

A HOSPITAL BASED PROSPECTIVE STUDY TO INVESTIGATE THE EPIDEMIOLOGICAL PROFILE OF INFRATENTORIAL SPACE OCCUPYING LESIONS IN INFANTS AND CHILDREN UP TO 15 YEARS OF AGE AT TERTIARY CARE CENTER

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Abstract

Background: ICSOL (Intracranial space occupying lesion) is common among children and leads to morbidity and mortality in absence of early diagnosis and treatment. The present study was designed to investigate the epidemiological profile of infratentorial space occupying lesions in infants and children up to 15 years of age in our institution. **Materials and Methods:** This is a prospective study done in the Department of Neurosurgery, Jay Arogya group of hospitals, Gajra Raja Medical College, Gwalior from December 2018 to December 2020. Only patients with the proven histopathological diagnosis were included in the study. In addition to the location and histological types of the tumor, patient demographics including age and sex were also recorded. All histopathological diagnoses were made according to the 2007 World Health Organization classification system. We analysed the data using Chi-square test and test of proportions (Z-test) were used wherever necessary to test for statistical significance. **Results:** Among 27 patients (62%) have posterior fossa mass up to 15 years of age. Male to female ratio was 2.33:1 in our study. Most of patients (74 %) presented with headache followed by vomiting (44%) and difficulty in walking (63%). 24 patients (88%) presented with hydrocephalus. Patients underwent definitive surgery in form of suboccipital craniectomy followed by Transvermian (71%) / Telovelar (13%) / Transcortical approach (16%) for tumor excision depending upon tumor location and extension. Out of four recurrence, two were medulloblastoma. Ependymoma and ATRT, one each case completes the list. One patient also developed drop metastasis in follow up. Radiotherapy was not offered to the patient of ATRT due to his young (10 months) age. Out of 24 operated cases, there were three mortalities, 2 medulloblastomas and one ATRT. **Conclusion:** Majority of the infratentorial space occupying lesion were medulloblastoma (43%) and 88% of patients with infratentorial space occupying lesion presented with hydrocephalus in which predefinitive surgery was done in the form of VP shunt. Every patient presenting with signs of raised ICP and motor system involvement should be thoroughly investigated for intracranial space occupying lesions and managed medically or surgically as needed to decrease morbidity and mortality.

INTRODUCTION

Infratentorial compartment extends from tentorium cerebelli superiorly to foramen magnum inferiorly, containing the cerebellum and most of the brainstem, specifically the pons and medulla. 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] Common posterior fossa tumors of childhood are medulloblastoma, ependymoma, and astrocytoma. Among infective etiologies Tuberculomas still constitute a large percentage of intracranial space occupying lesions of childhood. It can be seen at any site. Infratentorial tuberculomas are more common in children, two third are located in cerebellum and the rest in cerebrum.^[2] Brain abscess can occur in

children of any age but are most common between 4 and 8 years. The causes of infratentorial brain abscess include embolization due to congenital heart disease with right to left shunt (especially tetralogy of Fallot), meningitis, chronic otitis media and mastoiditis, soft tissue infection of the face or scalp, penetrating head injuries, and immunodeficiency states. Approximately 80% of cases are divided equally between the frontal, parietal, and temporal lobes. Brain abscesses in the occipital lobe, cerebellum, and brain stem comprise about 20% of the cases.

Vascular Lesions like arterio-venous malformation, Sturge – Weber Disease, Von Hippel Lindau (VHL) Syndrome are also identified. Congenital or development Lesions like tuberous Sclerosis, neurofibromatosis, Fahr’s disease, arachnoid cysts may present with headache, seizures, psychomotor retardation and raised intracranial pressure secondary to either the cysts themselves or secondary to hydrocephalus (up to 90% in dandy walker malformation).^[3]

Miscellaneous causes of ectopic calcification in infratentorial calcification include endocrine diseases like hyperparathyroidism, hypo - parathyroidism, pseudo hypoparathyroidism and toxic substances like Lead Poisoning, vitamin D intoxication, hypercalcemia are also been explained. Posterior fossa is considered as critical region of brain because it contains vital brainstem nuclei and masses in this region often cause hydrocephalus by blocking cerebrospinal fluid (CSF) outflow pathways which results in raised intracranial pressure. Symptoms usually start with headache often worse in the morning followed by vomiting and eventually gait disturbance, but in pediatric population the symptoms are confusing so it 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.^[4]

Aims of definitive surgery in posterior fossa masses is usually gross total resection without causing further neurological deficit, but attachment of tumor to vital structures in this region (floor of 4th ventricle) limits gross total resection of tumor. Another aim of surgery is histopathological diagnosis, so that adjuvant therapy can be started. Each tumor has different prognosis and response to treatment depending on the histopathology. The present study was designed to investigate the epidemiological profile of infratentorial space occupying lesions in infants and children up to 15 years of age in our institution.

MATERIALS AND METHODS

This is a prospective study done in the Department of Neurosurgery, Jay Arogya group of hospitals, Gajra Raja Medical College, Gwalior from December 2018 to December 2020.

Inclusion Criteria

Patients up to 15 years of age and CT/MRI brain suggestive of infratentorial space occupying lesions.

Exclusion Criteria

1. Previously operated case / recurrence
2. Patients not giving consent to be included in study.
3. Patient with significant co morbidity.

Methods: Only patients with the proven histopathological diagnosis were included in the study. In addition to the location and histological types of the tumor, patient demographics including age and sex were also recorded. Patients with metastatic tumor, tumor-like cystic lesion (arachnoid cysts, epidermoid cysts and colloid cysts), the secondary tumor, space occupying lesion of infectious etiology and vascular malformation were excluded from the study.

All histopathological diagnoses were made according to the 2007 World Health Organization classification system.⁵ We analysed the data using Chi-square test and test of proportions (Z-test) were used wherever necessary to test for statistical significance.

RESULTS

Our study showed that 44 patients of brain tumor (supratentorial + infratentorial). Out of 27 patients (62%) have posterior fossa mass up to 15 years of age.

Among 27 patients 4 patients (15%) were in age group of 0-5 years, and 12 patients (45%) were in age group of 5-10 years and in 10- 15 years of age group, there were 11 patients (40%). Male to female ratio was 2.33:1 in our study. Most of patients (74 %) presented with headache followed by vomiting (44%) and difficulty in walking (63%); visual disturbance was found in 19% of patients; since seizures are uncommon in posterior fossa tumors, only one patient presented had seizure without any supratentorial extension of tumor or pathology [Table 1].

Out of 27 patients studied 24 patients (88%) presented with hydrocephalus and remaining 3 patients didn't have hydrocephalus [Table 1].

Most common sign found in patients was ataxia in 67%; truncal ataxia was found in 12 patients (45%) and appendicular ataxia was found 78 in 6 patients (22%); dysdiadochokinesia was found in 64%; nystagmus and hypotonia in 37%, rebound phenomenon, past pointing and pendular knee jerk were found in 35% of patients; scanning speech in 25% and tremors were found in 22% of patients; lower cranial nerves involvement were seen in 10 % and titubation was found in 7% of patients. 70% patients were having papilledema on examination [Table 1].

Cerebellar signs (i.e. Ataxia dysdiadokokinesia) are most common findings on clinical examination followed by signs of raised intracranial tension. Brain

stem involvement was the least common finding on clinical examination [Table 1].

[Table 2] showed that VP shunt was done in 19(70%) patients before definitive surgery, and 24 (89%) patients underwent definitive surgery as suboccipital craniectomy with excision of mass (gross total / near total excision) done and 3(11%) patients underwent ventriculoperitoneal shunt followed by adjuvant therapy. 2 patients of brain stem glioma were sent for chemoradiotherapy and 1 patient of tuberculoma was given antitubercular medication after ventriculoperitoneal shunt [Table 2].

Patients underwent definitive surgery in form of suboccipital craniectomy followed by Transvermian (71%) / Telovelar (13%) / Transcortical approach (16%) for tumor excision depending upon tumor location and extension [Table 2].

Out 24 patients operated for posterior fossa mass, gross total resection was achieved in 12 patients (50%), in 10 patients (41%) near total excision and in 2 patients (9%) subtotal excision was done. Near total and subtotal excision was done in cases where the tumor was inseparable from the fourth ventricle floor or was densely adhered to neurovascular structures. Histopathological examination of 24 patients revealed, 10 patients (43%) had medulloblastoma; pilocytic astrocytoma in 6 patients (25%), 4 patients (16%) had ependymoma and; 1 patient (4%) each of epidermoid, abscess, brain stem glioma and ATRT. Two patients with radiological diagnosis of brain stem glioma were sent for chemoradiotherapy and one patient of tuberculoma was put on antitubercular regimen after ventriculoperitoneal shunt [Table 3].

Table 1: Demographic profile & signs and symptoms of patients.

Demographic profile	No. of patients (N=27)	Percentage (%)
	Age group (yrs)	
0-5 years	4	15%
5-10 years	12	45%
10-15 years	11	40%
	Sex	
Male	19	70%
Female	8	30%
	Symptoms	
Headache	20	74%
Vomiting	12	44%
Difficulty in walking	17	63%
Visual disturbances	5	19%
Siezuers	1	4%
	Presentation with hydrocephalus	
Hydrocephalus	24	88%
Without Hydrocephalus	3	12%
	Signs	
Ataxia (Truncal) (Appendicular)	18 (12) (6)	67% (45%) (22%)
Papilledema	19	70%
Dysdiadochokinesia	17	64%
Nystagmus	10	37%
Hypotonia	10	37%
Rebound phenomenon	9	35%
Past pointing	9	35%
Pendular knee jerk	9	35%
Scanning speech	7	25%
Tremors	6	22%
Lower Cranial Nerve Involvement	3	10%
Titubation	2	7%

Table 2: Demographic profile & signs and symptoms of patients

	No. of patients (N=27)	Percentage (%)
	Treatment	
VP Shunt done before definitive surgery	19	70.37%
Definitive surgery	24	88.88%
Shunt alone followed by adjuvant therapy	3	11.11%
	Definitive Surgical approach (N=24)	
Transvermian	17	71%
Transcortical	4	16%
Telovelar	3	13%

Table 3: Extent of resection (on various histology)

Sr. no.	Histopathological report	Gross total	Near total	Subtotal	Total patients
1	Medulloblastoma	8	2		10
2	Ependymoma		4		4
3	Epidermoid		1		1
4	Pilocytic Astrocytoma	4	2		6
5	Abscess	1			1

6	Brain stem glioma			1	1
7	Atypical teratoid/rhabdoid tumor			1	1
	Total	13	19	2	24

Table 4: Post-operative complications

Complications	No. of patients	Percentages
Pseudomeningocele	3	12%
Pulmonary Complications (Atelectasis Pneumonia, Respiratory failure)	3	12%
	1	4%
	1	4%
	1	4%
Lower Cranial Nerve Palsy	2	8%
CSF Leak	1	4%
Meningitis	1	4%
Wound infection	1	4%
Postoperative Hydrocephalus	0	0%
Cerebellar mutism	1	4%

Table 5: Mortality

Sr.no.	Histopathological Diagnosis	Mortality	Percentage
1	Medulloblastoma (10)	2	21%
2	Ependymoma (4)	0	-
3	Pilocytic Astrocytoma (6)	0	-
4	Atypical teratoid/rhabdoid tumor (1)	1	100%
5	Epidermoid (1)	0	-
	Total	3	

Pseudomeningocele developed in three patients (12 %) in postoperative period which was managed by compression dressings and shunt chamber compression. Pulmonary complications developed in three patients (12%); out of three patients who developed pulmonary complications, one patient developed atelectasis, one patient developed pneumonia which was managed successfully by higher antibiotics, high flow oxygen support and nebulization. Unfortunately, one patient (4%) went into respiratory failure and died. 1 patient had CSF leak from incision site which was managed by serial lumbar punctures; later on, that patient developed meningitis which was managed using sensitive higher antibiotics and supportive treatment. Lower cranial nerve palsies were also found in two patients (8%) which was managed conservatively with ryles tube feeding and physiotherapy. None of patient in present study developed post-operative hydrocephalus and one of the patients developed superficial wound infection which was managed by aseptic dressing and culture-based antibiotics. One of the patients developed cerebellar mutism which was managed conservatively and patient recovered over 4 months [Table 4].

Out of four recurrence, two were medulloblastoma. Ependymoma and ATRT, one each case completes the list. One patient also developed drop metastasis in follow up. Radiotherapy was not offered to the patient of ATRT due to his young (10 months) age. Out of 24 operated cases, there were three mortalities, 2 medulloblastomas and one ATRT [Table 5].

DISCUSSION

Infratentorial space occupying lesions constitute one of the most important problems in infancy and childhood. When malignancies are considered in

children, brain tumors rank only second to leukaemia. 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 84, 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 and adjuvant therapy. The present study was designed to determine the epidemiology of posterior fossa masses in children less than 15 year of age.

In present study, 5-10 years of age group was commonest age of presentation which correlates with Ahmed et al,^[6] (2007) found most cases in age group 5-9 years & Prasad KSV et al,^[7] (2017) revealed that 18 patients (49 %) were in the age group of 0-5 years. Our study revealed male predominance with over all male to female ratio of 2.33:1 which is almost similar to previous studies done by Zakrzewski K et al,^[8] (2003) male to female ratio was 1.35:1, Sudha Iyengar et al,^[9] (2016) found 1.9:1 and Prasad KSV et al (2017) reported male to female ratio of 1.4:1.^[10] Though the ratio is more in our study but it is in correlation with other studies. The male predominance is probably due to increased number of male patients being investigated, which is important social feature in Indian society and also due to the small sample size of our study.

Symptoms of posterior fossa mass usually result from either due to raised intracranial pressure or due to compression of vital structures. Most common presenting symptom in the present study was

headache in 74% of cases followed by difficulty in walking 63 % and vomiting in 44%, which is almost similar to previous studies except for the study by Tabatabaei SM et al,^[10] which means that clinical feature of raised intracranial pressure predominate over cerebellar symptoms. Cerebellar symptoms appear late. Gaur S et al,^[11] (2015) observed that 93.10% patients presented as headache, and vomiting. Prasad KSV et al,^[7] (2017) also reported similar presentation with headache in 80 % of cases. In present study hydrocephalus was found in 88% of cases which is at par with previous studies. Poor socioeconomic condition and delayed presentation may contribute to it. Due-Tønnessen B.J et al,^[12] 2007 found that 79 % of posterior fossa masses presented with hydrocephalus.

Present study revealed hydrocephalus in 88% of patients which was managed by placement of Ventriculoperitoneal shunt as an emergency procedure. According to study of Santos de Oliveira R et al (2008),^[13] it is not a standard practice to perform permanent CSF diversion procedure with placement of a ventriculoperitoneal shunt or endoscopic third ventriculostomy prior to tumor resection, because perhaps only one third of patients will require permanent CSF diversion.

Surgical excision of posterior fossa mass was done in 24 patients, remaining two patients of brain stem glioma were directly subjected to chemoradiotherapy (diagnosis made by imaging only). One patient of tuberculoma (diagnosis made by imaging only) received ATT after VP shunt for hydrocephalus. All the operated (definitive surgery) patients were approached via sub occipital craniectomy, Transvermian approach was used in 17(71%) patients followed by transcortical approach in 4(16%) patients and telovelar approach in 3(13%) patients. Dattatraya Muzumdar et al,^[14] (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 in his series. Sherise D et al,^[15] (2018) studied 16 pediatric patients of posterior fossa mass and found that transvermian approach was used in 11 (69%), and a telovelar approach was used in only 2 (12%) patients.

In the present study gross total resection was achieved in 12 patients (50%), near total resection was achieved in 10 patients (41%) and subtotal in 2 patients (9%). 80 % of medulloblastoma underwent gross total excision while remaining 10 % underwent near total excision. All four patients of ependymoma and a patient of epidermoid underwent near total resection. Reason for incomplete excision was its propensity to infiltrate the vital structures in the floor of 4th ventricle. Four patients of pilocytic astrocytoma underwent gross total resection. Similar results found by Dattatraya Muzumdar et al,^[14] (2011) & Winkler EA et al (2016).^[16]

Medulloblastoma was the most common tumor in the present study found in 43% of cases followed by

astrocytoma (25%) and ependymoma (16 %). Brain stem glioma was found in 1 (4%) cases. Medulloblastomas predominate in present study because present study because of small sample size and astrocytoma, ependymoma had almost similar incidence as compared to previous studies such as Sutton L et al (1989),^[17] Packer et al (1990),^[18] Sudha Iyengar et al,^[9] (2016) & Prasad KSV et al (2017).^[7] In present study incidence of pseudomeningocele was 12% and all patients were managed conservatively by means of compressive dressings and acetazolamide only. The mechanisms of formation of pseudomeningocele is not clear, but may be due to inadequate dural and wound closures. Small pseudomeningoceles may respond well to pressure bandages, needle aspiration, and lumbar CSF drainage. However, pseudomeningoceles may be a manifestation of hydrocephalus and in some cases may require CSF diversion to control. Smith GA et al,^[19] (2016) studied on 681 operated cases of posterior fossa surgery and found that midline posterior fossa surgery had incidence of 16.5 % and retrosigmoid posterior fossa surgery had an incidence of 11.9 %.

Incidence of CSF leak in present study is less than previous studies because we placed VP shunt preoperatively in 88% of patients and did water tight dural closure in all cases.^[20,21]

In present study one patient (4%) out of 24 who underwent surgery, developed postoperative meningitis, results were comparable favorably with study of Chen Chen et al,^[22] (2014) who also reported 8.6% risk of postoperative meningitis.

According to Russell D et al,^[23] (1989) the diffuse histological variant of medulloblastoma has a worse prognosis than the juvenile one but is not predictive of recurrence. In the present study, 2 patients (19%) of medulloblastoma, 1 patient (25%) of ependymoma and 1 patient (100%) of atypical teratoid and rhabdoid tumor had recurrence over the period of 2 years follow up.

In present study mortality was 20% (2 out of 10) in medulloblastoma subgroup, lesser than the study of Navaporn Nalita et.al (2018).^[24] Both the mortalities were in the age group of less than 3 years (both were deferred from radiotherapy because of their age). Less age is also an independent poor prognostic factor. The mortality was less because of small sample size and a shorter follow up period.

Pierre Kahn A et al,^[25] (1983) mortality of 16% and Dhammika P et al,^[26] (1995) 60% mortality, these all 60% patients underwent subtotal resection. Sverre J. Mark et al,^[27] (1977) reported mortality of 25 % . Vaidya et al,^[28] (2012) in a retrospective study of 43 patients reported 26% (11/43) mortality. In our study there was no mortality in ependymoma subgroup due to small sample size and short follow up period.

CONCLUSION

Majority of the infratentorial space occupying lesion were medulloblastoma (43%) and 88% of patients with infratentorial space occupying lesion presented with hydrocephalus in which predefinitive surgery was done in the form of VP shunt. Every patient presenting with signs of raised ICP and motor system involvement should be thoroughly investigated for intracranial space occupying lesions and managed medically or surgically as needed to decrease morbidity and mortality.

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