MANAGEMENT OF HYDROCEPHALUS IN POSTERIOR CRANIAL FOSSA TUMORS

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Abstract- Treatment of hydrocephalus in posterior fossa tumors in children is still a matter of controversy and different centers have their own routines. In this regard, hospital records of all children with posterior fossa tumors treated in our center during the interval of 1985-1995 were reviewed. Patients' demographic and diagnostic data were analyzed and the frequencies of shunting procedures were determined. Fisher exact test was employed to compare the frequency of postoperative complications in different groups. A total of 108 patients with age ranging from 3 months to 18 years and a male to female ratio of 1.5 comprised the study population. Ninety-nine cases had hydrocephalus at the time of diagnosis and 81 patients underwent preoperative shunting. Of the remaining 18 patients, 13 underwent external ventricular drainage at tumor operation session plus preoperative corticosteroid therapy. The rest of the patients got no primary treatment for hydrocephalus. Three of these 5 patients had postoperative shunting after tumor removal, but the other 2 remained shunt free. The rate of postoperative complications including cerebrospinal fluid leakage and septic meningitis were significantly lower in patients with preoperative shunting. The results of this study are in favor of those that approve the effect of preoperative shunting in decreasing postoperative complications. This is well established when the tumor size is big or when the diagnosis of posterior fossa tumor is made in later stages or when hydrocephalus is severe. It could be concluded that preoperative shunting can decrease the rate of postoperative complications.

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INTRODUCTION

Treatment of hydrocephalus in posterior cranial 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

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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). 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 is advisable (9, 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 cerebrospinal fluid (CSF) leakage, and septic meningitis were among the factors that increase the risk of postoperative need for shunting procedures (16, 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.

MATERIALS AND METHODS

Hospital records of all infants and children with posterior cranial fossa tumor that were treated in Shariati Hospital, Tehran, during the period of 1985-1995 were reviewed. Computerized axial tomography was recruited in 1984 for the diagnosis of intracranial tumors in this center. 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 one of the authors, and the shunting technique did not change during the study period.

Descriptive analysis was performed to determine the frequency of shunting procedures and postoperative complications in different groups. Comparison between the rates of postoperative complications in different shunting groups were made by Fisher exact test using SPSS for windows version 10. The outcome variables used in this study were postoperative complications including CSF leak, septic meningitis, persistent hydrocephalus and pseudomeningocele. 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 shunting 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 108 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 48 cerebellar astrocytoma (CA), 29 medulloblastoma (MB), 14 brainstem glioma (BSG), 12 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.

Among 108 patients, 9 did not have hydrocephalus and 99 had hydrocephalus. The patients without hydrocephalus comprised 5 BSG, 1 cerebellar dermoid cyst, 1 CPA tumor and 1 cerebellar cavernous hemangioma. Those with hydrocephalus were composed of 47 CA, 29 MB, 9 BSG, 12 ependymoma, 1 CPA tumor and 1 cerebellar dermoid cyst.

Table 1. Tumor definition by type

Tumor	Frequency (number)	Male: Female ratio	Mean age (year)
Cerebellar astrocytoma	48	1.3	9.4 ± 4.4
Medulloblastoma	39	3.1	8.7 ± 5.0
Brainstem glioma	14	1.0	7.4 ± 2.8
Ependymoma	12	2.0	9.1 ± 5.0
Miscellaneous	5	0.3	8.8 ± 44
Total	108	1.5	8.9 ± 4.4

Abbriviations: M, male; F, female.

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 99 hydrocephalic patients, 81 underwent preoperative shunting and 18 did not. Of these preoperative shunting procedures, 77 were ventriculo-peritoneal (VP) and 4 were ventriculoatrial (VA) shunts. Of those 18 patients who did not have preoperative shunting, 13 needed preoperative external ventricular drainage (EVD) 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 (one CA and one BSG) 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 pseudomeningocele in the postoperative period in different shunting groups. Persistent hydrocephalus or pseudomeningocele in the preoperative shunting group was significantly lower that 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 4 cases had shunt infection, all of whom needed a replacing VP shunt. None of the patients with preoperative shunting developed upward cerebellar herniation and 5 out of 79 patients had some degree of intracranial hemorrhage; none of them were severe enough to necessitate emergent hematoma evacuation.

EVD was performed in 13 cases with hydrocephalic posterior fossa tumors: 3 had MB, 3 had CA, 2 had ependymoma and 5 had BSG. Regrettably, the incidence of deleterious early complications in the selected group was unproportionately higher than the preoperative shunting group (Table 3).

Table 2. Table of general complications*

	CSF leakage	CSF infection	Persistent hydrocephalus	Pseudomeningocele
Preop Shunting (n= 81)	6	5	2	2
EVD (n= 13)	2	3	3	3
Neither (n=5)	2	1	2	2
Fisher Exact test	0.052	0.079	0.001	0.010

Abbreviations: EVD, external ventricular drainage.
*Data are given as number of patients.

Table 3. Major early complication in EVD patients*

Tumor type	EVD complicated	EVD uncomplicated	Total
BSG	3	2	5
MB	2	1	3
CA	1	2	3
Epen	1	1	2
Total	7	6	13

Abbreviations: EVD, external ventricular drainage; BSG, brainstem glioma; MB, medulloblastoma; CA, cerebellar astrocytoma; Epen, ependymoma.
* Data are given as number of cases.

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 (15). A study of BSG showed that hydrocephalus is present in 20-30% of cases (22); other studies showed a higher incidence of hydrocephalus in brainstem tumors (23, 24). Studies on cerebellar and fourth ventricular tumors show invariable presence of hydrocephalus (24). 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, 18).

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, 6, 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, 26, 27).

Some disadvantages have been mentioned for preoperative shunting in the literature (11). These disadvantages are upward cerebellar herniation (20), tumor hemorrhage (25) and dissemination of tumor cells through shunting systems (20). However, the exact definition and real association of these phenomena to shunting procedure has been questioned in other studies (1, 25) particularly in a developing country, where most of the cases are diagnosed with delay and when a severe hydrocephalus is present.

Some studies have suggested that preoperative shunting can be encouraged (18). In our limited study group, the majority of patients had hydrocephalus (99 of 108 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. A VA shunt or preferably a VP shunt 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 diagnostic findings when operating field is decompressed. Our survey emphasizes some hints regarding shunting procedures:

1) It significantly abates 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.

2) 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.

3) The rate of long-term shunt malfunction and infection were not statistically significant in our study (16 of 18). We missed the number of peritoneal seeding in the VP shunt group as the records did not recite autopsy findings. Nevertheless, there was no report of death due to macroscopic abdominal metastasis.

4) A gradual decrease in the ICP for the posterior fossa tumors is more favorable than an abrupt change. The preoperative mortality rate was considerably lower in patients with hydrocephalus

who underwent preoperative shunting 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. Prophylactic shunting in non-hydrocephalic patients who undergo preoperative radiotherapy and/or multi-agent chemotherapy is not warranted. In conclusion, the results of this study also showed that the group of patients with preoperative shunting had significantly less postoperative complications, and a considerable percentage of those without preoperative shunting needed it later in the course of their disease. Therefore, these results are in favor of preoperative shunting.

Conflicts of interests

We have no conflicts of interests.

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