Temporary ventricular drainage and emergency radiotherapy in the management of hydrocephalus associated with germinoma

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Object. The authors used an alternative strategy to avoid shunt placement for hydrocephalus associated with germinoma, and the ensuing complications.

Methods. Between 1998 and 2000, five patients presenting with germinomas of the pineal area and symptomatic obstructive hydrocephalus were treated with a novel strategy. On arrival, they underwent ventriculostomy placement and one of several surgical procedures to obtain tissue for diagnosis. Within several days of the initial diagnosis, stereotactically guided fractionated radiotherapy was started. All patients experienced rapid tumor shrinkage and resolution of hydrocephalus, allowing discontinuation of external ventricular drainage without the need for permanent shunting of cerebrospinal fluid. To date, follow up reveals 100% radiographically and clinically confirmed tumor control.

Conclusions. Prompt resolution of hydrocephalus and absence of complications make this a potentially valuable therapy for control of germinomas and their symptoms.

KEY WORDS • germinoma • hydrocephalus • ventriculoperitoneal shunt • radiotherapy

Intracranial germinomas are rare tumors that account for less than 1% of all intracranial neoplasms. Approximately two thirds of germinomas arise in the pineal region, with most of the remainder in the suprasellar space. These tumors are most frequently seen in young adolescents and occur more frequently in males.

Signs and symptoms on presentation of pineal region germinomas include headache, papilledema, nausea, vomiting, and lethargy caused by obstructive hydrocephalus. In addition, pressure on the dorsal midbrain may produce Parinaud syndrome, which is characterized by upgaze paralysis, pupillary light-near dissociation, and retraction-convergence nystagmus. Hypothalamic and pituitary dysfunction, including diabetes insipidus, growth failure, and delayed or precocious puberty, are frequently seen with suprasellar tumors.

The treatment of intracranial germinomas has changed substantially over the past several decades. Improvements in surgical technology have led to the routine use of tissue diagnosis, with specimens usually obtained through stereotactic biopsy sampling, but also through open craniotomy and endoscopic procedures. These lesions are highly curable with radiotherapy, although debate continues about the dosage, the use of craniospinal radiotherapy, and the role of adjuvant chemotherapy.21

Placement of a VP shunt is frequently used to alleviate hydrocephalus in these patients. The small but well-documented incidence of poor outcomes associated with shunt-disseminated metastasis led us to try temporary ventricular drainage and emergency radiotherapy as an alternative.

Clinical Material and Methods

Between April 1996 and April 1998, five patients with intracranial germinomas and symptomatic obstructive hydrocephalus were seen at the University of Florida. Presenting symptoms included headache, diplopia, nausea, and imbalance. One patient presented with two lesions (one in the pineal and one in the suprasellar region); he had panhypopituitarism, including diabetes insipidus. All patients underwent temporary ventricular drainage with ventriculostomy, and one of several surgical approaches was performed to obtain tissue samples for diagnosis—either magnetic resonance imaging-guided stereotactic biopsy (in three) or endoscopic biopsy (in two). All patients then underwent emergency radiotherapy; no patient received chemotherapy in this series. Case details, including the radiotherapy regimen, are listed in Table 1.

Results

All patients experienced complete resolution of hydrocephalus, allowing removal of the ventricular drain 2 to 14 days after initiation of therapy. No patient experienced a complication related to prolonged ventricular drainage. After completion of radiotherapy, all patients experienced resolution of the presenting symptoms. At the time of last follow up (19–58 months posttreatment), all patients had...
Hydrocephalus and germinomas

Discussion

Hydrocephalus is frequently associated with intracranial germinomas and is often treated with VP shunt placement. More than 35 cases of "shunt metastasis" have been reported to date.1,3,4,12,22,27,29 In many of these cases, peritoneal dissemination could not be controlled with further therapy and led to a poor outcome in what is generally regarded to be a highly curable disease. For this reason, we elected to try the alternative approach outlined here, which was specifically conceived to avoid the need for a shunt, and, thereby, the small risk of shunt-disseminated metastasis. There is a small risk of infection from prolonged ventricular drainage.11,14,16 Of course, shunts are also associated with other complications, including intracranial hemorrhage, infection, and device malfunction. These also might be avoided with the strategy described in this study.

Several decades ago, the risk from surgery (even a biopsy procedure) was sufficiently high that many patients were treated presumptively, with radiotherapy.6 Improvements in surgical technology have yielded highly effective open, stereotactic, and endoscopic methods8,21 for obtaining tissue samples for diagnosis in germinomas; tissue for diagnosis was obtained in all cases in this series.

Radiotherapy has long been the mainstay of treatment for these highly radiosensitive tumors;1,4,12,22,26,28 cure rates of better than 90% are typically reported. Although radiotherapy is clearly very effective, it is associated with delayed side effects, including impaired growth, cognition, and pituitary function. For this reason, in many centers the use of lower radiotherapy doses has been advocated more recently,13 along with the elimination of craniospinal radiotherapy,17 unless magnetic resonance imaging of the spine or cytological studies reveal spinal metastasis. All patients in this series were treated with a combination of relatively low-dose cranial radiotherapy, including ventricular volumes and a margin, and a stereotactically guided fractionated boost to the visible disease areas.7,18

Extracranial germinomas are very sensitive to chemotherapy. In addition, the agents commonly used for germinomas (cisplatin, vinblastine, bleomycin, cyclophosphamide, and so on) appear to penetrate the blood–brain barrier well, probably because of breakdown of the barrier associated with the tumor. For this reason, in some centers adjuvant chemotherapy has been recommended more recently as a method of reducing the dosage of radiotherapy, delaying it in younger children, or eliminating the need for it altogether.1,5,10,19,20,22 Nevertheless, the ability to produce a cure reliably in 90% of cases has not been attained with chemotherapy alone, and we did not use chemotherapy in this series.

Recent improvements in endoscopic technology have led to renewed interest in third ventriculostomy as a treatment for obstructive hydrocephalus, such as that seen in germinomas.9,23 In addition, tissue samples for diagnosis can frequently be obtained endoscopically during the same procedure. Third ventriculostomy does, however, carry a small risk of serious side effects.15 In addition, the potential for this procedure to cause dissemination of this disease through cerebrospinal fluid is unknown.

Conclusions

In this small series, external ventricular drainage and emergency radiotherapy resulted in prompt resolution of hydrocephalus as well as long term tumor control. This approach may be considered an alternative to VP shunt placement or third ventriculostomy for control of hydrocephalus.

References


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* As of their last follow-up visit, all patients were cured. Abbreviations: CS = craniospinal; frac = fractions; SRT = stereotactic radiotherapy; VB = ventricular boost.

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