Epidemiology Of Posterior Fossa Tumors In Paediatric Age Group: An Institutional Study

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ABSTRACT

Aim: To analyse the epidemiology of posterior fossa tumors in paediatric age group.

Material and Methods: The present study was conducted in the department of neurosurgery, among all the diagnosed cases of posterior fossa tumours attending and being admitted and treated to our institute during the study period were included in the study. During the study period, 480 cases of CNS tumours were reported, out of which 336 and 144 were supratentorialand infratentorial respectively.

Results: Incidence of posterior fossa tumors (<12 Years) was reported among 96 subjects (20%), out of which 50 subjects were recruited for the present study according to the inclusion and exclusion criteria. Medulloblastoma, ependymoma, JPA and glioma was diagnosed among 48%, 24%, 20% and 8% of the subjects respectively. Complications viz. hydrocephalus, postoperative mutism, cerebrospinal fluid leaks, wound infection, cerebellar hematoma and CN palsies was found among 12%, 12%, 10%, 8%, 4% and 4% of the subjects respectively in our study. Mortality was reported among 8% (4) of the subjects.

Conclusion: Early diagnosis of posterior fossa tumours is vital to prevent potential risks of Brain stem compression, herniation, hydrocephalus and death. With rapid advancement in radiology and advent of modern therapeutic modalities, early diagnosis and treatment reduced the morbidity and mortality rate and improved prognosis among the patients.

Keywords: Pediatric, Tumor, Posterior Fossa

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INTRODUCTION

Tumors of the brain are regarded as one of the most devastating group of neurological diseases-they are associated with significant neurological morbidity, they lead to progressive physical, cognitive and emotional dysfunction and are frequently fatal. The term brain tumor is used to describe both primary tumors that originate from the brain, cranial nerves, pituitary gland or meninges and secondary tumors (metastases) that arise from organs outside the nervous system[1]. Brain tumor is one of the most devastating forms of human illness, especially when

occurring in the posterior fossa and involving the brainstem. Cushing probably was the first to report a large series of posterior fossa tumors[2].

The posterior fossa is the commonest site of primary intracranial tumours in children. Posterior fossa tumours account for 45-60% of all pediatrics tumours[3]. The posterior fossa is the most frequent location of brain tumors in children in contrast to adults who typically have supratentorial lesions. The median time interval between symptom onset and tumor diagnosis is two months[4]. Neurology, anatomical localization, extension, histology, treatment options, short and long term survival and prognosis as well as functional outcome differs significantly comparing children with adults concerning neoplastic posterior fossa lesions. In young children (0-10years), posterior fossa tumors are more frequent than supratentorial lesions, in older children (>10 years) the incidence of supratentorial tumors gradually increases with age[5].

The most common tumours include juvenile pilocytic astrocytoma (JPA) which is the most common pediatric benign cerebellar neoplasm, medulloblastoma, ependymoma, and brain stem glioma are also common tumours, oligodendroglioma is a rare pediatric low grade, slowly growing cerebral hemispheric tumour and account for only about 1%.[6,7] Medulloblastoma is the most common malignant cerebellar tumours in pediatric, these tumours have aggressive course with tendency of CSF dissemination and fatal outcome. Ependymoma is the third posterior fossa tumours and arising from the floor of the fourth ventricle with a peak incidence between 3 to 5 years[8,9].

Using current treatments, 80%–90% of those without disseminated disease can be cured; however, treatment for this disease often results in significant endocrinology and intellectual sequelae. Local data are not available. The 5-years survival rates exceed 60% for all patients and 80% for certain good risk individuals with posterior fossa tumors. In the cases of pilocytic cerebellar astrocytoma, the 25-year survival rate exceeds 94%. According to one recent study, about 90% of the patients had satisfactory surgical outcome after surgical removal[10].

In view of the available background of this topic this study is very much relevant to know the epidemiology and outcome of posterior fossa SOL in paediatric age group in tertiary care neurosurgery centre in Eastern India. Hence the present study was conducted to analyse the epidemiology of posterior fossa tumors in paediatric age group.

MATERIAL AND METHODS

The present non-randomized prospective observational study was conducted in the department of neurosurgery among all diagnosed cases of posterior fossa tumours attending and being admitted and treated to our institute during the study period were included in the study. Ethical clearance to carry out the study was obtained from the ethical committee of the institute. The study protocol was explained to the patient/guardian and a written informed consent was obtained from each subject to be enrolled in the study.

INCLUSION CRITERIA

a) Age equal or less than 12 years.
b) Evidence of posterior fossa tumor in neuroimaging studies.

EXCLUSION CRITERIA

a) Age more than 12 years.
b) Prior surgery in another centres.
c) Parent's unwillingness to underwent surgery.

CASE SELECTION

During the study, a total of 50 patients were recruited. The data was collected by a preformed structured interviewer-administered questionnaire that was pretested with modifications made prior to its use in the study. The patients were interviewed that requests for the demographic, socioeconomic status, medical history and previous history of taking any medications and supplements.

DATA COLLECTION

After written informed consent the following variables were recorded in all patients: Age and sex distribution, duration and nature of symptoms, whether a pre or postoperative ventriculo-peritoneal (V-P) shunt was inserted, tumor type on histological examination and surgical outcome and complications.

The clinical follow-up observation was carried out by outpatient review. The patients were regularly followed up at 3, 6, 12 months after operation. All the data was recorded in a pre-designed and pre-tested proforma.

PARAMETERS TO BE STUDIED

Details of the patient in terms of clinical history taking, a thorough physical examination with special focus on central nervous system and relevant radiological investigations were recorded. Various manifestations of the disease were recorded too.

STUDY TOOLS

(A) Clinical: History taking and clinical examination with help of pre-designed and pre-tested proforma.

(B) Investigation:

[1] Routine hemaematological biochemical tests.

[2] Radiological investigation:

a. NCCT Brain

b. MRI Brain (P+C) with spinal screening

[3] Histopathology.

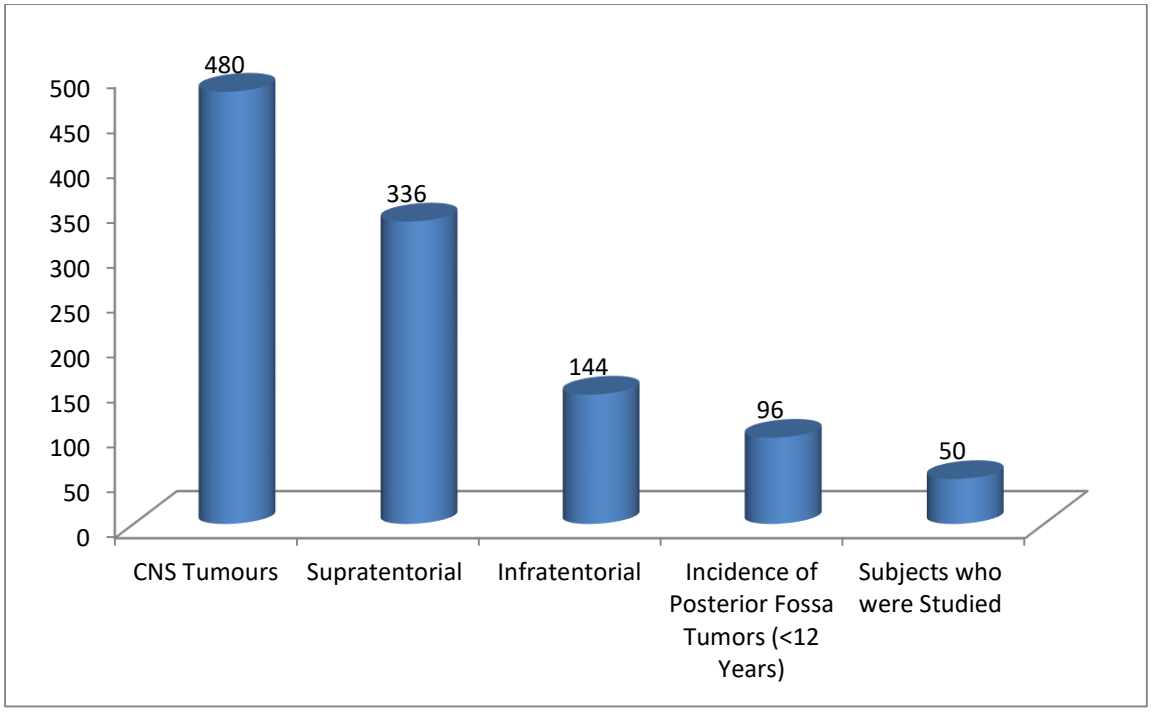
STATISTICAL ANALYSIS

Data so collected was tabulated in an excel sheet, under the guidance of statistician. The means and standard deviations of the measurements per group were used for statistical analysis (SPSS 22.00 for windows; SPSS inc, Chicago, USA). The level of significance was set at $p < 0.05$.

RESULTS

During the study period, 480 cases of CNS tumours were reported, out of which 336 and 144 were supratentorial and infratentorial respectively. Incidence of posterior fossa tumors (<12 Years) was reported among 96 subjects (20%), out of which 50 subjects were recruited for the present study according to the inclusion and exclusion criteria (graph 1). In our study, there is male dominance i.e. out of 50 subjects, there were 32 males (64%) and 18 females (36%). 6%, 34% and 60% of the subjects were having age <1, 1-5 and 5-12 years respectively as shown in table 1.

Headache and vomiting was the most common complaint reported among 82% of the subjects. Visual complaints, ataxia and increased head size was revealed among 28%, 8% and 6% of the subjects respectively. Most common location of posterior fossa tumour was fourth ventricle (72%) followed by cerebellum (20%) and brainstem (8%) as shown in table 2.



Graph 1: Incidence of posterior fossa tumors among the study subjects during the study period

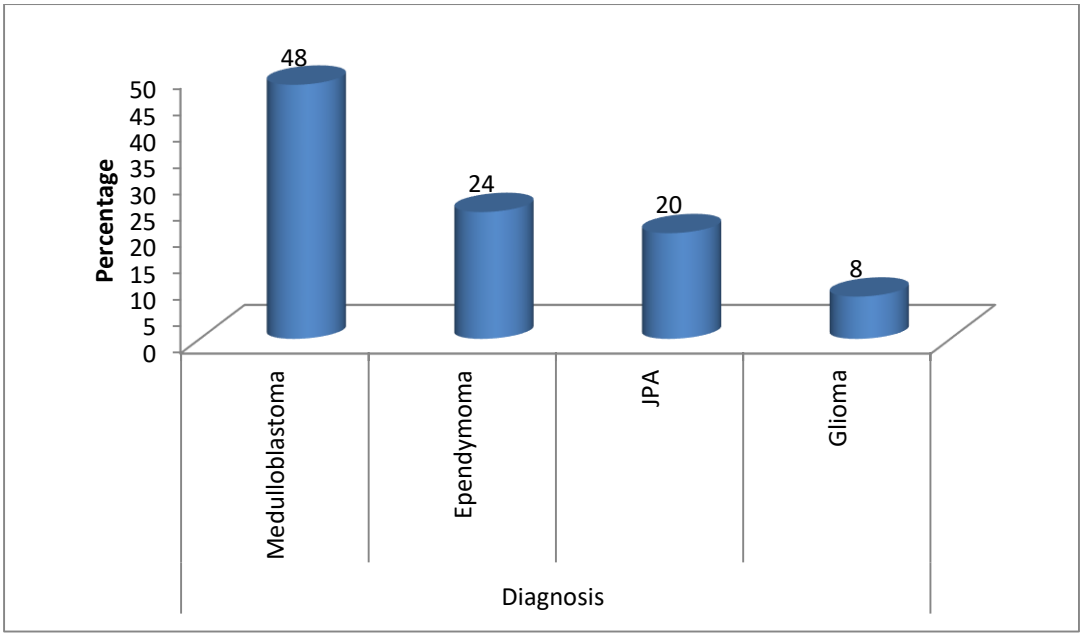
Table 1: Gender and age distribution among the study subjects

Gender	**N**	**%**
Male	32	64
Female	18	36
Age (in years)		
<1	3	6
1-5	17	34
>5-12	30	60
Total	50	100

Table 2: Complains, Posterior Fossa Tumours based on location and brainstem involvement among the study subjects

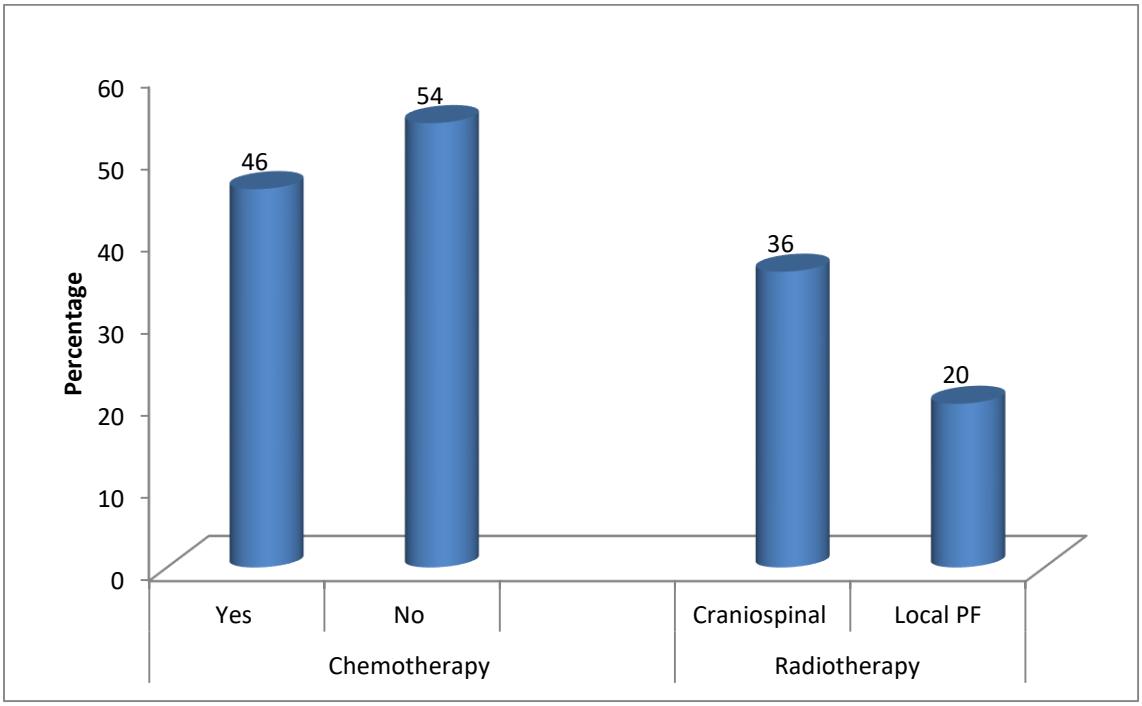
Complaints	**N**	**%**
Headache and Vomiting	41	82
Visual Complains	14	28
Ataxia	4	8
Increased Head Size	3	6
Site/Location		
Fourth Ventricle	36	72
Cerebellum	10	20
Brainstem	4	8
Brainstem Involvement		
Yes	22	44
No	28	56
Total	50	100

In the present study; medulloblastoma, ependymoma, JPA and glioma was reported among 48%, 24%, 20% and 8% of the subjects respectively (graph 2).



Graph 2: Diagnosis among the study population

46% of the subjects were given chemotherapy while radiotherapy viz. craniospinal and local PF was given to 36% and 20% of the subjects respectively (graph 3). Complications viz. hydrocephalus, postoperative mutism, cerebrospinal fluid leaks, wound infection, cerebellar hematoma and CN palsies was found among 12%, 12%, 10%, 8%, 4% and 4% of the subjects respectively.



Graph 3: Treatment among the study subjects

In this study, overall mortality was reported among 8% (4) of the subjects. When we analysed mortality according to different types of posterior fossa tumor, mortality was revealed among 4.17%, 8.33% and 50% of the subjects having medulloblastoma, ependymoma and glioma respectively (table 3).

Table 3: Surgical outcome of posterior fossa tumor during hospital stay

Diagnosis	N	Mortality During Hospital Stay	%
Medulloblastoma	24	1	4.17
Ependymoma	12	1	8.33
JPA	10	0	0
Glioma	4	2	50
Total	50	4	8

DISCUSSION

Posterior cranial fossa tumours in children differ from adults in their clinical presentation, behaviour, management and prognosis. Pilocytic Astrocytoma and Medulloblastoma are most prevalent tumours in childhood, and schwannomas and meningiomas were most frequent in adults. Medulloblastomas have varied prognosis and long-term survival rates are generally lower for in patients 3 years or younger. Posterior fossa tumours warrant surgical management to achieve decompression of the posterior fossa, relieve pressure on the brain stem, release intracranial pressure and avert the risk of herniation. Surgery is also mandatory for histopathological diagnosis of the tumour and to determine further plan of management depending on the nature of thetumour[11].

Approximately 90% of patients had a successful surgical result following surgical removal[12], according to one recent report. This research is very important to understanding the epidemiology and outcome of posterior fossa SOL in the paediatric age group in the tertiary care neurosurgery centre in Eastern India, taking into account the available context of this subject. Therefore, the latest research was carried out in the paediatric age group to investigate the epidemiology of posterior fossa tumours.

During the study period, 480 cases of CNS tumours were reported, out of which 336 and 144 were supratentorial and infratentorial respectively. Incidence of posterior fossa tumors (<12 Years) was reported among 96 subjects (20%), out of which 50 subjects were recruited for the present study according to the inclusion and exclusion criteria. These findings were little lower as compared to study done by kalyani et al[13] who reported 40.9% of posterior fossa tumors in their institute. Gubbala et al[14] in their study revealed that 80 cases of posterior fossa tumours were analysed and these constituted 69% of total CNS tumours in children below 5 years reported in their institute. In adults, posterior fossa tumours constituted from 32.5% of total CNS tumours in fifth decade to 24.1% in fourth decade. Intracranial posterior fossa tumours constituted 37.03% of total CNS tumours in their study. Similar studies done by Srilakshmi et al[15] (21.8%) and Meenakshi Sundaram et al[16] (60%) showed great variation in prevalence probably related to demographic attributes.

In our study, there is male dominance i.e. out of 50 subjects, there were 32 males (64%) and 18 females (36%). Salami A et al[17] in their study found male to female ratio of 1.57:1. Dawood MA et al[18] in their study found that 60% and 40% of the subjects were male and female respectively. Majority of the tumours in posterior Fossa had male predilection with M:F ratio of 1.6:1 as mentioned by Gubbala et al[14] in their study. These findings are similar to our study. Similar male dominance was reported by Srilakshmi et al[15] and Kalyani et al[13] in their study.

6%, 34% and 60% of the subjects were having age <1, 1-5 and 5-12 years respectively in the present study. Ahmad KB et al[19] in their study showed that posterior fossa tumours are frequently seen in pediatric age groups most of the malignant tumours were seen in first two decades of life with mean age of 12.69 yrs. Piscione PJ et al[20] in their study revealed mean age of 11.4years.

Most common location of posterior fossa tumour was fourth ventricle (72%)followed by cerebellum (20%) and brainstem (8%) in the present study. Salami A et al[17] in their study mentioned that more than half of the tumours (56.94%) arose from the cerebellar

hemispheres and ten cases (18.89%) from the 4th ventricle. Dawood MA et al[18] in their study revealed that all cases of pilocytic astrocytoma were seen located near the mid line arising from cerebellum as well-circumscribed, cystic like masses with a soft tissue mural nodule.

The difference in the rate of tumour location in the vermis and fourth ventricle may probably be reflected in the more frequent occurrence of medulloblastomas and Pilocytic astrocytoma, which have a predilection for the vermis and fourth ventricle, in the paediatric population. This difference might be probably related to demographic attributes.

In our study, brainstem involvement was reported among 44% (22) of the subjects. Piscione PJ et al[20] in their study found brainstem involvement in 30% of the subjects.

In the present study; medulloblastoma, ependymoma, JPA and glioma was diagnosed among 48%, 24%, 20% and 8% of the subjects respectively. Rami A et al[21] in their study found that medulloblastoma was the most common posterior fossa tumour. Dawood MA et al[18] in their study reported that MRI study revealed 16 cases diagnosed as medulloblastoma (40%), 8 cases as Ependymoma (20%) (Case 6), 8 cases as juvenile epilocytic astrocytoma (20%), 6 cases as pontine Glioma (15%) and 2 case as Schwannoma (5%). Similarly in a study by Piscione PJ et al[20]; medulloblastoma, cerebellar astrocytoma, ependymoma and brainstem glioma among 12 (40), 13 (43.3), 2 (6.7) and 3 (10) subjects respectively.

Complications viz. hydrocephalus, postoperative mutism, cerebrospinal fluid leaks, wound infection, cerebellar hematoma and CN palsies was found among 6 (12%), 6 (12%), 5 (10%), 4 (8%), 2 (4%) and 2 (4%) of the subjects respectively in our study. In a study by Rami A et al[21], general complication rate was 41.29%. Hydrocephalus, postoperative mutism, cerebrospinal fluid leaks, wound infection, cerebellar hematoma and CN palsies was found among 21, 19, 12, 11, 7 and 6 subjects respectively.

In the present study, overall mortality was reported among 8% (4) of the subjects. When we analysed mortality according to different types of posterior fossa tumor, mortality was revealed among 4.17%, 8.33% and 50% of the subjects having medulloblastoma, ependymoma and glioma respectively. Ahmad KB et al[19] in their study revealed intraoperative and postoperative mortality was 6.77%. The overall mortality rate related to surgery was 6.47% occurring in 13 patients as reported by Rami A et al[21] in their study.

This study has few limitations. First, a quantitative analysis could not be established due to great heterogeneity in the included studies, different patient populations of different ages, and multiple tumour subtypes. Hence we suggest more detailed appraisals for each encountered histological pattern and clinical manifestation. The diagnostic approach for brain tumors is changing significantly.

CONCLUSION

Posterior fossa tumours are critical brain lesions with significant neurological morbidity and mortality. Early diagnosis of posterior fossa tumours is vital to prevent potential risks of Brain stem compression, herniation, hydrocephalus and death. With rapid advancement in radiology and advent of modern therapeutic modalities, early diagnosis and treatment reduced the morbidity and mortality rate and improved prognosis among the patients.

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