

Surgical Outcome of Posterious Fossa Tumors in Children

ABDUL AZIZ KHAN, MUMTAZ ALI, KHALID KHANZADA

Muhammad Ali, Fazli Ghaffar

Department of Neurosurgery, Lady Reading Hospital, Peshawar

ABSTRACT

Posterior fossa tumors are most common in children than adults accounting for 54 to 70% of all childhood brain tumours. Out of them 30% are brain stem tumors. Most common posterior fossa tumors are Medulloblastoma, Astrocytoma, Ependymoma, and Brain Stem tumours. While dermoid, epidermoids and teratoma are the rare tumours. All focal and Cystic brain stem tumors in posterior fossa show better results with radical surgery than more diffused tumours that had stereotactic biopsy.

Key words: *Posterior Fossa Tumors.*

INTRODUCTION

The posterior cranial fossa containing roughly one fourth of the intracranial contents, is an important site of intracranial tumours in children. Approximately half of the childhood brain tumours arise in posterior cranial fossa. These tumours are medulloblastoma, cerebellar astrocytoma, ependymoma, cerebellar anaplastic gliomas, brain stem gliomas and choroids plexus papillomas, dermoid / epidermoid posterior fossa tumours. These tumors cause brain stem or cerebellar dysfunction and often associated with the blockage of fourth ventricle and hydrocephalus¹.

In children the commonest clinical features are headache, vomiting, gradual deterioration of vision, lower cranial nerve palsies and cerebellar dysfunction. Upward herniation of superior vermis of cerebellum through the tentorium may compromise diencephalic structures and downward displacement may compromise medulla resulting in alteration in level of consciousness or cardiorespiratory arrest².

The delay in the diagnosis is the hallmark of patients with posterior fossa tumours in our country and majority of them have already lost their vision by the time they reach neurosurgeon³. Early diagnosis and adequate surgical excision of these lesion followed by radiotherapy and chemotherapy can provide a better and bright future for these patients⁴.

MATERIAL AND METHODS

This study was carried out at Neuro Surgery Department, Government lady Reading Hospital Peshawar

from 24th May 2006 to 23th May 2007. It was descriptive study conducted on 30 patients with inclusion criteria as age under 14 years, both sexes and exclusion criteria as age more than 14 years, cases operated else where, patient not willing for surgery, Patient not fit for surgery, brain stem tumors. which are inoperable.

RESULTS

Sex Incidence

Out of total (n = 30) patients 15 were male and 15 were female giving male to female ratio 1:1.

Age Range

The mean age of the patient in our study was 9.2 years SD \pm 3.37. With pre-dominance between the age 7 to 10 years (40%).

Histopathology

The incidence of various posterior fossa tumors in our series as shown in Figure 2. Medulloblastoma n = 12 (40%), Astrocytoma n = 10 (33.3%), Ependymoma n = 6 (20%), Teratoma n = 1 (3.3%), Dermoide n = 1 (3.3%).

Complications

Similarly the incidence of various post operative complications are Cerebellar Mutism, with Cerebellar Ataxia n = 4 (13.3%), Meningitis n = 2 (6.6%), CSF

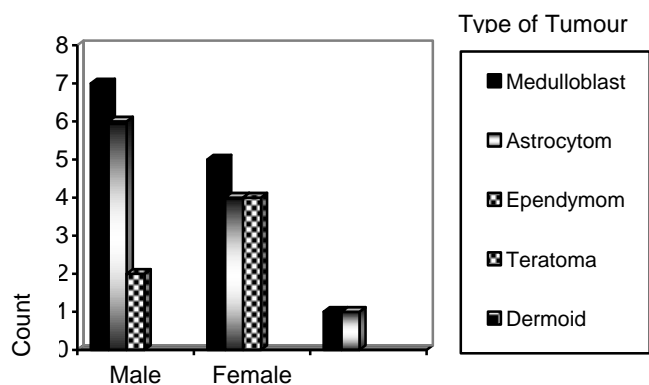


Fig. 1: Incidence of Various Tumors in Age Group Tumors.

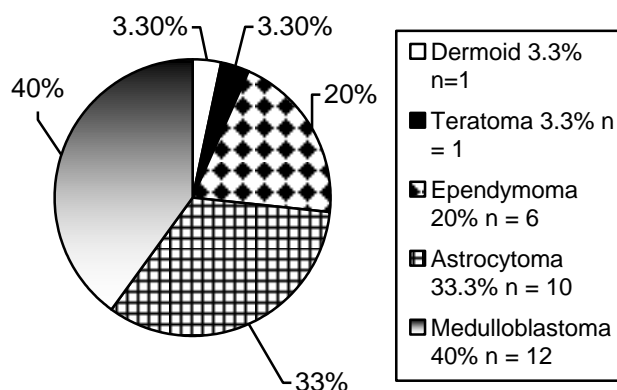


Fig. 2: Frequency of Various Tumors.

Table 1: The Occurrence of complications in different tumors (n = 30).

		Complications						Total
		Meningitis	CSF Leak	Cerebellar Mutisim	Cranial Nerve 7 th Palsy	Hydrocephalus	Mortality	
Type of Tumour	Medulloblastoma	1		2		1		12
	Astrocytoma		1	2				10
	Ependymoma		1		1	1	1	6
	Teratoma							1
	Dermoid	1						1
Total		2	2	4	1	2	1	30

ANALYSIS OF PRE AND POST treatment

Leak n = 2 (6.6%), Post operative Hydrocephalus n = 2 (6.6%) and Cranial Nerve Palsy n = 1 (3.3%) as shown in Table 5. While 2 patients died in 6 months follow up.

NEUROLOGICAL OUTCOME

The following grading procedure was employed for assessment of the patients on follow up.

Grade I: Excellent: Asymptomatic with no neurological deficit.

Grade II: Good : Has been left with mild neurological deficit.

Grade III: Fair: Partially dependent, can look after himself.

Grade IV: Poor: totally dependent.

Neurological out come discussed here is the status of the patients on (1st week to 6 months).postoperatively.

Twelve patients out of 30 were in grade IV pre-operatively, 7 were in grade III and 11 were in grade II. Postoperative and after completion of radiotherapy (03 – 06) months after surgery), 08 patients were in grade III, 11 patients were in grade II and 8 patients were in grade I. 2 patient expired. 1 patient was in grade IV.

Table 2: Neurological Status in Grades.

Tumour Type	No of Cases	PRE TREATMENT				POST TREATMENT				Grade V Mortality
		Grade I	Grade II	Grade III	Grade IV	Grade I	Grade II	Grade III	Grade IV	
Medulloblastoma	12	1	6	2	3	5	2	2	1	2
Astrocytoma	10	0	2	2	6	0	6	4	0	
Ependymomas	6	2	2	2	2	1	2	2	0	1
Dermoid	1	0	1	0	0	0	1	0	0	
Teratoma	1	0	0	1	0	1	0	0	0	
Total	30	1	11	7	11	8	11	8	1	3

Four patients with decreased vision had marked improvement in the visual acuity after the intracranial pressure was relieved.

Two patients were blind on presentation. They had no improvement after treatment. Sixteen patients out of 20 who were unable to walk initially started walking after completion of radiotherapy. Five patients out of 20 had relief of the symptoms of diplopia.

The analysis of pre and post treatment neurological status is shown in Table below.

Grade I : Normal with no neuro deficit.

Grade II: Has been with mild neurological deficit.

Grade III: Partially dependent, can look after himself.

Grade IV: Totally dependent.

Note: 2 Patients died post operatively in 6 month follow up.

DISCUSSION

Brain tumors are the second most common malignancy to occur during child hood. Posterior fossa is the most common region of the brain affected whose diagnosis and treatment is complex as well as challenging.⁵

Posterior fossa tumors can be divided into three groups. Cerebellar tumors, 4th ventricular tumors and brain stem tumors⁶.

In our study brain stem tumors were excluded. While 4th ventricular tumors and cerebellar tumors

were included. Brain tumours is the most common malignancy secondary to leukemia. Posterior fossa is constituting about half of paediatric brain tumours⁷. Total number of 69 patients of brain tumors both infratentorial and supratentorial in age group 0 – 13 years were operated in the study period. Thirty out of them were posterior fossa tumours, so almost the same incidence is seen in our study, which shows 43.5% of posterior fossa tumors occurred in children, with equal male to female incidence. Out of 30 patients 15 were male, and 15 were female. The international study carried out by Zakrzewski shows male predominance¹ with male to female ratio of 1.6 : 1. Study carried out at children hospital Lahore also shows male predominance⁸. Age distribution in our patients was 03 – 13 years with average age of 6.46 years. The peak incidence of medulloblastoma, was at the age of 8 years comparable with the incidence of medulloblastoma noted by slampa and his colleague in study carried out between 1997-2005⁹.

The survival of the patients with medulloblastoma varies from poor to excellent depending upon several variables. The period of risk of recurrence in general can be calculated from Collin's law. The period of recurrence is the age of the patients at the time of diagnosis plus 9 months¹⁰. But freedom from relapse after 8 years is considers as a cure by some authors¹¹. In our study we could appreciate early recurrence in 3 cases of medulloblastoma in 6 months follow up in whom total resection was impossible because of the location.

The second most common tumor in our study is Pilocytic Astrocytoma $n = 10$ (33.3%), and third most common is Ependymoma $n = 6$ (20%). Study carried out in Syria between 1993 – 2002¹² showed astrocytoma and ependymoma to be the second and third commonest tumors. The incidence of Teratoma was 3.3% in our study while the incidence mentioned in literature review of case report by Das – 2007 Jan¹³ shows 3% incidence.

Dermoid cyst also has got 3.3% incidence in our study. Study shows 0.7 to 1% occurrence¹⁴. So probably due to small size of sample the percentage of dermoid is higher in our study.

Majority of the patients presented with very late presentation of symptoms with 2-3 month duration. They go to general practitioners, paediatricians and physicians. By the time they reach us they are very late. Two patients were almost blind on presentation. The same delay in presentation was noted in a study carried out in India by Kumar⁴.

The common presenting complaints were headache, vomiting difficulty in walking and cranial nerve involvement and papilloedema.

The symptoms of headache and vomiting were the highest in study carried out by Vohra et al¹⁵.

Various diagnostic modalities were used in our series. Plain X-Ray skull, which showed calcification (teeth) in case of teratoma and two cases of ependymoma. Similarly signs and symptoms of increased intracranial pressure were there in 46% of the cases in form of sutural diastasis. Zaman reported an evidence of increased intracranial pressure is 40% of patients with posterior fossa tumors on X-Ray skull.³ Second line of investigation was C.T Brain with Special cuts for posterior fossa. Which gave us 90% sensitivity but 60% specificity. While in international study its 100% sensitivity 84.7% accuracy¹⁶ has been reported.

MRI brain was considered the ultimate decision about the location and radiological diagnosis of the lesion. Study of Sarkar et al show 100% sensitivity and 99% specificity was reported¹⁶.

Surgical resection and decompression of masses of the posterior fossa demand the best neurosurgical skills and neuroradiological localization.

All surgical procedures were carried out in prone position. Park also feels comfortable on operating patients 44 in prone position than sitting position¹⁷, which is associated with higher risk of embolism and uncontrolled CSF flow. Craniectomy was done, in all cases for exposure of posterior fossa. While Ganalingham

also found craniectomy Most safe and easy procedure for his study¹⁸.

Safty burrhole (Frezeir Burrhole) x was always made for EVD Dura opened in Y – Shaped manner and maximum safe total excision of tumor was done depending upon tumor type, location consistency. Dura was closed in water tight fashion. As most of the patients presented very late with hydrocephalus. So in fifteen patients preoperative shunting was done to deal with emergency, as most of them presented with gradual deterioration of vision, cranial nerve palsies and papilloedema.

Owing to the complications of shunting some studies suggest that preoperative shunting for posterior fossa tumor is not indicated¹⁹. While in developing countries like us, the disease is diagnosed in later stage and preoperative shunting is advisable²⁰. The same conclusion was derived from the study carried out at Pakistan institute of medical sciences Islamabad²¹.

Ten patients in our study EVD was placed preoperatively while in remaining 5 patients no CSF diversion procedure was carried out preoperatively or peroperatively. Out of these 5 patients, 2 develop CSF leak and 2 develops post operative hydrocephalus while in one of the study carried out by Abdullah Hus-sain. Out of 5 non-shunted patients, 3 develop hydrocephalus, 1 develop CSF leak²². Another study carried out at the department of neurosurgery and pediatric Turkey shows similar percentage (7.4%) of post operative complication.²³

Similarly 10 patients had EVD preoperatively out of 10 patients 2 develop meningitis, while in the study of Abdull ul Hassaini, 3 patients with EVD develop infection out of 13 patients. Another well established complication in our study was cerebellar mustism with ataxia. 4 patients 13 % out of 30 suffered cerebellar mustim, although 2 of these were transient and covered after a week. While in international study upto 8% cases have been mentioned. This little increase of incidences is due to delayed presentation.²⁴

In the mean while 1 patient died due to post operative hematoma. While another one died due to Meningitis and blockage of shunt. During 6 months follow up. Study carried out at Turkey show 14.8% mortality in long term follow up and the cause was recurrence. While in our study only 5 cases 3 medulloblastoma and 2 ependymoma showed early recurrence because of the sub total resection. So non of the patients died due to recurrence in 6 months follow up.

CONCLUSIONS AND RECOMMENDATION

Posterior fossa tumor in children comprise almost half of the childhood brain tumours with almost equal sex distribution.

The common presentation is gait disturbances, Head ache, Vomitting, Diplopia, ataxia and long tract signs. Medulloblastoma, Astrocytoma and Ependymoma are the commonest tumors of the posterior fossa. However dermoid and teratoma can also occur rarely. C.T Brain, and MRI are the investigations of choice.

To avoid most of the complications preoperatively ventriculo peritoneal Shunt is preferable in our setup as most of the patients present very late with hydrocephalus and visual deterioration. The external ventricular drain needs proper nursing care to avoid complications like infection. With proper backup in the form of neuro navigation and proper ICU facilities the per operative and post operative complications and mortality can be reduced to minimum.

More over with post operative radio therapy and Chemo therapy the survival of the patient with Medulloblastoma and Ependymoma can be improved in long term follow up.

So I recommend that any child with persistent head Ache, vomiting should be investigated for posterior fossa lesion before the complication are developed. There should be a proper liaison between pediatrician, neuro surgeon and oncologist to decided the future treatment of the patients.

ICU facilities should be enhanced and the working staff should be trained properly in post operative care. Awareness of the neurosurgical patients among the general practionar is the need of the hours so that they could refer the patients to the neuro surgical centre before irreparable brain damages done.

*For correspondence contact:
Dr. Abdul Aziz Khan
Medical Officer Neuro Surgery
Lady Reading Hospital, Peshawar
Contact No : 0300-9176146*

REFERENCES

1. Zakrzewski K, Fiks T, Polis L, Fiks T, Liberski PP. Posterior Fossa tumour in children and adults. A clinical pathological study of 216 cases. *Folia neuropathol* 2003; 41 (4): 251-2.
2. Changeof. Recent advances in childhood brain tumour. *KHJ Paediatr* 2006; (11): 3-12.
3. Zaman KU. posterior fossa tuomour in children: twenty months experience at P.I.M.S. *J Surg* 1990 Vol (1): 18-20.
4. Kumar R. Scenaria of pediatric Cys tumour. *India: JK Science (Elective)* 2006; 8-190.
5. Vernon-Levett P, Geller M. Posterior fossa tumors in children: A case study. *AACN Clin Issues* 1997; 8: 214-26.
6. Taveras JM. *Neuroradiology*. Baltimore, Williams and Wilkins, 1996; 676-84.
7. Badh PB, Chauhan PP, Mehta NK, Brainstem, Gliomas-A. Clinicopathological study of 45 cases with P53-immunohistochemistry. *Indian: J Canar* 2004; (41): 170-4.
8. Qureshi AA, Rehman MU. Posterior fossa masses in children, imaging features with emphasis on MR findings. *Pak J Pathol Sep* 2001; 12 (3): 135-8.
9. Slampa, P, Parelka Z, Dusek L. Long Term treatment results of child hood medulloblastorrla by craniospinal irradiation in supine position. *Neoplasma* 2007; 54 (1): 62-70.
10. Brown WD, Tavare CJ, Sobel EL, Gilles FH, Medulloblastoma and Colin's law: A critical review of the concept of a period of risk for tumor recurrence and patients survival. *Neurosurgery* 1995; 36: 691-7.
11. Belza MG, Donaldson SS, Steinberg GK, Cox RS, Cogen PH. Medulloblastoma: Freedom from relapse longer than 8 years-a therapeutic cure? *J Neurosurg* 1991; 75: 575-82.
12. Kadnri H, Mewla AA, Murad L. Incidence of childhood brain tumors in Syria (1993-2002). *Pediatr Neurosurg* 2005; 41 (4): 173-7.
13. Das S, Muro K, Goldman S, Rajaram V, Dipatri AJ. Medulloblastoma arising from immature Teratoma of posterior fossa case report. *J Neurosurg* 2007; Jan 106 (Suppl): 61-4.
14. Layadi F, Lauhab N, Lmejjah M, Amiba K, Aitelquadi A, Aitbenali S. Cerebellor dermoid cyst with occipetal dermal sinus report of two Pediatric cases. *Pediatr Neurosurg* 2006; 42 (6): 387-90.
15. Vohra AH, Ahmad M, Ali IR. Medulloblastoma clinical presentation and management. *Specialist, Pak J Med. Sci:* 1993; 10: 1: 49-54.
16. Sarkar S, Hossain MA, Mazunder U, Rehman KM, Akamda NL. Clinico Pathological study in Large Posterior Fossa midline Tumors. *Mymensingh Meds* 2007 Jan; 16 (1): 43-9.
17. Park TJ. Simple Technique for posterior fossa Craniotomy in adult. *J Korean Neurosurg* 2006; (40): 206-9.
18. Gnanolingham KK, Lafuonte J, Thompson D, Harkness W, Hayward R. Surgical Procedure for posterior fossa tumors in Children. Does Craniotomy lead to fewer complication Than Craniectomy. *J Nurosurg* 2002; (97): 821-6.
19. Whither AG. Preoperative Shunts for Posterior fossa tumors. *J Neurosurg* 1993; 7 (4): 395-9.

20. Griwan MS, Sharma BS, Mahajan RK, Kak VK. Value of Precraniotomy Shunts in Children with posterior Fossa tumors. Childs Neurosyst 1993 Dec; 9 (8): 462-5.
21. Ghani E, Zaidi IG, Nadeem M, Rehman L, Noman AM, Zaman KU. Role of CSF Diversion in Posterior Fossa tumor Surgery. J Coll Physicians surg Pak Jun 2003; 13 (6): 333-6.
22. Abdollahzadeh SM, Hosseini, Rezaishiraz H, Allah Dini F. Management of Hydrocephalus in Posterior Cranial fossa tumors. Acta Medical Iranica 2006; 44 (2): 89-94.
23. Akay MK, Izci Y, Baysefer A, Atabey C, Kismet E, Timurkaynak E. Surgical Outcomper of Cerebellar tumors in children. Pediatr Neurosurg 2006; 40: 220-5.
24. Doxey D, Bruce D, Sklar F, Swigr D, Kenshapir. Posterior Fossa Syndrome: Identifiable risk factor and irreversible complication. Pediatric Neurosurgery 1999; (31): 131-6.