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# CLINICOPATHOLOGICAL STUDY AND MANAGEMENT OF PAEDIATRIC POSTERIOR CRANIAL FOSSA TUMOURS

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## ABSTRACT

**Introduction:** Paediatric brain tumours are one of the leading causes of death among solid tumours of childhood. Posterior fossa tumours are more common in children in first decade of life. Brain stem compression, herniation and death are some of the important complications of these tumours. Most of the patients are critically ill at the time of presentation and need an emergency operative procedure, especially if they present with acute symptoms of brain stem involvement or herniation. **Materials and methods:** A total number of 52 cases of posterior fossa tumours who attended the neurosurgery department during the period from November 2015 to February 2018 were studied. Detailed clinical profile of the patient including presenting signs and symptoms were recorded. Children with features of raised intra cranial pressure were further investigated to rule out obstructive hydrocephalus. Almost all children were treated surgically followed by adjuvant therapy. **Results:** Maximum number of cases were found in between 6 to 10 year age group with slight female preponderance. The most common tumour encountered in this study was pilocytic astrocytoma(32.7%) followed by medulloblastoma(25%). CSF diversion procedure was done in most cases (86%). About 90% of patients underwent definitive surgery. Overall recurrence rate was 12.76% and mortality rate was 26.92%. **Conclusion:** Posterior cranial fossa tumours are more common in children than adults. Hydrocephalus is common, occurring in 71-90% of paediatric patients. Patients older than 3 years have relatively long term survival rates of approximately 85% in comparison to children less than 3 years of age.

**KEYWORDS:** Brain tumours, Paediatric, posterior fossa.

## INTRODUCTION

Tumours of the Central Nervous System, are the second commonest childhood tumours (20%) after leukemia (37%) and are the most common solid paediatric tumours comprising 40%-50% of all malignancies.[1,2] Their incidence varies between 1 and 3 per 100,000 in different series.[3] The location depends on the age. In infants there is a predominance of supratentorial tumours, while in children older than 4 years infratentorial tumours are more frequent. Overall, posterior fossa tumours account for 45–60% of all brain tumours,[4] being more common in children (45–60%) than adults (15- 20%).[5] Most are seen in the age group between 1 to 10 years.[6] Common complications are brain stem compression, herniation and death. Common posterior fossa brain tumours in children include juvenile pilocytic astrocytoma, medulloblastoma, ependymoma and brainstem gliomas. less frequent are atypical rhabdoid/teratoid tumour, hemangioblastoma, dermoids, schwannoma of the 8th cranial nerve, cerebellar gangliocytoma, meningioma and high grade glioma.[7,8]

Tumours in the posterior fossa are considered to be critical, because of the involvement of the vital brainstem nuclei. Presentation with acute symptoms of brainstem involvement often needs an emergency operation.[7]

The present study is aimed at, to analyse the various paediatric posterior cranial fossa tumours with reference to age and sex groups, there types, pathology, clinical features, incidences, sites of involvement, association of neuroradiological features and tumour type/ behaviour predictability, also to record the follow up data and patient outcome in our institution during the study period.

## MATERIALS AND METHODS

This prospective study includes paediatric patients (0 to 15 years of age group) with clinically suspected and

radiologically confirmed cases of posterior cranial fossa tumours who attended the Department of Neurosurgery, SCB Medical College, Cuttack, between November - 2015 to February – 2018.

Patient particulars were noted down by a pro forma, which included age, sex, chief complain with duration, history of present illnesses like mode of onset, chronology of symptoms of headache, vomiting, blurred vision, double vision, gait disturbances, movement disorders, speech and personality changes and relevant past history. A thorough general examination of the patients was done. Systemic examination with specification to the Central Nervous System examination were done. Routine laboratory investigations were conducted.

The radiological examination included, skull X-ray PA view & lateral view, X-ray chest PA view, plain & contrast CT scan, plain & contrast MRI. Otological and Ophthalmological examination were done as a routine procedure. All children with features of increased intracranial pressure /or decreased alertness were subjected to urgent CT brain to rule out obstructive hydrocephalus.

Patients having obstructive hydrocephalus and features of raised intracranial pressure were treated with emergency Ventriculo Peritoneal shunt. Almost all children (except few intrinsic brain stem lesions) were treated surgically. The resected tumour masses were sent to the Department of Pathology, for histopathological examination and a conclusive diagnosis, for proper planning of management.

Postoperative External Ventricular Drainage or shunting was based on patient's condition during the postoperative period. Almost all the patients with posterior fossa tumours (except pilocytic astrocytoma) were subjected to adjuvant therapy. Children less than 3 years of age were treated with chemotherapy till they reached 3 years of age. After 3 years of age, they were subjected to radiotherapy which included 54 Gy given in fractionated doses to posterior fossa along with 23.4 Gy to entire craniospinal axis in fractionation.

**OBSERVATION**

Out of the total number of 242 cases admitted during the study period, there were 90 paediatric cases. 52 out of the 90 cases of posterior fossa tumours were included in this study The frequency of childhood brain tumours of all brain tumours is 37.2% (chart-1). About 57.8% of all childhood brain tumours belong to posterior fossa.

All the patients with posterior fossa tumours were studied under three age groups: group 1 (0-5years), group 2 (6-10years), group 3 (11-15years). The mean age of the patients included in the study was 6.63 years. Maximum number of patients were in the group 1 and group 2. This implies that posterior fossa tumours are more common below 10 years of age. The sex distribution showed slight female preponderance (male – 25, female – 27). Pilocytic astrocytoma and medulloblastoma were common in the age group 6 – 10 years and ependymoma was common in the age group 0 – 5 years (chart-2).

Most of the posterior fossa tumours 43(83%) among the study population presented with symptoms of increased intracranial tension secondary to obstructive hydrocephalus (chart-3). The second most common symptom included cranial nerve palsies including third, fourth, sixth, seventh and lower cranial nerve palsies. Other symptoms were– torticollis, failure to thrive, obtundation, recurrent respiratory tract infection, unsteadiness and weakness**.**

Various MRI characteristics of the posterior fossa tumours were studied. They were grouped according to: Tumour consistency: a) Cystic, b) Solid, c) mixed; Tumour location: a) Midline, b) Midline with lateral extension. The pilocytic astrocytoma more commonly appeared as cystic mass in the MRI (fig-1) whereas medulloblastoma (fig-2) and intrinsic brain stem tumours almost always presented as solid lesions. Ependymomas presented as solid as well as mixed lesion in equal proportions. Medulloblastoma and intrinsic brain stem tumours almost exclusively occurred in the midline, whereas pilocytic astrocytoma and ependymoma showed higher probability of lateral cerebellar hemispheric extension.

Out of the total 52 patients, 43 (82.7%) had obstructive hydrocephalus (chart-3). Among those 43 patients, 28 had undergone pre-operative shunting and 7 had intra-operative shunt and 2 patients had intra-operative External Ventricular Drainage and the rest 6 patients were not treated with any form of CSF diversion procedure.

Except children with diffuse intrinsic brain stem lesions (5 out of 52), all the other children in the study group were subjected to surgical resection. Maximum extent of tumour resection was done for 28 patients (16 – near total; 12 –total excision). Resection was defined as biopsy (< 10% resection) or as partial (10-50%), subtotal (51-90%), near-total (> 90%), or total resection. A tumour removal that constituted more than 90% was considered a radical resection.[45] In this study, 5 patients having intrinsic brainstem lesion, were not operated; instead they were directly subjected to radiotherapy, but unfortunately all the 5 cases died during the study period. Rest 47 patients underwent definitive surgery. The most common post operative complication was lower cranial nerve palsy followed by aspiration pneumonitis and CSF leak (chart-4).

Post-operatively during the followup period, 2 patients received radiotherapy alone, 11 patients received chemotherapy alone, 22 patients received both chemo-

and radiotherapy. In rest 12 patients, 11 patients did not require any adjuvant therapy and 1 patient did not agree for any form of chemotherapy or radiation.

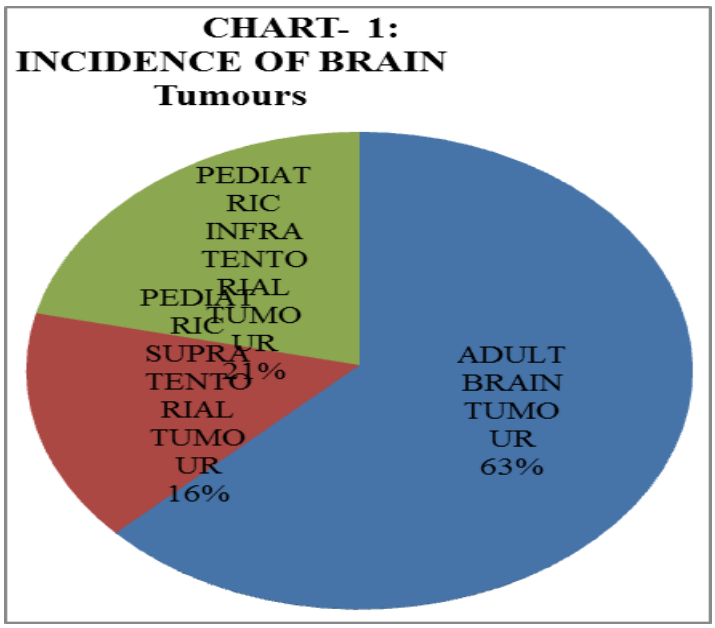
Out of 47 patients who underwent definitive surgery, there was recurrence in 6 patients. 5 patients were posted for redo operation; but parents of one 8 month old patient did not allow us for second look operation, who ultimately died along with 2 redo cases. Another 6 non-recurrent cases had also died during the follow-up period. Rest 38 patients are doing well during this study period and to be followed up subsequently.

Out of the total number of 52 cases studied, pilocytic astrocytoma (fig-3) was the commonest (17cases), followed by medulloblastoma (fig-4) (13cases), ependymomas (12cases) and least number of cases were diffuse astrocytoma (2 cases). Maximum mortality (50%) was seen in case of diffuse astrocytoma excluding intrinsic brain stem tumours (mortality 100%). Survival was best in pilocytic astrocytoma (chart-5).

Maximum recurrence was seen in case of pilocytic astrocytoma (17.65%) and almost in a similar manner in ependymoma (16.66%) with a overall recurrence rate of 12.76% (chart-6). Mortality is analysed in more detail using statistical data and chi- square tests.

When the tumour involvement of the brain stem was analysed as a predictor of mortality using chi square test, it was found to be statistically significant (p= 0.01). When the tumour involved brain stem, the probability of mortality was high with poor survival (chart-7).

When the WHO grading of tumours was analysed as a predictor of outcome using chi square test, it was found to be statistically significant (p=0.001). With high grade of tumours, the probability of mortality is high (chart-8).



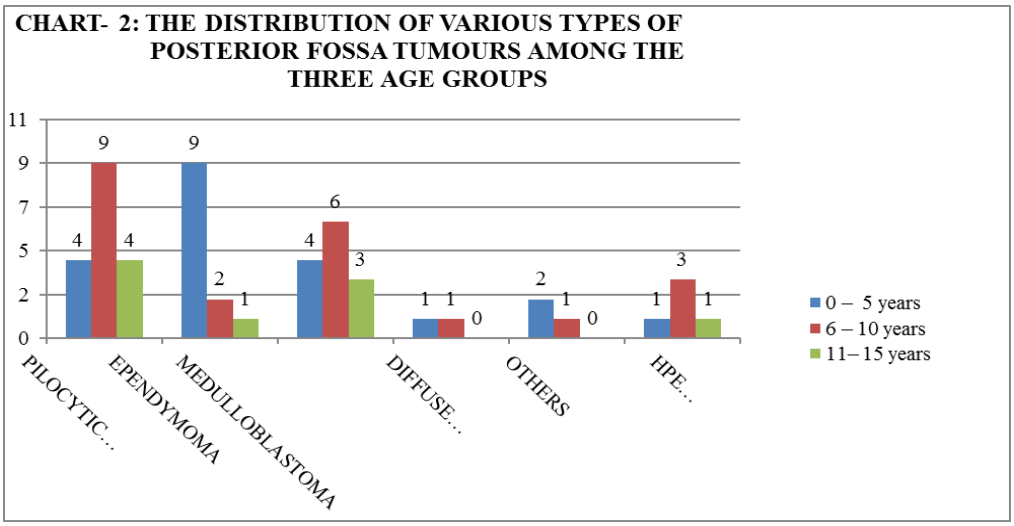



CHART-3: DISTRIBUTION OF OBSTRUCTIVE HYDROCEPHALUS IN THE TOTAL STUDY POPULATION

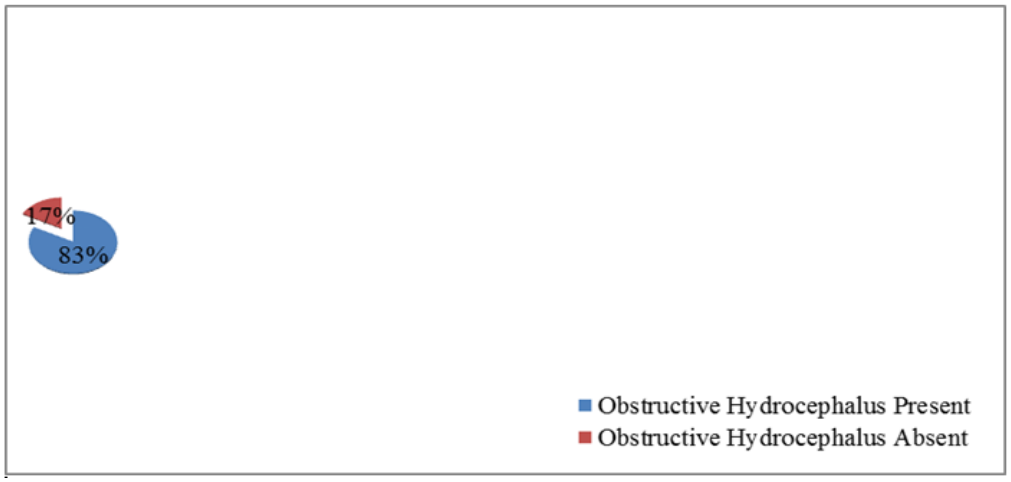



CHART-4: FREQUENCY OF VARIOUS COMPLICATIONS AMONG THE STUDY POPULATION

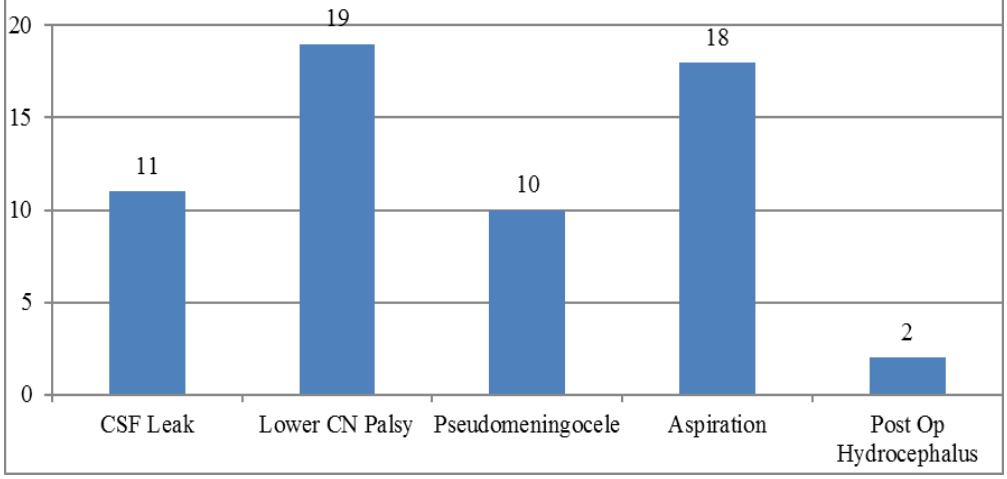
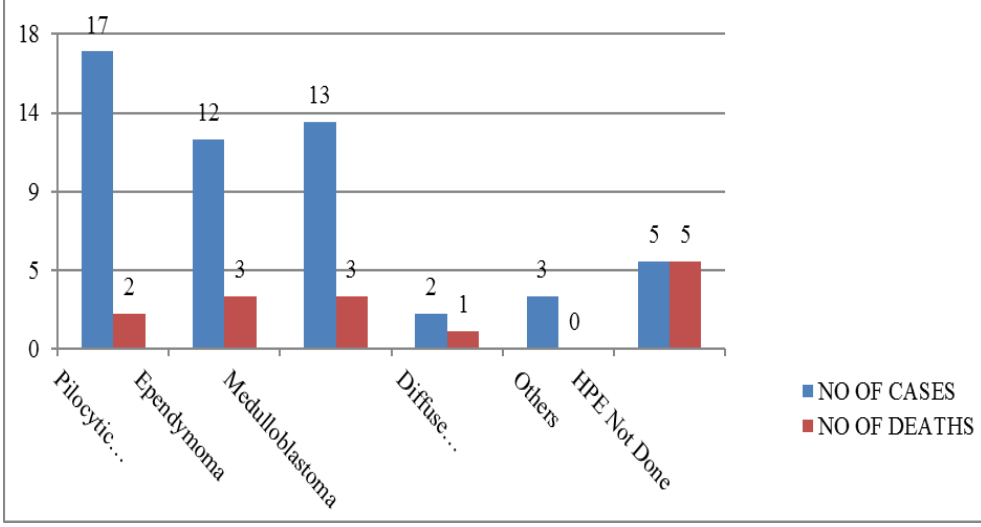



CHART- 5: MORTALITY AMONG HISTOLOGICAL VARIETIES



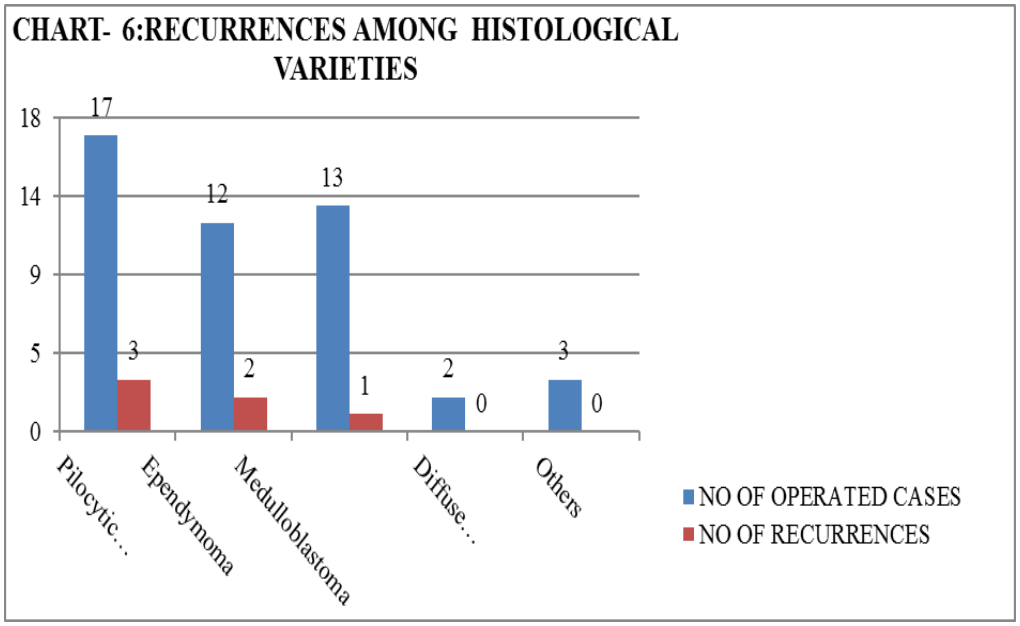


CHART- 6:RECURRENCES AMONG HISTOLOGICAL VARIETIES
18
14
9
5
0
17
3
12
2
13
1
2
0
3
0
Pilocytic…
Ependymoma
Medulloblastoma
Diffuse…
Others
NO OF OPERATED CASES
NO OF RECURRENCES

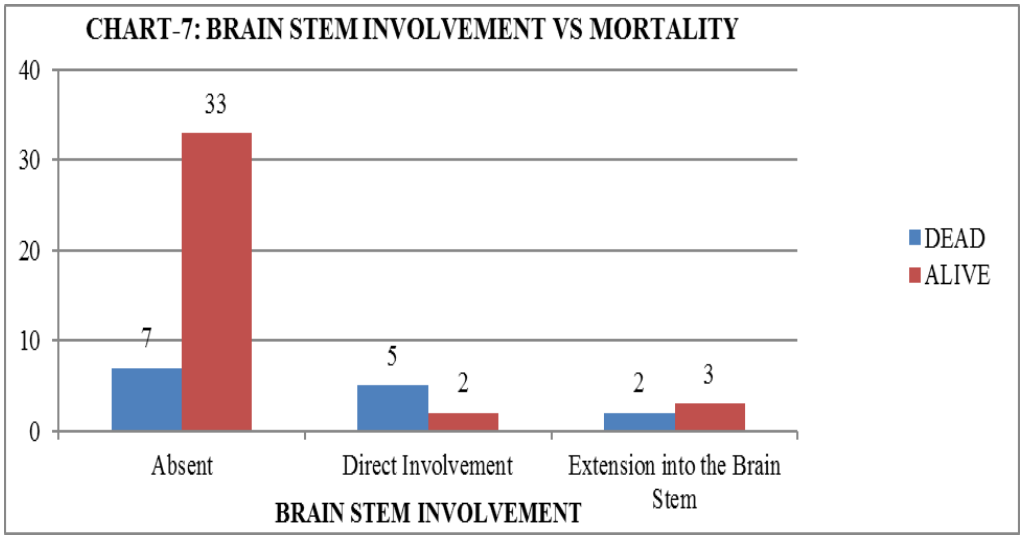


CHART-7: BRAIN STEM INVOLVEMENT VS MORTALITY
40
30
20
10
0
7
33
5
2
2
3
Absent
Direct Involvement
Extension into the Brain Stem
BRAIN STEM INVOLVEMENT
DEAD
ALIVE

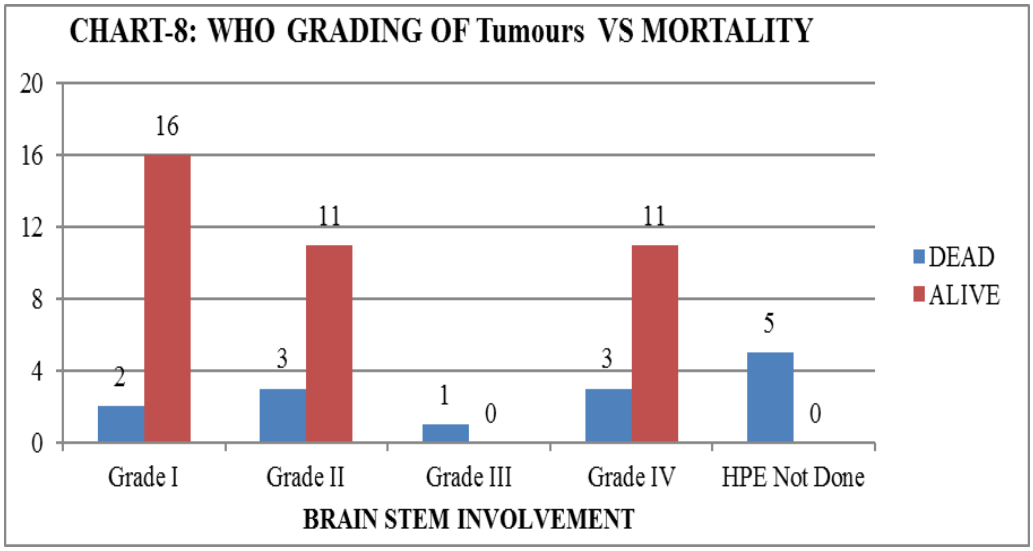


CHART-8: WHO GRADING OF Tumours VS MORTALITY
20
16
12
8
4
0
2
16
3
11
1
0
3
11
5
0
Grade I
Grade II
Grade III
Grade IV
HPE Not Done
BRAIN STEM INVOLVEMENT
DEAD
ALIVE

**Pilocytic Astrocytoma**

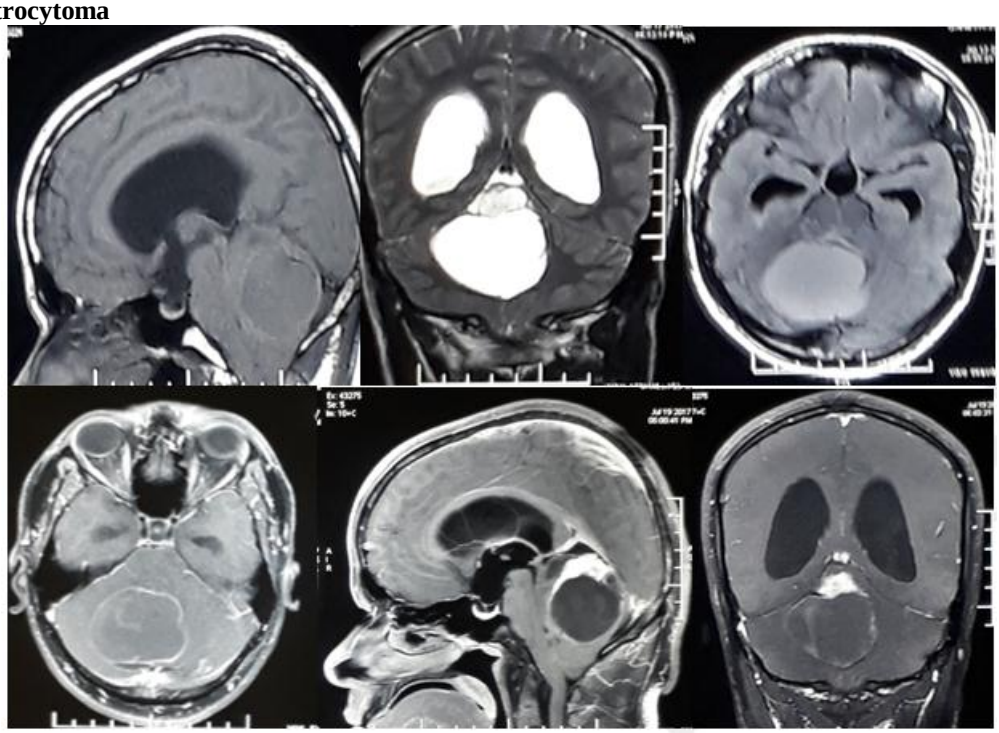


**Fig. - 1 a) Plain and Contrast MRI Brain showing Posterior fossa, right medio-cerebello-vermian region intra-axial, predominantly cystic lesion.**

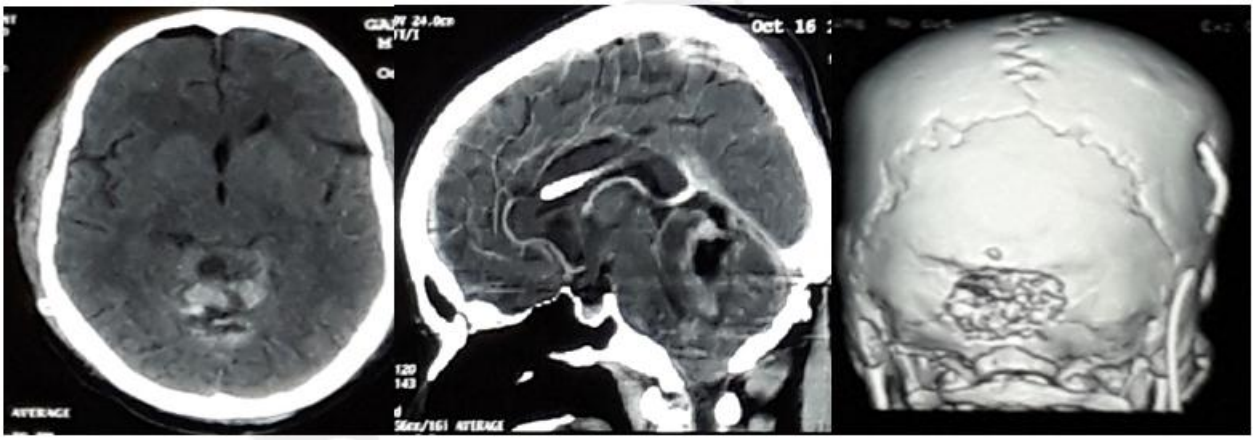



**Fig. - 1 b) Post-operative Plain and Contrast CT with 3D-Reconstruction.**

**Medulloblastoma**

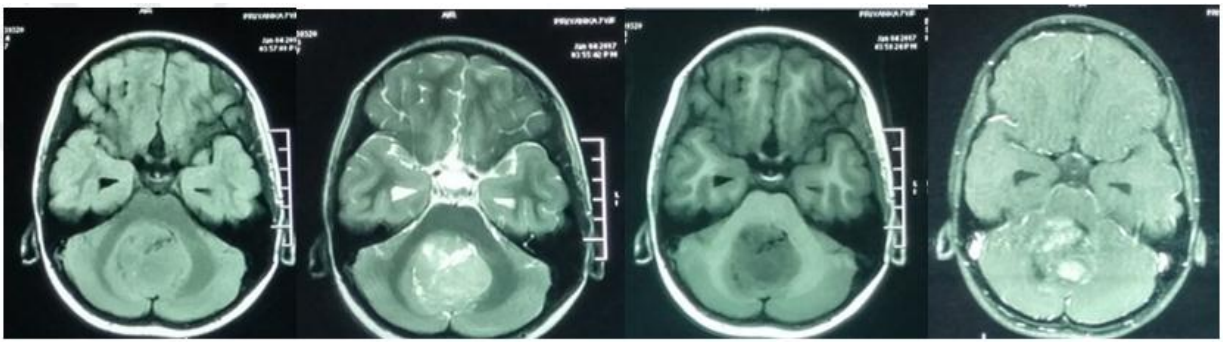


**Fig. - 2 a) Plain and Contrast MRI Brain showing mid-cerebellar mass causing obstructive hydrocephalus.**

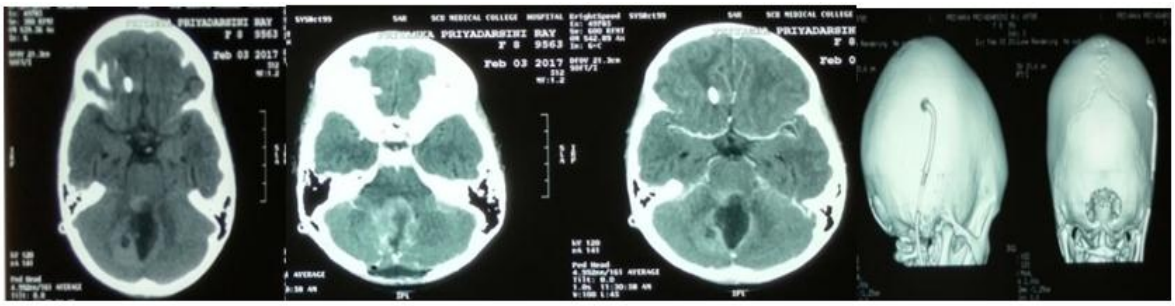


**Fig. - 2 b) Post-Operative Plain & Contrast CT with 3D- reconstruction.**

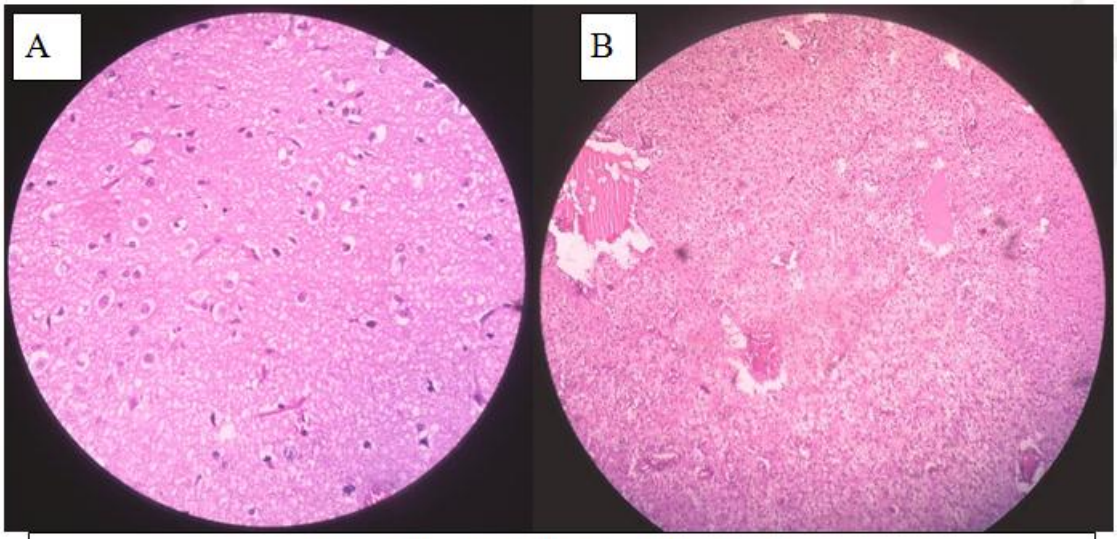
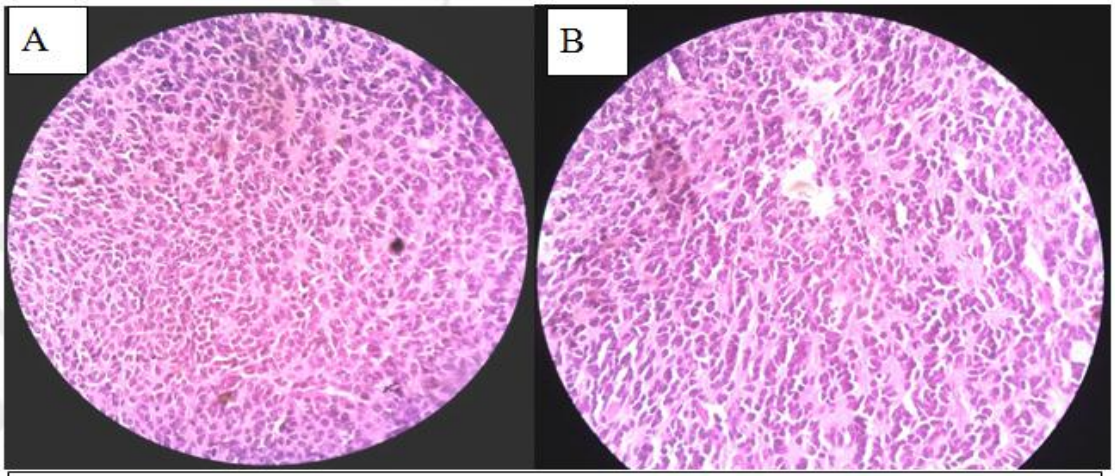



**Fig -3 Showing Pilocytic Astrocytoma.**

A) Showing Bipolar(Piloid) cells in dense fibrillary matrix, Rosenthal fibres and eosinophilic intra-cytoplasmic bodies.
B) Showing Micro-cystic formations.




**Medulloblastoma**

**Figure-4A Showing closely packed undifferentiated cells with round to oval hyper-chromatic nuclei and scanty cytoplasm.**
**Figure-4B Showing groups of tumour cells with attempted perivascular pseudo-rosette formation.**

## DISCUSSION

Posterior cranial fossa tumours in children differ from that of adults in many aspects, like clinical presentation, behaviour, management and prognosis. Certain tumours are common in particular age group. Ahmed N, et al.[9] reviewed 81 cases, of which 58 were male and 23 female (ratio 2.5:1) The maximum number of cases were observed in the age group 5-9 years. Our study is comparable to this study. In our study also, the maximum number of cases [22 (42%) cases] was found in group 2 (6 to 10 years) but with slight female preponderance. Development of posterior fossa surgery was one of the hallmarks of Harvey Cushing contribution to paediatric branch of neurosurgery.[10] Prior to that, posterior fossa tumours were considered inoperable and bony decompression alone was done without attempting tumour removal.

The symptoms were compared and analysed with those of others. Out of the total number of 177 patients studied by Raimondi AJ et al[11], 143 patients had hydrocephalic symptoms. In our study also, the hydrocephalic symptoms (60.6%) were more common than the non-hydrocephalic symptoms (50%).

The CSF diversion procedure in the present case was compared with other authors. Leland Albright L et al[12] studied 86 patients of posterior fossa tumours with secondary hydrocephalus. His conclusion was that the peri-operative mortality was less in the shunted group compared to non-shunted group. In our study, around 66.7% (4 cases out of 6) of the patients who were not subjected to any CSF diversion procedure died after posterior fossa surgery. This implies that any CSF diversion procedure significantly reduces the peri-operative mortality.

The imaging characteristics were compared with histological types and mortality rates. Arle JE et al[13] obtained radiological data via MRI in 33 children with posterior fossa tumours. He was able to predict the tumour pathology in 87.8% of patients when other factors like age, sex and spectroscopy were added to the imaging characteristics. But both the tumour appearance (solid / cystic /mixed) as well as tumour location on imaging was not correlating with mortality in our study population.

The postoperative complications in different study groups were analysed. The results of Riva-Cambrin J et al[14] showed approximately 30% of paediatric patients with posterior fossa tumours exhibited hydrocephalus after tumour resection. In our study, only 2 (3.8%) of the cases had post-operative hydrocephalus.

Only few studies in the literature analysed the predictors of outcome of posterior fossa tumours. Sarkar C et al[15] studied the new methods of risk stratifications among medulloblastoma. The most important factors predicting the outcome were CSF diversion (p=0.047) procedure before definitive surgery, brain stem involvement of the tumour (p= 0.01), extent of tumour resection (p= 0.000) and pathological grade of the tumour (p= 0.001). Figarella-Branger D et al[16] retrospectively assessed the prognostic factors in intracranial ependymomas in 37 children. Our study was comparable to this study because when the extent of resection was maximal, the frequency of post-operative mortality was low. But the location of the tumour was not correlating with the better outcome in our study (p= 0.371).

## CONCLUSION

Paediatric posterior fossa brain tumours are one of the leading causes of death among solid tumours of childhood. The classical clinical presentation and imaging findings are clues to the most likely diagnosis. This study provides a base line data on the incidences, clinical presentations, diagnosis, therapy, complications, tumour types, recurrences, mortality rates and outcome of these tumours. Further researches on understanding tumour biology, improvement in diagnostic techniques, advancement in surgical procedure and postoperative management will help reduce the mortality and improve patient survival.

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