Original Article

Clinical, Histologic, and Therapeutic Pattern of Posterior Fossa Tumors in Children in Cameroon: A Cross-sectional Study

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Abstract

Introduction: Posterior fossa tumors are significant in pediatric neurooncological populations due to their frequency and morbimortality. We convey a 10-year experience managing pediatric posterior fossa tumors at two reference centers in Cameroon. **Materials and Methods:** We conducted a cross-sectional study with data collected retrospectively in the Neurosurgery Department of the Central and General Hospitals of Yaounde from January 2010 to December 2019. Included in the study were all patients aged 0–15 years who underwent surgery for posterior fossa tumors. **Results:** We retained 43 pediatric posterior fossa tumor files, representing 47.89% of pediatric brain tumors. The male/female sex ratio was 0.86, with a mean age of 6.93 ± 4.345 years. The predominant clinical presentations were headaches + vomiting (97.7%), visual impairment (67.4%), and altered consciousness (14%). The vermis was the most frequent site affected (46.5%). Hydrocephalus was found to be associated with the tumor in 30 patients (69.8%). Tumor excision was total in 90.7% of cases, with cerebrospinal fluid diversion performed within the surgery in 83% of cases. Pilocytic astrocytomas represented 37.2%, medulloblastomas 30.25%, ependymomas 6.9%, and brainstem gliomas 4.6%. The postoperatory mortality rate was 20.9%. The survival rate at 5 years for medulloblastomas was 43%. **Conclusion:** Pediatric posterior fossa tumors are frequent. Pilocytic astrocytomas are the most common histologic type, followed by medulloblastomas. Radical surgery is the leading standard of care. Adjuvant treatment remains limited.

Keywords: Cameroon, clinical presentation, histology, posterior fossa tumors, surgery

Introduction

Posterior cerebral fossa (PCF) tumors in children portray a group of neoplasia developed from the posterior skull base structures. These tumors are paramount amongst pediatric tumors due to their frequency, which varies between 50% and 70% of all pediatric brain tumors.[1-4] They represent the second cause of neoplasia in children after leukemia. PCF tumors generally present with signs of intracranial hypertension associated with gait disturbances, cerebellar syndrome, and cranial nerve palsy. Studies show that 20%–90% of cases are associated with hydrocephalus.[5-7] The severity of these tumors is related to not only the location, but also type and histological grade, with; WHO Grade 1 pilocytic astrocytoma having a 10-year survival rate of 95%–100%, WHO Grade 4 medulloblastoma a 5-year survival rate of 50%–75%, WHO Grades 2–3 ependymoma a 5-year survival rate of 40%–45%, and WHO Grades 2 and 3 brainstem gliomas, a 5-year survival rate of <50%.[8] A diagnosis of this disease is increasingly frequent in Cameroon, with the advent of new medical imaging techniques, such as computed tomography (CT), magnetic resonance imaging (MRI), and the creation of neurosurgery centers. However, the management revolves mainly around surgical excision or palliative therapy, adjuvant treatment (radiotherapy) being scarce in our setting. Hence, we set out to examine the clinical, anatomopathological characteristics, treatment modalities, and survival of pediatric PCF tumors at two reference centers in Yaoundé.

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Materials and Methods

Type of study

This was descriptive cross-sectional study.

Sites of study

Our study took place in the Neurosurgery Departments of the Central and General Hospitals of Yaounde which are reference hospitals with a qualified personnel and an adequate technical platform. These hospitals receive the majority of the neurosurgery cases in the center region and numerous referred neurosurgery cases from the other regions of Cameroon.

Duration and period of study

The period of data collection was from January 2010 to December 2019, giving a retrospective interval of 10 years.

Population of study

Included in the study were patients aged 0–15 years who had surgery for posterior fossa tumor with confirmed histologic diagnosis. We collected files of 57 cases of posterior fossa tumors amongst which 14 were excluded (with ages over 15 years) and 43 retained. We analyzed the following: age, sex, circumstances of discovery, clinical presentation, radiologic aspects, surgical treatment, and the evolution (postoperative mortality and survival rate of medulloblastoma).

Procedure of data collection and analysis

- Recruitment of patient medical booklets having as diagnosis intracranial expansive processes
- Responding to the inclusion criteria
- The filling of the pre-established technical sheet
- The data collected entered and analyzed using the SPSS: Statistical Package for the Social Sciences by IBM and Microsoft Excel 2010.

Ethical considerations and administrative procedures

Our study was carried out in strict compliance with the principles of ethics in medical research, particularly:

- The principle of the interest and benefit of research
- Confidentiality.

The anonymity of the subjects was respected. The results obtained in our study will be used exclusively for scientific purposes.

Results

Ninety pediatric brain tumor patients were managed within this period. Posterior fossa tumors represented 47.89% of all pediatric brain tumors. There were 23 females and 20 males giving a male/female sex ratio of 0.86. The mean age was 6.93 ± 4.345 years and the median was 6 years, ranging from 2 to 15 years. The most represented age group was 5–10 years (44.18%) [Figure 1]. Seventy-two percent (31 patients) were schooling. The mean time interval between the onset of symptoms and neurosurgical consultation was 2 months. The leading symptoms were headache + vomiting (97.7%), followed by visual impairment (67.4%) and altered consciousness (14%) [Table 1].

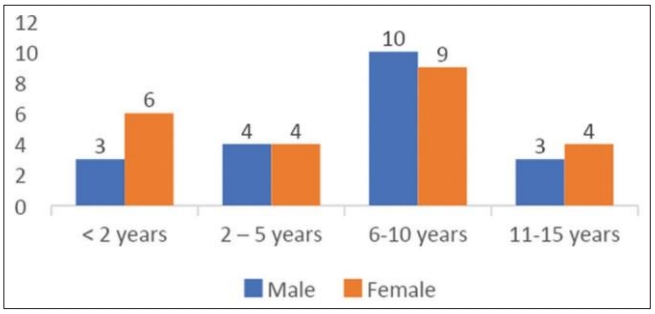


Figure 1: Age distribution of patients

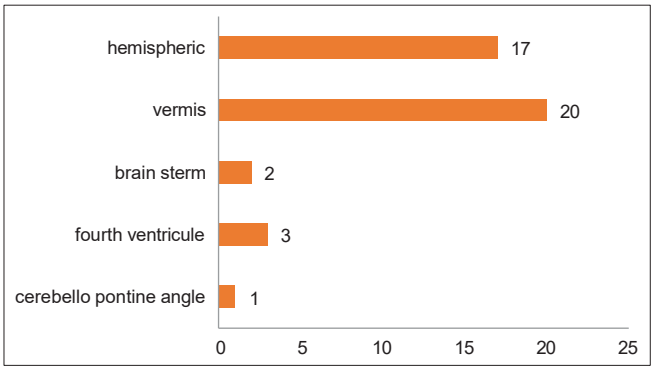


Figure 2: Distribution of tumors by site

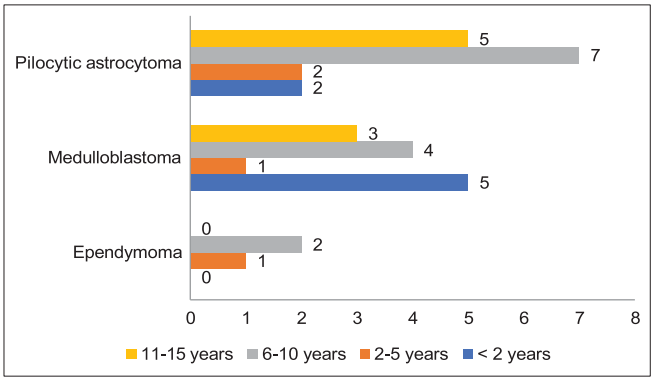


Figure 3: Distribution of main histologic types by age

A cerebellar syndrome was found in 76.7% and cranial nerve palsies in 39.5% of cases.

Out of the total of 43 patients, 38 of them (88.4%) had a brain CT scan, while only 5 patients (11.6%) underwent MRI. Tumors were primarily located in the vermis in 20 patients (46.5%), followed by the cerebellar hemispheres in 17 patients (39.53%). In addition, tumors were found in the 4th ventricle (V4) in 3 patients (6.98%), the brainstem in 2 patients (4.65%), and the cerebellopontine angle in 1 patient (2.32%) [Figure 2]. Thirty cases (69.8%) of tumors were associated with hydrocephalus; the vermis location was responsible in 53.3% of cases, followed by hemispheric tumors and V4 tumors, in 30% and 10%, respectively.

The interval from diagnosis to surgery was 6 days, ranging from 2 to 15 days. Thirty-six patients (83%) underwent tumor

Table 1: Clinical presentation

Clinical presentation	*n* (%)
Headache + vomiting	42 (97.67)
Visual impairment	29 (67.4)
Abnormal gait	13 (30.2)
Disorders of balance	15 (34.8)
Convulsions	2 (6.9)
Neck stiffness	3 (2.3)
Altered consciousness	6 (14)
Macrocrania	6 (13.95)

Table 2: Frequency of the histologic types

Histologic type	*n* (%)
Astrocytoma	16 (37.2)
Medulloblastoma	13 (30.2)
Ependymoma	3 (6.9)
Glioma	2 (4.6)
Hemangioblastoma	1 (2.3)
Meningioma	1 (2.3)
Subarachnoid cyst	4 (9.3)
PNET	2 (4.6)
Tuberculoma	1 (2.3)

PNET: Primitive neuroectodermic tumor

excision and cerebrospinal fluid (CSF) diversion per surgery (a one-stage procedure). Seven patients (16.3%) presented signs of high intracranial pressure due to hydrocephalus. We performed urgent CSF diversion before tumor excision within the same month (two-stage procedure). CSF diversion consisted of ventriculoperitoneal shunting (VPS) for patients with a 2-stage procedure. For those with a one-stage approach, external ventricular drainage (EVD) was done in 16 patients (37,20%) and VPS in 7 patients (16.27%). Thirteen patients (30.33%) did not undergo CSF diversion. Gross total resection was achieved in 39 patients (90.7%), subtotal in 2 cases, and partial in 2 patients. The postoperative mortality rate was 20.9% (9 cases), of which 9.3% (4 cases) underwent CSF diversion, and 11.6% (5 cases) did not undergo CSF diversion.

The most common histological findings were pilocytic astrocytomas (Grade I) in 37.2% (16 patients), medulloblastomas (Grade IV) in 30.25% (13 patients), and ependymomas (Grade II) in 6.9% (3 patients) [Table 2]. The age range of children diagnosed with astrocytomas was 1–15 years, with an average age of 10.06 years, with an equal male/female ratio. Regarding medulloblastomas, the mean age was 6.15 years, ranging from 1 to 13 years, with a male/female sex ratio of 1.16. We observed 2 age peaks for medulloblastomas: Before 2 and between 6 and 10 years [Figure 3]. Ependymoma cases had a mean age of 4.65 years, ranging from 2 to 6 years, with the male/female sex ratio of 1/2.

Seven patients (16.27%) of the 13 with medulloblastoma could get adjuvant treatment. Radiotherapy was done for all aged 6–13 years, and one patient received complementary

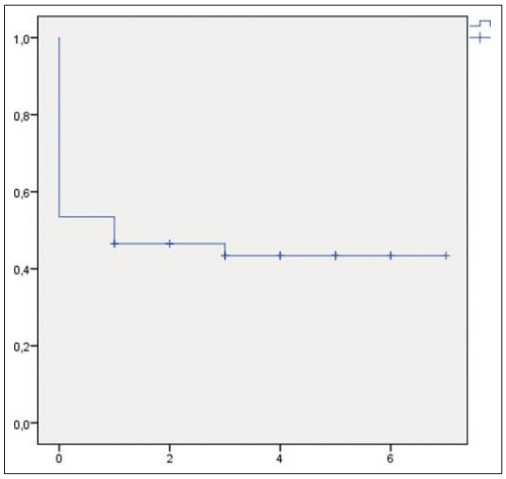


Figure 4: Survival curve for medulloblastomas

chemotherapy. The 5-year survival rate for medulloblastoma was evaluated using the Kaplan curve. It showed a survival rate of 43% at 5 years [Figure 4].

Discussion

Posterior fossa tumors are frequent in children. The subtentorial localization of tumors in children varies from 53% between 6 and 12 months, 42% between 2 and 16 years, and 74% between 1 and 2 years.[8] In our series, the frequency of posterior fossa tumors with respect to other pediatric tumors was 47.89%. Twenty-four per cent of patients were older than 2 years, conforming with the literature.[8] There was a slight female predominance with frequency (53.5%). Wanyoike[9] in Kenya also found a female predominance with frequency of 64.86%. Mbaye *et al*.[10] found a male predominance (57.3%) in Dakar. Several other authors in literature have equally reported male predominance.[1,4,11,12] The sex ratio in literature varies across different studies. The mean global age was 6.93 ± 4.34 years with range from 2 to 15 years. With respect to the histologic type, the mean age was 10.06 years for those with astrocytomas, 6.15 years for those with medulloblastomas and 4.65 years for those with ependymomas. These results are not different from those reported in literature. The mean age is between 10 and 16 years for astrocytomas, within the first decade for medulloblastomas and around 4 years for ependymomas.[8] Sid'Elhadj *et al*.[6] in their series in Nouakchott, found a mean age of 7.6 years with a majority (43%) of medulloblastoma cases.

Majority of the patients were received at consultation after 2 months from the onset of symptoms. This time delay is quite long. In effect, the specialized services are located in the two big cities of Yaounde and Douala. The diagnosis of posterior fossa tumors is not rapidly suspected in our milieu. At the time of diagnosis, 67.4% presented with visual impairment (severe impairment of visual acuity) and 14% with altered consciousness. Mbaye *et al*.[10] found a longer mean time interval in Dakar before consultation of 6.3 months.

They attempted to explain this delay with certain factors such as diagnostic errors, low socioeconomic statuses and, sociocultural beliefs. These factors were equally present in our setting. The signs that reveal a posterior fossa tumor are frequently signs of raised intracranial pressure, generally secondary to the tumoral volume and/or hydrocephalus resulting from compression or obstruction of the either the V4 or the foramina of Lushka.[13,14] In our series, signs of raised intracranial pressure (headache + vomiting) represented 97.7%. We observed a macrocrania in 6.94%.

The tumor was localized in the vermis in 46.5% of cases followed by the cerebellar hemispheres in 39.23% [Figure 2]. The vermis is the frequent site for medulloblastomas which are often found at the roof of the V4. Medulloblastomas are the main differential diagnosis to ependymomas which on their part are often found at the floor of the V4. Pilocytic astrocytoma is frequently hemispheric. In our study, all the cases of ependymomas were located at the floor of the 4th ventricule, vermis was the site of all the cases of medulloblastomas, 68.75% of cases of astrocytomas were at the hemispheric position, and 31.25% were located on the vermis. Hydrocephalus was associated to the tumors in 30 patients (69.8%). These results are comparable to those in literature concerning the presence of hydrocephalus.[4-7,15,16] Hydrocephalus associated with posterior fossa tumors is generally present in 20%–90% of cases.

The treatment of posterior fossa tumors is surgical. The removal of the tumor has to be complete with pilocytic astrocytomas and maximal in medulloblastomas and ependymomas. Tumor excision is difficult to perform in the presence of significant hydrocephalus. Therefore, a prior CSF diversion is classically performed alongside the suboccipital craniotomy.[13,17,18] Endoscopic ventriculo-cisternostomy (EVC) is the preferred method. In our context, the materials necessary to perform EVC are not always available. EVD was carried out during the same surgery alongside tumor excision in the majority of patients. However, in recent years, we directly carried out VPS to prevent infections. Half of the cases of hydrocephalus were associated with vermis tumors which is the frequent location of medulloblastomas in children <2 years. In our series, we noticed a peak incidence of medulloblastomas in children under 2 years. Within this age group in our setting, Djientcheu Vde *et al*.[19] explained why the place of VPS is more important compare to EVC in the management of hydrocephalus in infants. Le Fournier *et al*.[5] in France, found no difference between VPS, EVC, and EVD in the evolution of children operated upon for posterior fossa tumors with hydrocephalus. According to Mbaye *et al*.[10] in Dakar, on a series of operated intracranial tumors regardless of the histologic diagnosis, the mortality rate was 19.5%. In our series which focused mainly of posterior fossa tumors, the mortality rate was 20.9% (9 patients). Among the deceased, no explanation was retrieved even for 4 patients (9.6%) with CSF diversion and for 5 patients (11.6%) with no CSF diversion.

Concerning the histological types, pilocytic astrocytoma (Grade I) was the most frequent tumor (37.2%) followed by medulloblastoma (30.2%) and ependymoma (6.9%). These results are similar to those in literature.[8,10,11,20,21] Piloctytic astrocytoma (frequent between 10 and 20 years) represents 27%–40% of posterior fossa tumors. Medulloblastomas whose frequency by age varies according to the histological subtype (classical, desmoplastic, nodular, and anaplastic) represent 20%–30% of posterior fossa tumors. Ependymomas represent only about 9% of posterior fossa tumors.

Concerning radiotherapy, the management of medulloblastoma (Grade 4 malignant tumor) is radical surgery followed by whole-brain radiation therapy and a booster dose on the tumor bed.[22] Access to radiotherapy is limited in our context, especially children. The 5-year survival rate for medulloblastoma was evaluated using the Kaplan curve. This highlights the survival of operated patients for medulloblastoma (who is a World Health Organization (WHO) Grade 4 malignant tumor) in our limited conditions. It showed a low survival rate of 43% at 5 years and illustrated our difficulties concerning specific management of medulloblastoma.

Conclusion

Pediatric posterior fossa tumors represent almost half of the pediatric brain tumors in our setting. The waiting time before specialized management is still very long. The clinical presentation is dominated by signs of raised intracranial pressure and hydrocephalus is present in about two-thirds of cases. The histological profile is similar to that in literature with pilocytic astrocytomas and medulloblastomas occupying the top positions. Radical surgery is the basis for treatment and adjuvant therapy is still very limited in our setting.

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Conflicts of interest

There are no conflicts of interest.

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