ORIGINAL ARTICLE

Surgical Outcome of Cerebellar and Fourth Ventricle Tumors

Abdel Halim A. Moussa, Mohamed A. Ellabbad, Al Azzazi R. Al Azzazi, Muhammad A. A. Al-Sabbagh *

Department of Neurosurgery, Faculty of Medicine for Boys, Al-Azhar University, Cairo, Egypt

Abstract

Background: **The posterior cranial fossa extends from the tentorium down to the foramen magnum and contains the cerebellum, brainstem, fourth ventricle and the cranial nerves. Intra-axial tumors of the posterior fossa arise from the cerebellum, fourth ventricle or the brainstem.**

Aim and objectives: **To evaluate the surgical outcome of cerebellar and fourth ventricle tumors regarding management, including decision making, using cerebrospinal fluid(CSF) diversion or not, surgical approaches(techniques) and postoperative complications in order to establish the optimum way for management of cerebellar and fourth ventricle tumors.**

Patients and methods: **Both a retrospective analysis and a prospective study were carried out. The retrospective study covered a 2-year period from January 2020 to December 2021 and involved 25 patients with cerebellar and fourth ventricle tumors. The prospective study was done between January 2022 and June 2023 at Al-Azhar University hospitals (Department of Neurosurgery), El Sheikh Zayed Specialized Hospital and El-Sahel Teaching Hospital in Cairo.**

Results: The **most frequent tumors observed in our study are medulloblastomas 42%) then ependymomas 18%) then pilocytic astrocytomas (16%). Our study found a 12% incidence of newly-discovered postoperative hydrocephalus after posterior fossa surgery. Headache, nausea, vomiting, abnormal gait, palsy of the abducens nerve, or, in younger children, a swollen skull and bulging fontanelle are all symptoms of postoperative hydrocephalus. Shunting the CSF is the method of treatment.**

Conclusion: **The telovelar approach demonstrated superior progression-free survival, while the transvermian approach exhibited the best overall survival rates. Additionally, pre-excision CSF diversion(when needed) appeared to correlate with improved progression-free survival.**

Keywords: Ventricle tumors; Surgical outcome; Cerebellar tumors

1. Introduction

Most pediatric brain cancers, including medulloblastoma, ependymoma, and pilocytic Astrocytoma, are found in the posterior cranial fossa, which is in or near the midline. However, in adults, malignancies in the posterior fossa are commonly seen in the cerebellar hemispheres, away from the midline. When it comes to adult cerebellar masses, the two most prevalent and significant differential diagnoses are metastases and hemangioblastomas.[1]

The use of state-of-the-art neuroimaging techniques allows for earlier disease diagnosis. Although magnetic resonance imaging (MRI) is the preferred method for diagnosing malignancies in the posterior fossa, computed tomography (CT) may identify them in over 95% of cases.[2]

Surgeons should aim for the safest possible tumor resection when performing surgery. A radical or complete removal, within the bounds of safety, should be carried out if possible. Deaths and complications from surgery must be kept to a minimum. Thanks to the significant advancements in microsurgical techniques (including technology, instrumentation, and neuroanesthesia) during the past 30 years, neurosurgeons now have more confidence in their abilities and can do more complex procedures.[3]

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* Corresponding author at: Neurosurgery, Faculty of Medicine for Boys, Al-Azhar University, Cairo, Egypt.
E-mail address: mash_mohamed@yahoo.com (M. A. A. Al-Sabbagh).

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Changes in CSF flow dynamics after surgery, problems with surgical manipulation, and adverse effects of anesthesia are among the potential risks of posterior fossa surgery. Serious illness or death may result from brainstem injuries, whether they are direct or vascular in origin. Another complication of prolonged retraction after surgery is cerebellar edema.[4]

The aim of this study is to evaluate the surgical outcome of cerebellar and fourth ventricle tumors regarding management including decision making, using CSF diversion or not, surgical approaches(techniques) and postoperative complications in order to establish the optimum way for management of cerebellar and fourth ventricle tumors.

2. Patients and methods

Combined retrospective and prospective study including 25 patients diagnosed with cerebellar and fourth ventricle tumors between 2020 and 2021 and 25 patients diagnosed with the same tumors between 2022 and 2023 at Al-Azhar University hospitals (Department of neurosurgery), El Sheikh Zayed Specialized Hospital and El-Sahel Teaching Hospital in Cairo.

Inclusion criteria:

Irrespective of age and gender selection, patients with primary and secondary tumors in the cerebellum, fourth ventricle and brainstem, and cases with/without ventriculomegaly during presentation.

Exclusion criteria:

Patients with cerebellopontine angle tumors, patients with foramen magnum tumors, and recurrent cases.

Pre-operative assessment:

Personal history(age, sex, address), family history, past history(developmental milestones, past history of the same disease, other diseases and past surgical procedures), history of present illness; manifestations of increased intracranial pressure(headache, vomiting and blurring of vision), manifestations of cerebellar affection(gait disturbance), manifestations of cranial nerve affection(with special attention to optic nerve affection as decreased visual acuity or impaired visual field, six nerve palsy and lower cranial nerves affection as nasal regurgitation, hoarseness of voice or difficulty of swallowing, and manifestations of long tracts affection(especially para- paresis in patients with CSF seeding) were taken from all patients.

All patients were assessed for physical status, mental status, speech, neck rigidity and vital signs. Also, they were assessed for; conscious level, cranial nerve deficits with special attention to; optic nerves(examination of visual acuity, field of vision and fundus examination for detection of presence of papilledema or optic nerve atrophy), eye movement and presence of 6th nerve palsy, examination of pupils for of size, shape and reaction to light, and examination of facial symmetry, lower cranial nerves, motor system, sensory system and cerebellar functions.

Every patient had the standard pre- and post-operative lab work done, including a complete blood count (CBC), a hemoglobin profile (PT, PC, INR, PTT), and tests for kidney and liver function (ALT, AST, serum urea, serum creatinine, serum sodium, and potassium).

CT of the brain without contrast: was done for all cases focusing on assessment of ventricular size, periventricular permeation (hydrocephalus), and tumor calcification. MRI without and with IV Gadolinium contrast: was done for all cases to assess the details of the fourth ventricular tumor including size, borders, different imaging characteristics, enhancement, possible extension to CPA and invasion of brain stem.

Pre-operative preparations:

Management of brain edema: Mannitol 25% or corticosteroids were given pre-operatively to decrease peri-tumoral edema and consequently help in decreasing ICP. Pre-operative CSF diversion: Pre-operative CSF diversion is either through ventriculoperitoneal shunt(VPS), external ventricular drainage(EVD), or endoscopic third ventriculostomy(ETV).

Operative Procedures:

Position:Prone position was the position used in this study. Approach: Suboccipital craniectomy or craniotomy was used in this study according to the location and extent of the tumor.

Operative techniques:

Splitting of the vermis(trans-vermian), cerebello-medullary fissure approach(telo-velar), and trans-cerebellar. Tumor Excision: Gross total resection(no visible tumor was left), debulking, and biopsy. Closure of dura: Direct closure, and dural graft: auto graft, synthetic graft.

Postoperative Assessment and follow-up(6-months):

For conscious level, cranial nerve deficits(abducens, facial and bulbar nerves), long tract affection, and later for CSF collection or leak, and posterior fossa syndromes, whether complete profound syndrome “Cerebellar mutism” or partial, with determination of the component involved(linguistic, neuro-behavioral, and motoric components).

Radiologically:

Non-contrast CT brain was done for all cases within 24hours of the operation to exclude the presence of hematoma in the operative bed, significant pneumocephalus, and to assess the ventricular size. MRI brain without and with contrast was done within the first postoperative 48hours in order to assess the extent of resection and not to be confused with the surgical artifacts that maximize thereafter. MRI brain without and with contrast was done after six months postoperatively.

Pathological examination of the excisional biopsy:

Pathological examination of the biopsy material and grading according to the World Health Organization (WHO) classification were performed in all cases.

Adjuvant therapy:

Adjuvant therapy was then addressed according to the patient risk stratification and the histopathological type and grade of the tumor.

Outcome:

Outcome of the patients was assessed and estimated on the following occasions: On admission, immediately postoperative, on discharge, one-month postoperatively, and after 6-months postoperatively.

Statistical analysis:

Data was analyzed statistically with the help of IBM SPSS Statistics version 27, which was developed by IBM Corp. in Armonk, NY. The median and interquartile range are used to display skewed numerical data, while the Mann-Whitney U-test is used to compare differences across groups. Categorical data is shown as percentages and counts, and Fisher's or Fisher-Freeman-Halton's exact test is used to compare differences. Ordinal data is compared using linear-by-linear association. A p-value less than 0.05 is deemed to have statistical significance.

3. Results

Table 1. Demographic data of the studied patients.

	RETROSPECTIVE (N=25)		PROSPECTIVE (N=25)	
AGE(YEARS)	Child<18	Adult>18	Child<18	Adult >18
NO OF PTS(%)	15(60%)	10(40%)	13(52%)	12(48%)

The age of the patients ranged between(3 and 64) years for the retrospective study and(1.5 and 64) for prospective study. There is no statistically significant difference in age between the two groups,(table 1; figure 1).

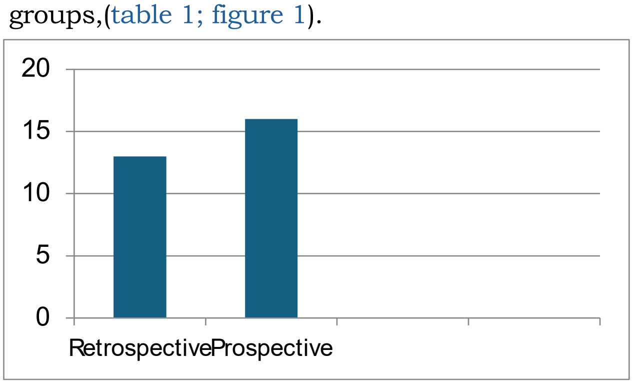

Figure 1. Median age of retrospective and prospective studies.

Table 2. Histopathology.

HISTOPATHOLOGY	RETROSPECTIVE (N=25)		PROSPECTIVE (N=25)		P-VALUE 0.203
MEDULLOBLASTOMA	8	32.0%	13	52.0%	
EPENDYMOMA	7	28.0%	2	8.0%	
ASTROCYTOMA	4	16.0%	5	20.0%	
HEMANGIOBLASTOMAS	3	12.0%	0	0.0%	
GLIOMA	1	4.0%	1	4.0%	
TERATOID/RHABDOIND TUMORS	0	0.0%	1	4.0%	
CHOROID PLEXUS PAPILLOMA	0	0.0%	1	4.0%	
CEREBELLAR METASTASIS	2	8.0%	2	8.0%	

The most common tumors in this study were medulloblastomas 21-patients(n=50) (42% of cases) followed by astrocytomas and ependymomas 9-patients(18% of cases). Medulloblastoma is the most common histological type in both cohorts, with a higher prevalence in the prospective group(52% vs. 32%). Astrocytoma represents 20.0% in the prospective group and 16% in the retrospective group. Ependymoma is significantly less frequent in the prospective group(8.0%) compared to the retrospective group(28.0%).

There are some discrepancies between the two cohorts in terms of tumor types, with certain tumors(e.g., hemangioblastomas, teratoid/rhabdoid tumors, choroid plexus papilloma) being present in one cohort but not the other. The overall difference in histopathological distribution between the two groups is not statistically significant(p-value=0.203),(table 2; figure 2).

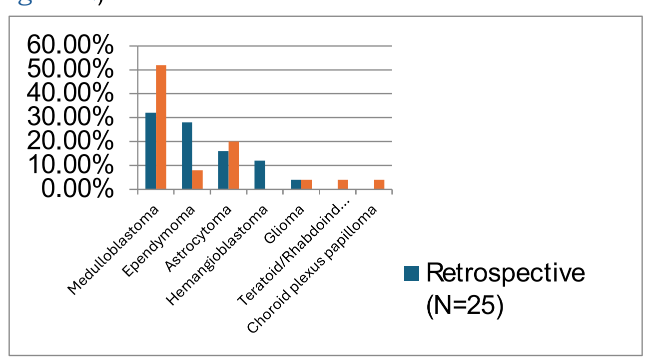

Figure 2.Histopathology.

Table 3. Postoperative complications.

POSTOPERATIVE COMPLICATIONS	RETROSPECTIVE (N=25)		PROSPECTIVE (N=25)		P-VALUE
DISTURBED LEVEL OF CONSCIOUSNESS	1	4.0%	2	8.0%	>0.999
CEREBELLAR MUTISM	1	4.0%	0	0.0%	>0.999
ACUTE HYDROCEPHALUS	2	8.0%	4	16.0%	0.667
CSF COLLECTION/LEAKAGE	2	8.0%	7	28.0%	0.138
LONG TRACT AFFECTION	0	0.0%	1	4.0%	>0.999
CEREBELLAR HEMORRHAGE	1	4.0%	0	0.0%	>0.999
WOUND INFECTION	3	12.0%	2	8.0%	>0.999

CSF collection/leakage is the most common complication, with a higher incidence in the prospective group(28% vs. 8%). Acute hydrocephalus is also a significant complication, occurring in 8% of the retrospective group and

16% of the prospective group. There are no statistically significant differences in the occurrence of any specific complication between the two groups,(table 3; figure 3).

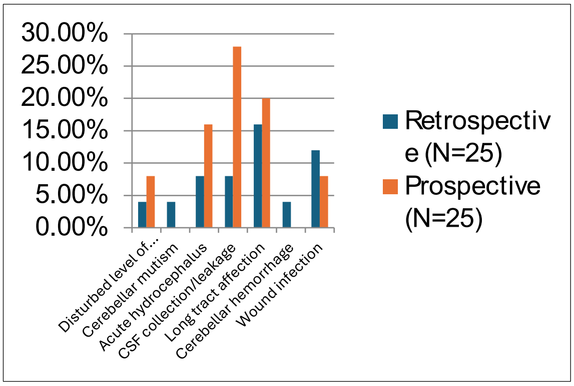

Figure 3. Postoperative complications.

Table 4. Correlation between surgical approaches in our study and development of post-operative complications.

	SURGICAL APPROACH								
	Telovelar (N=27)		Transvermian (N=8)		Transcerebellar (N=12)		Retrosigmoid (N=3)		
VARIABLE	n	%	n	%	n	%	n	%	p-Value†
POSTOPERATIVE COMPLICATIONS									
DISTURBED LEVEL OF CONSCIOUSNESS	1	3.7%	0	0.0%	1	8.3%	1	33.3%	0.216
CEREBELLAR MUTISM	0	0.0%	1	12.5%	0	0.0%	0	0.0%	0.005
ACUTE HYDROCEPHALUS	2	7.4%	0	0.0%	2	16.7%	2	66.7%	0.037
CSF COLLECTION/LEAKAGE	4	14.8%	4	50.0%	0	0.0%	1	33.3%	0.019

Tranvermian approach showed the highest percentage(50%) for CSF collection/leakage.The statistical significance (p-value) is reported as significant for CSF collection/leakage(p=0.019), Cerebellar mutism(p=0.005), and Acute hydrocephalus (p=0.037),(table 4; figure 4).

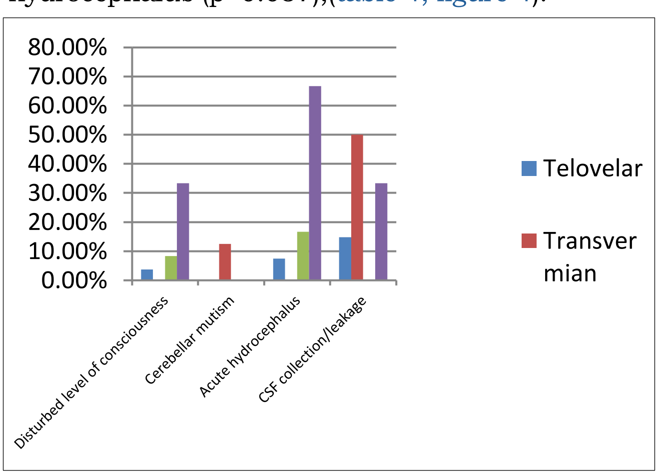

Figure 4. Incidence of postoperative complications by surgical approach

Table 5. Correlation between surgical procedures in our study and development of post-operative complications.

	SURGICAL PROCEDURE						
	Gross total resection (N=30)		Biopsy (N=3)		Debulking (N=17)		
VARIABLE	n	%	n	%	n	%	p-Value†
POSTOPERATIVE COMPLICATIONS							
DISTURBED LEVEL OF CONSCIOUSNESS	1	3.3%	2	66.7%	0	0.0%	0.007
CEREBELLAR MUTISM	1	3.3%	0	0.0%	0	0.0%	>0.999
ACUTE HYDROCEPHALUS	1	3.3%	2	66.7%	3	17.6%	0.007
CSF COLLECTION/LEAKAGE	5	16.7%	1	33.3%	3	17.6%	0.703
LONG TRACT AFFECTION	1	3.3%	0	0.0%	0	0.0%	>0.999
WOUND INFECTION	1	3.3%	0	0.0%	4	23.5%	0.110

There is a significant difference in disturbed level of consciousness between gross total resection(GTR) and others(p-value=0.007). Patients who underwent gross total resection had a much lower rate(3.3%) of experiencing this complication compared to biopsy(66.7%).

There is a significant difference in cerebellar mutism between biopsy and others (p-value=0.007). None of the patients who underwent GTR or Debulking experienced cerebellar mutism, whereas two out of three patients who underwent Biopsy did.

There is no significant difference in acute hydrocephalus(p-value=0.703), CSF collection/leakage(p-value=0.703), or long tract affection(p-value>0.999) between the surgical procedures.

It is important to note that, the sample size for biopsy is very small(N=3), which could limit the reliability of the findings for this group,(table 5).

Table 6. Effect of CSF diversion on surgical outcomes.

	CSF DIVERSION				
	No CSF diversion (N=12)		CSF diversion (N=38)		
VARIABLE	n	%	n	%	p-Value†
HYDROCEPHALIC CHANGES	1	8.3%	7	18.4%	0.661
PNEUMOCEPHALUS	0	0.0%	2	5.3%	>0.999
TUMOR BED HEMORRHAGE	0	0.0%	4	10.5%	0.560
CEREBELLAR HEMORRHAGE	0	0.0%	1	2.6%	>0.999
SDH	0	0.0%	1	2.6%	>0.999
POSTOPERATIVE MRI					
RESIDUAL TUMOR	5	41.7%	20	52.6%	0.742
BRAIN MRI F/U(6-MONTHS)					
RESIDUAL TUMOR	4	33.3%	10	26.3%	0.718
TUMOR RECURRENCE	3	25.0%	7	18.4%	0.686
1-YEAR SURVIVAL					
PROGRESSION-FREE SURVIVAL	7	58.3%	27	71.1%	0.486
OVERALL SURVIVAL	11	91.7%	33	86.8%	>0.999

There were no statistically significant differences in the incidence of hydrocephalic changes, pneumocephalus, tumor bed hemorrhage, cerebellar hemorrhage, or subdural hematoma(SDH) between the two groups. Tumor-related outcomes: There were no statistically significant differences in the rates of residual tumor on postoperative MRI or Brain MRI F/U(6-months), or in tumor recurrence between the two groups.

While progression-free survival appeared slightly higher in the CSF diversion group, the difference was not statistically significant. There was no statistically significant difference in overall survival between the two groups,(table 6).

Case presentation:

A 4-year-old male child complained of headache for 4-6 weeks. Vomiting began 2-weeks later, followed by head tilt and unsteady gait. He exhibited mild right facial palsy, ataxic gait, and nystagmus, which was more pronounced when looking to the right.

Urgent VPS(right frontal) was inserted, followed by gross total excision of the tumor via a trans-vermian approach. Histopathology confirmed classic medulloblastoma, grade 4. Radiotherapy was initiated one month later. There has been no evidence of recurrence or CSF seeding.

Pre-operative CT brain: demonstrated a well-defined, homogeneous, hyperdense mass in the midline of the posterior fossa, surrounded by vasogenic edema with acute hydrocephalic changes.

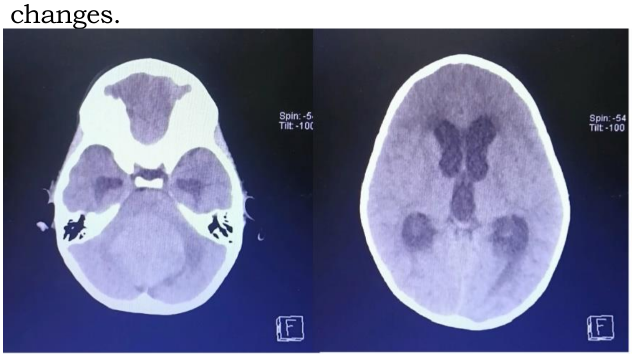

Figure 5. 2Axial cuts of CT brain without contrast.

MRI brain and whole spine: a well-defined midline posterior fossa SOL is seen centered on the 4th ventricle measuring 4×4.5×3.9cm, compressing the brain stem anteriorly. Normal contrast enhanced MRI study of the whole spine. Pre-operative MRI with contrast after VPS insertion. Contrast-enhanced T1-weighted MR images demonstrate intense, nearly homogeneous enhancement of the mass.

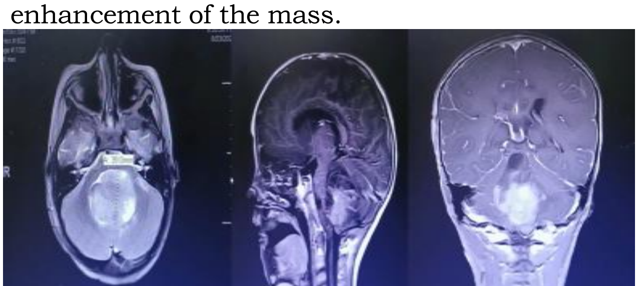

Figure 6. Axial, sagittal and coronal cuts.

Post-operative imaging(6months):

MRI brain and spine post-operative: No sizable residual or recurrent masses identified in the operative bed. Right frontal shunt tube in place with no evidence of hydrocephalic changes

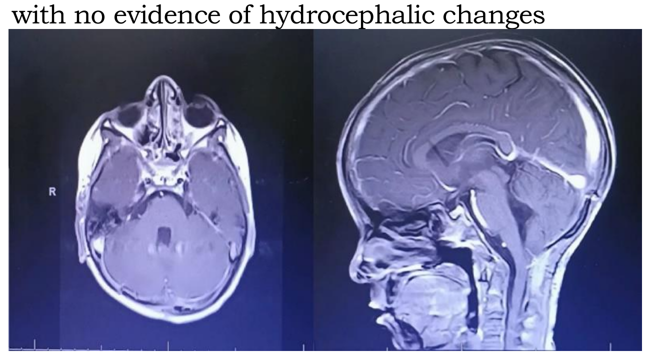

Figure 7. Axial and sagittal cuts MRI T1 with contrast.

4. Discussion

Cancers of the posterior fossa are among the deadliest diseases that humans might face. The morbidity and mortality that these malignancies produce make them important. It has also traditionally been difficult to surgically remove these tumors.[5]

In the present study, 32 patients (64%) who had preoperative hydrocephalus underwent VP shunt(29 cases), ETV (2 cases) and EVD (1 case).

Shih and Smirniotopoulos[6] found that permanent CSF diversion was needed in 33% patients. In another study by Bartlett et al.,[7] VP shunt was inserted in 53% patients.

In our study, the prevailing surgical approaches in both studies were the telovelar approach with 27 patients, the trans-cerebellar with 12 patients, the transvermian approach with 8 patients and the retrosegmoid approach was done for 3 patients.

The predominance of the telovelar approach could indicate its efficacy or preference among surgeons for specific types of cases.

In a study by Essa and Hamdan[8] Twenty-four patients, or 80% of the total, underwent telovelar excision, three patients, or 10% of the total, underwent trans-cerebellar excision, one patient, or 3.33% of the total, underwent trans-vermian excision, and one patient, or 3.33% of the total, underwent retrosigmoid excision.

In our study, the medulloblastoma was the dominant pathology(21-cases) in the both studies with 52% in the prospective study and 32% in the retrospective study, followed by ependymoma(9-cases) with 8% in the prospective study and 28% in the retrospective study, pilocytic Astrocytoma (8-cases) was 16% in both studies and cerebellar metastasis(4-cases) was 8% in both studies.

In the study by Khan and Iftikhar-ul-haq,[9] The following histological variants were found in the 53 cases: medulloblastoma(40.5%), ependymoma

(30.5%), pilocytic Astrocytoma (26.0%), and atypical teratoid/rhabdoid histological variants (3.1%).

In the study by Sajjad et al.,[10] histopathology findings included 133 cases of medulloblastoma (64.25%), 32 cases of ependymoma (15.45%), 28 cases of Astrocytoma (13.52%), and 3 cases of choroid plexus papilloma (1.44%). Found in 4 patients (1.93%).

In our study, the most common postoperative complications experienced by patients were, in decreasing order of frequency: cerebellar mutism, wound infection, disturbed level of consciousness, cerebellar hydrocephalus, acute hydrocephalus, cerebellar leakage, and long tract affection.

In a study by Essa and Hamdan, [8] A total of six patients, or 20% of the total, experienced post-operative hydrocephalus, which was surgically addressed with a ventriculoperitoneal shunt. Four patients, or 13.33% of the total, had cerebellar mutism, and three patients, or 10% of the total, had bulbar palsy. Tragically, two patients, or 6.666% of the total, passed away. Additionally, one patient, or 3.33% of the total, had a wound infection.

In a study by Sharafat et al.,[11] In 6.7% of instances, tumor bed hematoma occurred, whereas 4.9% of patients experienced wound infection and 4.9% had cerebrospinal fluid leak.

Based on our findings, CSF diversion is commonly done in both groups, usually before excision, and shunting is the method of choice. Shunting was the method of choice for CSF diversion. On the other hand, the prospective and retrospective groups do not differ significantly with respect to these activities.

In the study by Sajjad et al.,10 Postoperative wound infections occurred in 12 patients, or 5.79 percent. Unfortunately, meningitis developed in 9(4.3%) individuals and cerebrospinal fluid leaking occurred in 11(5.31%) patients. Cerebellar mutism was detected in 18 cases (8.69%).

In our study, 1-year progression-free survival in cases with CSF diversion(38/50) was 71.1% while in cases with no CSF diversion(12/50) was 58.3%.1-year overall survival in cases with CSF diversion was 86.8% while in cases with no CSF diversion was 91.7%.

In our study, regarding timing of CSF diversion, 1-year progression-free survival in cases with pre-excision CSF diversion(32/38) was 78.1% but in cases with post-excision CSF diversion(6/38) was 33.3%.1-year overall survival in cases with pre-excision CSF diversion was 90.6% while in cases with post-excision CSF diversion was 66.7%.

Our data on medulloblastomas shows that recurrence occurred in 23.8% of cases and postoperative residual tumor in 28.6%. There was an 81% overall survival rate and a 66.7% progression-free survival rate after 1 year.

The majority of patients (17.5%; 84.7% in children) in another investigation were found to have medulloblastomas. Recurrence occurred in 19.4% of cases, and residual tumors were detected in 43.0% of cases after surgery. In the first year following surgery, 30.5% of patients died.[12]

For ependymomas, our results showed that 22.2% had residual tumor after surgery, 11.1% had recurrence, and 88.9% were progression-free and survived the entire year.

In our study, regarding correlation between the surgical approach and survival rates, progression-free survival; the telovelar group demonstrated the highest rate(81.5%), while the retrosigmoid group had the lowest(33.3%). Overall survival; the transvermian group showed the highest rate(100%), and the retrosigmoid group the lowest(66.7%).

Our results suggest that the extent of surgical resection is associated with the development of postoperative complications. Patients who underwent less extensive procedures(biopsy and debulking) have a higher risk of hydrocephalic changes and residual tumor compared to those who underwent GTR. Patients who underwent GTR had the lowest rate of residual tumor(6.7%). Biopsy and debulking procedures resulted in significantly higher rates of residual tumor (66.7% and 58.8% respectively). GTR had a lower recurrence rate(10%) compared to debulking(41.2%).

In our study, medulloblastoma and Astrocytoma have the highest rates of residual tumor(28.6% and 33.3%, respectively). Hemangioblastomas have the lowest rate of residual tumor (0%). Hemangioblastomas have the highest rate of tumor recurrence(66.7%). Astrocytoma has the highest 1-year overall survival rate(100%). Ependymoma has the highest 1-year progression-free survival rate (88.9%).

4. Conclusion

The telovelar approach demonstrated superior progression-free survival, while the transvermian approach exhibited the best overall survival rates. Additionally, pre-excision CSF diversion(when needed) appeared to correlate with improved progression-free survival.

Disclosure

The authors have no financial interest to declare in relation to the content of this article.

Authorship

All authors have a substantial contribution to the article

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Conflicts of interest

There are no conflicts of interest.

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