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Original Article

Endoscopic considerations treating hydrocephalus caused by basal ganglia and large thalamic tumors

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Abstract

Background: Deep basal-ganglia and large thalamic (BGT) tumors may cause secondary hydrocephalus by compressing the lateral and third ventricles. The ventricular distortion, as well as the infiltrative nature and friability of these tumors, raise specific considerations and risks when treating these patients. Treatment goals may therefore focus on cerebrospinal fluid (CSF) diversion and tissue sampling, followed by nonsurgical treatment options. We present our experience in applying endoscopic techniques for the initial management of such patients.

Methods: Over a period of 15 months (January 2013 to April 2014), six patients with BGT tumors presented with signs and symptoms of increased intracranial pressure secondary to hydrocephalus. Data was collected retrospectively, including clinical, surgical, and outcome variables.

Results: Six patients aged 9–41 years (25.6 ± 12.5) were included. Endoscopic procedures included endoscopic third ventriculostomy (4), septum pellucidotomy (5), foramen of Monro stenting (2), and endoscopic biopsy (3). One patient underwent a ventriculoperitoneal shunt placement and another stereotactic biopsy. Indications for endoscopic treatment included the infiltrative nature of the tumor preventing a resective procedure, combined with clinical deterioration related to increased intracranial pressure secondary to hydrocephalus. Pathology results included anaplastic astrocytoma (3) and anaplastic oligodendroglioma (1). Pathological sampling was not possible in two patients. Five patients enjoyed a good clinical recovery with no associated morbidity. There was one perioperative death, secondary to preoperative herniation.

Conclusions: Endoscopic surgery may potentially play a significant role in the initial management of patients with large basal ganglia and large thalamic tumors causing obstructive hydrocephalus. Technical nuances and individualized goals are crucial for optimal outcomes.

Key Words: Basal ganglia, endoscopy, hydrocephalus, thalamus, tumor

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INTRODUCTION

Basal-ganglia and large thalamic (BGT) tumors cause ventricular distortion as well as medial compression, compressing the body of the third ventricle and the foramina of Monro and commonly produce obstructive hydrocephalus. Thus, in this subgroup of BGT tumors, presenting symptoms are attributed to focal signs combined with elevated intracranial pressure (ICP). These large deep-seated tumors mostly represent centrally located friable high-grade gliomas (HGG), and may not be amenable to radical resection. Accordingly, treatment options may be limited to cerebrospinal fluid (CSF) diversion and tissue diagnosis, followed by nonsurgical oncological treatments.

In contrast to hydrocephalus secondary to posterior third ventricular tumors, and small posterior-medial thalamic tumors, which compress the aqueduct or posterior third ventricle and cause triventricular symmetrical hydrocephalus, large BGT tumors cause distorted ventricular anatomy, often distorting the region of the foramina of Monro, and the anterior third ventricle. This distortion significantly increases the risks of surgical treatment.

We present our experience treating large BGT tumors causing hydrocephalus, focusing on the role of endoscopic surgery and discussing specific surgical nuances.

MATERIALS AND METHODS

This study was approved by the medical center IRB (approval number 0262-14-TLV). Between January 2013 and April 2014, six patients with obstructive hydrocephalus secondary to basal ganglia or large thalamic tumors underwent an endoscopic procedure at the Tel-Aviv Medical Center. Patient data, including clinical, radiological, surgical, and pathological records were retrospectively collected. Tectal tumors extending to the pulvinar region, and posterior thalamic tumors (compressing the aqueduct, but sparing significant third ventricular or foramina of Monro compression) were excluded from this cohort of patients. Two indications were recognized for endoscopic treatment: To alleviate increased ICP caused by hydrocephalus, and to achieve histological diagnosis by an endoscopic biopsy, if deemed medically necessary.

Surgical procedures

Several surgical procedures were performed, including endoscopic third ventriculostomy (ETV), septum pellucidotomy (SP), endoscopic biopsy (EBX), stereotactic biopsy (SBX), stent placement (St), and ventriculo-peritoneal shunting (VPS).

Navigation was used in all cases – identifying the entry point (according to the procedure plan), assisting in ventricular cannulation (according to the trajectory plan), and, in one case, for registering the endoscope as a navigation tool. Following dural opening, a peel-away (12F or 20F) was entered into the ipsilateral ventricle. Procedures were performed using a rigid endoscope (Oi HandyPro®, Storz, Germany [1 case], and Minop, Aesculap, Germany [5 cases]). When performing an EBX concurrently with an ETV or SP, CSF drainage procedures (ETV and SP) were performed prior to EBX.

In selected cases, a stent was inserted to maintain the patency of the foramen of Monro. This was always done after finishing the intraventricular procedures. The stents were placed using an intraventricular endoscope (NeuroPEN™, Medtronic PS Medical, CA, USA), and a ventricular catheter, with additional proximal holes placed through the relevant orifice and anchored to a subcutaneously--implanted Ommaya reservoir.

RESULTS

Six patients were included in this report (five males). Age at surgery ranged from 9 to 41 years (25.6 ± 12.5, mean ± SD). All patients presented with obstructive hydrocephalus, most having moderately enlarged ventricles (mild enlargement in one), and all having periventricular edema and diminished convexity subarachnoid space. All patients presented with symptoms related to increased ICP [Table 1]. Four had a right-sided pathology. Four patients harbored a thalamic tumor, one

Table 1: Patient data summary

Age at surgery (years)	Location	Presenting symptoms	Comments	Periventricular hyperintensity signal	Peripheral SAS
17	Rt/Th	HA, N/V		Yes	Diminished
38	Lt/Th	HA, MD		Yes	Diminished
20	Rt/Th	HA, N/V, Dr	GCS12	Yes	Diminished
29	Rt/BG+Th	HA, N/V, MD		Yes	Diminished
9	Rt/Th	HA, Dr	GCS14	No	Diminished
41	Lt/BG+fr	MD	Behavioral changes, just prior to surgery dilated pupils, somnolent	Yes	Diminished

Rt: Right, Lt: Left, BG: Basal-ganglia, Th: Thalamus, HA: Headache, N/V: Nausea/vomiting, MD: Memory decline, Dr: Drowsiness, GCS: Glasgow coma scale, SAS: Subarachnoid spaces

with a thalamic-basal-ganglia tumor, and one with a deep frontal-basal-ganglia tumor [Figure 1 and Table 1].

Endoscopic procedures included: ETV (4 patients), SP (5), EBX (3), and a transforaminal stent (2) [Table 2]. Besides the endoscopic procedure, one patient required a VPS placement, another patient required a temporary external ventricular drain (EVD), and a third patient subsequently underwent a SBX, performed at a later stage. In four cases, side of entry for the endoscopic procedures was contralateral to the tumor.

In total, one patient died shortly after surgery (as described below), the other five patients had an uneventful course, with complete resolution of their symptoms following the endoscopic procedure. One patient died several months after surgery (due to tumor progression).

A pathological specimen was obtained in four patients (three endoscopically and one with the stereotactic biopsy (SBX) at a separate setting). Results included anaplastic astrocytoma in three patients and an anaplastic oligodendroglioma in one. In one patient, who had presented with a hemorrhagic lesion, no biopsy was attempted. A sixth patient died shortly after surgery with no pathological diagnosis due to a technical inability to obtain an endoscopic specimen during the procedure.

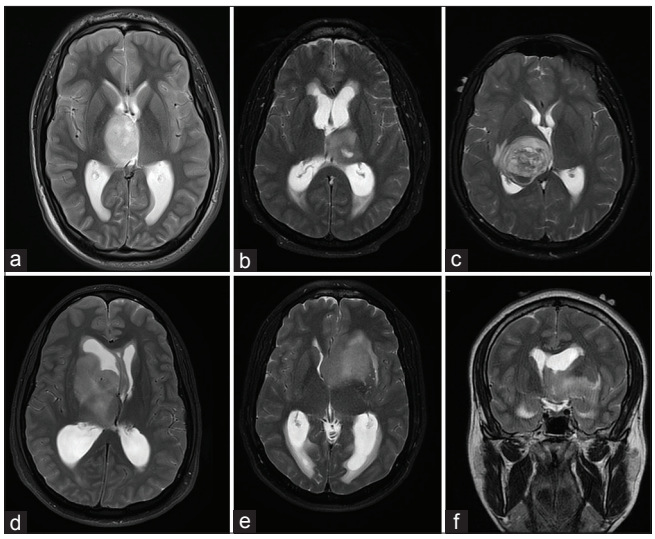

Figure 1: Representative T2 images corresponding to cases 1-4 (a-d) and case 6 (e-f)

Table 2: Surgical notes

Surgery	Side of endoscopic entry	Pathology	Follow-up in months (outcome)
ETV, SP, St, SBX (at 2nd stage)	Lt	AA	4 (alive)
ETV, SP, EBX	Rt	AO	15 (alive)
ETV, SP, St	Lt	No BX	11 (alive)
VPS, SP, EBX	Rt	AA	14 (alive)
ETV, SP, EBX	Lt	AA (→GBM)	9 (dead)
SP, EVD	Lt	No BX	1 week (dead)

Rt: Right, Lt: Left, ETV: Endoscopic third ventriculostomy, SP: Septum pellucidotomy, BX: Biopsy, SBX: Stereotactic biopsy, EBX: Endoscopic biopsy, St: Stent, EVD: External ventricular drain, VPS: Ventriculoperitoneal shunt, AA: Anaplastic astrocytoma, AO: Anaplastic oligodendroglioma, GBM: Glioblastoma

Two patients underwent additional surgery following the endoscopic procedure. A 9-year-old girl with rapid tumor progression underwent tumor resection and was found to have a glioblastoma multiforme (GBM; following an AA diagnosis obtained endoscopically). The second patient underwent a decompressive craniectomy shortly after the endoscopic procedure in a futile attempt to relieve uncontrolled increased ICP.

Four patients underwent subsequent oncological treatments (radiation + chemotherapy), based on the pathological diagnosis. Follow-up of five patients (the sixth died shortly after surgery) was 4–15 months (10 ± 4.4). One patient died of tumor progression. None of the remaining patients developed new CSF-related symptoms.

There was one intraoperative complication (leading to mortality). A 41-year-old male presented with behavioral changes over a course of several weeks. Following diagnosis, while awaiting surgery, he exhibited acute deterioration (secondary to herniation). During surgery, following introduction of the peel-away into the lateral ventricle, a SP was performed and the ventricles collapsed, eliminating any possibility of additional maneuvers. An EVD was inserted, but the patient failed to recover despite a subsequent decompressive craniectomy, and died a few days later.

DISCUSSION

Neuro-endoscopy has become a main stream of modern neurosurgery. In the past years, the scientific community is struggling to better understand pathophysiological mechanisms of ventricular pathology including tumors and hydrocephalus. We also try to improve the usage of the current available technology. Endoscopes are becoming cheaper and can be connected to many "towers" that are available in hospitals. With maturation of these new techniques and keeping the patient in the center, we can offer a new level of technology and safety. This study provides a detailed analysis of the surgical considerations when treating patients with obstructive hydrocephalus secondary to deep-seated tumors (basal ganglia or thalamic), focusing on the role of endoscopy.

Basal-ganglia tumors are relatively rare, and the literature regarding their surgical treatment is sparse.[2,3] Posterior thalamic tumors often cause obstructive hydrocephalus secondary to posterior third ventricular and aqueductal compression, with sparing of significant third ventricular and formina of Monro compression. These cases are

often treated with an ETV, sometimes combined with endoscopic biopsies.[7] Often, in large EBX series, there is no focus on anatomical considerations regarding the endoscopic technique, and many paraventricular locations are gathered together.[11] Large BGT tumors however, cause ventricular distortion as well as medial compression on the body of the third ventricle and the foramen of monro. These structural displacements in turn cause secondary obstructive hydrocephalus that is often asymmetrical.

Treatment of the hydrocephalus caused by BGT tumors depends on several factors; mainly tumor pathology, size, and tumor location relative to the ventricular system and cortico-spinal tract (including infiltration of these fibers). For example, focal, low-grade tumors such as pilocytic astrocytomas are often amenable to resection, with resolution of hydrocephalus and a potential cure.[6,10] Large metastatic tumors are also often resected, thus removing the cause of hydrocephalus. On the other hand, diffuse, infiltrative tumors may not be amenable to meaningful resection, especially in high-grade tumors, and treatment may be limited to a CSF-diversion procedure and histological diagnosis of the tumor. Smaller metastasis may also be treated with CSF-diversion surgery, with or without additional radiation therapy (such as stereotactic radiosurgery).

In the current report we present our experience treating patients with large thalamic and basal ganglia tumors causing obstructive hydrocephalus. Because preoperative imaging was suggestive of primary high-grade tumors, our treatment paradigm did not focus on tumor resection. Since the presenting symptoms were secondary to increased ICP, we elected to treat the CSF pathology and perform diagnostic biopsies only.

A common feature in these cases is that the ventricles were only mild-to-moderately enlarged despite clear symptoms of elevated ICP, which improved following CSF diversion. We deduced that ventricular compliance was low, and thus the smaller ventricular size underscores the intensity of the hydrocephalic process, increasing the risks of fast decompensation (as eventually happened in one of our cases). Two radiological signs that should alert the surgeon to an active hydrocephalic process include the presence of periventricular edema (on T2 sequences) and diminished supratentorial subarachnoid spaces on the convexity.

Surgical considerations

The primary goal of endoscopic surgery is to perform and optimize CSF diversion. Treatment options may include ETV, SP, and VPS, as a single procedure or in various combinations. There are several technical nuances to note regarding endoscopic procedures for these patients:

- The impact of a tight foramen of Monro region and the distorted anatomy must be addressed when planning the surgery
- Additional considerations to take into account include possible collapse of the ventricles immediately after cannulating the lateral ventricle (due to low ventricular compliance) as well as the risk of future tumor growth (which may completely obstruct the already small foramen of Monro). For these reasons, a SP should be seriously considered even if the foramen of Monro are initially not totally obstructed
- When performing a SP, it is important to anticipate contralateral engorged septal veins and coagulate the septum carefully, avoiding septal vein tear and a bleed that will hamper the endoscopic procedure[9]
- In general, we prefer an ETV to a VPS, as ETV provides a "physiological" solution to the obstructive hydrocephalus and ventricular collapse with proximal shunt occlusion is less likely to occur. However, if the anterior third ventricle is very small, a VPS is a valid option
- The risk of future tumor growth and complete subsequent obstruction of the foramen of Monro should provoke consideration on the use of a Monro stent, even when the foramen is still patent. This is accomplished through a ventricular catheter passing through the Monro, with added proximal holes above and below the foramen. The catheter is anchored using an Ommaya reservoir, which is positioned in a subcutaneous location. Correct placement of the stent may be optimized using an intraluminal endoscope (such as the NeuroPEN™, Medtronic PS Medical, CA, USA)[8]
- The entry side should be selected carefully. The ipsilateral ventricle is compressed, as is the ipsilateral Monro. If an ETV is planned, often the contralateral approach will be safer, through a larger Monro. A SP may be done from either side; however, if approached from the side contralateral to the tumor, the distance between the septum and the lateral wall of the ventricle on the side of the tumor should be expected to be shorter. A biopsy of a thalamic tumor may be easier when approaching from the contralateral side, through the third ventricle
- Navigation is important for several reasons. Planning the exact entry point (with a safe trajectory for an ETV, SP, and an EBX) is based on imaging rather than on surface anatomical landmarks. Also, cannulating the ventricle may be safer when using a navigation-calibrated peel-away. Once inside the lateral ventricle, regular ventricular anatomical structures lead the surgeon to the correct orientation. However, the septum may be thicker than in chronic hydrocephalus, and the septum superior–inferior span may be hard to estimate intraoperatively. Thus, continuous navigation with a calibrated endoscope is helpful in defining the safe location for a SP. Similarly, navigation may verify a location for an EBX, especially for subependymal tumors such as those in the presented series

- An important point to consider is whether to perform an EBX or a SBX. The advantage of an EBX is the fact that the endoscope is already in proximity to the pathological lesion. In two large series of 293 and 714 biopsies reporting on the hemorrhagic rate, the following results were noted. In the first series, moderate-to-severe intraventricular hemorrhage (IVH) occurred in 19%.[1] In the second series, 1.4% had an intratumoral hemorrhage and 2% had IVH.[5] The reason for this significant hemorrhagic rate difference may be explained by study population diversity, as well as different surgical techniques, and different tumor locations and pathologies. Regardless, these series included various ventricular pathologies, and did not focus on infiltrative BGT lesions with distorted and borderline foramen of Monro. In this specific patient subset, the safety boundaries are very slim due to the small and distorted ventricles as well as the low ventricular compliance. Tumor friability (such as in infiltrative high-grade astrocytomas) may pose an additional hemorrhagic risk. Additionally, although still a controversial observation, a sudden reduction of ICP (as happens following cannulation of the ventricles) may precipitate hemorrhage following EBX. Thus, in selected cases it would be advantageous to perform a SBX at a different setting following the CSF diversion procedure (staged procedure). The argument against a SBX would be the risk of parenchymal and tumoral hemorrhage, which could increase the pressure impact of the tumor on the ventricular system and CSF passages[4]
- CSF diversion procedures and EBX – what should be done first? We advocate starting with the CSF diversion procedure, such as the ETV and SP, and only then proceeding with EBX. Often, when cannulating the ventricles, CSF is drained and there is less space to safely maneuver the endoscope and perform the ETV or SP. As the most life-threatening aspect is associated with elevated ICP secondary to CSF accumulation, we finalize this part first, and only then recommend performing the EBX. Another reason is that often EBX are associated with some ventricular bleed. This bleed, although usually nonsignificant, may hamper safe maneuvers and performing the ETV or SP.

It is important to state, that even if an EBX is not performed at the same setting as the CSF diversion procedure, for instance because of a collapse of the ventricles, SBX may be done at a later stage.

Limitations

The rarity of large deep-BGT tumors, anatomical variability, and the retrospective nature of the data collection limit the ability to reach any solid conclusions regarding optimal management of these tumors. This limited series demonstrates the potential role of endoscopic surgery, analyzing the various surgical nuances and highlighting the specific anatomical distortions caused by these tumors.

CONCLUSIONS

Endoscopic surgery may potentially play a significant role in the initial management of patients with large basal ganglia and thalamic tumors causing obstructive hydrocephalus. As opposed to posterior thalamic tumors that compress the posterior third ventricle and the aqueduct, basal ganglia and large thalamic tumor cause significant ventricular distortion. Thus, endoscopic procedures may be associated with significant complications and morbidity that should be considered by the surgeon and patient. Technical nuances and individualization of surgical goals are crucial for reducing perioperative risks and optimizing patient outcome.

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