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Original Research Paper

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CLINICO-PATHOLOGICAL STUDY OF MIDLINE POSTERIOR FOSSA TUMORS IN INFANCY AND CHILDHOOD AT TERTIARY CARE CENTRE

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ABSTRACT Primary brain tumors are the most common solid tumors in the pediatric population, comprising 20% to 25% of all childhood cancers. The diagnosis is often difficult to establish in child because many of the signs and symptoms may mimic those of more common childhood illnesses. MRI brain and spine are standard modality for diagnosis. Definitive surgery is usually indicated in midline posterior fossa tumor for diagnosis, to established csf flow, and curative purpose. Adjuvant treatment in form of radiotherapy and chemotherapy is needed in selected cases. This study was planned to find out true incidence and clinicopathological profile of midline posterior fossa mass in infancy and child hood at tertiary care center.

KEYWORDS : Mid line posterior fossa tumor, Medulloblastoma, Ependymoma

1. INTRODUCTION

Primary brain tumors are the most common solid tumors in the pediatric population, comprising 20% to 25% of all childhood cancers. About 60% to 70% of all pediatric brain tumors originate in the posterior fossa.[1-4] Most common posterior fossa tumors of childhood are medulloblastomas, ependymomas, and astrocytomas.

The diagnosis is often difficult to establish in child because many of the signs and symptoms may mimic those of more common childhood illnesses. The diagnosis is suspected in the setting of irritability, loss of appetite, weight loss and failure to thrive. MRI of spine should be done to rule out ant drop metastasis because tumor like medulloblastoma, ependymoma and choroid plexus papilloma have propensity to spread via csf.

Hydrocephalus in pediatric patients with posterior fossa mass is matter of debate, Obstructive hydrocephalus is reported in 70% to 80% of children with posterior fossa tumors and is frequently the cause of clinical deterioration at the time of diagnosis.[5,6] Ventriculoperitoneal shunt is standard treatment for hydrocephalous but have certain complications like infection , blockage, peritoneal seeding and rare complications such as upward herniation. 15% to 30% of patients ultimately require permanent CSF diversion following resection of the tumor.[6,7]Aims of definitive surgery in posterior fossa masses is usually is gross total resection without causing further neurological deficit, but attachment of tumor to vital structures in this region limits gross total resection of tumor. This study was planned to find out true incidence and clinicopathological profile of midline posterior fossa mass in infancy and child hood at tertiary care center Gwalior.

3.MATERIAL AND METHODS:

1.PLACE AND PERIOD OF STUDY

This prospective study is carried out at Department of Neurosurgery, JA group of hospitals, Gajra Raja Medical College, Gwalior from December 2017 to November 2019.

2.STUDY DESIGN – Prospective study

3.STUDY POPULATION

Patients of midline posterior fossa mass up to 15 years of age who presented to neurosurgery department, JA groups of hospitals, G R Medical college, Gwalior M.P. over the period of 24 months.

4.INCLUSION CRITERIA

- All children up to 15 years of age and CT/MRI brain suggestive of midline posterior fossa mass.
- Midline in the posterior fossa mass include mass arising from vermis of cerebellum, 4th ventricle, brain stem and mass arising from cerebellar hemisphere approaching towards midline (within 1 cm of midline).
- Patients given consent to include in the study.

5.EXCLUSION CRITERIA

1. Previously operated case / recurrence
2. Patients not giving consent to be included in study.

6.STUDY PROCEDURE:

- All patients up to age of 15 years with midline posterior fossa mass who came to neurosurgery department assessed demographically including age, sex. In each groups males and females were separated.
- Patients having hydrocephalus underwent CSF diversion procedure like VP Shunt as emergency followed by Definitive surgery.
- Patients don't have hydrocephalus directly underwent definitive surgery as midline suboccipital craniectomy with excision of mass (Gross total excision / Near total excision- depending upon extension and infiltration of surrounding structures) with primary Dural repair / augmentation duraplasty
- After histopathological confirmation adjuvant radiotherapy / chemotherapy / both started depending upon HPE report.
- For radiotherapy and chemotherapy patient was referred to radiotherapy department, JA Hospital, GR Medical college, Gwalior.
- In case of medulloblastoma and ependymoma

craniospinal radiation was given.
- Optimal irradiation dose: 35–40 Gy to whole craniospinal axis +10–15 Gy boost to tumor bed , all fractionated over 6–7 wks.
- Children less than 3 years of age received chemotherapy used
- Lomustine (CCNU), cisplatin and vincristine (VCR Regimen)
- Patients of brain stem glioma directly underwent chemoradiotherapy.

7.FOLLOW-UP
- Minimum 2 months and Maximum 2 years.
- Patients was follow-up in neurosurgery opd after every 15 days up to 2 months and every month up to 6 months and every 2 months up to 1 year and every 3 months up to 2 years.
- Follow up CECT/ MRI brain (plain +contrast) to see recurrence done after 6 months, 1 year, and 1.5 year of surgery.

3.RESULTS:
Total 28 case of pediatric midline posterior fossa mass were included in study. Among 28 patients 5 patients (17%) were in age group of 0-5 years, and 8 patients (28%) were in age group of 5-10 years, and in 10-15 years of age group 15 patients (55%) were there.

Table 1: age wise distributon:

Sr.No.	AGE	NO. OF PATIENTS	PERCENTAGE
1	0-5 Years	5	17 %
2	5-10 Years	8	28%
3	10-15Years	15	55%
4	**Total**	**28**	

Table 2: GENDER WISE DISTRIBUTION: Among 28 patients included in study 16 were male constituting 60 % and 12 were females, constituting 40 %. Male to female ratio was 1.33 :1.

Sr No.	Gender	No. of patients	Percentage
1	MALE	16	60%
2	FEMALE	12	40%
3	**Total**	**28**	

3. Most of patients (75 %) presented as headache, followed by vomiting and difficulty in walking (67% each), visual disturbance was found in 7 % of patients, although seizures are uncommon in posterior fossa tumors, found in 7% of patients.

Table 3: Distribution of presenting complaints

Sr NO.	Symptoms	No. of patients	Percentage
1	HEADACHE	21	75%
2	VOMITING	19	67%
3	DIFFICULTY IN WALKING	19	67%
4	VISUAL DISTURBANCE	2	7%
5	SEIZURE	2	7%

3. SIGNS: Most common sign found in patients was ataxia in 67%, dydiadochakinesia in 64%, nystagmus, hypotonia, rebound phenomenon past pointing, and pendular knee jerk found in 35%, dysphasia and tremors found in 18 %, lower cranial nerves involvement were seen in 10 % and titubation was found in 7 %.

Sr no.	Sign	No. of patients	Percentage
1	Ataxia	19	67%
2	Pappilledema	17	60%
3	Dysdiadochokinesia	18	64%
4	Nystagmus	10	35%
5	Hypotonia	10	35%
6	Rebound phenomenon	10	35%
7	Past pointing	10	35%
8	Pendular knee jerk	10	35%
9	Dysphasia	8	18%
10	Tremors	8	18%
11	Lower Cranial Nerve Involvement	3	10%
12	Titubation	2	7%

4. TRAETMENT: Predefinitive surgery in the form of VP shunt was done in 17 (60%) patients, and 25 patients underwent definitive surgery in as midline suboccipital craniectomy with excision of mass (gross total / near total excision) done, and 3 patients of brain stem glioma directly sent for Chemo Radiotherapy.

Sr no.	Treatment	No. of patients
1	Predefinitive ventriculoperitoneal shunt	17
2	Definitive surgery	25
3	Direct chemoradiotherapy	03
4	**Total**	**28**

5.HISTOPATHOLOGY: Total 25 patients underwent definitive surgery, as three patients of brain stem glioma directly underwent chemoradiotherapy. On Histopathological examination of 25 patients, 15 patients (54%) were found medulloblastoma, 4 patients (15 %) each of ependymoma and pilocytic astrocytoma, 1 patient (3 %) each of epidermoid and dermoid tumor.

Sr no .	Hisopathology	No of patients	Percentage
1	Medulloblastoma	15	54%
2	Ependymoma	4	15%
3	Pilocytic astrocytoma	4	15%
4	Epidermoid	1	3%
5	Dermoid	1	3%
6	**Total**	**25**	

6. EXTENT OF RESECTION: Out of 15 patients of medulloblastoma ,13 patients underwent gross total resection and 2 patients underwent near total resection due to attachment to floor of 4th ventricle. All 4 patients of ependymoma underwent near total resection due to attachment to brain stem & floor of 4th ventricle. Patients of dermoid, epidermoid and pilocytic astrocytoma underwent gross total resection.

SR NO.	HISTOPATHOGICAL REPORT	GROSS TOTAL	NEAR TOTAL	SUBTOTAL	TOTAL PATIENTS
1	MEDULLOBLASTOMA	13	2	-	15
2	EPENDYMOMMA	-	4	-	4
3	DERMOID	1	-	-	1
4	EPIDERMOID	1	-	-	1
5	PILOCYTIC ASTROCYTOMA	4	-	-	4
6	**Total**	**19**	**6**		**25**

DISCUSSION
Posterior fossa tumors are most common solid pediatric tumors. Posterior fossa tumors comprise 54% to 70% of childhood brain tumors compared to 15%-20% in the adult population. The reason that pediatric brain tumors have a propensity to occur in the posterior fossa has not yet been elucidated. Cushing probably was the first to report a large series of posterior fossa tumors [8]. published information about 61 patients with cerebellar medulloblastoma (MB) with mostly fatal outcome. Now, the outcome is improving because of advances in the discovery of anesthesia, asepsis, neurological localization, and technique of tumor removal.

1. AGE AND GENDER WISE DISTRIBUTION (MALE TO FEMALE RATIO)
Our study revealed male predominance with over all male to

female ratio is 1.33 which is almost similar to previous studies. Sudha Iyengar et al (2016)[9] studied 32 patients of pediatric posterior fossa and found male to female ratio of 1.9: 1.

Present study revealed most of the patients in the age group of 10-15 years of age group which is slightly differ from previous studies. Ahmed et al.[10] (2007) who reported most cases in age group 5-9 years. Sudha Iyengar et al[9] (2016) reported that most common age group is 6-8 years. The male predominance probably due to increased number of male patients being investigated, which is an important social feature in Indian society.

2.SIGNS AND SYMTOMS: Symptoms of posterior fossa tumors usually result from either due to raised intracranial pressure or due to compression of vital structures. Symptoms and signs due to raised intracranial pressure were headache, nausea, vomiting and papilledema on fundus examination. Most common presenting symptom in our study was headache in 75% of cases followed by vomiting and difficulty in walking 67 % each, which is almost similar to previous studies. **Tabatabaei SM et al[11] (2012)** has reported Cerebellar symptoms were the most common cause of presentation (80.9%) followed by headaches (73.8%) and vomiting (38.1%). Visual problems were found in 7.14% of their patients probably due to late presentation. **Gaur S et al (2015)** [12] conducted a study on 58 patients and observed that 93.10% manifesting as headache, and vomiting. Ataxia was present in 41 %.

4.ASSOCIATION WITH HYDROCEPHALUS: Due to the anatomic relationships of these tumors to cerebrospinal fluid (CSF) drainage pathways, hydrocephalus is common, occurring in 71–90% of children with posterior fossa tumors,[13] and this may require emergent intervention with placement of an external ventricular drain (EVD) / VP shunt. Our study revealed that hydrocephalus association with posterior fossa mass is 60 % of cases which is slightly less than previous study. Due-Tønnessen B.J et al 2007 [14] found that 79 % of posterior fossa masses presented with hydrocephalus. Culley et al (1994)[15] studied 64 children with posterior fossa mass and found that out of 64 patients ,54 had hydrocephalus (84%).

5. DEFINITIVE SURGERY: Surgical excision of posterior fossa mass were done in 25 patients, remaining three patients were diagnosed as brain stem glioma and directly send for chemoradiotherapy. All the operated patients were approached via midline suboccipital craniectomy. Transvermian approach were used in 22 patients while telovelar approach were used in 3 patients. **Dattatraya Muzumdar et al (2011)** studied 211 cases of medulloblastoma over a period of 15 years and 93.4% of medulloblastoma approached via midline suboccipital craniectomy, and 5.6 % tumors approached via paramedian suboccipital craniectomy. **Sherise D et al (2018) studied 16** pediatric patients of posterior fossa mass and transvermian approach was used in 11 (69%), and a telovelar approach in only 2 (12%);

6.EXTENT OF RESECTION: Midline Posterior fossa masses usually arise from roof /floor of forth ventricle or vermis and attached to brainstem so gross total resection usually not possible in these cases. However in our study gross total resection was achieved in 19 patients (76%) and near total resection was achieved in 6 patients (24%). 86 % of medulloblastoma underwent gross total excision while remaining 14 % underwent near total excision. All the of patients (4) of ependymoma underwent near total excision, probable reason for incomplete excision was its propensity to infiltrate the vital structures over floor of 4th ventricle. All the patients of pilocytic astrocytoma, epidermoid and dermoid underwent gross total resection. **Dattatraya Muzumdar et al (2011)** studied 211 patients with medulloblastoma out of which 53% underwent gross total resection, 28.6% underwent near total resection and 17.6% underwent subtotal resection.

7.HISTOPATHOLOGY:
Medulloblastoma as most common tumor in our study found in 54% followed by astrocytoma and ependymoma 15 % each. Epidermoid and dermoid 3% each. **Sutton L et al (1989)** [16] fund medulloblastoma in about 36%, cerebellar astrocytoma in 28%, brainstem glioma 9%, and ependymoma in 4%. **Packeret al (1990)** also found predominance of medulloblastoma in 36% of patients followed by 28% cerebellar astrocytoma and 9% ependymoma. **Sudha Iyengar et al (2016)** studied 32 pediatric posterior fossa tumors, medulloblastoma were found in 16(56 %) of cases, 6(18%) patients had ependymoma while 3 (9.3%) patient were having brainstem glioma, which is almost similar incidence as in our study.

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