OPERATED PEDIATRIC POSTERIOR FOSSA TUMOURS IN BENGHAZI MEDICAL CENTER FROM 2015 TO 2016

Khalid Ali and Esam Alnajar

BACKGROUND:
Brain tumors in children have totally different clinical appearances, anatomical distribution, histopathological diagnosis, and clinical prognosis compared with adults.

Materials and Methods:
A retrospective study was conducted in the Department of Neurosurgery, Benghazi Medical Center which included all pediatric cases of posterior fossa tumor that presented or were referred to the Center and were considered for surgery from a period between the 1st Jan. 2015 to 31st Dec. 2016.

RESULTS:
Four children, two males and two females, with the mean age of 8.75 years (4–12) were registered. The presenting symptoms were cerebellar (3), headache (2), and vomiting (2). The four cases underwent ventriculoperitoneal shunt before tumor resection surgery. Gross total resection was performed in all cases. The thirty days mortality was 25. The most common microscopic anatomy diagnosis was primitive neuroectodermal neoplasm (PNET) (4). Adjuvant therapy was planned by the oncologist according to histopathology results.

CONCLUSION:
Further studies should be conducted in this field for the greater understanding of the clinical presentation and management of these tumors, and judicious use of modern neurosurgical techniques should lead to more efficacious results and outcome, followed by adjuvant therapy if necessary.

Keywords: tumour, children, surgery, posterior fossa

INTRODUCTION
Brain tumors are among the foremost devastating kinds of human health problems, particularly when occurring within the posterior fossa, representing 20% of all pediatric tumors. Brain-stem compression, herniation, and death are all risks in tumors that occur in this vital location. Tumors within the posterior fossa are considered serious brain lesions, primarily because of the restricted space inside the posterior fossa and also the potential involvement of important brainstem nuclei. The clinical presentation depends on the location of the neoplasm, biological behavior and aggressiveness of the neoplasm, and also the rate of growth. At the time of presentation, the patients are usually very unwell from severe headache, and frequent emesis because of associated hydrocephalus. The symptoms may be explained by focal compression of the cerebellum or brain stem centers and raised intracranial pressure. [1, 2, 3, 4]

No specific causes for posterior fossa tumors are known to exist. However, genetic factors, such as dysfunction of tumor suppressor genes (p53 gene) and activation of some oncogenes, could play a task in their development. [1] Environmental factors comparable to irradiation and toxins may also play a role. Tumor types that most often occur in the posterior fossa include medulloblastoma, ependymoma, cerebellar astrocytoma and brainstem glioma. Hippocrates, who probably performed it for headache, epilepsy, fractures, and visual defect, was the first to perform trephination. Cushing was the first to report an large series of posterior fossa tumors. He published information regarding sixty one patients with cerebellar medulloblastoma with largely fatal outcomes. [5] Advances in surgical operation for tumors primarily were because of discovery of anaesthesia, asepsis, neurologic localization, and also the ability to attain haemostasis.

MATERIALS AND METHODS
This retrospective study was conducted in the Department of Neurosurgery, Benghazi Medical Center. It included all pediatric cases of posterior fossa tumor that presented or had been referred to the Center and were considered for surgery from a period between the 1st January 2015 to the...

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31st December 2016. The demographic data including age, gender and tumor characteristics along with the location and pathological diagnosis were recorded. The surgical outcomes were assessed according to the pathological diagnosis.

Results
Our series consisted of four patients (two males, and two females). Cerebellar symptoms were the most common cause of presentation (75%) followed by headache (50%) and vomiting (50%), see charts, (chart 1, and chart 2). The youngest patient was four years old, two other patients were nine years old, and the fourth patient was twelve years old with a mean age of 8.75 years. The four patients underwent ventriculo-peritoneal shunting as an emergency procedure, during the preoperative preparation.

The intraoperative prone position was preferred and there were no perioperative complications with successful gross total resection achieved in all patients. The thirty days mortality was one patient (25%), most probably because of poor general condition before the tumour resection operation.

The four cases were diagnosed as having medulloblastoma (PNET) by histopathology examination and sent for further oncological management regarding adjuvant therapy.

Discussion
The posterior fossa is the commonest site of primary intracranial tumours in children. The most common neoplasms are pilocytic astrocytoma, medulloblastoma, ependymoma and brain stem glioma. In children over one year old, over two thirds of intracranial tumours arise from the cerebellum or brain stem, compared with adults. Survival rates of a number of these lesions have improved markedly over the last twenty years, thanks to advances in surgical techniques, chemotherapy, and radiation therapy. These tumours remain the main focus of intense analysis aimed not simply at prolonging survival, but, minimizing the impact of treatment on growth, psychological development, and long-term quality of life.

Gender Distribution
According to most published data, the gender ratio of brain tumors in infants is equal or slightly male predominated. The male to female quantitative ratio was 1.1 and 1.4 in studies by Chung et al.[9] and Mapstone,[8] respectively. Information collected from eleven series, with a total number of 1289 infants, revealed a slight male predominance of 53%. [6] Correspondingly, there was no distinction in gender dominance within the current study, however, unlike other studies Haddad et al. found a feminine predominance during this age group,[3] and Rickert et al. reported no gender preference.[4]

Clinical Manifestations:
The clinical presentation depends on the location, biological behavior and aggressiveness of the tumor. At the time of presentation, the patient may be very sick suffering severe headache or frequent emesis because of associated hydrocephalus with increasing the intracranial pressure.

Figure 1 MRI brain FLAIR axial image which reveals a large posterior fossa mass with cystic component, and acute obstructive secondary hydrocephalus.

Figure 2 MRI brain axial image with IV contrast, revealing an enhancing large posterior fossa mass with a cystic component wall.
Other symptoms may be caused by focal compression of the cerebellum or brain stem centers. Focal compression of the cerebellum includes characteristic eye findings and a vermis syndrome. Truncal ataxia may be a common finding in midline tumors, for example; medulloblastoma, ependymomas, and vermian astrocytomas. This is manifested by a tendency to fall frequently, and a wide based gait. A hemispheric syndrome manifests as limb ataxia, nystagmus, and dysmetria. Brainstem compression causes cranial nerve dysfunction that normally involves the nuclei or tracts of the third, fourth, or sixth cranial nerves, leading to ocular palsies and diplopia, and long tract signs such as hemiparesis. See Figures, (Figures 1,2 and 3 show large a posterior fossa mass compressing the brainstem and cerebellum with acute secondary obstructive hydrocephalus).

**MANAGEMENT AND SURGICAL APPROACH CONCERNS:**

**PREOPERATIVE MANAGEMENT:**
Medications, such as diuretics and corticosteroids, are administered before surgery to scale back the impact of edema on the surrounding structures. Some patients require and may tolerate an emergency operation without preoperative stabilization, particularly if they present with acute symptoms of brainstem involvement or herniation. The foremost operative approaches to the posterior fossa tumors are midline, paramedian, or retromastoid.

Preoperative clearance, together with cardiovascular, respiratory, renal, metabolic, and hematological assessments should be done. Management of the secondary effects of tumour and intracranial hypertension, most often by diversion of CSF either internally or externally is required. All of our patients in the current series had pre-operative ventriculo-peritoneal shunting. The International Society of Pediatric Neurosurgery (ISPNN) survey conducted by Di Rocco et al. revealed a 44% CSF diversion requirement. [11] Twenty percent of infants in Bognár’s study needed shunt implantation, as well. [11] Overall, it can be supposed that with appropriate management of external ventriculostomy, the rate of shunt requirement might decrease. However, this discussion is still open. Progressive neurologic deterioration necessitates emergency operation for tumor removal.

**INTRAOPERATIVE MANAGEMENT:**

- The prone position of the patient is a comfortable position for the surgeon. The head is fixed in a three points head fixation if the patient is older than two years. [13] The second preferred position is the sitting position, but it carries the risk of air embolism.
- A safety burr hole is placed within the occipital area. This may be employed in the case of acute hydrocephalus requiring ventricular drainage.
- A midline incision is made which extends from the inion to the upper cervical vertebra.
- The paracervical and suboccipital muscles are separated by diathermy.
- Cranietomy is performed consistent with the location and size of the tumour.

- The foramen magnum is opened, and the posterior arch of the atlas vertebra (Cl) is removed, particularly in tumors extending to, or beyond, the cranio-cervical junction.
- The dura is opened in a Y-shaped fashion, with the bottom upward.
- Tumors are removed using light suction, an ultrasonic surgical aspirator, or carbon dioxide optical laser. The last technique is employed solely sometimes. Mohanty et al. published a series of thirty one infants in 2013, 62% of them underwent a gross total or near total excision. [10] Accordingly, gross total resection was possible in all of our four patients. The increased rate of resection can

**Figure 3 MRI brain sagittal image with IV contrast, revealing an enhancing large posterior fossa mass with a cystic component.**

**Chart 1 Gender Distribution**

<table>
<thead>
<tr>
<th>Gender distribution</th>
<th>Male</th>
<th>Female</th>
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<tr>
<td></td>
<td>50%</td>
<td>50%</td>
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The extent of tumour removal must be weighed against the higher risk of complications, particularly with tumors adherent to the brainstem.

The dura is closed in a watertight fashion. Dural grafting may also be required for complete closure.

Surgical Mortality:
The outcome of surgery depends on the size of the tumor, histopathological diagnosis, and more importantly the infant's preoperative general condition. Fortunately, we had no perioperative mortalities. The operative mortality within the first thirty days of surgery was 25% in the current study, compared with 33% in Jooma's series,[7] and 13% in Boghár's series.[1]

Histopathological Diagnosis:
According to a worldwide study by Di Rocco, the ten most common types of brain tumors in infancy are, in descending order, astrocytoma, medulloblastoma, ependymoma, choroid plexus papilloma, primitive neuroectodermal tumor (PNET), teratoma, sarcoma, meningioma, ganglioglioma, and neuroblastoma.[11] In a recent study by Mohanty et al., choroid plexus tumors were the most common pathologies, followed by medulloblastoma.[10] Overall, astrocytoma and PNET account for the majority of pathological diagnoses in most other series. The most common tumor in a report by Mapstone and Warf was PNET (41%) followed by astrocytoma (36%).[8] Furthermore, in Rickerts' series of twenty two infants, PNET was the most frequent pathology.[4] In Haddad's study, however, astrocytoma slightly outnumbered PNET (seven versus six out of twenty two cases).[3] In a literature review by Larouche, astrocytic tumors represented the largest group (30.5%) followed by medulloblastoma (12.2%).[6] Oi et al. stressed ethical differences in the types of brain tumors in a report of five Far Eastern countries that had a higher incidence of medulloblastoma compared with the reports of Western countries.[12] In our series, all patients had medulloblastoma. Diverse ranges of tumor types in different series may be influenced by the subtypes of classifications used by different authors.

Postoperative Management:
Increasing ataxia, appearance of lower cranial nerve pathology, apnea, or other respiratory abnormalities are commonly expected as post-operative sequels, and should receive appropriate management.

Adjuvant therapy is advised by the oncologist according to the type of tumour.

Shortcomings and the need for further studies:
There are several limitations in the current study. The sample size was relatively small, and longitudinal follow-up was limited. The study design was retrospective and that imposed an inherent limitation on the data collection. The results of this study need to be replicated in a larger sample, preferably of prospective design, and with a longer duration of follow up.

Conclusion:
The diagnosis of brain tumors in the general pediatric population remains challenging. Most symptomatic children require several visits to a physician before the correct diagnosis is made. These patients are often misdiagnosed with gastrointestinal disorders. Greater understanding of the clinical presentation of these tumors and judicious use of modern neuroimaging techniques should lead to more efficacious therapies.

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None

Conflicts of interest
There are no conflicts of interest.

References


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