Surgical Management of Midline Posterior Fossa Tumours

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Abstract

Background: Tumors in the posterior fossa considered as a critical brain lesions. Radiation therapy may be the only or the main line of treatment as in diffuse brain stem gliomas or represents a complementary treatment after surgery as the condition in medulloblastomas and ependymomas.

Objective: to study clinical presentation, pathology, methods of investigations, operative techniques, complications of management of midline 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 midline posterior fossa tumors.

Patients and methods: This study was conducted on 30 patients at the neurosurgical department at SOHAG EL-HELAL health insurance, 6 OCT. between OCT 2012 and OCT 2014.

Results: the peak incidence of tumer was between 3 to 10 years old patients. The most common presenting features in this study were manifestations of increased ICP with predominance of headache and vomiting. 26.7% of cases had cranial nerves affection. 80% of cases in this study presented with surpratentorial hydrocephalus.The commonest post-operative complications in this study were cerebellar symptoms. It showed that CSF diversion decreased the risk of post-operative complications.

Conclusion: Patients with definite neurological symptoms and signs of increased ICP and/or cerebellar dysfunction and their CT brains were not conclusive should be investigated with MRI (with and without contrast) before exclusion of posterior fossa tumors.

Keyword: *Surgical Management, Midline Posterior Fossa, Tumours.*

INTRODUCTION

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.1

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.1

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.2 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.3

There are different protocols for management of posterior fossa tumors in pediatrics. Frequently, children with posterior fossa tumors are very ill at the time of presentation, with severe headache and vomiting, usually the result of obstructive hydrocephalus. Previously, the policy of initial management was to carry out ventriculo-peritoneal (V-P) shunting 7 to 10 days prior to definitive surgery followed by direct tumor attack. The recent policy, nowadays, is to manage those patients using pre-operative external ventricular drainage (EVD) in an attempt to avoid pre-resection shunting. The external drainage system is placed as an emergency in patients who present with severe symptomatic hydrocephalus; otherwise it is placed at the time of surgery and to be removed within 5 days post-operatively.4

Adjuvant therapy (mainly radiation therapy) plays an important role in management of posterior fossa tumors that are histologically malignant. Benign tumors in which brain stem invasion has prevented gross total removal can be followed clinically and radiologically (CT and/or MRI) for evidence of progression. Radiation therapy may be the only or the main line of treatment as in diffuse brain stem gliomas or represents a complementary treatment after surgery as the condition in medulloblastomas and ependymomas.5

PATIENTS AND METHODS

This study was conducted on patient with variant ages at the time of diagnosis, presenting with midline posterior fossa tumors, admitted and managed at the neurosurgical department at SOHAG EL-HELAL health insurance, 6 OCT. health insurance at Cairo and SOHAG university hospitals between OCT 2012 and OCT 2014.

All patients in this study were submitted to the following: History, Examination (General Examination and Neurological Examination), Investigations (Routine laboratory investigations, Radiological investigations and Histopathological examination), Pre-operative

Preparations, Operative Procedures, Post-operative follow- up and Post-operative complications.

STATISTICAL METHODS:

The data in this study was analyzed by SPSS version 11.0 statistical package (SPSS Inc., Chicago, IL, USA). Quantitative data were expressed as mean and standard deviation (X+SD). Qualitative data were expressed as number and percentage and analyzed by Fisher Exact test. Level of significance was set as P-value less than 0.05.

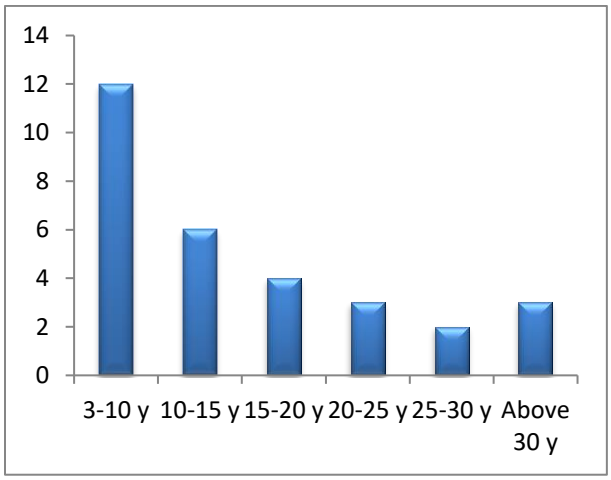
RESULTS

Table (1): Age distribution in this study

Age (years)	No.	%
3-10	12	40
10-15	6	20
15-20	4	13.3
20-25	3	10
25-30	2	6.7
Above 30	3	10
Total	30	100

Regarding the age distribution among 30 cases in this study, the peak incidence was between 3 to 10 years old (40% of patients) and the least incidence was 25 to 30 years old (5% of patients).

Graph (1): Age distribution in this study.



Most of patients in this study were males (21 patients, 70% of cases), while only (9 patients, 30% of cases) were females with male: female ratio (2.3:1)

Table (2): Presenting symptoms and signs in this study.

Clinical Presentations	No.	%
A- Conscious level assessment:		
Conscious.	27	90
Disoriented.	3	10
Total	30	100
B- Manifestations of increased I.C.P:	24	80
Headache	24	80
Vomiting	21	70
Papilledema	21	70
Increased head circumference	0	0
Tense fontanels	0	0
Impaired vision	3	10
C- Manifestations of cerebellar dysfunction:	21	70
Ataxia:	21	70
-Trunkal ataxia	12	40
- limb ataxia	9	30
Unsteady gait	18	60
Dysartheria	3	10
Nystagmus	6	20
Disdiadokokinesia	3	10

The most common presenting features in this study were manifestations of increased ICP (80% of patients) with predominance of headache (80%) and vomiting (70%) followed by manifestations of cerebellar dysfunction (70% of patients) with predominance of ataxia (70%) and unsteady gait (60%). Pre-operative deterioration of the conscious level was reported in only 3 patients (10%).

Graph (2): Manifestations of increased I.C.P.

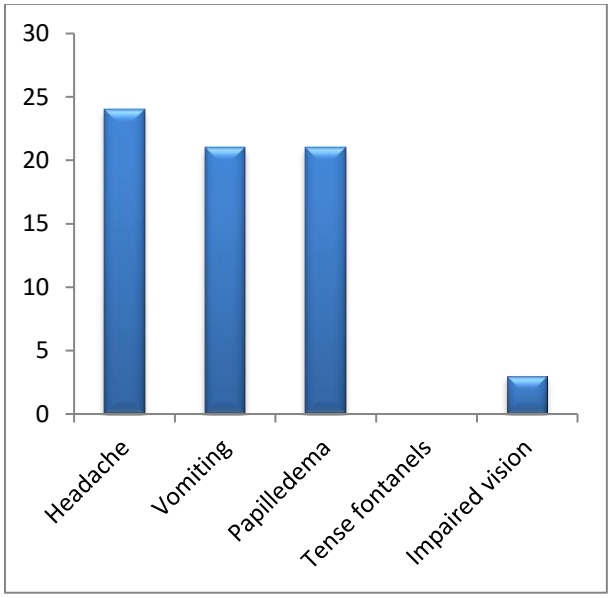


Table (3): Cranial Nerves Affection in this study

Cranial Nerves Affection	No.	%
(VI)	5	62.5
(VII)	2	25
(VII & Bulbar nerves)	1	12.5
Total	8	100

Regarding cranial nerves affection in this study, 8 patients (26.7% of cases) were involved, of which 7 cases (87.5%) with single cranial nerve and (12.5%) with multiple cranial nerves affection. The abducens (VI) nerve was the most common affected cranial nerve (62.5%) followed by facial (VII) nerve (25%).

Graph (3): Tumour size (in relation to posterior fossa diameter).

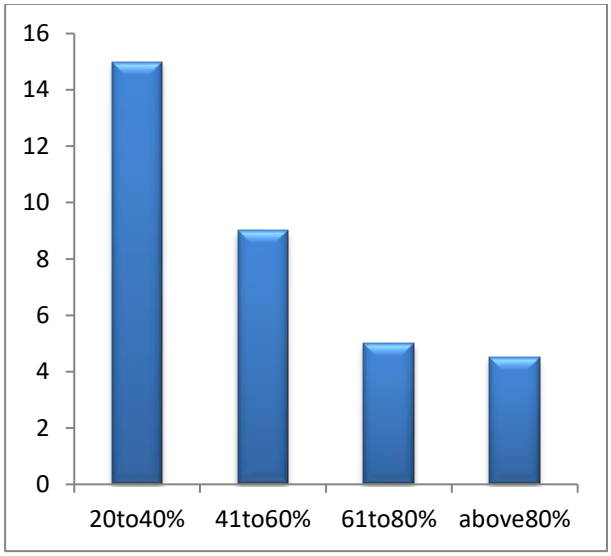


Table (4): Pre-operative hydrocephalus in this study and its management.

Pre-operative hydrocephalus and its management	No.	%
A. Pre-operative hydrocephalus:		
Present	24	80
Absent	6	20
Total	30	100
B. CSF diversion:		
Done	20	83.3
Not	4	16.7
Total	24	100
C. Type of CSF diversion:		
V-P shunt	15	75
Endoscopic 3rd ventriculostomy	2	10
E 3rd V followed by V-P shunt	3	15
Total	20	100

24 patients (80% of cases) in this study presented with surpratentorial hydrocephalus of which only 20 patients (83.3%) needed CSF diversion. It was done by V-P shunt in 15 cases (75%), endoscopic 3rd ventriculostomy in 2 cases (10%) and endoscopic 3rd ventriculostomy followed by V-P shunt in 3 cases (15%).

Table (5): Early post-operative complications in this study.

Post-operative complications	No.	%
A) Complications:		
Present	13	43.3
Absent	17	56.7
B) Type of complications:		
.Wound infection	2	6.7
.CSF leak	3	10
.pseudomeningocele	2	6.7
.Acute hydrocephalus	1	3.3
.Cerebellar symptoms	3	10
.Bulbar manifestation	2	6.7

The commonest post-operative complications in this study were cerebellar symptoms in 3 cases (10% of cases), CSF leak in 3 cases(10%) , bulbar manifestation in 2 cases (6.7%) , pseudomeningocele in 2 cases (6.7% of cases) ,wound infection in 2 cases (6.7%)and post-operative acute hydrocephalus in one case (3.3% of cases).

Table (6): Correlation between post-operative adjuvant therapy in our study and outcome of patients.

Adjuvant Therapy	Good No.=27		Poor No.=3		Fisher Exact Test (P)
	No.	%	No.	%	
- Received	21	77.8	3	100	1.000
- Not	6	22.2	0	0	1.000
Total	27	100	3	100	

The Table Showing the relation between post-operative adjuvant therapy and outcome of patients in our study. 21 patients (77.8% of cases) received post-operative adjuvant therapy had good outcome compared with 22.2% of patients without post-operative adjuvant therapy (statistically insignificant P> 0.05).

Graph (4): Post-operative adjuvant therapy in this study.

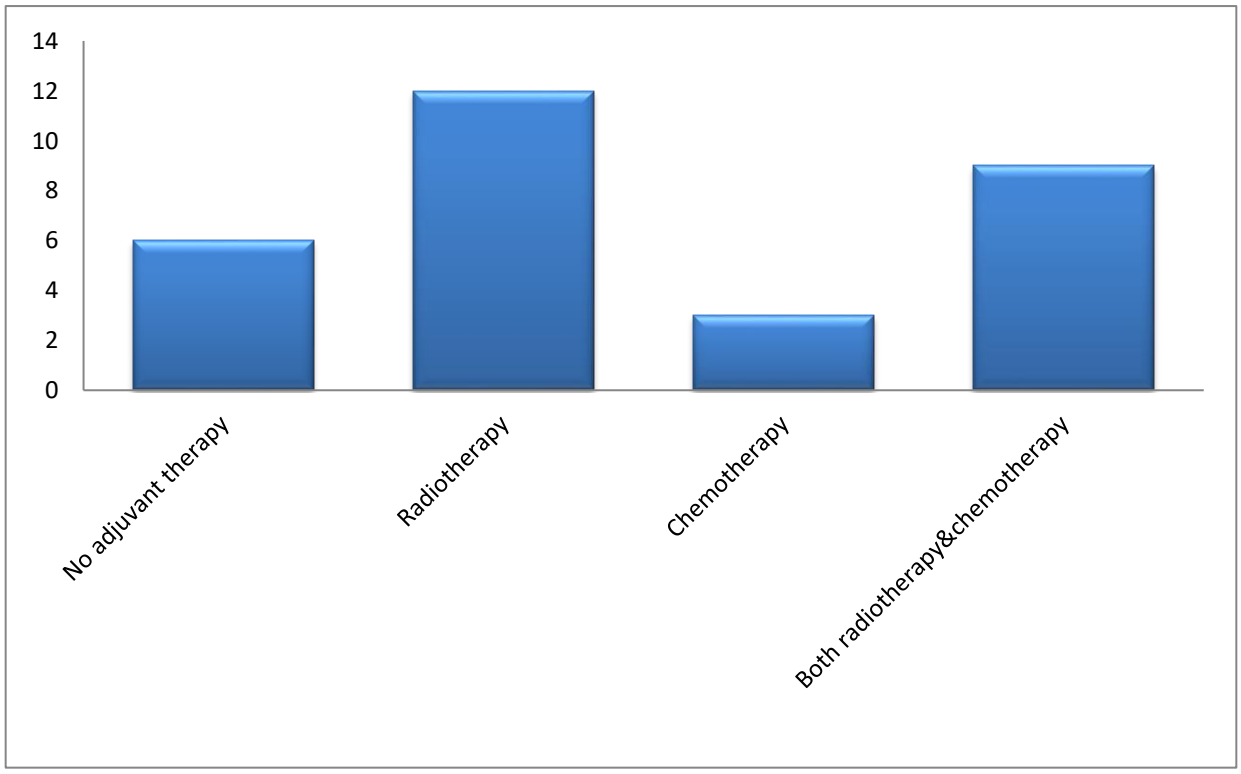


Table (7): Tumor recurrence in this study.

Tumor Recurrence	No.	%
A) Recurrence:		
.Present	5	16.7
.Absent	25	83.3
B) Type of tumor recurrence :		
- Medulloblastoma	3	60
- Low-grade astrocytoma	1	20
- Anaplastic astrocytoma	1	20
- Total	5	100

This table showed that Recurrence of the tumors in this study was reported in 3 cases of medulloblastomas during the follow-up period followed by astrocytomas in 2 cases.

Table (8): Correlation between CSF diversion in our study and development of post-operative CSF leak and/or pseudomeningocele.

C.S.F. diversion	Complications (CSF leak& pseudomeningocele)				X^2	P value
	Present No.=10		Absent No.=20			
	No.	%	No.	%		
- Done	3	60	17	89.5	2.47	0.35
- Not	2	40	2	10.5		0.15
Total	5	100	19	100		

The Table Showing the relation between CSF diversion and development of post-operative CSF leak and/or pseudomeningocele. It showed that CSF diversion decreased the risk of post-operative complications as 89.5% of cases with CSF diversion passed without complications compared with 10.5% of cases without CSF diversion.

CASES PRESENTATION

Case (1): Cerebellar Astrocytoma

Male patient five years oldpresenting with chronic headache, repeated vomiting with no improvement with medical treatment.

Photo (1): Pre V-P operation CT brain, showing midline posterior fossa mass seen within 4th ventricle.

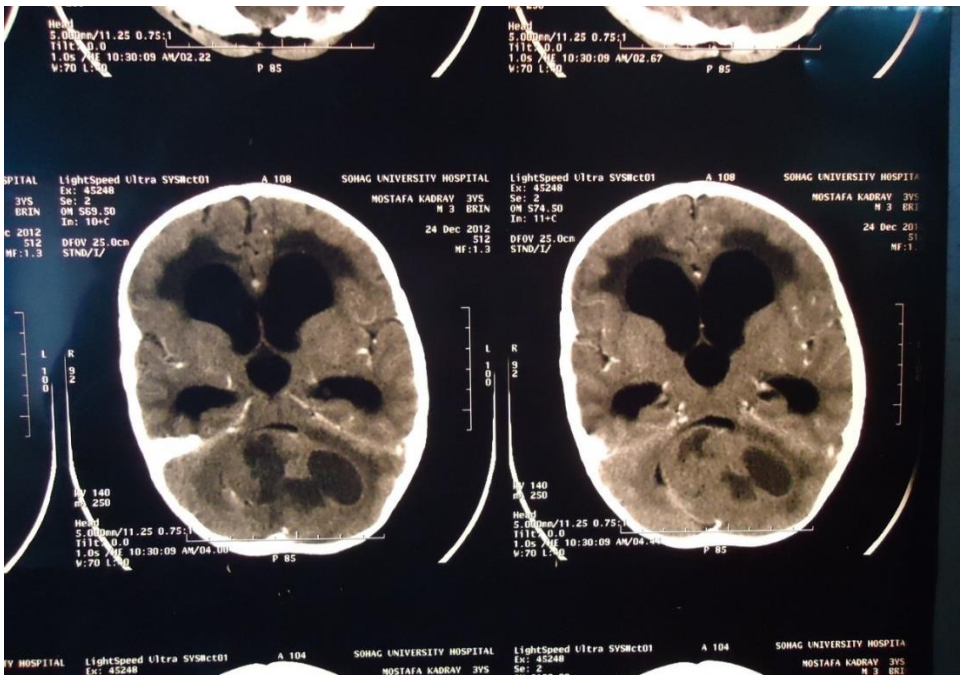


Photo (2): Pre-operative MRI brain showing midline posterior fossa tumour seen within 4th ventricle

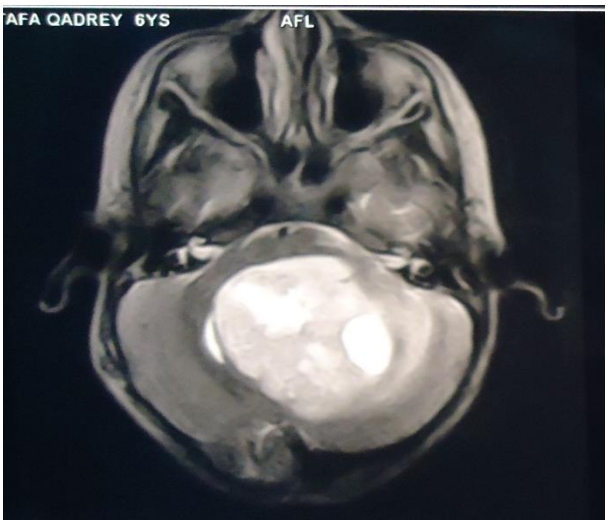
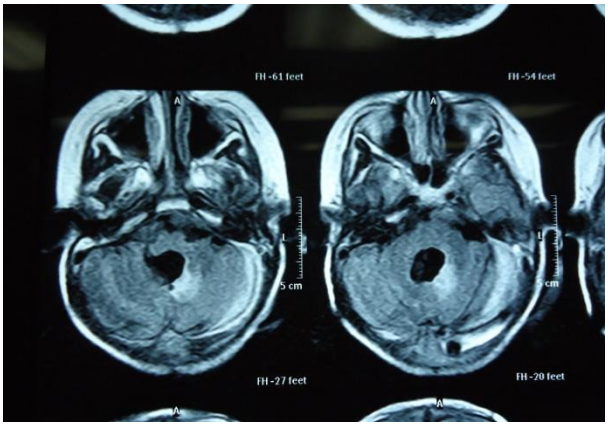


Photo (3): Post-operative MRI brain showing removal of the tumor.



Case (2): Medulloblastoma

Female patient13years old presenting with chronic headache, repeated vomiting, blurring of vision and unsteadiness.

Photo (4): Post shunt operation MRI of the brain, showing mid-line (vermian) tumor with heterogeneous intensity.

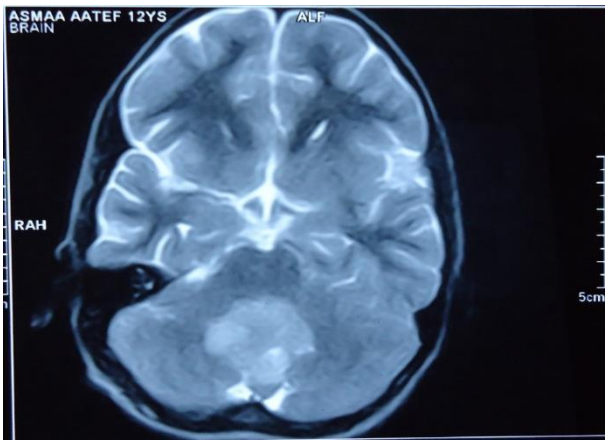


Photo (5): Post-operative MRI brain, showing minimal residual after excision of the tumor

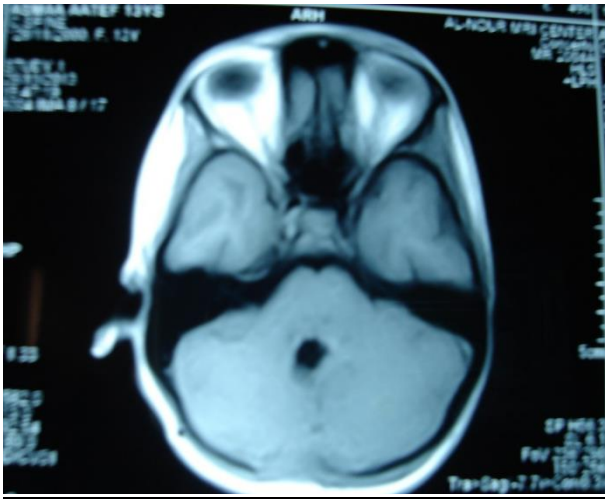
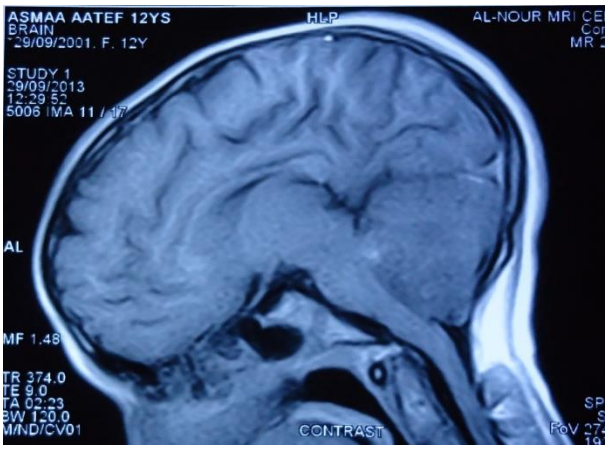


Photo (6): Post-operative MRI brain, sagittal view showing minimal residual after excision of the tumor



DISCUSSION

Tumors of the posterior fossa are more common in children than adults. In fact, between 54-70% of all childhood brain tumors originate in the posterior fossa compared with 15-20% in adults.2

In our study, the patient's age ranged from 3 to 40 years old. The age distribution figure revealed that the peak was between 3 to 10 years old ,The mean (average) age was 21.5 years while in Cochrane et al.6 series patient's age ranged between 2 months and 16 years old, the mean age was 7 years old.

In this study, there was no relation between patient's age at presentation and prognosis, and this is in agree with Pencalet.7 who didn't find any relation between age and outcome.8 while Sobel et al.9 concluded in his study that the younger the patients, the better the prognosis.

In our study, 30 patients were included, of which, there were 21 males and 9 females (male: female ratio 2.3:1) while in Zimmerman et al.10 series sex distribution for 141 patients was 85 males and 56 females (male: female ratio 1.5:1). Also, in Goldie et al.11 series 19 males were prominent over 12 females (male: female ratio 1.6:1).

In our study Cranial nerves affection were noted in 8 patients (26.7%), in 7 of them (87.5%), there was single nerve affection while multiple cranial nerves affection were noted in only one patient (both VII & bulbar nerves). Both headache and cerebellar ataxia were the main presenting symptoms and signs in this study as they were present in more than 70% of patients. But, at the same time, there was no relation between these two manifestations and outcome, while, altered level of consciousness had a remarkable relation to outcome of patients.

this is in agree with Pencalet et al.8 who concluded that headache was the most common symptom to be found, then, signs of cerebellar dysfunction constituted the second most common finding.

Also in Raimondi and Tomita's series.12 manifestations of increased ICP were present in 69.9% of patients, cerebellar dysfunction manifestations in 63%, cranial nerves affection in 25.2% of total number of their posterior fossa series consisted of 156 cases.

In our study, 24patients (80% of total cases) had pre-operative obstructive hydrocephalus of which only 18 cases (75%) were associated with manifestations of increased ICP .Neurosurgical opinions on the management of pre-operative hydrocephalus vary. There were authors that advocated pre-operative insertion of ventriculo-peritoneal shunt (V-P); others advocated direct tumor removal with insertion of external ventricular drainage (EVD) before dural opening to gain lax posterior fossa or placement of V-P shunt post-operatively according to patient's requirements.12,13

In this study, post-operative complications developed in 13 patients (43.3% of total cases). The most common complications in this study were CSF leak and/or pseudomeningocele (16.7%) ,cerebellar symptoms (10%), wound infection (6.7%),bulbar manifestation (6.7%) and post-

operative acute hydrocephalus (3.3%). In relation to the outcome, 63% of complicated cases had good outcome.

this is in agree with Cochrane et al.6 who reported 4 patients (5%) developed post-operative CSF leak, 2 of them required post-operative CSF diversion.

In correlation between the outcome of patients who received post-operative adjuvant therapy and those did not in our study, it was found that radiotherapy did not improve the outcome of patients or prevent recurrence of the tumor. As among patients received post-operative adjuvant therapy, only 77.8% had good outcome and this is in agree with Undjian and colleagues.14 and Sgouros et al.15 who did not find any improvement in prognosis after post-operative radiotherapy even when it was administered to patients with incomplete excision and brain stem involvement.

According to tumor pathology, recurrence occurred most commonly in medulloblastomas (60% of cases) followed by astrocytomas (40%). This result in agree with David et al.16 who reported recurrence in 37 patients (46%) with medulloblastomas.

In our study, it was found that recurrence of tumors was associated with poor prognosis as 66.7% of recurred tumors had poor outcome and this is in agree with Hayostek et al.17 and Morreale et al.18 who considered tumor pathology was the most important factor that determined tumor recurrence.

CONCLUSION

From this study, we could conclude that:

1-Proper management of posterior fossa tumors requires the following:

Careful general and neurological examination of patient with intractable headache and vomiting. They must be investigated by CT brain (with and without contrast) to exclude the possibility of posterior fossa tumors.

Pre-operative MRI (with and without contrast) is considered the most accurate imaging technique for the diagnosis of posterior fossa tumors. Pre-operative imaging of the spinal cord with MRI (with and without contrast) is required if the pre-operative MRI of the brain was highly suspicious of tumors which tend to metastasize into the spinal subarachnoid space as medulloblastomas and ependymomas. This helps in categorizing patients as high or low risk group and consequently helps in planning of the post-operative management.

Complete and precise knowledge of the gross and microsurgical anatomy and also, the surgical pathology of the posterior cranial fossa is mandatory in the management of posterior fossa tumors.

The presence of highly skilled neurosurgical team, expert neuroanaesthiologist, and special neurosurgical intensive care unit (ICU) is a primitive requirement for successful management of pediatric posterior fossa tumors.

Pre-operative shunting should be restricted to patients who had very bad general condition and/or patients with severe manifestations of increased ICP not fit for direct tumor excision.

Intra-operative ventriculostomy before dural opening to gain lax posterior fossa and smooth post-operative course should be used only if the classical medical measures to decrease ICP failed.

Post-operative CT or MRI brain (with and without contrast) helps in determining the extent of tumor resection. Also, urgent CT or MRI should be done in cases of any post-operative clinical deterioration.

Radiotherapy is an important adjuvant therapy in certain tumors as medulloblastomas, ependymomas and anaplastic astrocytomas and it is the main line of treatment of diffuse brain stem gliomas.

Chemotherapy has an important role in management of malignant posterior fossa tumors especially in children less than 4 years to avoid the hazards of radiation on the developing CNS.

2-Patients with definite neurological symptoms and signs of increased ICP and/or cerebellar dysfunction and their CT brains were not conclusive should be investigated with MRI (with and without contrast) before exclusion of posterior fossa tumors.

3-Most midline solid tumors were medulloblastomas, followed by ependymomas and cerebellar astrocytomas while, cerebellar astrocytomas were the most common lateral tumors.

4-Malignant posterior fossa tumors tend to affect infants and young children (less than 4 years old) than youth&elders.

5-Deteriorated level of consciousness, the younger the age of patients, pre-operative hydrocephalus, mid-line tumor location, incomplete tumor resection and post-operative pseudomeningocele appear to be important prognostic factors necessitating permanent shunt insertion.

6-A good outcome could be predicted.

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