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REVIEW

Management strategies for pediatric patients with tectal gliomas: a systematic review

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Abstract

Pediatric tectal gliomas generally have a benign clinical course with the majority of these observed radiologically. However, patients often need treatment for obstructive hydrocephalus and occasionally require cytotoxic therapy. Given the lack of level I data, there is a need to further characterize management strategies for these rare tumors. We have therefore performed the first systematic review comparing various management strategies. The literature was systematically searched from January 1, 2000, to July 30, 2020, to identify studies reporting treatment strategies for pediatric tectal gliomas. The systematic review included 355 patients from 14 studies. Abnormal ocular findings—including gaze palsies, papilledema, diplopia, and visual field changes—were a common presentation with between 13.6 and 88.9% of patients experiencing such findings. CSF diversion was the most performed procedure, occurring in 317 patients (89.3%). In individual studies, use of CSF diversion ranged from 73.1 to 100.0%. For management options, 232 patients were radiologically monitored (65.4%), 69 received resection (19.4%), 30 received radiotherapy (8.4%), and 19 received chemotherapy (5.4%). When examining frequencies within individual studies, chemotherapy ranged from 2.5 to 29.6% and radiotherapy ranged from 2.5 to 28.6%. Resection was the most variable treatment option between individual studies, ranging from 2.3 to 100.0%. Most tectal gliomas in the pediatric population can be observed through radiographic surveillance and CSF diversion. Other forms of management (i.e., chemotherapy and radiotherapy) are warranted for more aggressive tumors demonstrating radiological progression. Surgical resection should be reserved for large tumors and/or those that are refractory to other treatment modalities.

Keywords Tectal gliomas · Management · Systematic review · Pediatric

Introduction

Tectal gliomas are rare tumors of the midbrain often found in the pediatric population. Although tectal gliomas can encompass a variety of histologically distinct tumor types, the majority of these tumors are low-grade astrocytomas [4, 13, 14, 27, 29]. Due to the proximity of the tectal plate to the cerebral aqueduct, patients with tectal gliomas often suffer from obstructive hydrocephalus [4, 7, 11, 13, 27, 29, 32]. Accordingly, patients with tectal gliomas may present with increased intracranial pressure (headache, nausea, and vomiting) and/or Parinaud's syndrome, which is characterized by upgaze paralysis, light-near dissociation, convergence retraction nystagmus, and Collier's sign/lid retraction [2, 4, 7, 11, 13, 24, 27, 29, 32]. Given the indolent nature of tectal gliomas, management of most tumors involves radiological monitoring and potential treatment of symptomatic hydrocephalus via cerebrospinal fluid (CSF) diversion [2, 4, 10, 13, 27]. The most common methods for achieving CSF diversion are endoscopic third ventriculostomy (ETV) and ventriculoperitoneal shunt (VPS) [2, 7, 10, 11, 13, 24, 27, 32]. In cases of radiologically progressing tumors, other treatment modalities that target the tumor itself—such as radiotherapy, chemotherapy, or surgical resection—may be utilized [7, 14, 29]. Although there are studies in the literature that have attempted to investigate the outcomes of less utilized treatment options in the management of tectal gliomas, these studies are few in quantity.

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Given the rarity of tectal gliomas—especially the progressive, high-grade subset of tumors—a limited number of studies exist that compare the various tectal glioma management options including surgical resection, biopsy, chemotherapy, and radiotherapy in a pediatric population [10, 13, 14, 27]. Herein, we perform the first comprehensive systematic review of the literature to compare the presentation, frequency of management options, and outcomes in these patients. In addition to quantifying the frequency of use for each management option, we discuss indications for treatment selection and the outcomes of such treatments.

Methods

Data sources and search strategies

The systematic review was performed according to Preferred Reporting Items for Systematic Reviews and Meta-Analyses (PRISMA) guidelines [19]. We conducted a comprehensive search of several databases including MEDLINE via PubMed, Embase, and Scopus from 2000 to August 7, 2020. We only included studies available in English.

The search strategy was designed and conducted by an experienced librarian (C.J.B.) with input from the study's investigators (M.M.J.B., A.R.B). Controlled vocabulary supplemented with keywords was used to search for studies describing management options for pediatric tectal gliomas.

Study selection

Abstracts were independently screened by two study authors (M.M.J.B., C.R.) for studies that discussed the management of tectal gliomas with discrepancies resolved by two other study authors (A.R.B, C.Z.). Studies were excluded from our systematic review based on the following criteria: (1) data for which adult tectal gliomas were reported; (2) pediatric patients were included in the study, but not separately reported on; (3) no discussion of management options; (4) case series with less than 3 patients. Full-text articles were screened and abstracted for outcomes of interest by one study author (M.M.J.B).

A total of 96 abstracts were identified using the search strategy described above after removing duplicates, of which 44 were excluded because they did not report outcomes in our study population of interest. Among the 52 remaining abstracts, their full-text articles were assessed for eligibility and 38 were excluded because the authors did not investigate management options, report separate pediatric data, or had $n < 3$ (Fig. 1).

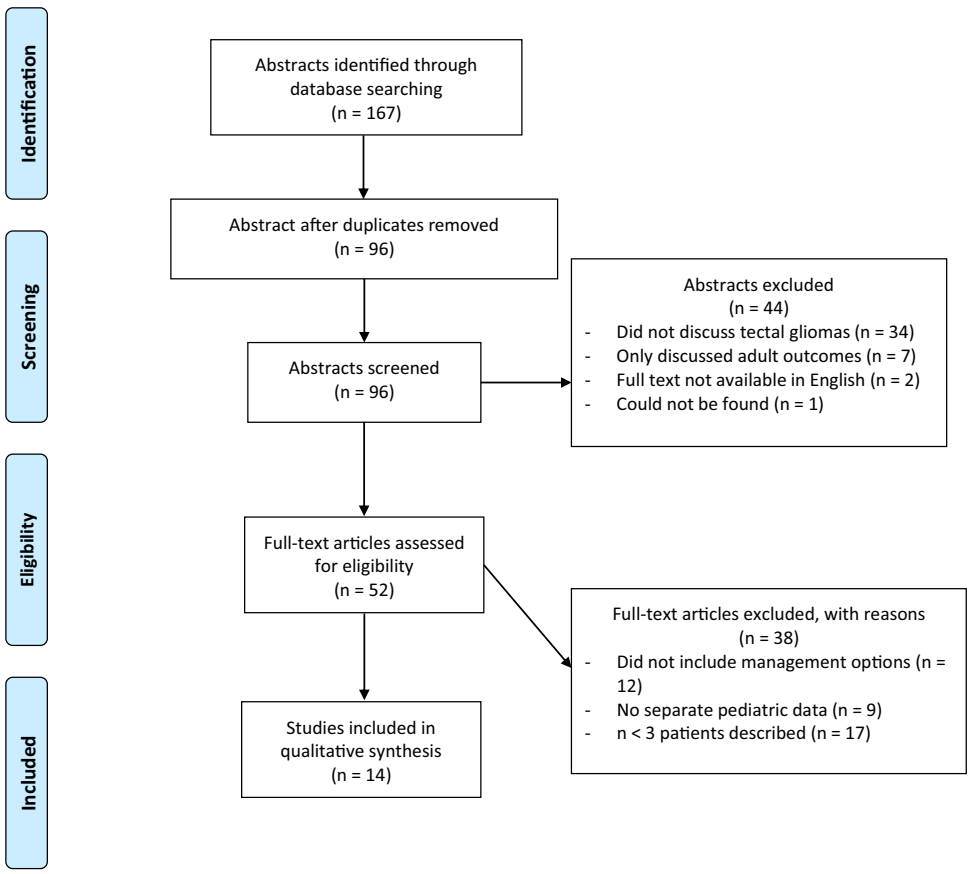

Fig. 1 Search strategy employed using the systematic review process to identify studies to be included in our study

Due to the heterogeneity of the included studies, a formal meta-analysis was deferred at this time. The results of the studies, including number of patients, histological findings, and types of procedures, were reported using descriptive statistics.

Results

Clinical presentation and radiographic features

A total of 14 manuscripts describing 355 patients were included in our study as shown in Table 1 [2, 4, 7, 10, 11, 13, 14, 16, 20, 23, 24, 27, 29, 32]. Study sizes ranged from 6 to 44 patients and included patients with ages ranging from 0 to 20 years old. Clinical presentation varied among the different cohorts. Abnormal ocular findings—including gaze palsies, papilledema, diplopia, and visual field changes—were identified in 129 patients (36.3%). Individual studies noted between 13.6 and 88.9% of patients experienced one or more ocular abnormalities. When examining radiological features upon initial diagnosis, there was variation between individual studies concerning the enhancement pattern of the tumors. A total of 78 patients (22.0%) were noted to have contrast enhancement on magnetic resonance imaging (MRI). Four studies reported no contrast enhancement within their cohort [4, 11, 23, 27]. In the remaining studies, tumor enhancement accounted for 14.3 to 77.8% of the tectal gliomas in individual cohorts.

CSF diversion

CSF diversion was utilized in 317 patients (89.3%). Within each individual study, the percentages of patients receiving CSF diversion ranged from 73.1 to 100.0%. In four studies, CSF diversion was the only procedure performed, and patients did not receive any tumor-directed treatment [4, 11, 16, 32]. However, multiple patients required additional CSF diversion surgeries as reported in nine studies. Within the systematic review, 85 patients were reported as needing a secondary CSF diversion procedure (23.9%). When examining individual studies, between 15.4 and 66.7% of the patient cohorts required correction to their prior ETV or VPS.

Tumor management strategies

Radiological monitoring (i.e., no surgery, chemotherapy, and/or radiotherapy) was utilized in the vast majority of patients. In the systematic review, 232 cases were radiologically monitored (65.4%). When examining individual cohorts, four studies only utilized radiological monitoring throughout the clinical course without any direct tumor treatment [4, 11, 16, 32]. On the other hand, two studies medically and/or surgically treated the tectal gliomas in all patients and did not employ radiological monitoring as a management strategy [20, 23]. Within the remaining studies, radiological monitoring as a management strategy was utilized in 22.7 to 90.9% of the patient cohort.

Three studies did not acquire biopsies from any of their patients [4, 16, 32]. In the remaining studies, biopsies were obtained from 9.1 to 100.0% of the patients within a cohort. A total of 117 tumors were histologically classified. The most common histological classifications of the tumors were pilocytic astrocytoma with 44 cases (37.6%), diffuse astrocytoma II with 30 cases (25.6%), and low-grade (non-pilocytic) glioma with 15 cases (12.8%). The frequency of the histopathological diagnosis greatly varied between the individual studies as shown in Table 1. Within individual studies, the frequency of pilocytic astrocytomas ranged from 20.5 to 100%. Low-grade astrocytomas were identified in four studies [11, 14, 23, 29] and constituted between 7.1 and 100% of the included tumors within the studies. Diffuse, infiltrating astrocytomas ranged from 16.7 to 100% of patients in the studies. Molecular analyses of the tectal gliomas were not reported in any of the included studies.

Compared to the other treatment options, resection showed the greatest variation in usage. A total of 69 patients received resection of their lesion (19.4%) as documented in eight studies. In the individual studies, the frequency of the resection ranged from 2.3 to 100.0%. On the other hand, only five studies utilized chemotherapy and had the lowest frequency of usage among the treatment modalities. In the systematic review, a total of 19 patients received chemotherapy (5.4%). When examining individual studies, the frequency of chemotherapy usage ranged from 2.5 to 29.6%. Similarly, radiotherapy usage ranged from 2.5 to 28.6% within the 10 patient cohorts. There was a total of 30 patients that received radiotherapy (8.4%), thereby making it the second most common treatment modality, behind resection.

Outcomes

Radiological progression was reported in 10 studies for a total of 104 patients (29.3%). In these studies, between 9.7 and 85.7% of the individual patient cohorts experienced radiological progression. Although nearly a third of all included patients experienced radiological progression, few patient deaths were reported. Four studies reported no patient deaths in their cohorts [2, 10, 27, 32]. Four studies reported death in 2.5 to 18.5% of their patient populations [13, 20, 23, 29] for a total of 9 patients. The remaining studies did not include details about the survival of their patient cohorts.

Table 1 Patient characteristics, procedures, and outcomes from the studies included in the systematic review

Ref	Number of Pxs	Range of Px age (years)	Enhancing tumors	Abnormal ocular findings	CSF diversion	Second CSF diversion	Radiological monitoring
Bowers [2]	$n=7$	3.3–16.7	$n=1$ (14.3%)	$n=4$ (57.1%)	$n=7$ (100.0%)	NR	$n=6$ (85.7%)
Daglioglu [4]	$n=9$	6–17	$n=0$	$n=8$ (88.9%)	$n=9$ (100.0%)	$n=6$ (66.7%)	$n=9$ (100.0%)
Gass [7]	$n=26$	1–17	$n=5$ (19.2%)	$n=7$ (26.9%)	$n=19$ (73.1%)	$n=5$ (26.3%)	$n=20$ (76.9%)
Greissenauer [10]	$n=44$	2–19	$n=8$ (18.2%)	$n=6$ (13.6%)	$n=36$ (81.8%)	NR	$n=40$ (90.9%)
Javadpour [11]	$n=6$	9–19	$n=0$	$n=4$ (66.7%)	$n=6$ (100.0%)	$n=4$ (66.7%)	$n=6$ (100.0%)
Kaufmann [13]	$n=71$	0.1–17.5	$n=25$ (35.2%)	$n=25$ (35.2%)	$n=63$ (88.7%)	$n=20$ (31.7%)	$n=41$ (57.7%)
Kershenovich [14]	$n=40$	0.3–18	NR	$n=16$ (40.0%)	$n=35$ (87.5%)	NR	$n=26$ (65.0%)
Li [16]	$n=31$	6 weeks–20	$n=8$ (25.8%)	$n=9$ (29.0%)	$n=31$ (100.0%)	$n=19$ (61.3%)	$n=31$ (100.0%)
Mottolese [20]	$n=27$	2–16	$n=21$ (77.8%)	$n=11$ (40.7%)	$n=20$ (74.1%)	NR	$n=0$
Ramina [23]	$n=7$	8–17	$n=0$	$n=4$ (57.1%)	$n=7$ (100.0%)	NR	$n=0$
Romeo [24]	$n=22$	4–18	NR	$n=7$ (31.8%)	$n=22$ (100.0%)	$n=5$ (22.7%)	$n=5$ (22.7%)
Stark [27]	$n=12$	4 weeks–16	$n=0$	$n=4$ (33.3%)	$n=12$ (100.0%)	$n=7$ (58.3%)	$n=9$ (75.0%)
Ternier 29]	$n=40$	0–17	$n=10$ (25.0%)	$n=18$ (45.0%)	$n=37$ (92.5%)	$n=17$ (45.9%)	$n=26$ (65.0%)
Wellons [32]	$n=13$	4–16	NR	$n=6$ (46.2%)	$n=13$ (100.0%)	$n=2$ (15.4%)	$n=13$ (100.0%)
Ref	**Biopsy**	**Histological findings**	**Resection**	**Chemotherapy**	**Radiotherapy**	**Radiological progression**	**Px death**
Bowers	$n=3$ (42.8%)	Infiltrating, non-pilocytic astrocytoma ($n=3$, 100.0%)	$n=0$	$n=0$	$n=1$ (14.3%)	$n=6$ (85.7%)	$n=0$
Daglioglu	$n=0$	NA	$n=0$	$n=0$	$n=0$	NR	NR
Gass	$n=3$ (11.5%)	Diffuse astrocytoma II ($n=1$, 33.3%); Pilocytic astrocytoma ($n=2$, 66.7%)	$n=0$	$n=4$ (15.4%)	$n=4$ (15.4%)	$n=6$ (23.1%)	NR
Greissenauer	$n=4$ (9.1%)	Pilocytic astrocytoma ($n=2$, 50.0%); Non-diagnostic ($n=2$, 50.0%)	$n=1$ (2.3%)	$n=2$ (4.5%)	$n=3$ (6.8%)	$n=14$ (31.8%)	$n=0$
Javadpour	$n=2$ (33.3%)	Low-grade astrocytoma ($n=1$, 50.0%); Non-diagnostic ($n=1$, 50.0%)	$n=0$	$n=0$	$n=0$	$n=0$	NR
Kaufmann	$n=39$ (54.9%)	Diffuse astrocytoma II ($n=20$, 51.3%); Pilocytic astrocytoma ($n=8$, 20.5%)	$n=15$ (21.1%)	$n=4$ (5.6%)	$n=11$ (15.5%)	$n=22$ (31.0%)	$n=2$ (2.8%)
Kershenovich	$n=14$ (35.0%)	Pilocytic astrocytoma ($n=10$, 71.4%); Low-grade glioma ($n=1$, 7.1%)	$n=14$ (35.0%)	$n=1$ (2.5%)	$n=1$ (2.5%)	$n=31$ (77.5%)	NR
Li	$n=0$	NA	$n=0$	$n=0$	$n=0$	$n=3$ (9.7%)	NR

Table 1 (continued)

Ref	Number of Pxs	Range of Px age (years)	Enhancing tumors	Abnormal ocular findings	CSF diversion	Second CSF diversion	Radiological monitoring
Mottolese	$n=24$ (88.9%)	Diffuse astrocytoma II ($n=4$, 16.7%); Pilocytic astrocytoma ($n=15$, 62.5%)	$n=24$ (88.9%)	$n=8$ (29.6%)	$n=4$ (14.8%)	NR	$n=5$ (18.5%)
Ramina	$n=7$ (100.0%)	Low-grade astrocytoma ($n=7$, 100.0%)	$n=7$ (100.0%)	$n=0$	$n=2$ (28.6%)	$n=1$ (14.3%)	$n=1$ (14.3%)
Romeo	$n=2$ (9.1%)	Pilocytic astrocytoma ($n=2$, 100.0%)	$n=1$ (4.5%)	$n=0$	$n=2$ (9.1%)	NR	NR
Stark	$n=5$ (41.7%)	Diffuse astrocytoma II ($n=2$, 40.0%); Pilocytic astrocytoma ($n=1$, 20.0%)	$n=3$ (25.0%)	$n=0$	$n=1$ (8.3%)	$n=3$ (25.0%)	$n=0$
Ternier	$n=14$ (35.0%)	Pilocytic astrocytoma ($n=5$, 35.7%); Low-grade glioma ($n=6$, 42.8%)	$n=14$ (35.0%)	$n=0$	$n=1$ (2.5%)	$n=14$ (35.0%)	$n=1$ (2.5%)
Wellons	$n=0$	NA	$n=0$	$n=0$	$n=0$	$n=4$ (30.8%)	$n=0$

NA not applicable, *NR* not recorded, *Px* patient

Discussion

There is a paucity of literature examining the different management options for tectal gliomas in the pediatric population. To the best of our knowledge, we have completed the first systematic review comparing the various options available for treating pediatric tectal gliomas along with the clinical presentation and outcomes of these patients.

Differential diagnosis and overall management of tectal glioma

Tectal gliomas are a rare tumor in the midbrain that usually occurs in the pediatric population. These tumors, alongside pineal region tumors, are defined as periaqueductal tumors [3]. However, recent studies have reported pure aqueductal tumors that are radiographically similar to tectal gliomas [25, 28]. Furthermore, since pure aqueductal tumors typically originate from the aqueductal region and present with obstructive hydrocephalus, the distinction between these different midbrain tumors can often be obscured. While tectal gliomas generally have an indolent course, some pure aqueductal tumors have shown aggressive clinical features that may warrant resection [25]. Despite both tumor types causing obstructive hydrocephalus, it is important to radiologically distinguish between tectal gliomas and pure aqueductal tumors. This diagnosis is often made through examination of the tectum as tectal gliomas will cause expansion of the tectum, while pure aqueductal tumors reside solely in the aqueduct and result in thinning of the tectum. Therefore, given the varying clinical course of midbrain tumors, it is essential to properly diagnose tectal gliomas to ensure appropriate medical treatment.

Studies have found that most tectal gliomas present with an indolent course and require only intervention to relieve symptoms from hydrocephalus [16, 32]. The majority of these lesions are low-grade and are histologically from pilocytic astrocytomas or infiltrating gliomas [23, 29]. On MRI, tectal gliomas are typically hyperintense on T2 and iso- to hypointense on T1, and do not enhance. Given the benign nature of most tectal gliomas, radiological monitoring is usually the first choice for pediatric patients [4, 11]. However, certain patients may present with radiographically "atypical" lesions in the tectum, which can represent other tumor types such as pilocytic astrocytomas or higher grade gliomas [7, 24]. Thus, patients with rapidly growing tumors may warrant intervention with more aggressive management, including radiotherapy, chemotherapy, and/or surgery.

Radiotherapy

From our systematic review, we found that radiotherapy is occasionally used in the management of tectal gliomas at a frequency of less than 30%. Traditionally, the use of radiotherapy in the management of pediatric tectal gliomas is often seen in cases where the tumor can no longer be managed through observation alone [20, 22, 23]. However, other researches have also suggested use of radiotherapy as a safe first-line treatment for tectal gliomas [6]. Compared to other management options including resection, radiotherapy has been largely reported as being a safe and successful option for tumors requiring treatment beyond observation [6, 13, 27]. Kaufmann et al. reported that of the patients receiving treatment for their tectal gliomas due to radiological or neurological progression, 82% of the group receiving radiotherapy had a 10-year progression-free survival (PFS), reflecting a higher success rate than the surgical group which reported only a 53% 10-year PFS [13]. In this study, the authors advocated for a non-surgical first-line treatment (i.e., radiotherapy and chemotherapy) in an effort to avoid the risks associated with surgery.

Focal radiotherapy in the form of stereotactic radiotherapy [2] and focal photon radiation [7, 23] have been common modalities for radiation treatment of tectal gliomas. In many of these patients, radiotherapy occurred following resection [23] or alongside chemotherapy [10]. Most patients were reported to have aggressive tumor growth, thus requiring the use of a multimodal therapy. On the other hand, only three studies reported patients that received stereotactic radiotherapy as their only treatment modality [2, 7, 13]. In this group, the majority of patients responded well to radiotherapy, thus suggesting that radiotherapy can be potentially effective in a patient population that demonstrates minor tumor progression. However, a recent study has also explored the use of gamma-knife therapy as a main treatment option for pediatric tectal gliomas [6]. In this study, tumor control and volume reduction were achieved in all cases with over half of the tumors in patients eventually disappearing [6]. Therefore, gamma-knife therapy may show promise as a first-line treatment modality in some patients and may prevent the need for CSF diversion procedures; however, given the limited literature about this technique, additional studies are needed to determine if gamma-knife surgery has routine use in the treatment of tectal gliomas.

When administering focal radiotherapy, studies reported doses range from 45 to 56.8 Gy with the exact dosage often depending on the age of the patient [2, 7, 13]. For younger patients, irradiation has been used at a lower dose compared to that used for older patients. One study limited their dosing to 45 Gy in children less than 5, while children older than 5 received 54 Gy; both groups had fractions of 1.8 Gy per day [13]. For studies utilizing gamma-knife surgery specifically, doses ranged from 11 to 14 Gy with all patients being above the age of 5 [6].

Overall, the use of focal radiotherapies in tectal gliomas is a relatively safe treatment option for progressive tumors that avoids radiation-related morbidities associated with other radiation modalities such as whole-brain radiation. There were no instances of secondary malignancies or radiation-induced vasculopathies discussed in the studies within the systematic review. The only complications encountered in the literature were four instances of cysts that developed post-radiation treatment as described in El-Shehaby et al. [6]. However, these cysts did not present any significant morbidity to the patients as two resolved and two remained stable. Therefore, our study did not find that focal radiotherapy modalities presented significant morbidity or mortality when used to treat pediatric tectal gliomas.

Chemotherapy

Similar to radiotherapy and resection, the indolent nature of pediatric tectal gliomas does not often warrant chemotherapy usage as a first-line treatment, unless there are indications of tumor progression or recurrence [2, 13, 14]. Chemotherapy may be chosen over radiotherapy when initially treating younger children with atypical tectal gliomas [13]. In our systematic review, we found that the low rates of chemotherapy usage were consistent across all institutions. Unlike radiotherapy, which was used in 10 studies, only five studies utilized chemotherapy as a treatment option. Carboplatin and vincristine were most commonly cited as the choice for chemotherapy [7, 13]. However, for one patient, the traditional carboplatin/vincristine combination administered at diagnosis was ineffective, thereby resulting in 11 courses of temozolomide following the first progression of the tectal plate lesion [7].

For many older patients, chemotherapy served as adjuvant therapy following tumor resection [13, 20] and occurred alongside radiotherapy treatment [7, 14, 20]. However, few patients received chemotherapy as their only treatment modality at the time of initial diagnosis [7, 13]. In one of these instances, chemotherapy was selected as the non-surgical first-line treatment for children under 8 as per the institutional preference [13].

Since few tumors progress among pediatric tectal gliomas, there is limited use for chemotherapy as a treatment option, and there are few studies in the current literature that address the usage of chemotherapy as an effective treatment. However, if adjuvant therapy is deemed necessary, physicians may still choose chemotherapy over radiotherapy for young children to avoid excessive brain radiation.

Surgical resection

Compared to other management options, our systematic review showed that utilization of surgical resection has the highest degree of variability in the literature. Only one study elected to pursue resection after receiving the histological diagnosis from the prior biopsy [14]. In all other cases, histological results were obtained at the time of resection. While some studies did not use resection whatsoever [7], other studies reported using resection for all the patients in their cohort [23]. This variability reflects an established discrepancy about the uses of resection as a treatment option for pediatric patients with tectal gliomas.

Previous research by Wang et al. examining resections of brainstem gliomas between 1986 and 1997 advocated for the use of surgical resection when treating tumors in this area of the brain [31]. However, the authors acknowledged the difficulty in resecting large tectal gliomas and described the instance of one pediatric patient who has post-operative complications after receiving a subtotal resection of an astrocytoma of the tectum. Ramina et al. presented a stronger stance on the issue and advocated for the use of surgical resection as a main option for tectal glioma management with the possibility of being potentially curative and having better outcomes compared to other treatment options [23]. On the other hand, many studies recognize the associated risk of neurological damage that accompanies the use of surgical resection [13, 17]. For instance, Liu et al. reported that two of the 3 patients receiving gross total resection developed significant neurological deficits resulting from strokes [17]. Now, more contemporary research indicates reserved use of surgical resection under special circumstances including the treatment of medium- and large-sized tumors or ones that show radiological progression, despite the previously established high complication rates [14, 29].

In many cases, the nature and size of the lesion dictate the amount of resection that can be safely performed. Often, resection is more readily performed on tumors that are larger in volume. Most studies that use size as a determining factor operated on tumors greater than 6 cm^3 on average [10, 14]. Furthermore, exophytic tumors, such as pilocytic astrocytomas, may allow for a gross total resection, while more infiltrative lesions may limit resection to partial debulking or a subtotal resection [23]. Some series have claimed successful gross total resections in the majority of patients [20, 23]. However, others preferentially performed subtotal resection at their institutions [13, 27]. Thus, individual tumor features strongly dictate the extent of possible resection in these patients.

The majority of studies reported the usage of adjuvant therapy (i.e., chemotherapy and/or radiation) for a portion of their patients following resection ranging from 7.1 to 33.3% [13, 20]. In the two case series that reported only one instance of resection, both patients required adjuvant treatment [10, 24]. Radiotherapy was the most commonly used adjuvant treatment following resection with only one study using chemotherapy as the follow-up adjuvant treatment [13]. Despite the apparent need for further adjuvant treatment following resection, some studies such as Ramina et al. report great successes using resection as there were no instances of tumor recurrence in individuals receiving gross total resection [23].

Although biopsy of the lesion can often be performed in conjunction with ETV surgery via an endoscopic approach, an open approach has been the standard for tectal glioma resection. Should resection be deemed necessary, it has been reported that a paramedian, infratentorial-supracerebellar, transcollicular approach provides safe access to the posterior and middle tentorial incisura [1, 12, 26]. However, recent success has been reported in use of a fully endoscopic transforaminal-transchoroidal approach to tectal plate [30]. This approach allowed for gross total resection of tectal gliomas in two adult patients through use of an endoscopic aspirator. These new, developing surgical techniques may allow for safer resection options in pediatric patients, thereby increasing the utility of resection as a treatment option. However, given the current literature, surgical resection should be limited to cases with large and/or progressive tumors causing refractory symptoms.

Management of hydrocephalus

Even though CSF diversion procedures do not treat the tumor itself, these surgeries are performed quite often in pediatric patients with tectal gliomas to treat the accompanying hydrocephalus that is found in the majority of these patients. Typically, the onset of hydrocephalus in these patients displays a slow time course due to progressive obstruction of the aqueduct by the tectal glioma. In a rare subset of patients, intratumoral hemorrhage may result in acute obstructive hydrocephalus, requiring immediate surgical intervention either through CSF diversion or tumor resection [21]. However, this condition is more common in diffuse intrinsic pontine gliomas and other highly malignant midbrain tumors [9], although can rarely be seen in the setting of more aggressive tectal gliomas. In our systematic review, there were no instances of intratumoral hemorrhage.

In our systematic review, we found that the vast majority of patients received at least one CSF diversion procedure during their clinical course (89.3%). Some studies reported using CSF diversion in all of their patients [2, 4, 11, 16, 23, 24, 27, 32]. In a subset of these studies, CSF diversion was the only procedure required and tumors were radiologically monitored for the entire clinical course without any need for tumor treatment [4, 11, 16, 32]. Despite successful treatment of hydrocephalus via CSF diversion, some individuals may

experience tumor progression, thereby resulting in recurrent hydrocephalus [2, 20]. In these individuals, tumor treatment in the forms of radiotherapy, chemotherapy, and/or resection may be warranted, as separately discussed above.

While CSF diversion is the most utilized procedure, it is still prone to failure. Despite an ETV success score generally predicting up to 90% ETV success rate, in cases of failure, patients may require a second ETV procedure or may be switched to a shunt [8, 15, 24]. The reports of failure in these procedures have been variable among the published literature. In Romeo et al., 77% of ETVs performed were a success with only four children out of 22 requiring a second procedure [24]. Similarly, Wellons et al. reported only two children out of 13 needing additional CSF diversion surgeries post-ETV, with all children being shunt-free at the time the study was conducted [32]. Despite these cited successes of ETV, there are multiple studies in the literature that report rates of ETV failure above 30%.

Many studies agree that the failure rate of ETV is lower than that of VPS [5, 14, 29]. In addition, studies have shown that ETV in children overall favors long-term reduction of ventricle size, often with the greatest reduction occurring at the 1-year mark [24, 32]. Although VPS may be considered the historical gold standard for CSF diversion, the high failure rates and potential for infection make this an undesirable option both for patients and physicians. Therefore, the current literature supports the notion that use of ETV has an overall higher success rate, as predicted by ETVSS, than VPS when managing obstructive hydrocephalus from tectal gliomas [8, 15]. Regardless of the failure rate in either procedure, CSF diversion still remains the most commonly performed procedure in pediatric patients with tectal gliomas and is an effective management strategy for treatment and symptomatic relief of associated hydrocephalus in these patients.

Additional procedures

Of the biopsies reported in the literature, many occur in conjunction with surgical resection and endoscopically during ETV procedures [11, 13]. In addition, cases with radiologically progressing or ambiguous tumors can warrant the use of isolated biopsies as a useful diagnostic tool or to test for mutations that may significantly impact treatment options [29]. Previous research has focused on developing radiological guidelines for accessing the need of a biopsy. These studies have suggested that tectal gliomas of an indolent nature often exhibit lack of enhancement and are small in size on radiographs, thus indicating that a biopsy is likely not required [4, 18]. However, since there is not an established protocol for obtaining a biopsy, the use of biopsy in pediatric tectal glioma patients depends on the treating physicians [11, 29].

Limitations

There are several limitations to the present study including (1) studies that were potentially missed during our literature search that exist in other scientific databases, and (2) study heterogeneity in our systematic review due to different standards of management at different academic medical centers. However, in being the first systematic review about the management options for tectal gliomas in the pediatric population, our study provides valuable insight for physicians treating these rare tumors.

Conclusions

For the pediatric population, tectal gliomas can be effectively managed through surveillance and treatment of hydrocephalus using CSF diversion if necessary. Since hydrocephalus is a common occurrence in these patients, there is often a higher frequency of hydrocephalus treatment than treatment of the tumor itself. Other forms of management, such as radiotherapy, may show promise as a treatment method for progressing tumors. There is still a need for further scientific investigation to establish the safety, efficacy, and use of surgical resection.

Author contribution M.M.J.B. and A.R.B conceived the research topic. C.J.B. collected the data. M.M.J.B., A.R.B., C.R.Z., and C.R. performed the systematic review. M.M.J.B. and A.R.B. interpreted the data, performed the analysis, and wrote the manuscript. R.M.N. and D.J.D. aided in interpreting the results and critically revising the manuscript.

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Declarations

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