



SPECTRUM OF MIDLINE POSTERIOR FOSSA TUMORS: AN INSTITUTIONAL STUDY

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ABSTRACT

The aim of this study is to assess the clinicopathological trends of posterior fossa midline mass lesions and to assess the outcome of the patients following surgery. Here we present an observational descriptive study done from January 2018 to December 2019 on the patients admitted with posterior cranial fossa midline mass lesions who underwent surgical treatment in department of Neurosurgery, Osmania general hospital, Hyderabad. Patients were evaluated with regard age, sex, clinical, radiological and pathological status at presentation, admission, during and after treatment and outcome of the surgery was evaluated and follow up for a period of 6 months done. Complications evaluated were cerebellar mutism, hydrocephalus, CSF leak, meningitis, posterior fossa syndrome, pseudo-meningocele, cranial nerve palsies and recurrence. Out of 32 patients, 4 (12.5%) developed obstructive hydrocephalous, 4 (12.5%) developed CSF leak, 2 cases (6.25%) developed meningitis, 3 cases (9.37%) developed cranial nerve palsy, 2 cases (6.25%) had respiratory failure, and 1 (3.12%) case developed pseudomeningocele. A total of 4 patients expired in the post-operative period (12.5%). Patients were discharged on day 14 or in hemodynamically stable condition (whichever was later) and were followed up after 1 month, 3 months and 6 months.

KEYWORDS : posterior fossa, midline tumors, mass lesions.**INTRODUCTION:**

Posterior fossa midline mass lesions, due to the limited space in posterior cranial fossa and potential involvement of vital brain stem nuclei, are considered as critical brain lesions. Being more common in children than adults, between 54% and 70% of all childhood tumours arise from posterior cranial fossa compared with 15-20% in adults. Anatomically lesions can arise in this location from, cerebellar vermis (most commonly), fourth ventricle and brain stem.

The clinical presentation associated with posterior fossa midline mass lesions is essentially similar in adults and children. In general, briefer history means likeliness of rapidly growing tumor arising in the axial midline of the posterior fossa, obstructing cerebrospinal fluid (CSF) flow. Early in the course of illness, nonspecific complaints of vague, intermittent headache, fatigue, and personality change may predominate while later manifestations include increased intracranial pressure, focal neurologic deficits secondary to compromise of brain stem or cerebellar tissue, and meningeal irritation. The clinical triad of increased intracranial pressure—headache, vomiting, and blurred or double vision—is the hallmark of an infratentorial tumor.

Contrast enhanced MRI is the investigation of choice, certain radiological findings which, when combined with the clinical information can narrow the differential diagnosis.

Although usually associated with high morbidity and mortality as arising from vital structures, advances in diagnostic imaging, micro surgical techniques, neuro anaesthesia and critical care medicine have reduced the operative morbidity and mortality associated with removal of posterior fossa lesions.

Aims & Objectives:

To assess the clinico-pathological trends of posterior fossa midline mass lesions.

To assess the outcome of the patients following surgery

METHODOLOGY:

We did a prospective observational study conducted from January 2018 to October 2019 in the department of Neurosurgery, Osmania

General Hospital, associated with Osmania Medical College on patients admitted with posterior cranial fossa midline mass lesions who underwent surgical treatment. All age group patients are included in the study. Patients with mass lesions arising on either side of the midline of posterior cranial fossa extending to midline and recurrent posterior fossa midline lesions were excluded.

Patients were evaluated with regard Age, Sex, Clinical, Radiological and Pathological status at presentation, during and after treatment and outcome of the surgery was evaluated and a follow up for a period of 6 months was included in the study. Patients admitted with above mentioned presenting complaints underwent detailed history, clinical examination and investigative protocol as follows

CT, MRI Brain Findings:

A full cranio-spinal pre- and post-contrast MRI scan is performed before surgery to ensure complete tumour staging before surgical contamination of the CSF by blood products.

1. Size of lesion
2. Hydrocephalus
3. Involvement of lower cranial nerves
4. Brain stem compression
5. Surgical Details
6. Site of origin
7. Resectability of tumour
8. Pre-op VP Shunt

Those patients diagnosed with the posterior fossa midline mass lesions were included in the present study.

The patients with raised ICP and hydrocephalus on imaging studies were treated with CSF diversion on emergency basis. All the other cases were operated through midline suboccipital craniotomy and excision of lesion. The patients were advised radiotherapy and chemotherapy in whom so ever required.

Patients were followed at 15 days, 1 month, 3 months and 6 months.

1. Post Op Complication If Any
2. Histological Feature

3. Outcome

Treatment:

High dose dexamethasone is commenced on presentation. Symptomatic hydrocephalus (headache, vomiting, papilledema and reduced level of consciousness) requires urgent surgical treatment. An external ventricular drain is inserted with a view to removing it after definitive tumour resection and resolution of the CSF obstruction.

RESULTS:

A total of 32 patients were included in our study. Age range varied from 1.5 to 60 years with maximum patients in age group 1 to 10 years.

| Age | Incidence | Percentage |
|-------|-----------|------------|
| 1-10 | 20 | 62.5% |
| 11-20 | 6 | 18.7% |
| 21-40 | 3 | 9.35% |
| 41-60 | 3 | 9.35% |
| Total | 32 | 100% |

| Location | Frequency | Percentage |
|-------------------|-----------|------------|
| Cerebellar vermis | 18 | 56.25% |
| Brainstem | 8 | 25% |
| Fourth Ventricle | 5 | 15.62% |
| Tentorium | 1 | 3.125% |

| Pathology | Incidence | Percentage |
|-----------------------|-----------|------------|
| Medulloblastoma | 15 | 46.9 |
| Low-Grade Astrocytoma | 5 | 15.6 |
| Ependymoma | 4 | 12.5 |
| High-Grade Glioma | 4 | 12.5 |
| Arachnoid Cyst | 1 | 3.1 |
| Hemangioblastoma | 1 | 3.1 |
| Meningioma | 1 | 3.1 |
| Epidermoid Cyst | 1 | 3.1 |
| Total | 32 | 100 |

Medulloblastoma was the most common pathology encountered, followed by low-grade astrocytoma, ependymoma, high-grade glioma and others.

| Clinical Features | Incidence | Percentage |
|-------------------|-----------|------------|
| Headache | 16 | 50% |
| Ataxia | 8 | 25% |
| Seizure | 7 | 21.9% |

Morbidity:

- Obstructive hydrocephalus -----4 cases
- CSF leak -----4 cases
- Meningitis -----2 cases
- cranial nerve palsy-----3 cases
- Recurrence -----3 cases
- Respiratory failure -----2 cases
- Pseudomeningocele -----1 case

| Complications | No. of Patients | Percentage |
|---------------------|-----------------|------------|
| Hydrocephalous | 4 | 12.5% |
| CSF leak | 4 | 12.5% |
| Meningitis | 2 | 6.25% |
| Cranial Nerve Palsy | 3 | 9.37% |
| Recurrence | 3 | 9.37% |
| Respiratory Failure | 2 | 6.25% |
| Pseudomeningocele | 1 | 3.12% |

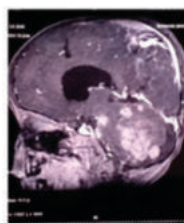
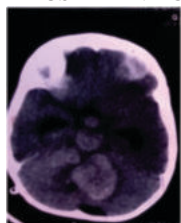
MORTALITY:

Out of 32 cases there were 4 deaths noted.

Follow up:

Follow-up- ranges from 1 month – 24 months. Average follow-up period being 12 months. 3 cases showed recurrence

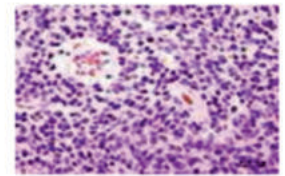
ILLUSTRATIVE CASE



A 3yrs male child brought to the hospital with the history of inability to walk and vomiting for 3months. CT brain and MRI shows SOL in the posterior fossa with dilated ventricular system suggestive of Medulloblastoma. VP shunt was done preoperatively. Patient operated through midline suboccipital craniotomy and near total excision of the lesion was done. In Postoperative period was patient developed CSF leak for which lumbar drain was placed. Patient improved and discharged after 10 days.



Intra Operative Image



HISTOPATHOLOGICAL IMAGE Showing a diffuse pattern of growth with poor cellular differentiation nuclear molding and minimal indistinct cytoplasm – classic Medulloblastoma

DISCUSSION:

midline posterior fossa mass lesions are more commonly observed in children than adults. In the present study 28.8% of lesions of posterior fossa occurred in the midline.

Age And Sex Incidence:

In our study 62.5% of midline posterior fossa mass lesions occurred in age group of up to 10 years (most common age of presentation) and 82.2% cases observed below 20 years.

Incidence in Males and females was found 1:1 in the present study.

Location:

In our study, posterior fossa midline mass lesions were located in cerebellar vermis most commonly, followed by brain stem and fourth ventricle regions.

Surgical excision of the posterior midline mass lesions is most often challenging and troublesome to cure because of location, infiltration of brain stem and surrounding structure, encasement of vessels and cranial nerves which make it difficult to radical excision.

In the Pre-Micro surgical era operative mortality was high ranging from 22% to 57%. The advent of newer neuro imaging modalities, microsurgical techniques and conservative radical approaches has resulted in reduction in morbidity and mortality. In the present study the reported mortality rate is 12.5% and we found three cases of recurrence.

The gross appearance of the lesions varied according to the pathological entity.

Medulloblastoma:

In our study Medulloblastoma was the most common pathological type and constituted 46.9% of lesions. It belongs to Primary Neuro Ectodermal Tumors(PNETs). Macroscopically is appeared as soft and friable lesion with areas of necrosis and focal hemorrhage and confined itself to vermis, fourth ventricle and adjacent cerebellar hemisphere in 50% cases.

Low Grade Astrocytoma:

In the present study low grade astrocytoma is second most common type of pathological type, constituting 15.6% of cases of midline posterior fossa lesions. Of the total 5 cases of low grade astrocytomas, 4 cases were paediatric brainstem lesions and 1 case was found in adult age group. The mean age of occurrence was < 20 years but they can occur at any age.

Ependymomas:

Ependymomas are third most common tumour in midline posterior fossa region and belong to PNETs with ependymal differentiation. In our study ependymoma represents 12.5% cases. They are common in children younger than 20 years of age as observed in our study but 1 odd case was found in elder age group. Macroscopically, Ependymoma appears as tan to greyish in colour, soft in consistency with areas of cysts, hemorrhage, necrosis and calcification. It frequently fills fourth ventricle.

High Grade Gliomas:

Brain stem gliomas constitute about 20% of paediatric brain tumors

and are uncommon in adults accounting for less than 1%. In our study 4 lesions were high grade gliomas, that is 12.5% of study population, all of them arising from brainstem. All cases were found in paediatric age group.

Arachnoid Cysts:

Posterior Fossa Arachnoid cysts (PFACs) are common in the midline and presents as discrete CSF collections that are clearly separate from the Fourth ventricle and valliculae. In our study it was found in 1 case, that is 3.1% of midline posterior fossa lesions in our study group.

Hemangioblastoma:

Haemangioblastoma is a WHO grade I tumor which is highly vascular and seen in the cerebellum and spinal cord. In our study 1 of midline posterior fossa lesions was hemangioblastoma, that is 3.1%, found in adult age group.

Hemangioblastomas are typically well circumscribed cystic lesion with small mural nodule on gross appearance. Its presentation may vary according to the solid component with central cyst.

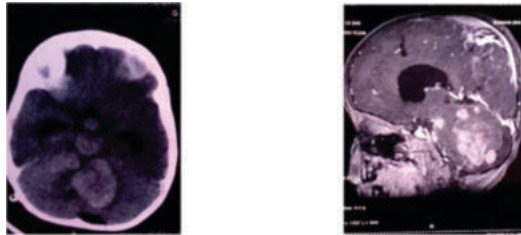
Meningiomas:

Midline Meningiomas belongs to clival, tentorial and Foramen magnum types of posterior fossa meningiomas. In our study 1 case was meningioma, that is 3.1%. It was arising from tentorium cerebelli. These lesions macroscopically appear as greyish white or reddish brown in colour, variable consistency with moderate to high vascularity with capsule.

Epidermoid Cysts:

These are rare in posterior fossa, if present they typically are located in midline. Its incidence was 3.1% of lesions in our study. Grossly appears greyish white in colour, variable consistency and may present with sinus tract in some cases.

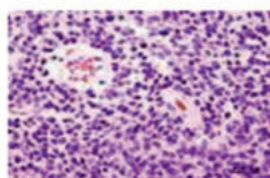
Illustrative Case:



A 3yrs male child brought to the hospital with the history of inability to walk and vomiting for 3months. CT brain and MRI shows SOL in the posterior fossa with dilated ventricular system suggestive of Medulloblastoma. VP shunt was done preoperatively. Patient operated through midline suboccipital craniotomy and near total excision of the lesion was done. In Postoperative period was patient developed CSF leak for which lumbar drain was placed. Patient improved and discharged after 10 days.



Intra Operative Image



HISTOPATHOLOGICAL IMAGE Showing a diffuse pattern of growth with poor cellular differentiation nuclear molding and minimal indistinct cytoplasm - classic Medulloblastoma

Surgical Treatment:

Surgical treatment is complete excision of the lesion whenever possible. This depends upon the location of the lesion, grade and extent of the tumour as assessed MRI. About 80% of posterior fossa lesions occur in and around the fourth ventricle, presenting with hydrocephalus. In the present study 40.6% of patients required CSF diversion preoperatively.

All posterior fossa midline mass lesions were approached through midline suboccipital craniectomy and involved splitting of vermis in large tumours, that is TRANSVERMIAN approach. The modification of this approach is TELOVELAR approach which involves dissection

of the cerebellomedullary fissure to reach Fourth ventricle without splitting cerebellar vermis. In our study five cases were operated through Telovelar approach.

Operative findings depend upon pathological type and grade of the lesion. High grade lesions may present with infiltration to surrounding structures and are not amenable to complete resection. The treatment of choice for Arachnoid cyst is complete removal, if not possible partial or complete excision of cyst wall or marsupialisation of cyst into sub arachnoid space.

In lesions where tumour capsule or tumour tissue is firmly adherent to brain stem and fourth ventricle can be left over to avoid post-operative morbidity and mortality. Partial or subtotal resection will result in recurrence at varied periods according to pathological type of lesion and is noted in three cases of our study. If the capsule is thin and is adherent to brain parenchyma it can be teased out slowly by sharp micro dissection techniques under high magnification. These are tightly packed solid tumors and do not easily give in for internal decompression.

Frequent thorough saline irrigation is very important to prevent CSF contamination, post-operative development of aseptic meningitis and hydrocephalus.

Immediate post-operative CT scan confirms the extent of surgical resection but even after near total resection of lesion shows hypodensity in tumour bed possibly due to long standing tissue deformation of the neuronal structures. Coagulation of the residual capsule has been advocated by some authors but is no longer practiced now.

Post-operative MRI is helpful to diagnose early recurrence or residual tumour also differentiate radiation necrosis from residual tumour.

Radiotherapy And Chemotherapy:

Adjuvant therapy for posterior fossa midline lesions plays a key role in control of disease, prevention of recurrence and improved survival rate in patients.

In our study post-operative cases of Medulloblastoma, Ependymoma, Hemangioblastoma, High grade Gliomas and lesions with infiltration to brain stem and surrounding structures were advised adjuvant therapy.

CONCLUSION:

- Posterior fossa midline mass lesions are more common in children than adults.
- There is higher incidence of malignant lesions in children when compared to adults as observed in our study.
- Incidence rate of midline mass lesions in posterior fossa is 28.8 %.

The common sites of origin of posterior fossa midline lesions are the vermis of cerebellum, brainstem and fourth ventricle.

- Most common pathological types are Medulloblastoma and low grade Astrocytoma.
- About 40.6% of posterior fossa midline lesions require CSF diversion prior to definitive surgery.
- Recurrence was noted in three cases; two in Medulloblastoma and one in brainstem astrocytoma.
- Mortality was noted in 12.5% cases of posterior fossa midline mass lesions.
- The postoperative cases, cases with partial resection and cases of high grade pathology with surrounding infiltration were advised adjuvant therapy.
- With advent of newer imaging modalities and micro neurosurgical techniques, it is possible to achieve total or near total resection whenever possible to avoid recurrence of the lesion resulting in better patient outcome

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