Childhood Posterior Fossa Tumours: Experience from a Tertiary Center in Nepal

Bikas Thapa, Amit Bahadur Pradhanang, Gopal Sedain, Sushil Krishna Shilpakar, Mohan Raj Sharma

Department of Neurosurgery, Tribhuvan University Teaching Hospital

Corresponding Author: Dr. Bikas Thapa, Email: bikasthapa1141@gmail.com
ORCID iD: https://orcid.org/0000-0001-9605-1694

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ABSTRACT

Background: Brain tumour is a common problem in paediatric age group and posterior fossa is the commonest location. Treatment of these tumours is difficult due to a confined space and potential involvement of brainstem. The aim of this study is to analyse the clinical features, radiological findings, pathological diagnosis, surgical management, postoperative complications and outcome in 3 months in paediatric patients with posterior fossa tumours.

Method: Retrospective chart review of 24 patients treated in the Department of Neurosurgery, TUTH, between 2019 April – 2023 April was done. The variables included were age, sex, clinical features, radiological findings, histopathological diagnosis, management and postoperative complications.

Results: Twenty-four patients (30%) out of 80 childhood tumors were located in posterior fossa. The most common presenting symptoms was headache in 83.3% and vomiting in 75%. The most common location of tumour was the fourth ventricle in 66.7% while most common tumour histology was Type IV medulloblastoma in 45%. The commonest mode of treatment in 90% of cases was Ventriculoperitoneal (VP) shunting followed by surgical excision of tumour.

Conclusion: Posterior fossa tumours are common tumour occurring in children. Majority of the patients presented with features of raised intracranial pressure and gait ataxia. Fourth ventricle and cerebellar vermis are the most common location. Most common histology types are medulloblastoma followed by pilocytic astrocytoma and ependymoma. Management involves addressing acute hydrocephalus followed by definitive surgery.

Keywords: Medulloblastoma, Paediatrics, Posterior fossa, Surgery, Ventriculoperitoneal shunt

INTRODUCTION

Brain tumours are the most common solid tumours and commonest cause of cancer death in infants and children aged 0–14 years, second to leukemia [1,2]. Posterior fossa (PF) tumours have higher incidence in paediatric age groups comprising between 54% and 70% of childhood brain tumours compared to 15%–20% in the adult population [3-5]. The majority of patients present with features of raised intracranial pressure. Tumours in this location are challenging due to the narrow area of posterior fossa and close proximity to critical structures such as lower cranial nerves, vertebral artery, basilar artery and brainstem. The most frequent histological pathological types are medulloblastoma, ependymoma, and cerebellar astrocytoma [6].

There is large burden of neurosurgical diseases in low- and middle-income countries (LMIC). However, the coverage of paediatric patients in cancer registry is poor [7]. Although there are many papers on this topic from neighbouring countries and Asia, there is paucity of literature from Nepal. There is no national database of brain tumours among paediatric population yet in Nepal [8]. The first operative series of childhood CNS tumour in Nepal was published in 2015 from our institute. We aim to analyse the clinical features, radiological findings, pathological diagnosis, surgical management, postoperative complications in paediatric patients with posterior fossa tumours.

METHODS

Permission from the Institutional Review Committee of our

Article information

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institute was obtained before the data retrieval. A total of 24 paediatric patients with posterior fossa tumours treated in the Department of Neurosurgery, Institute of Medicine, Nepal between 2019 April – 2023 April were analyzed. The inpatient records of patients were collected. The exclusion criteria in this study were those with incomplete medical records.

Management protocol

All patients with suspected PF tumour on computed tomography underwent routine MRI imaging of brain with contrast. Those presenting with acute hydrocephalus underwent CSF diversion procedure before definitive tumour surgery. In some patients, direct posterior fossa tumour surgery was done with external ventricular drain placement which was gradually weaned off in post-operative period. CSF routine analysis along with cytological assessment for malignant cells was done in all cases. Patient was managed in paediatric ICU and followed with routine post-operative care and discharged in 7-10 days in uncomplicated case. Histopathological categorization was done by pathologist of our center in accordance to WHO classification of brain tumours. Further treatment (Chemotherapy and radiotherapy), was dependent upon the grade and type of tumour and degree of resection by a routine MRI in 6 weeks after surgery. Those patients who had inoperable tumour or who declined surgery, symptomatic management was offered. As this study follow up was only for 3 months, describing tumour recurrence and long-term outcome is not the objective of our study.

We described the following variables in this patient population: age, sex, presenting features, radiological findings, the management strategy and postoperative complications. The demographic and clinical characteristics of patients and morphometric characteristics of paediatric posterior fossa tumour were reported using medians or means for continuous variables and frequencies for categorical variables.

RESULTS

Out of 80 children (aged 16 and less) with brain tumours, twenty-four patients (30%) had PF tumours. Twenty-one patients underwent surgery. In one patient with suspected medulloblastoma, surgery was declined by the guardian. In two cases of intrinsic brainstem gliomas, no surgery was offered. Hence this patient does not have histological confirmation of the tumour type.

Demographics: Patient's age ranged from 2-16 years of years with mean of 9 years. Majority of patients (15, 62.5%) were below 10 years of age. Fourteen (58.3%) patients were male.

Symptoms: The most frequent symptoms were headache (20, 83.3%) and vomiting (18, 75%) followed by gait ataxia (9, 37.5%). Two children with brainstem glioma presented with dysphagia. None of our patient presented in an unconscious state.

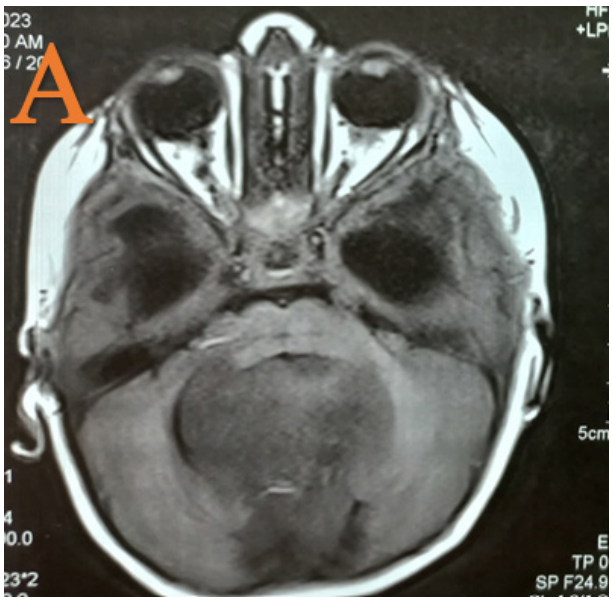
Diagnosis: Based on preoperative imaging, diagnosis included medulloblastoma (15, 62.5%), ependymoma (3,12.5%), glioma (3,12.5%), hemangioblastoma (2,8.3%) and pilocytic astrocytoma (1,4.1%).

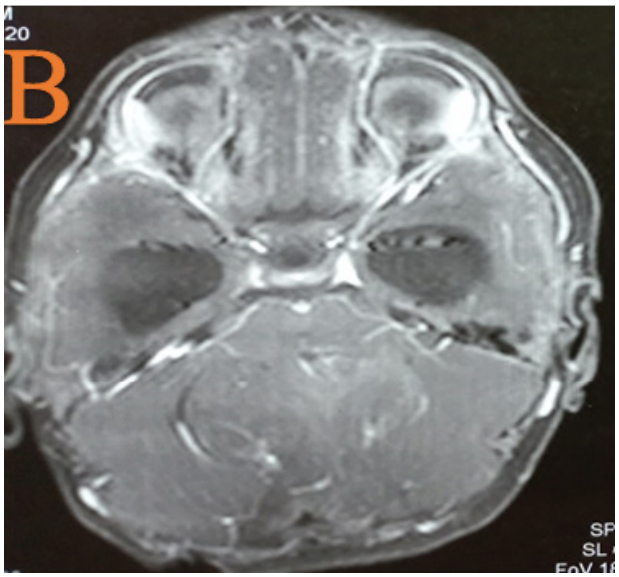
Final histopathological diagnosis was available in 21 patients [Table 1]. The most common diagnosis was medulloblastoma type IV (10, 47.6%). Concordance between preoperative diagnosis and histopathological diagnosis was observed in 15 out of 21 patients (71.42%).

Twenty patients underwent suboccipital craniectomy and tumour excision. All patients with fourth ventricle tumours were approached via transvermian route. Preoperative Ventriculoperitoneal (VP) shunting was done in 18 patients. One patient who had preoperative VP shunting developed bilateral subdural hygroma. One patient who did not have preoperative VP shunting required shunt placement in postoperative period for management of pseudomeningocele, while one patient required a temporary external ventricular drain for hydrocephalus. Two patients (10%) developed pseudomeningocele and one (5%) developed cerebellar mutism in postoperative period. Shunt infection was observed in one patient (5%). One patient with midbrain germinoma underwent endoscopic third ventriculostomy followed by radiotherapy

Table 1: Histopathological diagnosis

Tumor type	Number of patients (%)
Medulloblastoma	10(47.6%)
Pilocytic astrocytoma	5(23.8 %)
Ependymoma	4 (19.04%)
Hemangioblastoma	1 (4.7%)
Germinoma	1 (4.7%)





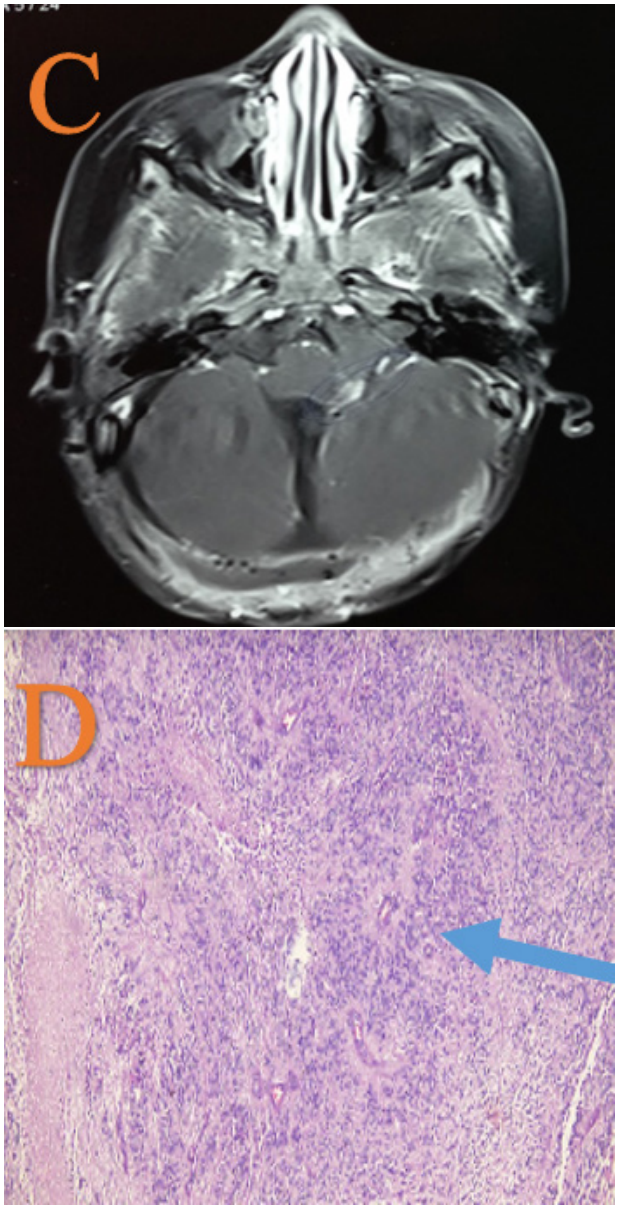


Figure 1: MRI, T1 axial Plain (A) and contrast enhanced (B) images of 13 months male showing heterogeneously contrast enhancing lesion in 4th ventricle. Post-operative MR scan (C) at 6 weeks shows gross total resection and Histopathological findings (D) of perivascular rossettee (blue arrow), suggestive of Ependymoma.

DISCUSSION

Posterior fossa tumours occur in childhood mainly between the age group of 1-10 (peak 4-6 years) with 81% detected in this age group [5, 6, 9]. In our series, 62.5% of the patients were below 10 years of age with 37.5 % incidence between 4-6 years of age. There is a male preponderance for this disease, which was seen in our series too (58.3%) [5,6,10]. This is concurrent with our previous publication that showed 62.5% males in overall paediatric brain tumours [11].

PF tumours present with signs and symptoms of increased intracranial pressure mainly due to the obstruction of cerebrospinal flow [10-12]. Headache is observed in 80-92% followed by vomiting (64%) and gait ataxia (57%) [5,13]. In our series, the presenting complaint was headache in 83.3% cases followed by vomiting in 75% and gait disturbances in 37.5%. Magnetic resonance imaging (MRI) of brain along with screening of spine remains the imaging of choice [14, 15]. All patients in our series underwent MRI in preoperative period.

Medulloblastomas constitute the majority of the tumours in posterior fossa in children [5,13]. Medulloblastomas constituted the highest number of cases in our series followed by pilocytic astrocytomas and ependymomas. The most common tumours in the study by Sharafat et al were medulloblastomas (33.1%), ependymomas (22.7%) and astrocytomas (19%) in descending order of frequency [13].

There is controversy regarding the optimal management of hydrocephalus in children with posterior fossa tumours [16-21]. Some recommend preoperative CSF diversion procedure with definitive surgery done later while others prefer tumour excision and external ventricular drain (EVD) [22]. A study by Picariello showed 69.6 % children being managed for hydrocephalus in preoperative period either by VP shunt or EVD [23]. In our institution, 18 (85%) out of 21 children with hydrocephalus underwent preoperative VP shunting. This was done for better preoperative optimization and avoid neurological deterioration. CSF analysis for cytology was negative for neoplastic cells in all cases. Shunt related complication included shunt infection in one patient and bilateral subdural hygroma in one patient.

The mainstay of treatment for posterior fossa tumour is gross total excision [24-25]. Although restoration of normal anatomical planes and preservation of posterior fossa contents can be provided by craniotomy, most of the centre prefer craniectomy [25]. Twenty patients in our center underwent suboccipital craniectomy and excision of tumour. Gross total resection was achieved in 12 out of 20 (60%). Following surgery, patients with medulloblastoma were sent for adjuvant radiotherapy in other institutions. The postoperative complication included pseudomeningocele (10%) and cerebellar mutism (5%).

Study Limitation: This is a retrospective study with small number of patients restricted to immediate postoperative outcome. A large sample size with long follow up of patients would give more robust analysis and conclusion.

CONCLUSION

Posterior fossa tumours are common tumour occurring in children. Majority of the patients presented with features of raised intracranial pressure and gait ataxia. Fourth ventricle and cerebellar vermis are the most common location. Most common histology types is medulloblastoma followed by pilocytic astrocytoma and ependymoma. Management involves addressing acute hydrocephalus followed by definitive surgery.

Author's Contribution: BT and MRS conceptualize the research, BT did data collection, analysis and prepare result, drafted the manuscript, and all authors reviewed the manuscript and agree to be accountable for all aspects of the research work, and BT is corresponding author. BT, and MRS are abbreviated name authors for Bikas Thapa, and Mohan Raj Sharma.

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Data Availability: The data that support the findings of

this study are available from the corresponding author upon reasonable request.

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