 1: (a) T1W coronal MRI section with contrast showing a large cystic lesion occupying the third ventricle. (b) After transcortical endoscopic biopsy and fenestration of the cystic craniopharyngioma, the cyst collapsed and has been stable for 4 years with no symptoms and no treatment.

with one having residual tumor. Endoscopic disconnection was performed for that patient and to another patient with excellent results. Surgical objective was initially achieved in 80% of hamartoma surgeries. As for pituitary tumors, one child with decreased vision secondary to a growing macroadenoma had partial resection, with his second surgery done for progression of the residual complicated by leak and decreased vision, although GTR was achieved. The surgery for Rathke's cyst was uneventful.

Pineal region and tectal plate lesions

There were 13 surgeries for lesions in pineal/tectal plate region. These were mostly associated with hydrocephalus and presented with gait disturbances (30%), headache (30%), and diplopia (30%). There were 6 tectal gliomas (46%), 4 germ cell tumors (GCTS) (31%), and 3 pineoblastomas (23%). Endoscopic techniques were used in all these lesions, where typical tectal gliomas were treated by ETV only (n = 3, 2%), but that were combined with endoscopic biopsy in all other lesions. Although the biopsy was inconclusive (small sample size) in one child with germ cell tumor, the diagnosis was achieved through markers, and the surgical objective was thus achieved in all patients (including treatment of associated hydrocephalus).

Lateral intraventricular tumors

The last group was lateral intraventricular tumors (n = 8, 8%) and was mostly choroid plexus lesions and ependymomas, presenting mostly with signs and symptoms of hydrocephalus including vomiting (37%), macrocephaly, ataxia, and syncope (25% each). These were treated microsurgically, achieving the surgical objective in all, but with a notable high rate of CSF-related complications (hygromas and persistent hydrocephalus) necessitating VPS placement (50%).

Overall, the surgical objective was achieved in 95% of the patients, and mild/moderate complications were noted in 17% of the patients, mostly related to CSF circulation and pituitary insufficiency. Although microsurgery is the mainstay of surgical strategy, endoscopic techniques have been frequently and successfully used, especially in tumors in the anterior third ventricle, pineal, and tectal plate regions (n = 23, 23%).

As for the long-term follow-up, and in view of the heterogeneous pathologies seen in our series of supratentorial pediatric brain tumors, we studied available data for one of the most common pathologies seen across the described locations, namely, low-grade glial and glioneuronal tumors. There were 28 such lesions in convexity, tectal, and diencephalic locations, and carried an excellent prognosis. Two children had neurofibromatosis (NF1). Long-term data were available on 22/28 children, with a mean follow-up of 61 months, and progression/recurrence seen in four tumors in diencephalic location, controlled with chemotherapy and/or radiation. There were no recurrences nor progression in the tectal nor convexity low-grade gliomas and gangliogliomas.

DISCUSSION

The breadth of tumors that can arise in the supratentorial region in children is extensive. These tumors can present with a variety of different sizes, locations, and histology, and each individual tumor has distinct features that favor one specific surgical approach over another. Radical surgery is not justified in all cases, especially with adjunct treatment modalities now readily available.

To the best of our knowledge, this is the first case series of pediatric supratentorial CNS tumors in Lebanon (99 children). In contrast, a series of 64 children with infratentorial tumors were recently reported at the same institution over an approximately similar period of time (2006–2018) and involving the same senior author, conferring that supratentorial pediatric brain tumors may be twice as common as their infratentorial counterparts.[44] Results from our 10-year study showed a slight predominance of males compared to females, with a male-to-female ratio of 2.3:1, and a median age of 9 years (in contrast to M: F ratio of 1.5:1 and mean age of 6 years in infratentorial tumors). Most commonly, these tumors were located in a convexity location (44%), followed by anterior third ventricle, sellar and suprasellar region (34%), pineal and tectal plate region (13%), and lateral intraventricular location in 8%. Presentation varied with location and the grade of the tumor; while seizures were a common presentation in benign convexity gliomas, their malignant counterparts presented more with headaches and vomiting. Anterior third ventricle and sellar tumors presented with hypopituitarism and visual symptoms, whereas pineal region and lateral ventricular tumors presented mostly with signs and symptoms of hydrocephalus.

The commonly encountered pathologies were high-grade glial tumors (18%), convexity low-grade glial tumors (15%), craniopharyngiomas (15%), hypothalamic, thalamic, and optic pathway gliomas (11%), ependymomas (8%), tectal plate gliomas (6%), hamartomas (5%), GCTs (4%), and choroid plexus tumors (4%) [Table 1].

The surgical objective was achieved in most surgeries overall (95%). Microsurgical resection of tumors such as convexity gliomas and lateral intraventricular tumors remains the mainstay of their management, and this was achieved in all these cases. Certain tumors, however, pose a more difficult surgical and therapeutic challenge: craniopharyngioma resective surgery may be associated with significant pituitary/hypothalamic morbidity, with a high recurrence rate. Optic pathway gliomas and GCTs have several nonsurgical therapies, and in tectal gliomas, the mere treatment of hydrocephalus may be sufficient. We will thus shed some insight into some of the controversies associated with the management of these lesions, in view of our results, and highlight the role of neuroendoscopic techniques as part of the neurosurgical armamentarium.

Craniopharyngiomas

Childhood-onset craniopharyngioma is often diagnosed late, manifesting with symptoms of increased intracranial pressure; other common manifestations include impairments in the endocrine system and vision.[46] These manifestations were also seen in our study, where 47% presented with visual symptoms, 20% with endocrinological deficits, such as failure to thrive and hypopituitarism, and 20% with symptoms indicative of increased intracranial pressure. Recurrence rate was also high with 33% of the patients presenting with progression on follow-up MRI.

The surgical management of craniopharyngiomas in children remains a controversial topic for neurosurgeons. Resection may be radical or limited. Theoretically, the benign histology implies that GTR should be sufficient to provide a cure. However, this strategy has been associated with high morbidity and mortality as well as a high recurrence rate.[15,28,67] Most treatment strategies nowadays aim to decompress optic structures, reduce mass effect, restore CSF flow, and facilitate tumor control keeping postoperative morbidity to a minimum.[39,47] Various grading systems and classifications may serve as a guide based on tumor location and its relation with the hypothalamus.[53] Patients may be selected for less than total resection followed by radiation therapy (RT) based on the neurosurgeon's evaluation that a GTR cannot be achieved with acceptable morbidity. Limited surgery can also be used to reduce the volume targeted by irradiation and better visualize the distance between the targeted and normal tissue fields.[20,39,41,53,57,64] RT is typically focal, including conformal or proton beam RT, and there are a number of published series reporting high rates of tumor control with long-term follow-up.[31,47] It carries a substantial risk of long-term complications, however, such as cognitive impairment, visual deterioration, and hypothalamic-pituitary dysfunction.[31]

Surgery for craniopharyngioma may also involve cyst drainage and/or management of hydrocephalus through endoscopic or catheter placement procedures when presenting with poor vision. Commonly, and especially in large cystic tumors, a catheter may be inserted into the cyst and permanently attached to an Ommaya reservoir in preparation for adjuvant therapy such as intracystic interferon-alpha, which has been shown to delay disease progression and has more of a favorable side effect profile as opposed to bleomycin or radiotherapy agents.[14,32] VPSs are preferably avoided as first-line treatment due to potential risk of dysfunction and overdrainage.[51] Different surgical approaches can be performed on the basis of the hypothalamic involvement with endoscopic transsphenoidal approaches for tumors with no involvement; endoscopic transsphenoidal or transcranial approaches for tumors in contact with the hypothalamus; and transcallosal, pterional, or unilateral/bilateral subfrontal transcranial approaches for tumors invading the hypothalamus.[2,11,16,37]

In our study, microsurgical debulking/near-total removal and radiation were done for four children with predominantly solid tumors. One patient had a cystic recurrence treated with intracystic interferon infusion, only to relapse few years later, where microsurgical total removal was complicated with hemiparesis. Another child also had a recurrence after 3 years, operated again by microsurgical excision. The cystic lesions were mostly treated through more conservative approaches with Ommaya reservoir placement and intracystic P^{32} or interferon in a total of three children. As intracystic treatment agents became less available, two children with large cystic craniopharyngiomas had transventricular endoscopic fenestration/biopsy, and Ommaya reservoir was placed in one of them. Both cysts collapsed into small sellar lesions, and one stayed in remission till the date of this report (4 years) with no other treatment and no symptoms or endocrine-related abnormalities, as illustrated earlier in [Figure 1], supporting the minimally invasive approach advocated for mostly cystic lesions. The other cyst collapsed for 2 years and then recurred, and as adjustment of the previously placed Ommaya reservoir failed, microsurgical removal of the cystic tumor was done with postoperative moderate hypothalamic dysfunction symptoms. Three children with mostly recurrent sellar cystic craniopharyngiomas and hypopituitarism were operated through endoscopic transsphenoidal debulking, followed by focal radiation, with good results.

In view of the high morbidity associated with surgical treatment of craniopharyngiomas, and the significant

associated recurrence rates, we propose an algorithm for the management of these tricky lesions. Different surgical approaches may be used depending on whether these lesions are predominantly cystic or solid, recurrent, or 1st time presentation, and whether they have pituitary insufficiency or not. A child who presents with a predominantly large solid lesion is best managed by microsurgical removal/debulking followed by RT. Solid recurrence is best managed by gross microsurgical removal and may be removed transsphenoidally if sellar in location, especially when hypopituitarism is already present. Transventricular endoscopic biopsy and fenestration of cystic lesions filling the 3rd ventricle are advised, especially when they are large and associated with hydrocephalus. The cyst usually slowly collapses away from hypothalamic wall into a small lesion, which can be observed (preserving pituitary function), or removed or irradiated on progression. If the cystic lesion is more inferior or sellar without hydrocephalus, it may be managed with Ommaya reservoir placement and intracystic treatment or transsphenoidal removal (especially if hypopituitarism is already present such as in cystic recurrences).

Pineal region tumors

The spectrum of tumor types that may arise in the pineal area necessitates a histologic diagnosis for optimal treatment planning. Tumors are divided into four categories: GCTs (50–75%), pineal parenchymal cell tumors (14–27%), glial cell tumors, and miscellaneous lesions.[19,27,38,40,65] CSF cytology and radiographic characteristics may provide insight into the diagnosis, but are not sufficiently sensitive to supplant tissue. Surgery in this region may be among the most arduous of microsurgical challenges. Tissue diagnosis can be achieved by either stereotactic or endoscopic transventricular biopsy or open surgical resection. Endoscopic transventricular biopsies are preferred for tumors extending to the posterior third ventricle, because this approach grants more access to the target tissue and may also treat hydrocephalus through ETV.[25,48] Large pineal tumors may only require stereotactic measures.[25]

In our study, and for seven children with pineal region tumors, ETV to treat hydrocephalus was done in six patients, with shunt placement performed in one patient where ETV was not possible. Endoscopic biopsy was attempted in all patients and was diagnostic in six children, with the diagnosis obtained through markers in one child where the specimen was too small for pathologic examination. GCTs accounted for 4/7 children (57%), whereas the rest were pineoblastomas (43%).

The management of pineal tumors depends on the histopathology of the tumor, as well as serum and CSF markers. Nongerminomatous GCTs secrete specific proteins, such as alpha-fetoprotein (AFP) and beta-human chorionic gonadotropin (b-hCG), which are beneficial in determining prognosis and response to treatment.[8,35,54] Nongerminomatous cell tumors, such as yolk sac tumors, mixed GCTs, and embryonic carcinomas, generally express AFP. While b-hCG is mainly expressed in choriocarcinomas, GCTs generally have low b-hCG expression levels but are often placental-like alkaline phosphatase-positive. Nongerminomatous GCTs are aggressive tumors which require a combination of chemotherapy followed by craniospinal irradiation.[33,55] Germinomatous GCTs, however, are radiosensitive but adjuvant chemotherapy is incorporated to reduce the long-term consequences of radiotherapy alone.[30,56] As such, surgery does not play a significant role in the management of GCTs except for the treatment of obstructive hydrocephalus associated with these GCTs, requiring ETV or insertion of a VPS.[24,60] There may be a role for second-look resection in patients with incomplete response to distinguish residual disease from growing teratoma syndrome.

Surgical resection, however, may be necessary for other classes of pineal region tumors, such as pineocytomas, meningiomas, hemangioblastomas, and other relatively benign tumors. Patients with pineocytomas can undergo GTR if the tumor is enlarging, but must be followed up on a regular basis if near-total resection is performed.[12] Although patients with aggressive pineoblastomas used to undergo surgical resection followed by chemotherapy and RT, more recent trends favor adjuvant treatment followed by possible resective surgery.[18,21,22,62] The two main surgical approaches for gaining access to the pineal region are the infratentorial supracerebellar approach and the occipital transtentorial approach. The former is preferred for smaller midline tumors located in the posterior third ventricle whereas the latter is favored for larger tumors extending to the pulvinar thalami.[23,59]

Since management of pineal region tumors is heavily dependent on their pathology, and since they present mostly with hydrocephalus, endoscopic techniques to treat hydrocephalus and obtain tissue diagnosis along with serum and CSF markers have become the mainstay surgical strategy for tumors in that location.[48,58] A VPS is not initially advised since the reduction of ventricular size will render endoscopic surgery more difficult but may be done through the same burr hole used for endoscopic biopsy, should ETV be technically not possible (i.e. tumor filling anterior third ventricle floor). ETV is also performed to treat hydrocephalus secondary to tectal gliomas with a very high success rate. A biopsy is not needed for the typical lesion, but may be done at the same time for some lesions to rule out a higher-grade tumor.

Optic pathway tumors

The optimal treatment of optic pathway gliomas (OPGs) continues to be an area of controversy in contemporary

literature. The main management decision is determining when and what type of therapy, if any, should be administered. OPGs can be managed by surveillance, surgical resection, RT, and/or chemotherapy. The decision to actively treat a patient is based on the presence of marked symptoms (i.e. proptosis or ocular pain) and/or deterioration of visual acuity and/or imaging findings. When active intervention is indicated, chemotherapy is usually administered first. Disagreement continues over the role of surgery in the management of OPG. Complete resection is only feasible when the tumor is confined to the optic nerve and associated with complete blindness. In chiasmatic and hypothalamic gliomas, radical surgery carries the risk of damage to the visual apparatus, hypothalamus, and vascular structures. Surgery for tumor partial tumor removal has been advocated when there are marked visual symptoms, hydrocephalus, or significant mass effect, and since GTR is usually not possible, adjuvant chemotherapy and radiation are usually necessary to stabilize residual disease as has been seen in our patient series.[4,10,29,34,36,42,63,66] Surgery may also be done when imaging is inconclusive for the diagnosis of OPG, and tissue diagnosis may be then needed. We have noted only one case of associated NF1 in our OPG series, as such as association, when present, indicates the diagnosis, and narrows the indications for surgery. A number of polychemotherapy regimens have been used.[3,50,52] In most series, the 5-year event-free survival (EFS) is 30–40%. As a consequence, many children require more than 1 line of chemotherapy. The role of radiotherapy has decreased overtime. This treatment is essentially considered as a salvage option, although some physicians still use this modality early in the management of older patients. Studies have reported 5-year EFS of 48–100%. These results usually compare favorably with chemotherapy. However, such comparison is flawed, due to the fact that the population groups differ significantly between RT and chemotherapy series (median age 8–10 years vs. 3–5 years, respectively).[5,13,44,61] The use of Gamma Knife radiosurgery has also been shown to be an effective treatment modality for OPG with EFS of 83% at 3 years of follow-up.[17] Nevertheless, there has yet to be a well-established and effective management approach for these tumors.

CONCLUSION

A variety of benign and malignant pathologies may affect the pediatric population in the supratentorial region of the brain. Different surgical strategies may be employed to remove, debulk, or biopsy these tumors depending on their location, suspected diagnosis, prognosis, and the need for treatment of possible associated hydrocephalus. After having reviewed our surgical series over 10 years, we have shed light on these various pathologies and their surgical management strategies, and have shown that we were able to achieve our set surgical goals in most of our patients and have proven that in addition to known microsurgical techniques for tumor removal, endoscopic procedures may be very valuable in achieving the desired surgical goals with minimal morbidity.

Strengths and limitations

To the best of our knowledge, this is the first case series in the region demonstrating surgical management strategies of supratentorial tumors in the pediatric population. We demonstrated that we were able to achieve our set surgical goals in most of our patients. However, this study has certain limitations. Our surgical goals were set according to tumor location and presumed pathology in line with the senior author's experience, and in view of the varied pathologies and the retrospective nature of the case series, were not assessed by an independent observer. While we narratively synthesized our findings, we were not able to perform a statistical analysis due to the nature of the data. There was also no standardized treatment or follow-up algorithm used in this study, but management decisions were made according to the senior author's medical expertise and judgment.

Declaration of patient consent

Institutional Review Board (IRB) permission obtained for the study.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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