Cerebrospinal Fluid Diversion Procedures in Children with Posterior Fossa Tumors: Ventriculoperitoneal Shunt Versus Endoscopic Third Ventriculostomy Versus External Ventricular Drainage

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Abstract
Treatment of hydrocephalus in posterior fossa tumors in children is still a matter of controversy. The child with hydrocephalus in a primary brain tumor may be considered to have two distinctly different diseases which complicate one another and contribute to the complex picture of increasing intracranial pressure (ICP): (1) tumor itself, and (2) hydrocephalus [1]. Changes in cerebral blood flow that result from an increase in ICP and ventricular dilatation must also be considered in the pathogenesis of disease.

To manage the complicating effects of hydrocephalus, some neurosurgeons, such as Cushing, used to place separate burr holes routinely in the operations on the posterior fossa tumors to drain the ventricles in the old days [2,3]. With improvements in the preoperative diagnosis of hydrocephalus and shunting procedures, neurosurgeons suggested shunting before tumor extraction [4-8]. Ventricular decompression may result in sudden decrease in ICP, and some cases have been reported to develop epidural hematoma, which has an ominous consequence [9-11].

The percentage of preoperative shunting in hydrocephalic posterior fossa tumors has been quite variable in different studies and depends on the policy of the center where the study is done: 79% [12,13], 10% [14] and 91% [15,16].

The complications of shunting have raised the question of its application and some studies suggest that preoperative shunting makes the subsequent tumor excision even more difficult and hazardous and causes several problems, so they suggest that preoperative shunting for posterior fossa tumors is not frequently indicated [17].

Endoscopic third ventriculostomy is a plausible choice for the emergency control of severe hydrocephalus caused by posterior fossa tumors. It can...
quickly eliminate symptoms and in addition, it eliminates the risk of the cerebrospinal fluid (CSF) infection related to external drainage and avoids complications of shunting like shunt infection and failure [18]. It has also been suggested that in developing countries, where the disease is usually diagnosed in the later stages and the increase in ICP is more severe at the time of diagnosis, preoperative shunting or ETV is advisable [15,18].

Some of the cases that have not undergone preoperative shunting will need it after the operation. By studying these cases it was found that young age, presence of CSF leakage, and septic meningitis were among the factors that increase the risk of post-operative need for shunting procedures [19-21]. It seems that CSF leakage has no relation to the degree of hydrocephalus, CSF diversion or methods of dural closure [7,22]. Here we present the result of our experience in the management of hydrocephalic posterior fossa tumors in children.

Material and Methods

Hospital records of all infants and children with posterior cranial fossa tumor that were treated in Children’s Cancer Hospital Egypt (CCHE), during the period of 2008-2011 were reviewed. All patients have computerized tomography (CT) scan of the brain, brain and whole spine magnetic resonance imaging (MRI) been done. Information regarding demographic data, diagnostic findings, therapeutic procedures and postoperative period of the patients were extracted from their hospital records. Most of the patients were operated by the second author, and the shunting and ETV technique did not change during the study period. Follow-up CT scan was performed after CSF diversion for all patients.

Descriptive analysis was performed to determine the frequency of CSF diversion procedures and postoperative complications in different groups. Comparison between the rates of postoperative complications in different CSF diversion groups were made by exact test using Statistical Package for the Social Science (SPSS) for windows version 15. The outcome variables used in this study were postoperative complications including CSF leak, septic meningitis, persistent hydrocephalus and pseudo-meningocele. We did not compare long-term survival in different shunting groups because it was mostly affected by the tumor type, tumor location and the extent of its resection; any associations made between CSF diversion procedure and long-term outcome of the patients could be confounded by these factors and would make the judgment difficult.

Results

A total number of 170 children with posterior cranial fossa tumors were studied. Ages ranged from 3 months to 18 years at the time of diagnosis. The mean age±SD of patients was 8.9±4.4 and male: Female (M:F) ratio was 1.5. The tumors composed of 87 medulloblastoma (MB), 48 cerebellar astrocytoma (CA), 30 ependymoma, 2 cerebello-pontine angle (CPA) tumors, 2 cerebellar dermoid cysts and 1 cerebellar cavernous hemangioma. For each of these tumors, the M:F ratio and the mean age is given in the (Table 1).

![Table 1: Tumor definition by type.](image)

Among 170 patients, 9 did not have hydrocephalus and 161 had hydrocephalus. The patients without hydrocephalus comprised 6 CA, 1 cerebellar dermoid cyst, 1 CPA tumor and 1 cerebellar cavernous hemangioma. Those with hydrocephalus were composed of 87 MB, 42 CA, 30 ependymoma, 1 CPA tumor and 1 cerebellar dermoid cyst.

The most frequent clinical findings in non-hydrocephalic patients were sixth cranial nerve palsy and diplopia followed by headache, papilledema and ataxia. In the hydrocephalic patients the most frequently encountered clinical findings were headache, nausea, vomiting and papilledema followed by ataxia, diplopia and pyramidal signs.

Of 161 hydrocephalic patients, 81 underwent preoperative shunting, 62 underwent preoperative ETV and 18 did not undergo either of them. All of these preoperative shunting procedures were ventriculoperitoneal (VP) shunts. Of those 18 patients who did not have preoperative shunting or ETV, 13 needed external ventricular drainage (EVD) at tumor operation session and 5 did not. Five cases diagnosed to suffer from hydrocephalus prior to definitive surgery did not undergo preoperative shunting or EVD. Corticosteroids were administered at least a week preoperatively.

General complications are shown in (Table 2). Two of 5 cases who had CA and did not undergo any shunting procedure met major postoperative morbidity (hemorrhage in the operative bedroom, CFS leak, septic meningitis). Another CA patient had a multitude of postoperative complications
and a resultant delayed persistent hydrocephalus in the follow-up period, which needed a VP shunt.

Two patients received prophylactic postoperative shunting prior to radiotherapy to hinder a probable future hydrocephalus.

We also determined the percentage of CSF leak, septic meningitis, persistent hydrocephalus and pseudo-meningocele in the postoperative period in different CSF diversion groups. CSF leakage and septic meningitis in the preoperative shunting group and preoperative ETV group were significantly lower than the other two categories. During the follow-up visits of the 84 patients who had a permanent shunt, 81 preoperative and 3 postoperative, 12 patients had shunt malfunction and 5 cases had shunt infection, all of whom needed a replacing VP shunt. None of the patients with preoperative shunting developed upward cerebellar herniation and 3 patients had some degree of intracranial hemorrhage; none of them were severe enough to necessitate emergent hematoma evacuation.

Preoperative ETV was performed in 62 cases with hydrocephalic posterior fossa tumors: 32 had MB, 17 had CA, and 13 had ependymoma. The ETV resolved the increased ICP before posterior fossa surgery in all cases. One case was complicated with infection. Nineteen cases developed postoperative hydrocephalus and were treated by VP shunt insertion.

EVD was performed in 13 cases: 5 had MB, 5 had CA, and 3 had ependymoma. Regrettably, the incidence of deleterious early complications in the selected group was unproportionately higher than the preoperative shunting group or preoperative ETV group (Table 3).

Table (2): General complications.

<table>
<thead>
<tr>
<th>Tumor type</th>
<th>Preoperative shunting (n=81)</th>
<th>Preoperative ETV (n=62)</th>
<th>External ventricular drainage (n=13)</th>
<th>Neither (n=5)</th>
<th>Exact test</th>
</tr>
</thead>
<tbody>
<tr>
<td>CSF leakage</td>
<td>6</td>
<td>1</td>
<td>2</td>
<td>2</td>
<td>0.005</td>
</tr>
<tr>
<td>CSF infection</td>
<td>5</td>
<td>1</td>
<td>3</td>
<td>1</td>
<td>0.017</td>
</tr>
<tr>
<td>Persistent hydrocephalus</td>
<td>2</td>
<td>19</td>
<td>3</td>
<td>2</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>Pseudo-meningocele</td>
<td>2</td>
<td>19</td>
<td>3</td>
<td>2</td>
<td>&lt;0.001</td>
</tr>
</tbody>
</table>

Table (3): Major early complications in EVD patients.

<table>
<thead>
<tr>
<th>Tumor type</th>
<th>EVD complicated</th>
<th>EVD uncomplicated</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td>MB</td>
<td>3</td>
<td>2</td>
<td>5</td>
</tr>
<tr>
<td>CA</td>
<td>2</td>
<td>3</td>
<td>5</td>
</tr>
<tr>
<td>Ependymoma</td>
<td>2</td>
<td>1</td>
<td>3</td>
</tr>
<tr>
<td>Total</td>
<td>7</td>
<td>6</td>
<td>13</td>
</tr>
</tbody>
</table>

Fig. (1): Sagittal T1-weighted MR images of the brain after gadolinium injection. Left: Preoperative image showing posterior fossa ependymoma and hydrocephalus. Right: Postoperative image after ETV and gross total tumor excision showing resolution of hydrocephalus.

Fig. (2): (a,b) Axial CT brain cuts showing posterior fossa medulloblastoma and hydrocephalus. (c) Axial CT brain cut after insertion of VP shunt showing resolution of hydrocephalus. (d) Sagittal T1-weighted MRI of the brain after gadolinium injection showing that despite gross total tumor excision the patient remains shunt dependent.
Discussion

The association of hydrocephalus with brain tumors is well known but its incidence is not clear. A study showed that CA results in hydrocephalus in 50% in midline location and 20% in hemispheric locations [22]. Studies on cerebellar and fourth ventricular tumors show invariable presence of hydrocephalus [23]. Studies in developing countries also show a very high incidence of hydrocephalus in brain tumor patients that may be due to delay in diagnosis of the disease [15,24].

In most instances, the complicating hydrocephalus is responsible for the symptoms and signs that brings the child with a brain tumor to the neurosurgeon [1,25]. Papilledema and visual impairment, seizure and impaired consciousness are among the problems caused by hydrocephalus. Papilledema that has been described in association with hydrocephalus in many studies [2,11,15] responds very well to preoperative shunting [23]. Some studies have shown the association between seizure and hydrocephalus and its amiable response to CSF drainage [23,24,26].

Some disadvantages have been mentioned for preoperative shunting in the literature [10]. These disadvantages are upward cerebellar herniation [21], tumor hemorrhage [10] and dissemination of tumor cells through shunting systems [25]. However, the exact definition and real association of these phenomena to shunting procedure has been questioned in other studies [1,24] particularly in a developing country, where most of the cases are diagnosed with delay and when a severe hydrocephalus is present.

Endoscopic third ventriculostomy in the management of hydrocephalus secondary to posterior fossa tumors was proposed, for the first time, by Chumas et al., in 1995 [27] and its efficacy was reviewed by Sainte-Rose et al., in 2001 [16]. Sainte-Rose et al., reviewed 67 ETVs performed before tumor removal in patients with severe hydrocephalus. In this series there were no deaths, no permanent morbidity related to the procedure, a 98.5% rate of immediate symptomatic resolution and a 94% rate of shunt-free patients after tumor removal. In CCHE, we found that patients with ependymomas and gliomas, with totally excised tumors, are better candidates for ETV than those with medulloblastomas [28] (Fig. 1).

Some studies have suggested that preoperative shunting can be encouraged [4]. In our study group, the majority of patients had hydrocephalus (161 of 170 cases) with tumor. These tumors constitute a heterogeneous group of disorders with diverse living and disease free survivals, histological type, extent of resection of the primary tumors, and the patient’s age at presentation. It looks as if increased ICP demonstrates a different entity in these tumors and hence calls for an exclusive approach. In the case of concomitant hydrocephalus, our findings are in favor of performing a preoperative shunting procedure (Fig. 2) or ETV. A VP shunt or ETV can significantly decrease the pressure of the tense posterior fossa and cater for a more appropriate approach for the tumor removal. The surgeon’s interpretation of total or near total extraction of the tumor bulk is more closely correlated with postoperative CT or MRI diagnostic findings when operating field is decompressed.

Our survey emphasizes some hints regarding shunting and ETV procedures:

- They significantly abate the rates of postoperative CSF leak and formation of pseudomeningocele. These adverse effects make the patient prone to septic meningitis and protracted surgical wound healing.

- There was no discrepancies in cases of contusion, upward cerebellar herniation and life-threatening intracranial hemorrhage between those who received preoperative shunting and those who did not.

- The rate of long-term shunt malfunction and infection were not statistically significant in our study (17 of 84). There was no report of death due to macroscopic abdominal metastasis.

- A gradual decrease in the ICP for the posterior fossa tumors is more favorable than an abrupt change. The perioperative mortality rate was considerably lower in patients with hydrocephalus who underwent preoperative shunting or ETV contrasted with those who had either EVD or no shunt at all despite hydrocephalus. Increased ICP in posterior cranial fossa tumors implies a chronic process which should be addressed independently of that for the native primary tumor.

ETV should be considered as an alternative procedure to VP shunt in controlling severe hydrocephalus, related to posterior fossa tumors while patients await their definite tumor excision. The use of pre-resectional ETV was found to be an effective and safe procedure with a high success rate. However, ETV can not always prevent postoperative hydrocephalus in all cases of posterior fossa tumors, the thing that makes using postoperative VP shunt an alternative [28].
Conclusion:

The results of this study showed that the group of patients with preoperative shunting and preoperative ETV had significantly less postoperative complications, and a considerable percentage of those without preoperative shunting needed it later in the course of their disease. Endoscopic third ventriculostomy proved to be successful in immediate and long-term control of hydrocephalus both clinically and radiologically with minimal risk and complications.

References


