

Endoscopic Reconstruction of CSF Pathways in Ventricular Tumors

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Abstract Neuroendoscopy is now considered to be a minimally invasive surgical approach for expanding lesions bulging into the ventricle, and it is also considered to be a relevant tool for performing biopsy procedures, fenestration of cystic walls, or for performing tumor removal in selected cases. Furthermore, the use of neuroimaging and the accurate follow-up of brain tumor patients have allowed the documentation of tumoral and pseudotumoral cystic areas that cause the obstruction of cerebrospinal fluid (CSF) pathways. Neuroendoscopic procedures enable the fenestration of cystic lesions, in addition to enabling third ventriculostomy or septostomy to restore CSF pathways. We analyze our experience regarding 77 patients affected by brain tumors arising from the wall of the third or lateral ventricle. In all cases hydrocephalus or obstruction of CSF flow was present. With an endoscopic technique, septostomy, cystostomy, endoscopic third ventriculostomy (ETV), and tumor resection were performed to control intracranial hypertension.

ETV was performed in 53 patients with noncommunicating hydrocephalus. In 4 patients with low-grade astrocytoma ETV was definitely the only surgical treatment. In 12 cystic tumors, cystostomy and marsupialization into the ventricle solved a relevant mass effect with clinical intracranial hypertension syndrome. In 10 patients, neuroendoscopic relief of CSF pathways was possible by performing septostomy with the implantation of an Ommaya reservoir or one-catheter shunt. In 5 colloid cysts and 2 cystic craniopharyngiomas, removal was possible by restoring CSF flow without other procedures. After intracranial hypertension control, in 13 malignant gliomas and 5 leptomeningeal metastases, the patients' quality of life improved sufficiently to provide for tumor adjuvant therapy.

In this series, endoscopy, due to its minimally invasive characteristics and reduced complications, was found to be safe and effective, without any relevant postoperative morbidity, gained by avoiding major surgical approaches.

Based on these results and on the increasing number of series described in the literature, we believe that endoscopic techniques should be considered a selected approach for treating CSF obstructions caused by para-intraventricular tumors. The result of using neuroendoscopy is the reconstruction of CSF pathways that bypass the tumor occlusion. This surgical procedure is not only limited to the relief of noncommunicating hydrocephalus, but it is also useful for tumor removal or biopsies and the evacuation of cystic lesions. In patients affected by malignant tumors, neuroendoscopy can be performed to control intracranial hypertension before the patients start adjuvant chemotherapy or radiotherapy.

Keywords Intraventricular tumor • Endoscopic biopsy • Endoscopic third ventriculostomy • Hydrocephalus

Introduction

Because of the central location of intra- and paraventricular tumors, the commonly employed open surgical approaches have relatively high potential morbidity and mortality [7, 21]. Notably, the increased use of neuroimaging and the accurate follow-up of brain tumor patients have now frequently allowed the documentation of tumoral and pseudotumoral cystic areas that cause the obstruction of cerebrospinal fluid (CSF) pathways [8, 18]; these tumors are often associated with dilated ventricles and intracranial hypertension [19, 20]. For this reason, even today, microsurgical removal is considered the best therapeutic option in selected cases. However, due to the deep location of intra- and paraventricular tumors, this removal remains challenging and is fraught

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with potential complications, which may be functional and cognitive or even life-threatening. Neuroendoscopy is now considered a minimally invasive surgical approach for expanding lesions bulging into the ventricle, as well as being considered a relevant tool for performing biopsy procedures, discontinuation of cystic walls, or performing tumor removal in selected cases [2, 4, 5, 14]. Furthermore, neuroendoscopic procedures can be used to reconstruct alternative CSF pathways and treat intracranial hypertension by the fenestration of cystic lesions, in addition to being used for endoscopic third ventriculostomy (ETV) or septostomy [6, 12, 13]. In addition to the use of endoscopic procedures for diagnostic sampling, patients with primary cystic tumors (colloid cyst, epidermoid cyst, cystic craniopharyngioma, Rathke's cleft cyst) are optimal candidates for endoscopic procedures, given the ease of cyst aspiration and cyst ablation or resection. In selected cases, partial or total tumor removal is possible to improve CSF circulation, the success of which is dependent upon the tumor consistency, a feature that may be difficult to predict preoperatively [9, 17]. Thus, many patients will benefit by treatment of the tumor manifestations of non-communicating and compartmentalized hydrocephalus through the use of endoscopic procedures. These procedures can also be coupled with the accurate placement of catheters for intracavitary therapeutic purposes [10] or sequential aspiration. Thus, in patients in whom the disease will be treated primarily by nonsurgical means (primary central nervous system [CNS] germ cell tumors, primary CNS lymphoma, disseminated metastatic disease, malignant glioma, tectal glioma) endoscopic CSF pathway relief can offer a distinct benefit by avoiding a more extensive intracranial procedure.

Material and Methods

From 2002 to 2014, 77 patients with solid or solid cystic tumors arising from the wall of the ventricles underwent neuroendoscopic procedures. These patients ranged in age from 8 to 79 years (median 55 years.). There were 42 males and 35 females, with seven children. All patients were symptomatic: 23 (30%) patients presented with a classical intracranial hypertension syndrome. The others presented with focal neurological signs, as well as ataxic gait, cognitive disorders, and headache with papilledema. The patients had a median presurgical Karnofsky performance score (KPS) of 55 (range 30–70). At preoperative magnetic resonance imaging (MRI), tumor sites were: 31 (40%) in the third ventricle, 27 (35%) in the lateral ventricle, 2 in the fourth ventricle, 4 in the Sylvian aqueduct, and 8 in the brain stem, while 5 showed leptomeningeal diffusion. Ventricular dilation with hydrocephalus or obstruction of CSF flow was present in all cases. Depending

on the location of the tumor and the ventricle size, unilateral access (mainly right side) was performed in all cases. The endoscope's trajectory was planned in only 5 cases, with the help of a neuronavigation system (BRAINLab system; Feldkirchen, Germany). Septostomy, cystostomy, ETV, or tumor resection was performed with rigid (25%) or flexible (75%) endoscopes to control intracranial hypertension. If needed to obtain a diagnosis for further oncological treatment, a tumor biopsy was performed during the same endoscopic CSF relief procedure. We used a thulium (Tm) diode pumped solid state (DPSS) laser (Revolix LISA Laser Products, Katlenburg, Germany) for the shrinkage or tissue desiccation of thick tissue and for the hemostasis of highly vascularized tumors [11, 15].

Results

The CSF obstruction was associated with tumors of different histologies (Table 1). In 34 patients, new diagnosis tumor endoscopic biopsy was performed, combined with procedures to reconstruct CSF pathways. After diagnosis, 10 patients were operated on for microsurgical removal. From 2009, in 24 procedures the Tm laser was used for the ablation and cutting of tumor tissue, hard synechiae, and cyst membranes. With Tm laser septostomy, cyst fenestration, tumor resection, and foramen of Monro and aqueduct opening were feasible to promote the reestablishment of ventricular anatomy and hydrodynamic patterns. In 5 patients, total removal of colloid cysts was possible, and in 2 patients with cystic craniopharyngiomas subtotal removal was possible by restoring CSF flow without other procedures. In 53 patients with noncommunicating hydrocephalus, ETV was performed successfully. In 10 of these patients, the hydrocephalus was due to posterior cranial fossa tumor, while in 5 it was secondary to radionecrosis. In 12 cystic tumors cystostomy and marsupialization into the ventricle solved a relevant mass effect with clinical intracranial hypertension syndrome. In 10 patients the relief of CSF pathways was possible by performing septostomy associated with the implantation of an Ommaya reservoir or one-catheter ventriculoperitoneal (VP) shunt. In 2 patients with epidermoid cysts with postoperative entrapped fourth ventricle after aqueductoplasty and ETV, reconstruction of CSF flow was obtained (Fig. 1a, b).

Complications

No mortality or morbidity due to the procedures was present. Mild or severe bleeding was successfully controlled with the Tm Laser. Following ETV failure in six cases, a one-catheter VP shunt was implanted. In two patients with

Table 1 Histopathological diagnoses

Diagnosis	No of patients
Glioma; low grade	9
Glioma; high grade	13
Tectal glioma	4
Malignant teratoma	2
Colloid cyst	5
Radionecrosis	7
Craniopharyngioma	7
PNET	5
Lymphoma	4
Metastases	9
Leptomeningeal metastases	5
Epidermoid cyst	2
Nonspecific tumor	5

PNET primitive neuroectodermal tumor

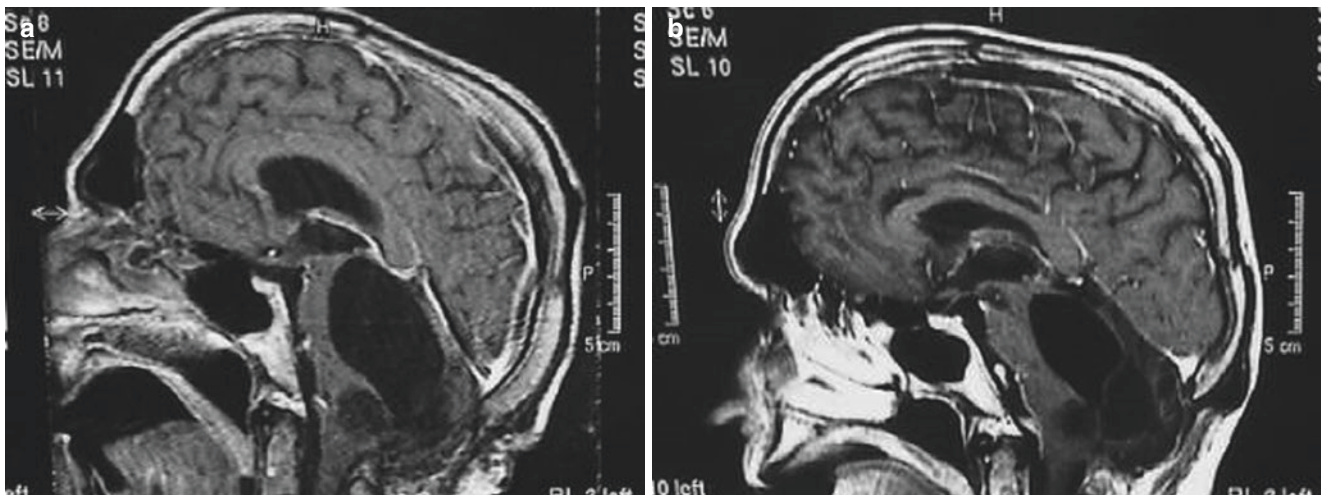


Fig. 1 (a) Pre-endoscopic magnetic resonance imaging (MRI), showing the entrapped fourth ventricle as a complication of microsurgical removal of an epidermoid cyst; (b) post-endoscopic MRI, showing

reduction of the cystic fourth ventricle after aqueductoplasty and endoscopic third ventriculostomy (ETV)

entrapped ventricle the fenestration was redone, while two cystic tumors were treated microsurgically.

quality of life and overall survival. Patients with tumors responding to therapy or radionecrosis showed a longer overall survival.

Follow-Up

The reconstruction of CSF pathways increased the median KPS > 80 (range 60–100). The follow-up ranged from 1 to 10 years, with periodic clinical and MRI examinations. In 4 tectal low-grade astrocytomas ETV was definitely the only surgical treatment. After intracranial hypertension control was achieved, in 13 malignant gliomas, 5 leptomeningeal metastases, and other malignant tumors, specific chemotherapy and/or radiotherapy was administered, improving

Discussion

Ventricular tumors represent a heterogeneous group in terms of histology and therapy, but they often present a common clinical history and common radiological aspects [14, 19]. The most frequent type of clinical presentation is a syndrome arising from intracranial hypertension. In the present series this syndrome was present in 30% of our patients, accompanied by ventricle dilation due to blockage of the

CSF pathways with hydrocephalus or entrapped ventricle. In these cases the patients' clinical features ruled out the possibility of performing other therapies as an alternative to surgery, which is elective both for diagnosis and for relieving intracranial hypertension. Some tumors (i.e., lymphoma or germinoma) are radiosensitive and their surgical removal is excluded [3].

Intraventricular endoscopic surgery has an integral and expanding role in the management of patients with brain tumors. Established applications exist for tumor biopsy, concordant CSF diversion, tumor cyst decompression, and colloid cyst removal. This surgery also offers the possibility of employing ETV for the treatment of associated hydrocephalus, instead of VP shunting, a procedure that may play a role in the dissemination of some tumors, such as pineoblastomas and germ cell tumors, into the peritoneum [14].

In accordance with the literature [1, 16], in the present series endoscopy was found to be safe and effective—without any relevant postoperative morbidity—for reconstructing CSF pathways and restoring CSF flow, and for providing ETV, septostomy, tumor resection, or other endoscopic procedures. In patients affected by malignant tumors, neuroendoscopy can be performed to control intracranial hypertension before the patients start adjuvant chemotherapy or radiotherapy. Because of the tumor site these patients are frequently admitted with intracranial hypertension and a low KPS, which creates a challenge for any kind of treatment. After endoscopic procedures the reduction of intracranial pressure and improvement of the KPS is made possible, depending on the histology, adjuvant therapy, or microsurgery. Furthermore, in tectal glioma or radionecrosis ETV can be employed as the only surgical procedure without any other therapy.

Based on these results and on the increasing number of series described in the literature, we believe that endoscopic techniques should be considered as a selected approach to treat CSF obstructions caused by para-intraventricular tumors. This surgical procedure is not only limited to the relief of noncommunicating hydrocephalus, but is also useful for tumor removal or biopsies and the evacuation of cystic lesions.

Conflict of Interest Statement The authors declare that they have no conflicts of interest.

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