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# PAEDIATRIC POSTERIOR FOSSA TUMORS : AN EPIDEMIOLOGICAL AND ANALYTICAL STUDY AT TERTIARY CARE CENTRE IN EASTERN INDIA

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| **Dr. S. I. Sadique** | Associate Professor, Dept. of Neurosurgery, B.I.N, I.P.G.M.E & R, Kolkata, India. |
| **Dr. Md. Shahid Alam*** | Senior Resident,Dept. of Neurosurgery, B.I.N, I.P.G.M.E & R, Kolkata, India.*Corresponding Author |
| **Dr. S. Chatterjee** | Professor, Dept. of Neurosurgery, B.I.N, I.P.G.M.E & R, Kolkata, India. |
| **Dr. S. Ghosh** | Professor, Dept. of Neurosurgery, B.I.N, I.P.G.M.E & R, Kolkata, India. |

**ABSTRACT** **Introduction:** Posterior fossa is the commonest site of primary intracranial tumors in children, accounting for 45-60% of all pediatric tumors[1]. The aims and objectives of the study is to analyse the incidence, clinical features, surgical outcome and complications in paediatric patients with posterior fossa tumor.

**Material and Methods:** The present study is a non-randomized prospective observational study, conducted in the department of neurosurgery, Bangur Institute of Neurosciences (B.I.N), IPGME & R, Kolkata from January 2019 to December 2020. Sample size is 50.

**Observations & Results:** Out of 480 cases of total CNS tumors who presented in the study period, 96 cases(20%) were of paediatric posterior fossa tumors. Male dominance was seen i.e. 32 cases(64%). Most of them were in the age group 5-12 years i.e. 30 cases(60%). Headache and vomiting was the most common presenting complain present in 41 cases(82%). Fourth Ventricle was the most common location, 30 cases(60%) with Medulloblastoma being the most common tumor, 24 cases(48%). Brainstem involvement was seen in 22 cases(44%). Post-op hydrocephalus and cerebellar mutism were seen in 6 cases(12%) each. Overall mortality was 8%(4 cases).

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

**KEYWORDS :** Posterior fossa tumors, Medulloblastoma, Hydrocephalus.

## INTRODUCTION:

Brain tumor is one of the most devastating forms of human illness, especially when occurring in the posterior fossa and involving the brainstem. They are associated with significant neurological morbidity. They lead to progressive physical, cognitive and emotional dysfunction and are frequently fatal. The posterior fossa is the commonest site of primary intracranial tumors in children. Posterior fossa tumors account for 45-60% of all pediatric tumors[1]. The median time interval between symptom onset and tumor diagnosis is two months[2]. 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. Juvenile Pilocytic Astrocytoma (JPA) is the most common pediatric benign cerebellar neoplasm. Among the malignant tumors Medulloblastoma, Ependymoma, and brain stem glioma are also common tumors. Medulloblastoma is the most common malignant cerebellar tumors in children. These tumors have an aggressive course with tendency of CSF dissemination and fatal outcome. Ependymoma is the third most common posterior fossa tumors arising from the floor of the fourth ventricle with a peak incidence between 3 to 5 years[3,4]. The spectrum of posterior fossa neoplasm includes Cerebellar Astrocytoma (30-35%), Medulloblastoma (20-25%), Brainstem gliomas (20-25%) and Ependymoma (10-15%). This series of tumors accounts for approximately 80- 90% of all posterior fossa tumors within the first decade of life. Genetic factors, such as dysfunction of some tumor suppressor genes especially; p53 gene and activation of some oncogenes, may play a role in their development. Environmental factors such as irradiation and toxins may also play a role[5]. Posterior fossa tumors often present with clinical manifestations of hydrocephalus and raised intracranial pressure. More aggressive tumors present with a shorter history. The most prevalent symptoms include headache, nausea and vomiting. Raised intracranial pressure may also cause drowsiness, neck stiffness, sixth nerve palsy and visual disturbances. Papilloedema is common in patients presenting with long-standing progressive symptoms. A head tilt may be a reflection of tonsillar herniation or a fourth nerve palsy related to a diffuse brainstem tumor. Young children with progressive hydrocephalus demonstrate macrocephaly, with fullness of the fontanelles and increased separation of calvarial sutures. Ataxia arises from vermis and cerebellar hemisphere involvement, brainstem dysfunction and chronic hydrocephalus. Patients who present with posterior fossa tumors undergo surgery for three reasons; (1) Tumor decompression and cytoreduction; (2) To diagnose the tumor based on histopathology, (3) To determine the further plan of management with radiotherapy and/or chemotherapy depending on the nature of the tumor and histological grading. Gross total resection is appropriate surgical treatment in all posterior fossa tumors. With aggressive surgery, craniospinal radiotherapy and chemotherapy, more than 50% of children with medulloblastoma can be expected to be free of disease 5 years later[6]. Using current treatments, 80%–90% of those without disseminated disease can be cured; however, treatment for this disease often results in significant endocrinological 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 cerebeller 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[7].

## AIMS AND OBJECTIVES:

To analyse the incidence, clinical features, surgical outcome and complications in patients with paediatric posterior fossa tumor in our institute.

## MATERIAL AND METHODS:

The present study was conducted in the department of Neurosurgery, Bangur Institute of Neurosciences (B.I.N) & SSKM Hospital, IPGME & R, Kolkata. BIN caters patient from various districts of West Bengal as well as adjoining states like Bihar, Jharkhand, Orissa and North-East India. The study was conducted from January 2019 to December 2020. The study design was non randomized prospective observational study. Total sample size was 50.

## INCLUSION CRITERIA:

1) Age equal or less than 12 years.
2) Evidence of posterior fossa tumors in neuroimaging studies.

## EXCLUSION CRITERIA:

1) Age more than 12 years.
2) Prior surgery in another centre.
3) Parent's attendant not giving consent for surgery.

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 patients were regularly followed up at 3, 6, 12 months after operation.

**Study Tools:**

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

**2) Investigation:**

a) Routine hematological and biochemical tests.
b) Radiological investigation: NCCT Brain, MRI Brain (P+C) with MR Spectroscopy with spinal screening.
c) Histopathology.

All patients who were fit in Pre -Anaesthetic Checkup were undertaken for surgery after witten & informed consent.

## OBSERVATIONS & RESULTS:

During the study period, 480 cases of CNS tumors 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 as shown in Table 1.

**Table 1: Incidence of posterior fossa tumors among the study subjects during the study period.**

| Variables | N | % |
|---|---|---|
| CNS Tumors | 480 | 100 |
| Supratentorial | 336 | 70 |
| Infratentorial | 144 | 30 |
| Incidence of Posterior Fossa Tumors (<12 Years) | 96 | 20 |
| Subjects who were Studied | 50 | |

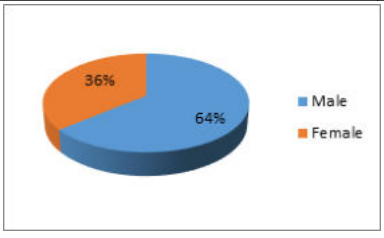


**Figure 1: Gender distribution among study group.**

There was male dominance i.e. out of 50 subjects, there were 32 males (64%) and 18 females (36%) as shown in Figure 1.

6%, 34% and 60% of the subjects were having age <1, 1-5 and 5-12 years respectively. 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 as shown in Table 2.

**Table 2: Complains among the study subjects**

| Complaints | N | % |
|---|---|---|
| Headache and Vomiting | 41 | 82 |
| Visual Complains | 14 | 28 |
| Ataxia | 4 | 8 |
| Increased Head Size | 3 | 6 |

Most common location of posterior fossa tumor was fourth ventricle (60%) followed by cerebellar hemisphere (20%), cerebellar vermis (12%) and brainstem (8%). Brainstem involvement was reported among 44% (22) of the subjects. Medulloblastoma, Ependymoma, JPA and Brainstem glioma was reported among 48%, 24%, 20% and 8% of the subjects respectively as shown in Table 3.

**Table 3: Diagnosis among the study population.**

| Diagnosis | N | % |
|---|---|---|
| Medulloblastoma | 24 | 48 |
| Ependymoma | 12 | 24 |
| JPA | 10 | 20 |
| Brainstem Glioma | 4 | 8 |
| Total | 50 | 100 |

46% of the subjects were given chemotherapy while radiotherapy viz. craniospinal and local posterior fossa boost was given to 36% and 20% 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 as shown in Table 4.

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 Brainstem glioma respectively as shown in Table 5.

**Table 4: Complications among the study subjects.**

| Complications | N | % |
|---|---|---|
| Hydrocephalus | 6 | 12 |
| Postoperative Mutism | 6 | 12 |
| Cerebrospinal Fluid Leaks | 5 | 10 |
| Wound Infection | 4 | 8 |
| Cerebellar Hematoma | 2 | 4 |
| CN Palsies | 2 | 4 |

**Table 5: 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 |
| Brainstem Glioma | 4 | 2 | 50 |
| Total | 50 | 4 | 8 |

## DISCUSSION:

Intracranial posterior fossa tumors are heterogenous group of neoplasms more common in children than in adults. In view of limited space within the posterior fossa and the potential involvement of vital brain stem nuclei, the posterior fossa tumors are considered critical brain lesions. Brainstem compression, herniation, and death are all risks associated with these tumors. The clinical presentation depends on the site of the tumor, biological behaviour, aggressiveness of the tumor and the rate of growth. Symptoms are caused by focal compression of the cerebellum or brain stem centres and the increased intracranial pressure with resultant hydrocephalus. Hydrocephalus is common in children, occurring in 71-90% of paediatric patients and approximately 10-40% demonstrates persistent hydrocephalus even after posterior fossa tumor resection.[8] Posterior cranial fossa tumors in children differ from adults in their clinical presentation, behaviour, management and prognosis. Pilocytic Astrocytoma and Medulloblastoma are most prevalent tumors in childhood. Medulloblastomas have varied prognosis and long-term survival rates are generally lower in patients 3 years or younger. Posterior fossa tumors warrant surgical management to achieve decompression of the posterior fossa, relieve pressure on the brainstem, release intracranial pressure and avert the risk of herniation. Surgery is also mandatory for histopathological diagnosis of the tumor and to determine further plan of management depending on the nature of the tumour[9]. Majority of patients has a favourable outcome following complete surgical removal.

## STATISTICALANALYSIS:

### Incidence of Posterior Fossa Tumors:

Incidence of posterior fossa tumors in total and in paediatric age group(<12years) was reported among 144 subjects (30%) and in 96 subjects (20%) in our study. These findings were little lower as compared to study done by kalyani et al[11] who reported 40.9% of posterior fossa tumors in their institute. Gubbala et al[12] in their study revealed that 80 cases of posterior fossa tumors were analysed and these constituted 69% of total CNS tumors in children below 5 years reported in their institute. Intracranial posterior fossa tumors constituted 37.03% of total CNS tumors in their study.

### Gender:

In our study, there is male dominance i.e. out of 50 subjects, there were 32 males (64%) and 18 females (36%) with male to female ratio of 1.78:1. Dawood MA et al[13] in their study found that 60% and 40% of the subjects were male and female respectively. Majority of the tumors in posterior fossa had male predilection with M:F ratio of 1.6:1 as mentioned by Gubbala et al[12] in their study. These findings are similar to our study. Salami A et al[14] in their study found male to female ratio of 1.57:1. Similar male dominance was reported by Kalyani et al[15] in their study.

**Age:** 6%, 34% and 60% of the subjects were having age <1, 1-5 and 5-12 years respectively in the present study. In a study by Dawood MA et al[13], 65% of the subjects were having age between 5-14 year which is similar to the present study. Rami A et al.[16] in their study showed that 61.7% of posterior fossa tumors involved children <14years with 23.9% in those <5years.

**Complaints:**
In our study; 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. Dawood MA et al[13] in their study revealed similar finding i.e. the most common clinical presentation in their group of patients was headache and vomiting. Rami A et al[16] too reported that headaches and vomiting were the most common presenting symptoms. In a study by Gubbala et al[12] majority of patients presented with headache and vomiting irrespective of age and gender. Hydrocephalus was the second most common clinical presentation in the children and cerebellar signs with Gait abnormalities in the adults. Cranial nerve palsies were reported in 16.3% of cases. In study done by Kalyani et al.[15] Cerebellar signs was the most common presentation and hydrocephalus in study done by Meenakshi Sundaram et al[17]

**Location:** Most common location of posterior fossa tumor was fourth ventricle (60%) followed by cerebellar hemisphere (20%), cerebellar vermis (12%) and brainstem (8%) in the present study. Salami A et al[14] in their study mentioned that more than half of the tumors (56.94%) arose from the cerebellar hemispheres and ten cases (18.89%) from the 4th ventricle.

**Brainstem Involvement:** In our study, brainstem involvement was reported among 44% (22) of the subjects. Piscione PJ et al[18] in their study found brainstem involvement in 30% of the subjects.

**Diagnosis:** In the present study; Medulloblastoma, Ependymoma, JPA and Brainstem glioma was diagnosed among 48%, 24%, 20% and 8% of the subjects respectively. Rami A et al[16] in their study found that Medulloblastoma was the most common posterior fossa tumor. Dawood MA et al[13] in their study reported that MRI study revealed 16 cases diagnosed as Medulloblastoma (40%), 8 cases as Ependymoma (20%), 8 cases as Juvenile Pilocytic Astrocytoma (20%), 6 cases as Pontine Glioma (15%) and 2 case as Schwannoma (5%). Similarly in a study by Piscione PJ et al[18]; Medulloblastoma, Cerebellar Astrocytoma, Ependymoma and Brainstem glioma among 12 (40), 13 (43.3), 2 (6.7) and 3 (10) subjects respectively.

**Complications:**
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[16], 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.

**Mortality:**
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 Brainstem 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[16] in their study.

**CONCLUSION:**
Posterior fossa tumors are critical brain lesions with significant neurological morbidity and mortality. Early diagnosis of posterior fossa tumors 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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