

Posterior Fossa Tumor in Children

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Seyed Mahmoud TABATABAEI MD¹,
Afsoun SEDDIGHI MD²,
Amir Saied SEDDIGHI MD²

1. Professor of Neurosurgery,
Mehrad Hospital, Functional
Neurosurgery Research Center of
ShohadaTajrish Hospital, Shahid
Beheshti University of Medical
Sciences, Tehran, Iran
2. Assistant Professor of
Neurosurgery, ShohadaTajrish
Hospital, Functional Neurosurgery
Research Center of Shohada Tajrish
Hospital, Shahid Beheshti University
of Medical Sciences, Tehran, Iran

Corresponding Author:
Seddighi A. MD
Shohada Tajrish Hospital, Functional
Neurosurgery Research Center of
ShohadaTajrishHospital Tehran,
Iran.
Tel: +982188265188
Cell Phone: +98 9121852917
Email: afsounseddighi@gmail.com

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Abstract

Objective

Primary brain tumors are the most common solid neoplasms of childhood, representing 20% of all pediatric tumors. The best current estimates place the incidence between 2.76 and 4.28/100,000 children per year. Compared with brain tumors in adults, a much higher percentage of pediatric brain tumors arise in the posterior fossa. Infratentorial tumors comprise as many as two thirds of all pediatric brain tumors in some large series. Tumor types that most often occur in the posterior fossa include medulloblastoma, ependymoma, cerebellar astrocytoma and brainstem glioma.

Materials & Methods

All pediatric cases of posterior fossa tumor that were considered for surgery from 1981 to 2011 were selected and 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 pathological diagnosis.

Results

Our series consisted of 84 patients (52 males, 32 females). Cerebellar symptoms were the most common cause of presentation (80.9%) followed by headache (73.8%) and vomiting (38.1%). The most common histology was medulloblastoma (42.8%) followed by cerebellar astrocytoma (28.6%), ependymoma (14.3%), brainstem glioma (7.2%) and miscellaneous pathologies (e.g., dermoid, and tuberculoma) (7.2%).

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 for gastrointestinal disorders. Greater understanding of the clinical presentation of these tumors and judicious use of modern neuroimaging techniques should lead to more efficacious therapies.

Keywords: Posterior, Fossa; Tumor; Surgery

Introduction

Posterior fossa tumors constitute about two thirds of pediatric tumors in large scale studies (1). This pattern is seen especially in children younger than 10 years of age. This is in contrast with adult series. Medulloblastoma, ependymoma, cerebellar astrocytoma and brainstem glioma are the most common posterior fossa tumors

in children. Hemangioblastoma, meningioma, sarcoma, chordoma and metastasis—mainly neuroblastoma, Wilms' tumor and embryonal rhabdomyosarcoma—are other scarce posterior fossa tumor types (3).

Due to the raised intracranial pressure caused by hydrocephalus or tumor mass, children with posterior fossa tumors generally present with symptoms such as headache, nausea, vomiting, restlessness and poor feeding (4). Infants may present with macrocephaly and bulging fontanel. Cerebellar symptoms, particularly ataxia (difficulty with balance and coordination), are a frequent complaint of patients with posterior fossa tumors that invade or exert mass effect on the cerebellum or on the cerebellar peduncles. As a result of recumbency and rise of PaCO₂, ICP increases leading to worsening of symptoms in the morning. In cases with delayed diagnosis, cerebellar symptoms are more commonly detected. On physical examination, cerebellar manifestations and nystagmus are common in posterior fossa tumor patients. In patients with hydrocephalus, papilledema may be observed. Continuous evaluation of the altered level of alertness is necessary (5, 6).

In this paper we intend to report the 30-year experience and long-term assessment of surgical outcome on a large series of patients. An important feature of our study is that patient selection and all the surgical procedures and follow-up studies were performed by a single surgical team.

Materials & Methods

From March 1981 to December 2011, all pediatric patients who were referred to the neurosurgical clinic and were operated for posterior fossa tumor were evaluated. Inclusion criteria were age equal or less than 14 years and evidence of posterior fossa tumor in the neuroimaging studies. The exclusion criteria were age more than 14 years, prior surgery in another center and the parents' unwillingness to perform surgery. The recorded data included age and sex distribution, duration and nature of symptoms, whether a pre or postoperative ventriculoperitoneal (V-P) shunt was inserted, tumor type on histological examination and surgical outcome. The statistical analysis was performed by Wilcoxon & Fisher methods and a p-value less than 0.05 was considered statistically significant.

Results

The study included 84 patients (52 males, 32 females; M/F ratio, 1.62; Mean age, 8.6 years; range. 1-14 years; standard deviation, 3.4 years).

In our series, 50%(42 cases) of pediatric posterior fossa tumors occurred in children less than five years, 30% (25 cases) in the 6-10 year age group and 20% (17 cases) occurred in older children (11-14 years).

The duration of symptoms from the beginning to the time of surgery ranged from 3 days to 4 years. The average value was 16 months and the standard deviation was 32 months.

Cerebellar symptoms were the most common cause of presentation (80.9%) followed by headaches (73.8%) and vomiting (38.1%). Visual problems were found in 7.14% of our patients probably due to late presentation. Neck stiffness was described in 2.3% of our patients. The most commonly involved cranial nerve was abducens nerve palsy which occurred mainly in cases with hydrocephalus (Table 1).

In our operated cases, the most common histology was medulloblastoma (42.8%) followed by cerebellar astrocytoma (28.6%), ependymoma (14.3%), brainstem glioma (7.2%) and miscellaneous pathologies (such as dermoid and tuberculoma) (7.2%) (Table 2).

In 25 patients, (30%) ventriculoperitoneal (VP) shunts were inserted before tumor resection due to severe hydrocephalus and poor general condition of the patients. In four patients (4.7%), a VP shunt was inserted post-operatively while the other patients (66.3%) did well without a shunt procedure.

In 70%, gross total resection of the tumor was performed. In 24%, the tumor was resected subtotally and in 6%, we performed partial tumor resection due to severe adhesion to the brain stem and involvement of the cranial nerves. The mean follow-up period was 78 months (range, 6 months-18 years).

The most common complication was superficial wound discharge that occurred in nine cases (10.7%). Twelve patients (14.3%) developed pseudomeningocele which responded to either medical treatment or VP shunt insertion. Cerebellar mutism or posterior fossa syndrome occurred in 14 patients (16.6 %) after a posterior fossa tumor resection. Early postoperative mortality occurred in two cases (2.4 %). One (1.2%) was due to

postoperative edema and the other (1.2 %) occurred after protracted meningitis.

Discussion

Tumors of the central nervous system are the second most common childhood tumors (20%) after leukemia (37%) and the most common solid pediatric tumors comprising 40-50% of all tumors (7,8).

Posterior fossa (infratentorial) tumors comprise 54% to 70% of childhood brain tumors compared to 15%-20% in the adult population (9). Worldwide, the most common pediatric posterior fossa tumors are medulloblastoma (20%), astrocytoma (15%), brainstem glioma (15%) and ependymoma (8%) (10).

The estimated incidence of brain tumors in children is 2-3.5 per 1 00,000 (11). The mean age at diagnosis has decreased from about 13 years in 1930 (12), nine years in 1970 (13) to the current figure of 6.5 years in 2010 (9). In our study, the mean age at diagnosis was 8.6 years. The clinical presentation of a child with posterior fossa mass is dependent on the anatomical location, histology type and the presence or absence of hydrocephalus (14). Because posterior fossa is only one tenth of the intracranial volume and contains structures that are responsible for vital functions, tumors in this region are quite challenging for the health care team.

Symptoms result from either compression of vital structures or raised intracranial pressure. Common presenting features include cerebellar symptoms, multiple cranial nerve palsies, headaches, vomiting and blindness due to raised intracranial pressures (15).

Medulloblastoma is the most common primary neuroectodermal tumor and also the most common solid childhood tumor. It comprises approximately 20% of posterior fossa tumors in childhood (16). In our study the tumor constituted 42.7% of the cases. They are extremely malignant tumors and are characterized by the ability to seed along cerebral spinal fluid pathways. The tumors are diffusely blue in color on staining and are often called blue tumors. They are treated surgically and in most cases, complete macroscopic removal is possible. Controversy exists on whether to perform pre or post operative shunting procedure. Adjuvant therapy involves chemotherapy or radiotherapy. Radiotherapy however is not given to children less than 4 years due to

its side effects.

Most cerebellar astrocytomas are low grade pilocytic astrocytomas. In our study, this tumor was seen in 28.6% of our patients. These are mostly cystic with a mural nodule. Solid astrocytomas usually found in the vermis are more aggressive with a poor prognosis depending on the histological grade. However, of all childhood tumors, cerebellar astrocytoma has the best prognosis (15).

Ependymomas arise from the ependymal lining of the ventricles (4th ventricle in the posterior fossa). They are mostly benign but fast growing tumors (13). They recur frequently and tend to seed along cerebral-spinal fluid pathways. They are reddish grey, lobulated tumors and microscopically have a diagnostic feature of rosette formation.

Brainstem gliomas account for 25% of tumors arising in the posterior fossa in children; however, in our series only 7.2% of the patients had this pathology. They present with multiple cranial nerve palsies, signs of long tract involvement and imbalance of movement.

Classically, they present with diplopia due to involvement of the 4th and 6th cranial nerves. Blockage of the aqueduct leads to signs of raised intracranial pressure. Brain stem gliomas are classified into diffuse tumors, which balloon the brainstem and involve all layers. These are malignant with a very poor prognosis. They involve several cranial nerves particularly the fifth, sixth, seventh and the tenth cranial nerves. The other group are the focal tumors that lie within the brainstem and finally exophytic tumors that arise from the brainstem, but extend into the cavity of the fourth ventricle.

Both focal and exophytic brainstem tumors are usually pilocytic and surgery is the treatment of choice, unlike the diffuse type which is malignant and only chemotherapy and radiotherapy can palliate (16).

The discrepancy between tumor types in our series compared to other studies could be explained by the fact that our study was based only on operated cases in our institute not an epidemiologic survey.

Although the correlation between posterior fossa tumors and hydrocephalus has been established, its incidence is not clear. It has been reported that in midline tumors, hydrocephalus is seen in 50% of the cases and when the location is hemispheric, hydrocephalus is seen in 20% of these cases (17). Twenty to thirty percent of brain stem

gliomas showed hydrocephalus in a study conducted by Hojer et al. (18). In other studies, a higher incidence of hydrocephalus has been reported in brainstem tumors (19,20). In cerebellar and fourth ventricular tumors, hydrocephalus has been detected permanently (20). In developing countries, a very high incidence of hydrocephalus has been mentioned in brain tumor patients possibly indicating the diagnostic delay.

Complicated hydrocephalus, presenting with vision defect, papilledema, seizure and a decreased level of consciousness, is the predominant problem that sends the child with brain tumor to the neurosurgeon (22,23).

The drawbacks mentioned for preoperative shunting are namely tumor cell spreading through shunting systems, upward cerebellar herniation (24, 25) and tumor hemorrhage (23). In other studies carried out in developing countries, in which diagnostic delay and severe hydrocephalus are seen, there has been some doubt regarding the precise definition and actual relationship between the above mentioned circumstances and the shunting procedure (22,23).

In some studies, the fact that preoperative shunting may be beneficial has been suggested (21). In our limited study group, 34.5 % of the patients had hydrocephalus. It seems that the rise in ICP needs to be approached exclusively as it introduces a new entity in these tumors. A preoperative shunting procedure should be performed in case of concomitant hydrocephalus in patients with poor general and neurologic conditions. A shunt may decrease ICP and relax the cerebellum and make the tumor resection more feasible. The other benefit of shunt is reduction of the risk of postoperative CSF leak,

pseudomeningocele and meningitis. In our patients there were no differences in the occurrence of contusion, upward cerebellar herniation and fatal intracranial hemorrhage between those who received preoperative shunting and those who did not. We had no evidence of death due to abdominal metastasis.

One of the most important issues in surgery of posterior fossa tumors is posterior fossa syndrome or cerebellar mutism. Cerebellar mutism was first reported in 1979 by Hirsh also known as posterior fossa syndrome (26). After posterior fossa surgery, the patients manifestations include cerebellar dysfunction, apraxia, emotional lability and mutism. The cause may be dentatothalamic tract injury (27). It occurred in 15% of our cases. In our study, factors associated with the development of mutism were an age less than 10 years, midline tumor location, cerebellar vermal incision and large tumor size (>5cm) and medulloblastoma.

In conclusion, primitive neuroectodermal tumor (including medulloblastoma) and gliomas are the most common tumors in our children. The children presenting to us come very late in the course of their illness, when tumors attain very large size and become inoperable at times. These patients are often misdiagnosed for gastrointestinal disorders. Increased attention of pediatricians, teamwork and multidisciplinary approach to the complaints along with advances in neuroimaging techniques are prerequisite for the early diagnosis and the best surgical outcome of posterior fossa tumors in children.

Table 1. Clinical Presentations, Frequency and Their Mean Duration in Patients

Clinical Presentations	Frequency (Percent)	Mean Duration (Month)
Headache	62 (73.8%)	8.5
Vomiting	32 (38.1%)	3.4
Neck Stiffness	2 (2.3%)	1.2
Cerebellar Signs	68 (80.9%)	5.8
Cranial Nerve Palsy	14 (16.6 %)	3.8
Visual Problems	6 (7.14%)	2
Speech Disturbance	4 (4.7%)	2.4

Table 2. Pathological Types of Posterior Fossa Tumor in Patients

Pathological Types	No (Percent)
Medulloblastoma	36 (42.7 %)
Cerebellar Astrocytoma	24(28.6 %)
Ependymoma	12(14.3 %)
Brain Stem Glioma	6 (7.2 %)
Miscellaneous	6(7.2 %)
Total	84 (100%)

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