Panacea Journal of Medical Sciences 2022;12(1):198–203

Content available at: https://www.ipinnovative.com/open-access-journals

Panacea Journal of Medical Sciences

Journal homepage: http://www.pjms.in/

Original Research Article

Posterior fossa midline mass lesions clinicopathological study and management

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ARTICLE INFO

Article history:
Received 13-03-2021
Accepted 14-05-2021
Available online 30-04-2022

Keywords:
Posterior fossa midline mass lesions
Medulloblastoma
cerebellar vermis

ABSTRACT

Introduction: Some of the most severe brain cancers are tumors in the posterior fossa. This is due to the small space available in the posterior fossa, as well as the possibility of essential brainstem nuclei involvement.

Aims: To assess the clinicopathological trends of posterior fossa midline mass lesions and to assess the outcome of the patients following surgery.

Materials and Methods: This is a prospective study done in the patients admitted with posterior cranial fossa midline mass lesions who underwent surgical treatment in department of Neurosurgery. Patients were evaluated with regard Age, Sex, Clinical, Radiological and Pathological status at presentation, admission, during and after treatment.

Results: Incidence rate of midline mass lesions in posterior fossa is 34.6%. Common age group of occurrence of posterior midline lesions is less than 10 years. Posterior fossa midline lesions are more common in males. The common sites of origin of posterior fossa midline lesions are the Vermis of Cerebellum and fourth ventricle. Most common pathological types are Medulloblastoma and Pilocytic astrocytoma. About 29.6% of posterior fossa midline lesions require CSF diversion prior to definitive surgery. Recurrence is noted in two cases one in brainstem astrocytoma and another in medulloblastoma. Mortality is noted in 18.5% cases of posterior fossa midline mass lesions.

Conclusion: Prognosis is good in patients with total excision. This drives the surgeon to go for total excision.

1. Introduction

posterior fossa lesions in the midline of the brain are classified as critical. This is due to the restricted space in the posterior cranial fossa and the likelihood of essential brain stem nuclei being involved. Children are more likely than adults to develop midline lesions of the posterior cranial fossa. In children, between 54 percent and 70 percent of tumors originate in the posterior cranial fossa, compared to 15-20 percent in adults. [1]

Most commonly the tumours are from cerebellar vermis. Most of the histopathological varieties of posterior fossa midline mass lesions have been reported at any age, certain types of including primitive neuro-ectodermal tumours (PNET), medulloblastomas, ependymomas, astrocytoma's of cerebellar vermis and brain stem more commonly effect infants and children's. Most common type is medulloblastoma of vermis, cystic lesion of fourth ventricle and tumours of brain stem. Certain types of glial tumours including mixed gliomas are common in children. These are more frequently located in cerebellum (67%) and are usually benign histologically. As a rule astrocytoma tumours of adult most commonly display the histological features of malignancy than those of children certain tumours including metastatic lesions, hemangioblastoma

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https://doi.org/10.18231/j.pjms.2022.037

and lymphoma are more commonly affect adults than children.[2]

In adults and infants, the clinical appearance of posterior fossa midline mass lesions is approximately the same. In general, the shorter the patient's history, the more likely the tumor is to be rapidly developing and emerging in the axial midline of the posterior fossa, obstructing CSF flow. Increased intracranial pressure, focal neurologic defects secondary to compromise of brain stem or cerebellar tissue, and meningeal discomfort are typical late in the course of illness, while nonspecific symptoms of vague, irregular headache, exhaustion, and personality change may predominate early in the course of illness.

MRI has generally super ceded computed tomography for nearby all types of posterior fossa mass lesions. However while neither MRI nor CT scan distinguish one tumour type from another type with certainty. Certain trends exists which, when combined with the clinical information can narrow the differential diagnosis. Posterior fossa midline mass lesions 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.[3]

2. Materials and Methods

This is a prospective study done from September 2013 to February 2016 on the patients admitted with posterior cranial fossa midline mass lesions who underwent surgical treatment in department of Neurosurgery, Osmania General Hospital. Patients were evaluated with regard age, sex, clinical, radiological and pathological status at presentation, admission, during and after treatment.

2.1. Inclusion criteria

All age group patients are included in the study with Mass lesions arising from midline posterior cranial fossa.

2.2. Exclusion criteria

Mass lesions arising on either side of the midline of posterior cranial fossa extending to midline, recurrent posterior fossa midline lesions, patients admitted with the features of headache, vomiting, ataxia, motor weakness and cranial nerve palsy are evaluated with the investigative protocol. Those patients diagnosed with the posterior fossa midline mass lesions were included in the present study.

Clinical information, follow up data of cases collected which includes age, gender, location, symptomatology and histopathological diagnosis were included. 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.

2.3. Routine Blood Investigations

Complete blood picture, Blood sugar levels, Renal parameters, Electrolytes, Coagulation profile, Viral screening, Liver function tests, Ultrasound abdomen and pelvis Chest x-ray,X-ray DL spine, CT Brain plain and contrast MRI brain plain and contrast CT Angiogram.

Treatment is determined by the patient's condition at the time of admission. On presentation, high-dose dexamethasone is initiated. This also leads to a decrease in symptoms. Symptomatic hydrocephalus, on the other hand, necessitates immediate surgical care (headache, vomiting, papilledema, and reduced level of consciousness). After conclusive tumor resection and resolution of the obstruction to CSF flow, an external ventricular drain is inserted with the intention of removing it. Before surgery, a full craniospinal pre- and post-contrast MRI scan is performed to ensure complete tumor staging and to prevent surgical contamination of the CSF with blood products.

2.4. Surgical technique

A midline suboccipital technique is used to resect tumors in the posterior fossa. The patient is lying down with his or her head raised and flexed. A midline longitudinal incision is made from the external occipital protuberance to the level of the posterior arch of C1 to expose the squamous occipital bone. The muscular and ligamentous attachments to C2 are not exposed; preserving these attachments is vital because subsequent radiotherapy and surgery can result in progressive cervical instability. The C1 posterior arch is visible but intact. A craniotomy of the posterior fossa is superior to a craniectomy. This reduces post-operative pain and allows better restoration of CSF flow around the foramen magnum post-operatively.

The foramen magnum is used to extend the craniotomy. The dura is opened in a Y shape, keeping in mind that an occipital sinus descending in the midline from the torcula may need ligation in most young children. The microscope is brought into the field after the dura has been reflected, and the arachnoid at the craniocervical junction is opened..

After the tumor has been removed, the cavity's walls are carefully re-examined to ensure that no tumor has been left behind. If the capsule of a pilocytic astrocytoma enhances on post-contrast MRI, it should be resected as well. Without the use of oxidized methylcellulose or other haemostatic agents, haemostasis is achieved; however, these agents appear to enhance on MRI and can be misinterpreted as residual tumor. A patch of suturable dural replacement is used to close the dura in a watertight manner. Ependymomas

also reach into the cerebellopontine angle, requiring a lateral extension of the standard midline suboccipital craniotomy. This allows both the fourth ventricular and retrosigmoid corridors to be used during the primary operation, increasing the chances of achieving gross total resection. The cranial nerves, the brainstem, and the arteries of the posterior fossa are all infiltrated by these tumors. Gentle dissection is needed, using low-pressure microsuction in conjunction with a microdissector, in a lateral to medial direction along the cranial nerves. These nerves are delicate and elongated, as are the perforating arteries from the basilar artery. In most cases, patient dissection enables full tumor resection with no lower cranial nerve palsy or brainstem infarction. The analysis found complications.

3. Results

Table 1: Demographic distribution in study

Age in years	Incidence	Percentages
1-10	13	48
11-20	4	15
21-40	7	26
41-60	3	11
Gender		
Male	15	55.6
Female	12	44.4

Age range varied from 1yr to 60 yrs, Youngest was 14 months and oldest 60 years. The sex distribution of posterior fossa midline mass lesions in this study showed of incidence in male cases is 15 (55.6%), female cases is 12 (44.4 %).Table 1

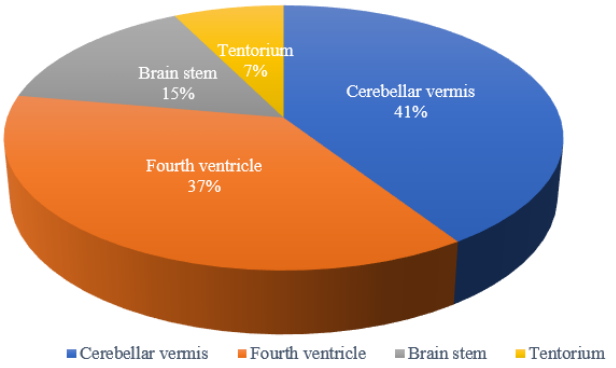

Fig. 1: Showing the location of lesions in mid line of posterior fossa

The most common locations are the cerebellar vermis, lobe followed by the fourth ventricle and then the brainstem.Figure 1

Majority of the tumors are Medulloblastomas, Pilocytic astrocytoma and ependymomas.

Table 2: Pathological distribution of lesions

Pathological distribution	I ncidence	Percentage
Medulloblastoma	7	25.9%
Pilocytic astrocytoma	5	18.5%
Ependymoma	4	14.8%
Arachnoid cyst	3	11.8%
Hemangioblastoma	2	7.4 %
Meningiomas	2	7.4 %
Epidermoid cyst	1	3.7%
AT/RT	1	3.7%
Others	2	7.4%

Table 3: Symptoms and signs in present study

Clinical Features	Incidence	Percenta ge
Headache	17	62 %
Vomiting	5	18.5%
Ataxia	3	11.1%
Seizure	1	3.7%
Cranial Nerve Palsy	1	3.7%

The most common presenting symptom is raised intracranial pressure with headache and vomiting.

In our study out of 27 cases 8 cases were required pre operative emergency CSF diversion for who admitted with features of raised intra cranial pressure. These patients were improved post operatively.

Total no. of cases -27 (100%).

Preoperative CSF diversion is done in 8 cases (29.6%).

All cases were operated through Midline suboccipital craniotiomy. Suboccipital craniotomy was preferred to craniectomy. In 7 cases partial resections was done and in 18 cases complete or near total resection.

Table 4: Complications in study

Complications	Number o f Patients
Hyrdocephalus	2
CSF leak	3
Meningitis	2
Cranial nerve palsy	1
Recurrence	2

Out of 27 cases there were 5 deaths noted. Follow-up- ranges from 6 months – 40 months. Average follow-up period being 36 months. Two cases of recurrence were reported so far.

4. Discussion

Posterior fossa lesions are more common in children than adults. About 54% to 74% of all childhood brain tumors arise in the posterior fossa when compared with 15% to 20% in adults. In our study midline posterior fossa mass lesions are more commonly observed in children than adults. In the present study 34.6% of lesions of posterior fossa occurred in the midline. Most common age of presentation is < 10

years of age. 61.2% of posterior fossa midline lesions are observed before the age of 20 years. According to Albright, most of the posterior fossa tumors occur in the age group between 1 and 10 years.[4]

In our series, male participants are 15(55.6%) in number and female participants are 12(44.4%). The male predominance may be due to increased number of male patients being investigated, which is a social feature in Indian society. May PL et al showed slight male predominance was seen.[5]

The most common location of posterior fossa midline mass lesion in present study is Cerebellar vermis followed by Fourth ventricle. Cerebellar vermis is 40.7%, Fourth ventricle is 37%, Brain stem is 14.8% and Tentorium is 7.4%. According to data from the Children's Hospital, Philadelphia, MBs range about 36%, cerebellar astrocytoma in 28%, brainstem glioma 9%, and ependymoma in 4%.[6]

In a study from the Hospital for Sick Cjildren in Toronto, MB range about 27%, astrocytomas 23%, brainstem glioma in 9%, and ependymoma in 8% of cases.[7]

In Kadali Satya Vara Prasad et al study MB form about 34%, astrocytomas 27%, ependymoma 18%, tuberculomas 10%, brainstem glioma 5%, meningiomas in 2%, and choroid plexus papilloma 2% were observed.[8]

In the present study posterior fossa midline mass lesions are located in cerbellar vermis, followed by fourth ventricle and brain stem regions. The gross appearance of the lesions varies according to the pathological entity. In our study Medulloblastoma is most common pathological type occurred and constitutes more than 25.9% of posterior fossa midline lesions. Is commonly observed in males and belongs to Primary Neuro Ectodermal Tumors (PNETs). Macroscopically is appeared as soft and friable lesion with areas of necrosis and focal hemorrhage and confine itself to vermis ,Fourth ventricle and adjacent cerbellar hemisphere in 50% cases.

In the present study Pilocytic astrocytoma is second most common type of pathological type constitutes 18.5% of cases of midline posterior fossa lesions. Of the total 5 cases of Pilocytic astrocytomas, 3 cases were brain stem high grade lesions. The mean age of occurence is < 20 years but can occur at any age. The high grade lesions grows rapidly, highly vascular and infiltrates surrounding structures at the time of diagnosis and associated with poor outcome, recurrence and mortality.

Ependymomas are third most commonest tumour in midline posterior fossa region and belong to PNETs with ependymal differentiation. In our study ependymoma represents 14.8% cases. Common children younger than 10years age observed in our study and 50% cases in children less than 4years. Macroscopically Ependymoma appear as tan to greyish in colour, soft in consistency with areas of cysts, hemorrhage, necrosis and calcification. It frequently fills Fourth ventricle.

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 valleculae. In our study 11.8% of cases of midline posterior fossa lesions are Arachnoid cysts.

Haemangioblastoma is WHO grade I tumor, highly vascular and seen in the cerebellum and spinal cord .In our study 2 of midline posterior fossa lesions are hemangioblastomas, more common in adults and rare in children. Hemangioblastomas are typically well circumscribed cystic lesion with small mural nodule on grass appearance. Its presentation may varies according to the solid component with central cyst.

Midline Meningiomas belongs to clival, tentorial and Foramen magnum types of posterior fossa meningiomas. In our study 7.4% cases of posterior fossa midline lesions are meningiomas. These lesions macroscopically appears as greyish white or reddish brown in colour, variable consistency with moderate to high vascularity with capsule.

These rare in posterior fossa if present they typically locate in midline and represented 3.7% of lesions in our study. Grossly appears greyish white in colour, variable consistency and may present with sinus tract in some cases. Is rare tumour and represented 3.7% cases in our study. They are macroscopically indistinguishable from PNETs and diagnosed by histopathological examination. In present study two rare cases were noted one is cysticercosis and another is soft tissue tumor.

In our study majority of midline posterior fossa mass lesions occurred in age group up to 10 years and nearly 64% cases are observed below 20 years. Males out number females in incidence in the present study. Clinical presentation of posterior fossa midline mass lesions varies according to the location of tumour, pathological type, rate of tumour progression and mass effect. Common presenting symptoms include headache, vomiting, swaying while walking and seizures. Clinical findings includes ataxia, cerebellar dysfunction, cranial nerve palsies and hemi paresis when brainstem is compressed.

Posterior fossa midline mass lesions includes Medulloblastoma, Astrocytomas, Ependymoma, Arachnoid cyst, Hemangioblastoma etc. In our study one case of Cysticercosis and one case of soft tissue sarcoma are found. The Posterior fossa midline mass lesions require MRI of brain and spinal cord to rule out leptomeningeal spread preoperatively for planning of management. CT scan of the brain is also useful to diagnose some lesions alone with or without contrast.

Tumours with higher cellularity eg; medulloblstoma appears as higher density on CT or hypointense onT1sequence of MRI when compare to pilocytic astrocytoma or ependymoma. Solid portion of the pilocytic astrocytoma are hyperintense to CSF on T2 sequence of MRI in 50% cases. Ependymoma typically extends

laterally or inferiorly from Foramen of Luschka, Foramen of Magendie respectively. Epidermoid cysts MRI they are hypodense in T1w1 and hyperintense like CSF in t2wi because of their solid nature but on CT scan appears hypodense mass lesion irregular shaped that does not show contrast enhancement.

Diffusion Weighted Images (DWI) and flair images are helpful in differentiating cystic lesions. Imaging characteristics often overlap or atypical and diagnosis based on traditional imaging, Diffuse Weighted Imaging (DWI) and Magnetic Resonance Spectroscopy (MRS) can enhance the diagnostic predictive value. Hydrocephalus may be found in> 30% cases may significant and requires a CSF diversion procedure pre-operatively results in improvement in patient status. [1]

Surgical treatment is complete excision of the lesion whenever possible. This depends upon the location of the lesion, grade and extent of the tumour which can be assessed accurately pre operatively by MRI. About 80% of posterior fossa lesions are occured in and around the fourth ventricle will presents with hydrocephalus. In present study 29.6% of patients required CSF diversion preoperatively.

In our present study all posterior fossa midline mass lesions approached thorough midline suboccipital craniotomy and it involves splitting of Vermis of Cerebellum for large tumours. The modification of this approach is TELOVELAR approach is involves dissection of the cerebellomedullary fissure to reach Fourth ventricle without splitting cerebellar vermis. In our study two case were operated through Telovelar approach.

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

In lesions where tumour capsule or tumour tissue firmly adherent to Brain stem and Fourth ventricle can left over to avoid post operative morbidity and mortality. Partial or subtotal resection will results in recurrence and is noted in two cases of our study. The capsule is thin and is adherent to brain parenchyma but 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. Sub total or partial resection leads to recurrence at varied periods according to pathological type of lesion. Immediate post operative CT scan confirms the extent surgical resection but even after near total resection of lesion shows hypo density 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. Obstuctive hydrocephalus 2 cases, CSF leak 3 cases, Meningitis 2 cases, Cranial nerve palsy 1 case. Recurrence is seen in 2 cases. Mortality is noted in 5 cases. In the present study five of our patients expired due to various post operative complications. 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. Qwallner et al. [9] found that pilocytic astrocytomas are associated with improved survival. Bruce et al. [10] reported 5.6% of recurrence. Pencalet et al. [11] also showed that it is a sole independent factor that significantly detects the tumor recurrence. Total resection influences surgical outcome and quality of life. Brainstem infiltration and nonpilocytic variant of astrocytomas were associated with recurrence.

In the Pre-Micro surgical era operative mortality was high ranged from 22% to 57%. With the advent of newer neuro imaging modalities, microsurgical techniques and conservative radical approaches resulted in morbidity and mortality. In the present study the reported mortality rate is 18.5%. In our study the follow up period is very small and we found two cases of recurrence in residual tumours.

Adjuvant therapy for posterior fossa midline lesions plays a key role in control of disease, prevention of recurrence and improve survival rate in patients. In our study post operative cases Ependymoma, AT/RT, Hemangiblastoma, high grade Gliomas and lesions with infiltration to brain stem and surrounding structures advised adjuvant therapy. [12]

5. Conclusions

Posterior fossa tumors are predominantly seen in children with peak incidence in the first decade. Majority of the tumors are Medulloblastomas, Pilocytic astrocytoma and ependymomas. The most common presenting symptom is raised intracranial pressure with headache and vomiting. The most common locations are the cerebellar vermis, lobe followed by the fourth ventricle and then the brainstem. Out of 27 cases there were 5 deaths noted. Two cases of recurrence were reported.

The postoperative cases, cases with partial resection and cases of high grade pathology with surrounding infiltration were advised adjuvant therapy. With advent 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 total cure of the patient. Prognosis is good in patients with total excision.

This drives the surgeon to go for total excision.

6. Conflict of Interest

No conflict of interest.

7. Source of Funding

None.

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Cite this article: Rajamohan B, Rao PN. Posterior fossa midline mass lesions clinicopathological study and management. *Panacea J Med Sci* 2022;12(1):198-203.