Surgical Options for Treatment of Posterior Fossa Tumors with Hydrocephalus

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Abstract: **Background:** A brain tumor is one of the most devastating forms of human illness, especially when occurring in the posterior fossa. Brainstem compression, herniation, and death are all risks in tumors which occur in this critical location. **Aim of the work:** The aim of this work is to review clinical presentation, pathology, methods of investigations, operative techniques, complications of management of posterior fossa tumors and eventually prognosis. Also, this study aims at comparing the results of different types of management in order to establish the optimum way for management of posterior fossa tumors. **Subjects and Methods:** The admission, progress and outcome data of 24 patients with of posterior fossa tumors with hydrocephalus are collected in this series. Patients were grouped into 2 groups. The first group will be managed by direct surgical excision of the tumor and the second group will be managed by ventriculo-peritoneal (V-P) shunting, EVD or ETV prior to definitive surgery followed by direct tumor attack. **Results:** briefly in our study: 6 patients (60%) were completely cured of hydrocephalus without recurrence after total tumor excision in group (1) and 9 patients (64%) were completely cured of hydrocephalus without recurrence after total tumor excision in group(2). Only 3 patients 42% out of the 7 patients having external ventricular drainage in group 2 needed permanent CSF diversion. **Recommendation:** Decision of CSF diversion in posterior fossa tumor should be individualized according to patient condition and is not aroutine.
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1. Introduction

Tumors in the posterior fossa considered as a critical brain lesions. This is, primarily, because of the limited space within the posterior fossa and the potential involvement of vital brain stem nuclei.

Posterior fossa tumors are more common in children than the adults.

Between 54% and 70% of all childhood brain tumors originate in the posterior fossa.

About 15-20% of brain tumors in adults occur in the posterior fossa. Certain types of posterior fossa tumors, such as medulloblastoma, pineoblastoma, ependymomas, primitive neuroectodermal tumors (PNETs), and astrocytomas of the cerebellum and brain stem, occur more frequently in children. **(Ghani, et al 2003)**

Some glial tumors, such as mixed gliomas, are unique to children. They are located more frequently in the cerebellum (67%) and are usually benign. Genetic factors, such as dysfunction of some tumor suppressor genes (p53 gene) and activation of some oncogenes, may play a role in their development. **(Sutton, et al. 1989)**

Environmental factors such as irradiation and toxins may also play a role. The clinical presentations of posterior fossa tumors are remarkably constant and originate by two mechanisms: the first mechanism is obstruction of the normal cerebrospinal fluid (CSF) pathway, while direct infiltration and/or mass effect on the surrounding structures (cerebellum, cranial nerves or their nuclei and long tracts) represents the second mechanism responsible for manifestations of posterior fossa tumors. **(Bouramas, et al 2012)**

Patients who present with posterior fossa tumors undergo surgery for the following goals:

1-To decompress the posterior fossa for the purpose of relieving pressure on the brain stem and/or to release intracranial pressure and avert the risk of herniation.

2-To diagnose the tumor based on histopathology.

There are different protocols for management of posterior fossa tumors, this study aims at comparing the results of different 2 types of management in order to establish the optimum way for management of posterior fossa tumors.

2. Patients and Methods

The admission, progress and outcome data of 24 patients with of posterior fossa tumors with hydrocephalus are collected retrospectively and prospectively. Referrals from district general hospitals were included in this series.

Preoperative assessment involved thorough clinical history taking, general examination, and neurological examination. Preoperative investigations included laboratory and radiological assessment.

Patients will be grouped into 2 groups

*the first group (10 patients) will be managed by direct surgical excision of the tumor.

*the second group (14 patients) will be managed by ventriculo-peritoneal (V-P) shunting, pre-operative external ventricular drainage (EVD) or third ventriculostomy prior to definitive surgery followed by tumor attack.

I- History

A) Personal history:

Name, age, sex, occupation, residence and marrital state.

B) Complaint:

Including one or more of the following;

-Increased intracranial pressure: headache, vomiting, blurring of vision or disorders of eye movement (double vision).

-Expanding head size in infants with unfused cranial sutures and fontanels.

-Focal neurological deficits including:

a) Cerebellar deficits as ataxia and nystagmus.

b) Other deficits (according to site and nature of tumor) as 7th nerve palsies, bulbar palsy, or long tracts involvement in tumors arising from or invading the brain stem (floor of the fourth ventricle).

C) Present history:

Including analysis of the complaints regarding mode of onset, course and duration as well as any associated conditions.

D) Past history:

Of previous operations and medications.

II- Examination

A) General examination:

Including vital signs and other system affection.

B) Neurological examination:

Including conscious level assessment by Glasgow coma scale (GCS), intellectual functions and speech, cranial nerve examination, examination of motor power and reflexes, examination of the sensations and sphincteric function.

c) Ophthalmological examination:

To detect the presence or absence of papilledema

III- Investigations

A) Radiological investigations:

CT scan without and with contrast, in addition to MRI. A routine post-operative CT scan with and without contrast was done in the first 24 hours unless clinical deterioration occurred earlier, a control MRI brain with contrast in 6 weeks duration then annually thereafter.

B) Laboratory investigations:

Blood picture and bleeding profile, liver and kidney function, blood glucose and electrolytes.

3. Results

Regarding the results of group (1)

Which included 10 patients who have undergone direct posterior fossa surgery without any procedures for dealing with hydrocephalus, we found that 6 patients (60%) were cured of hydrocephalus while the other 4 patients (40%) had recurrence or persistence of hydrocephalus after tumor excision.

In this group, 2 patients (20%) had acute onset of deterioration of conscious level due to post-operative hydrocephalus evidenced by CT scan, were taken immediately to the OR and a CSF diversion was done (v-p shunt). The remaining 2 patients (of the 4 who developed hydrocephalus postoperatively), 1 patients had almost slower onset of hydrocephalus with no rapid or marked conscious level deterioration and they had (V-P shunt) insertion in due time, the remaining 1 patient developed post-operative pseudomeningocele, which was repeatedly aspirated, and they didn't respond after several aspirations and required insertion of v-p shunt.

Total excision of the tumor was done for 8 patients (80%) of group (1) while subtotal excision done for the remaining 2 patients (20%). All 2 patients with subtotal excision had post-operative hydrocephalus.

****Regarding the results of group (2)**

Out of the 14 patients of this group, 9 patients (64.3%) were completely cured of hydrocephalus without recurrence after total tumor excision and the follow up period (average 6 months).

- 7 patients had external ventricular drainage, 4 patients had v-p shunt and 3 patients had endoscopic third ventriculostomy.
- Only 3 patients 42% out of the 7 patients having external ventricular drainage needed permanent CSF diversion.

(1 of them after total tumor excision and the other 2 patients after subtotal tumor excision)

- 1 patient presented with post-operative hydrocephalus after one month from tumor excision due to ventriculo-peritoneal shunt obstruction after subtotal tumor excision. patient underwent shunt revision
- 1 patient presented with post-operative hydrocephalus after failed endoscopic third ventriculostomy followed by subtotal tumor excision. Then v-p shunt was inserted for permanent CSF diversion.
- Total excision of the tumor was done for 10 patients (71.4 %) of group (2) while subtotal excision done for the remaining 4 patients (28.5%). All 4 patients with subtotal excision had post-operative hydrocephalus.

briefly in our study:

* 6 patients (60%) were completely cured of hydrocephalus without recurrence after total tumor excision in group 1.

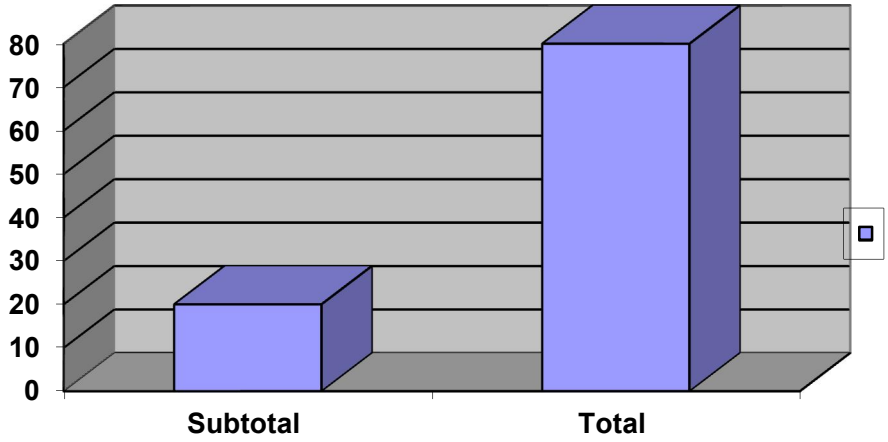
* 9 patients (64%) were completely cured of hydrocephalus without recurrence after total tumor excision in group 2.

* Only 3 patients 42% out of the 7 patients having external ventricular drainage in group 2 needed permanent CSF diversion.

Extent of tumor removal

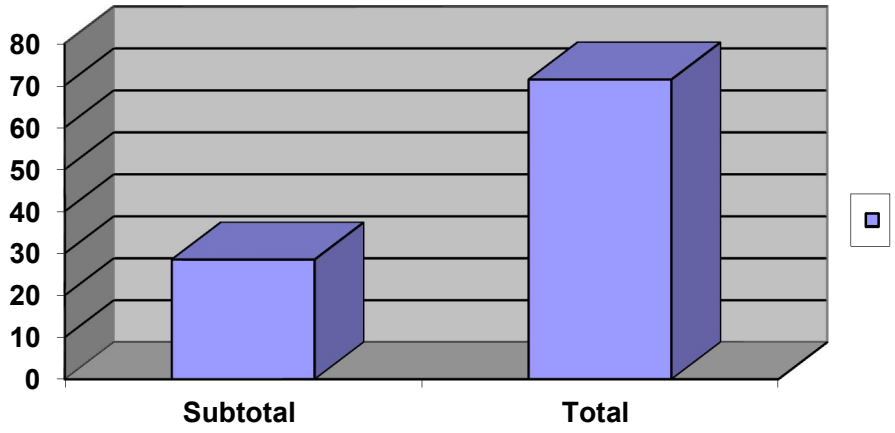
Extent of tumor removal in group 1

Extent of tumor excision	Count	Table N %
Subtotal	2	20 %
Total	8	80%



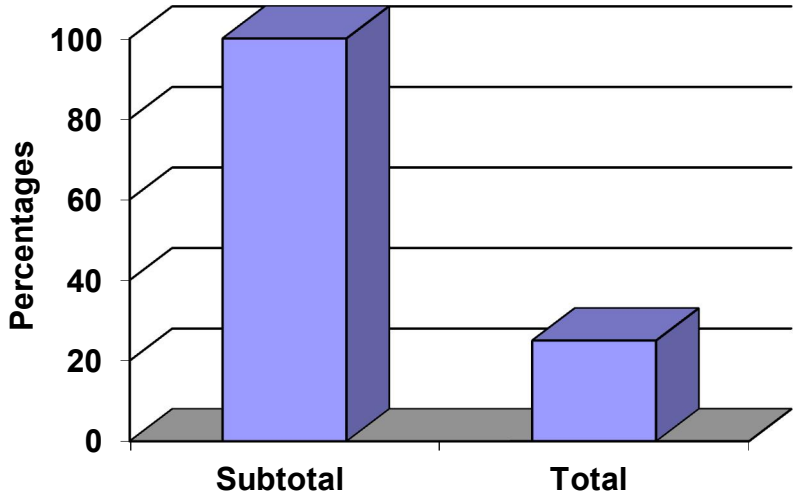
Extent of tumor removal group 2

Extent of tumor excision	Count	Table N %
Subtotal	4	28.5 %
Total	10	71.4%

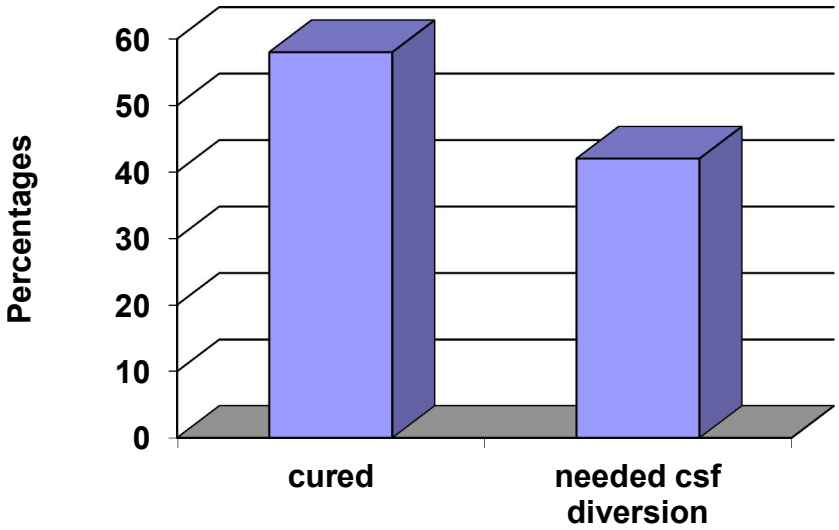


Extent of tumor removal compared to the need of post-operative shunting in group 1

100% of the patients who had subtotal tumor excision needed postoperative CSF diversion while only 25% of patients who had direct total tumor excision in group 1 required a CSF diversion



Extent of post-operative need for permanent CSF diversion after external ventricular drainage



4. Discussion

Posterior fossa tumors constitute a serious problem by their mass effect and create a secondary problem of CSF pathways obstruction. So, dealing with these tumors necessitates dealing with CSF pathways. (Wong TT et al, 2011).

In our study 6 patients (60%) were completely cured of hydrocephalus without recurrence after total tumor excision in group 1 and 9 patients (64%) were completely cured of hydrocephalus without recurrence after total tumor excision in group 2.

And only 42% of patients having external ventricular drainage in group 2 needed permanent CSF diversion.

- So we found no significant difference whether we do a direct tumor attack or CSF diversion first.

It is preferable to do CSF diversion first when we lack close observation to patient or in high risk patient which presents with signs of marked increased CSF pressure. As sudden increase in CSF pressure may be life threatening. However diversion may expose patient to extra hazards that may complicate CSF diversion surgery and infection is one of the most serious complications that may happen.

- 100% of the patients who had subtotal tumor excision needed postoperative CSF diversion, therefore subtotal tumor excision as well as extremely bloody surgeries, almost always lead to the

development of postoperative hydrocephalus, hence careful meticulous posterior fossa surgeries aiming at total tumor excision are extremely recommended.

- Regarding the results of group (1) which included 10 patients who undergone direct posterior fossa surgery without any procedures for dealing with hydrocephalus, we found that 6 patients (60%) were cured of hydrocephalus while the other 4 patients (40%) had recurrence or persistence of hydrocephalus after tumor excision.
- In this group, 2 patients (20%) had acute onset of deterioration of conscious level due to post-operative hydrocephalus evidenced by CT scan, were taken immediately to the OR and a CSF diversion was done (either v-p shunt or ETV according to the availability of an endoscopy-trained neurosurgeon on call), these 2 patients underwent subtotal tumor removal.
- The remaining 2 patients (of the 4 who developed hydrocephalus postoperatively) had almost slower onset of hydrocephalus with no rapid or marked conscious level deterioration and they had V-P shunt insertion in due time, these 2 patients underwent total tumor removal.
- Total excision of the tumor was done for 8 patients (80%) of group (1) while subtotal excision done for the remaining 2 patients (20%).
- From the data mentioned above, we found that direct tumor attack without addressing the hydrocephalus may jeopardize the lives of an appreciable number of patients (40% of patients), although 60% of patients of group (1) had been cured, but the considerable risk of morbidity or even mortality that we exposed patients to, deserves to attract our attention towards the dangerous aspect of this kind of practice, however we can avoid this risk by doing intraoperative(EVD) and close careful post-operative observation and follow up.
- We also found that risk of developing post-operative hydrocephalus is considerable in mid line tumors however this does not apply in special cases with posterior fossa tumors located away from CSF pathways like cerebellar astrocytomas and haemangioblastomas, as in such cases there is very low or almost no risk of developing acute hydrocephalus following tumor excision (Kamal HM, 2003).
- Regarding the results of group (2) which included 14 patients who had an external ventricular drain fixed immediately before tumor excision, Endoscopic third ventriculostomy Or ventriculo-peritoneal shunt we found that, the use of CSF diversion facilitates intraoperative intracranial pressure control facilitating the exposure as well.

In case of external ventricular drainage (EVD) it remains for few days postoperatively (2-4 days) for drainage of post-operative debris, ICP monitoring to determine whether hydrocephalus will persist or resolve and at the same time it acts as a safety valve guarding against the development of postoperative acute hydrocephalus with subsequent morbidity and/or mortality (as faced in some patients of group 1).

- Out of the 14 patients of this group, 9 patients (64.2%) were completely cured of hydrocephalus without recurrence during the follow up period (average 6 months).
- Total excision of the tumor was done for 10 patients (71.4%) of group (2) while subtotal excision done for the remaining 4 patients (28.5%). All 4 patients with subtotal excision needed post-operative permanent CSF diversion either for patients who had pre-operative EVD (2 patients) or who had failed ETV or v-p shunt.
- In case of external ventricular drainage most neurosurgeons fear infection which can be easily minimized by proper intra and postoperative care and shortening the period of external drainage as much as possible (Kamal HM, 2003). Factors increasing the risk of infection in external ventricular drainage, as mentioned by (Hickman and colleagues, 1990) were; duration (most important), patient's age and general condition, the kind of monitor used, maintenance of a closed system, and environment in which the monitor is inserted and nursed.

Summary and Conclusions

By analyzing the results of this study, the conclusion in dealing with posterior fossa tumor patients can be enumerated in the following points:

- No significant difference between results in both 2 groups.
- Not all patients of posterior fossa tumors need post-operative permanent CSF diversion however direct tumor attack may be risky for some cases as they may develop post-operative hydrocephalus and it may be of acute onset.
- External ventricular drainage is not much risky as thought by many neurosurgeons, if properly fixed and handled for a period not exceeding 72 hours (maximum 4 days).
- Subtotal tumor excision as well as extremely bloody surgeries, almost always leads to the development of postoperative hydrocephalus, hence careful meticulous posterior fossa surgeries aiming at total tumor excision are extremely recommended because even if they will develop hydrocephalus mostly it will be delayed.
- The development of postoperative pseudomeningocele, increasing headache, 6th nerve palsy, deterioration of conscious level all are alarming signs of development of hydrocephalus and should draw the attention of the resident in charge.

During our study we made an observation about the onset of hydrocephalus. We found that the immediate post-operative hydrocephalus appears in a dramatic way and may cause loss of the life of the patient however the intermediate onset of hydrocephalus which appears few days or weeks after surgery usually comes in a more insidious onset and shows slower progression which allows proper management in due time.

Recommendations

Finally our recommended protocol for management of posterior fossa tumors with hydrocephalus:

Direct tumor attack with aim of total removal in the nearest surgical list as long as patient is clinically stable, otherwise (ETV) is indicated pre-operatively.

Close monitoring post-operatively both clinically and radiology until patient gets established with possible few shots of ivmanittol post-operatively, otherwise persistence of hydrocephalus is noted, (ETV) is indicated.

In case of subtotal removal achieved for any reason or other, end surgery with (EVD) and monitor ICP to decide necessity of another diversion procedure (V-P shunt or ETV).

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