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Management of hydrocephalus in children with posterior fossa tumors

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Case 1: A 2-year-old male with no prior medical history presented to the emergency room with a 3-week history of constant headache and daily vomiting. Computed tomography (CT) and subsequent magnetic resonance imaging (MRI) of the brain [Figure 1] showed a minimally enhancing mass in the fourth ventricle, which extended out through the foramen of Luschka on the left. There was associated supratentorial hydrocephalus. He had no evidence of spinal metastasis on MRI of the spine. There was no papilledema on the fundoscopic exam. He underwent placement of a right frontal external ventricular drain (EVD) and gross total resection of the tumor through a modified telovelar approach at the same time. The pathology was consistent with a grade II ependymoma. Postoperatively, the ventricular drain was unable to be weaned, and he underwent ventriculoperitoneal shunt placement without complication 1.5 weeks after initial surgery. He was eating and ambulatory after recovery. He went on to radiation therapy.

Case 2: A 9-year-old male with no prior medical history presented to an outside hospital emergency room with 2 weeks of progressive headaches and 1-day of vomiting. A CT of the head showed a posterior fossa mass. MRI of the brain [Figure 2] showed an enhancing fourth ventricular tumor with associated metastatic lesions throughout both cerebellar hemispheres and supratentorial hydrocephalus. There was no evidence of spinal metastasis. Fundoscopic exam was positive for papilledema. He underwent placement of a right frontal EVD and resection of the fourth ventricular mass through a modified telo-velar approach at the same time. The infiltrative lesions in the cerebellum were not resected. The pathology was consistent with medulloblastoma. Postoperatively, his EVD was weaned over the course of 2 weeks and removed. He did not require permanent cerebrospinal fluid diversion. He was discharged home after recovery and went on for adjuvant radiation therapy.

INTRODUCTION

Central nervous system tumors are the most common solid tumors in children, and they predominantly occur in the posterior fossa.[7] Due to the anatomic relationships of these tumors to cerebrospinal fluid (CSF) drainage pathways, hydrocephalus is common, occurring in 71–90% of children with posterior fossa tumors.[11] Hydrocephalus after tumor resection occurs in 10–36% of cases,[2,4] with a worldwide average of 30%.[10]

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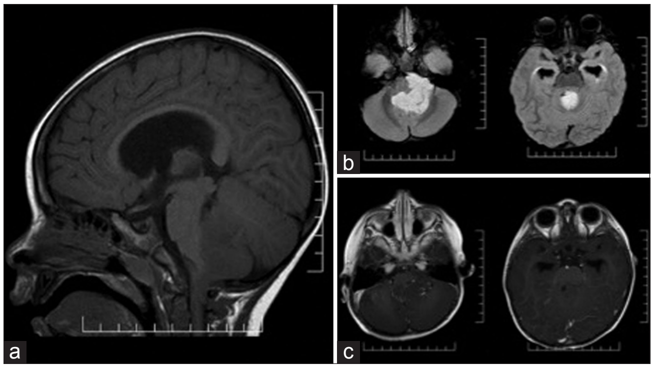

Figure 1: Magnetic resonance images of patient described in case 1. (a) Sagittal precontrast. (b) Axial fluid-attenuated inversion recovery. (c) Axial postcontrast

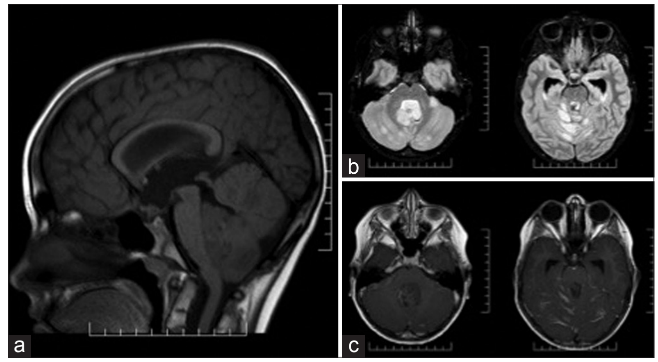

Figure 2: Magnetic resonance images of patient described in case 2. (a) Sagittal precontrast. (b) Axial fluid-attenuated inversion recovery. (c) Axial postcontrast

MANAGEMENT

The optimal management of hydrocephalus in a child with a posterior fossa tumor is a topic of debate.[13] The question of whether to place an external ventricular drain (EVD), insert a ventriculoperitoneal shunt (VPS), perform an endoscopic third ventriculostomy (ETV), or defer CSF diversion procedures before resective surgery depends on the clinical presentation and individual surgeon practice; there exists no class I evidence to guide management. In 2001, Sainte-Rose *et al*. reported that preoperative ETV was associated with a lower rate of postoperative hydrocephalus (27% vs. 6%) in a retrospective series of pediatric patients with posterior fossa tumors ($n = 196$).[11] Only a portion of these patients would have gone on to develop postresection hydrocephalus, so performing a preresection ETV in every case potentially exposes over 70% of patients to unnecessary surgery.[4,6]

Purported benefits of permanent preresection CSF diverting surgery, such as ETV of VPS other than the reduced incidence of postresection hydrocephalus, include the following: (1) Being able to delay resection surgery, thus avoiding resection under emergent conditions or allowing for preresection adjuvant therapy in certain circumstances;[3] (2) reducing the likelihood of needing external CSF diversion, which may carry risk of infection;[3] and (3) potentially reducing risk of postresection CSF leak or pseudomeningocele.[2] Purported disadvantages of permanent preresection CSF diversion surgery include the following: (1) Performance of a procedure that ultimately may not be clinically indicated, exposing patients to the risks of unnecessary surgery; (2) ETV may be less reliable in controlling intracranial pressure (ICP) and does not allow for ICP monitoring; and (3) no ability to externally drain spillage of blood products after the resection. The exact cause of postresection hydrocephalus is not completely characterized, with absorptive and obstructive processes implicated.

FACTORS PREDICTIVE OF POSTRESECTION HYDROCEPHALUS

Ideally, we would be able to predict which patients will develop postresection hydrocephalus. The benefits of early CSF diversion could be captured while simultaneously avoiding the harm of subjecting patients to unnecessary procedures. Many groups have attempted to analyze retrospective data looking for clinical factors associated with a need for postoperative CSF diversion. Culley *et al*. ($n = 117$, 1976–1990) found that age <3 years, midline tumor location, subtotal resection, prolonged EVD requirement, cadaveric (vs. autologous) dural grafts, pseudomeningocele formation, and CSF infections were statistically significant factors associated with the need for postoperative shunt placement.[2] Due-Tønnessen and Helseth ($n = 87$, 1990–2003) found that patients with medulloblastoma and ependymoma had much higher rates of postoperative shunt requirement than astrocytomas.[4] Kumar *et al*. ($n = 175$, 1983–1993) found age <3, ependymoma/medulloblastoma tumor histology, and subtotal resection to be risk factors.[8] Santos de Oliveira *et al*. ($n = 64$, 1990–2006) found younger age, midline location, and greater ventricular index at presentation to be risk factors.[12] Morelli *et al*. ($n = 160$, 1989–2004) found medulloblastoma histology and severe preoperative hydrocephalus to be risk factors.[9] Bognár *et al*. ($n = 180$, 1990–2000) found younger age, tumor histology, and presence of EVD to be predictive of postoperative need for CSF diversion, but they found that tumor location, extent of resection, and postoperative CSF leak or pseudomeningocele were not predictive.[1]

In 2009, Riva-Cambrin *et al*. used a cohort of 343 patients to develop a clinical prediction rule for postresection hydrocephalus and validated it against another cohort of 111 patients from another institution in an attempt to identify high-risk patients who would benefit most from prophylactic ETV.[10] The group analyzed demographic, clinical, and radiographic factors. They performed stepwise multivariate regression to determine which factors were associated with a greater risk of needing CSF diversion after tumor resection and assigned point values reflecting the relative weights. The final scale is out of 10 points, with 3 points given for age <2 years, 1 point given for papilledema, 2 points given for moderate or severe hydrocephalus, 3 points given for cerebral metastases, and 1 point given for ependymoma, medulloblastoma, or dorsally exophytic brainstem glioma pathology predicted by preoperative radiology report. A score of >4 points was chosen as the cut-off for "high-risk." Those with a score of 0–2 are predicted to have <20% chance of developing postresection hydrocephalus while the likelihood is >80% for those with a score of 7–10. High-risk (score 5–10) and low-risk (score 0–4) groups differed in posttest probabilities for developing postresection hydrocephalus by 48% (73% for high-risk, 25% for low-risk).[10] Foreman *et al*. later validated and modified Riva-Cambrin *et al*.'s predictive model, using fewer variables in a much smaller cohort (n = 99 patients): Age <2 years, moderate/severe hydrocephalus, preoperative tumor diagnosis per radiology report, and transependymal edema. These posterior fossa tumor patients were also stratified into high- and low-risk categories for development of postresection hydrocephalus.[5]

TREATMENT RECOMMENDATIONS

There exists no class I evidence in the literature to guide the management of hydrocephalus in children with posterior fossa tumors. It is possible to draw guidance from the extant data highlighted above. As the overall incidence of postresection hydrocephalus is typically 30%, any anticipated benefit should be weighed against exposing the patient to more surgery or permanent shunt implantation. It is noted that in lower resource settings, there may be other considerations, including the cost of care, access to the operating room and need to minimize the number of surgeries. In our practice, in cases where there is no hydrocephalus on presentation, preresection CSF diversion is not done. In cases where there is symptomatic hydrocephalus on presentation, preresection EVD, VPS or ETV should be applied as clinically appropriate. EVD is favored for its advantages of expedient placement, external control over drainage perioperatively, and egress of resection-related blood and protein products. In cases where the child possesses multiple described risk factors for the development of postresection hydrocephalus, preresection prophylactic CSF diversion may be considered. Overall, close observation is recommended, with a preference for expectant management, rather than prophylactic surgery, and postresection definitive CSF diversion procedures undertaken only as clinically necessary.

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