Brain Tectal Tumors: A Flexible Approach

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BACKGROUND AND IMPORTANCE: Mesencephalic tectal gliomas represent a subset of midbrain tumors, which are more frequent in children than in adults. They usually become symptomatic when causing hydrocephalus by occluding the aqueduct. Because of their slow progression, due to their benign histology, they are characterized by a relatively good prognosis, although hydrocephalus might jeopardize patients' prognosis. Treatment is usually represented by cerebrospinal fluid diversion associated or not with biopsy.

CLINICAL PRESENTATION: We report 2 illustrative cases of tectal gliomas in adults where endoscopic third ventriculostomy (ETV) and simultaneous endoscopic biopsy were obtained during the same operation by means of a single burr hole with a flexible endoscope.

CONCLUSION: We recommend using this overlooked neurosurgical tool for such cases, since it allows the surgeon to safely perform an ETV, then judge whether biopsy can be done or not, without harming the patient, and possibly achieving an important piece of information (histopathological diagnosis) to manage this subset of oncological patients.

KEY WORDS: Endoscopic third ventriculostomy, Flexible neuroendoscopy, Tectal plate gliomas

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Brainstem tumors represent 10% to 20% of paediatric and 1% of adult brain tumors.[1-3] A distinct subgroup of brainstem tumors is represented by tectal plate gliomas (TPG), involving the quadrigeminal plate of the midbrain. TPG usually present with hydrocephalus, focal neurological deficits being rare. TPG may be treated with surgical excision,[4-7] radiotherapy,[8] or stereotactic radiosurgery.[9] However, it has been shown that many TPG may remain stable for several years,[10] since the majority are low-grade astrocytomas.[11] Thus, a more conservative strategy has been recommended, namely long-term follow-up by means of serial magnetic resonance imaging (MRI).[2] Surgery becomes mandatory when cerebrospinal fluid (CSF) circulation is impaired; endoscopic third ventriculostomy (ETV) represents the standard option, although ventriculoperitoneal shunting might also be possible.[12] In this regard, we present 2 cases of TPG to underlie the role of flexible neuroendoscopy to simultaneously perform—if deemed safe—a biopsy right after ETV, with a single procedure.

ABBREVIATIONS: CSF, cerebrospinal fluid; **ETV,** endoscopic third ventriculostomy; **MRI,** magnetic resonance imaging; **TPG,** tectal plate gliomas

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CLINICAL PRESENTATION

Case A

A 24-yr-old man presented with stabbing headache, followed by amaurosis fugax lasting a few seconds; headaches become more frequent during the following week. A contrast-enhanced MRI of the brain revealed a non-communicating, triventricular hydrocephalus consequent to a non-enhancing small rounded cystic lesion, (containing a CSF-similar liquid), hypointense in T1 and hyperintense in T2; the lesion displaced the tectal plate, thus occluding the Sylvian aqueduct (Figure 1). Neurological examination was normal. The patient underwent ETV followed by a biopsy with a flexible endoscope (Karl Storz GmbH, Tuttlingen, Germany); a small bioptic sample was obtained from the tumor (see below); tumor histology was low-grade glioma. At the 3-mo follow-up MRI ventricles sizes were normal, the lesion remained stable, and all symptoms disappeared.

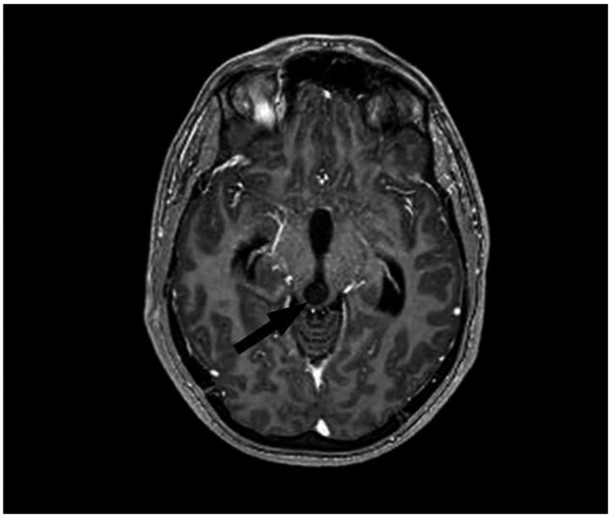

FIGURE 1. *Axial T1 MRI scan of patient #1; the arrow's tip indicates where the tectal lesion is. Please note the enlarged ventricles' size.*

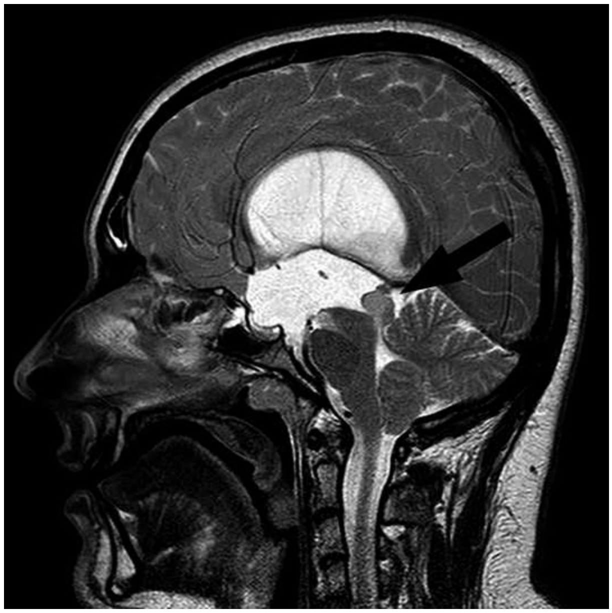

FIGURE 2. *Sagittal T2 MRI scan of patient #2; the arrows' tip shows the tumor originating in between the superior and the inferior colliculi.*

Case B

A 25-yr-old man presented urinary and fecal incontinence along with postural instability and dysaesthesias of lower limbs. Brain MRI revealed a triventricular hydrocephalus, consequent to a non-enhancing tectal lesion, hypointense in T1 and hyperintense in T2 (Figure 2). The patient underwent ETV with the flexible endoscope. The aditus aqueducti was seen, but the tumor was not reachable since it grew inside the aqueduct while no abnormal tissue was visible on the surface of the tectum. Thus, biopsy was deemed to be unsafe and surgery ended. Three months later the patient reported an improvement of his preoperative symptoms (likely due to the resolution of the hydrocephalus), and the MRI showed no tumor progression with still slightly enlarged ventricles.

Operative Procedure

The operative procedure is shown in the **Video, Supplemental Digital Content**. Patients expressed written consent to surgery and to using their anonymized data for publication. Institutional Review Board approval was not necessary, since we performed standard surgical procedures that are indicated for such neurosurgical conditions. A slightly curved incision is made (patient under general anesthesia, supine position, neuronavigation [BrainLab AG, München, Germany]) on the Kocher point (right-hand side). A burr hole is performed, then the dura is opened and a flexible endoscope is introduced into the right lateral ventricle. Then the endoscope is pushed into the third ventricle through the Monro's foramen. The region between the mammillary bodies and the infundibulum is coagulated and the floor of the third ventricle is fenestrated. The fenestration is enlarged with a 2F Fogarty balloon (Edwards Lifesciences LLC, Irvine, California). The flexible endoscope's tip is then oriented backward towards the aqueduct (Figure 3). The lesion protruded from the inside of the aqueduct (lamina quadrigemina), dilating the aditus. A biopsy is performed by means of dedicated forceps (specimens are sent for histopathological analysis).

DISCUSSION

TPG in adults are rare entities, accounting only for a small percentage of central nervous system tumors.[1-3] However, because it causes hydrocephalus, TPG have been described as "the smallest tumor […] that can lead to death."[13] Focal deficits are rarely observed and are mostly reversible after CSF diversion.[14,15] The typical appearance on an MRI is tectal fullness, or a focal tumor in the tectum of the midbrain, isointense/hypointense on T1-weighted and hyperintense on T2-weighted images, and no enhancement following Gadolinium-DTPA administration.[2,16]

Historically, the management of TPG has followed a conservative approach, due to the generally indolent nature of these tumors, which sometimes can behave more aggressively despite of their radiological appearance.[10] Therefore, the primary goal was to minimize the consequences of increased intracranial pressure by diverting CSF. ETV has proven to be highly effective for the control of hydrocephalus in these patients, with a success rate ranging around 95%,[17] with a low morbidity and mortality, together with lower chances of re-obstruction compared to shunting.[2] Surgical resection is not recommended; it might

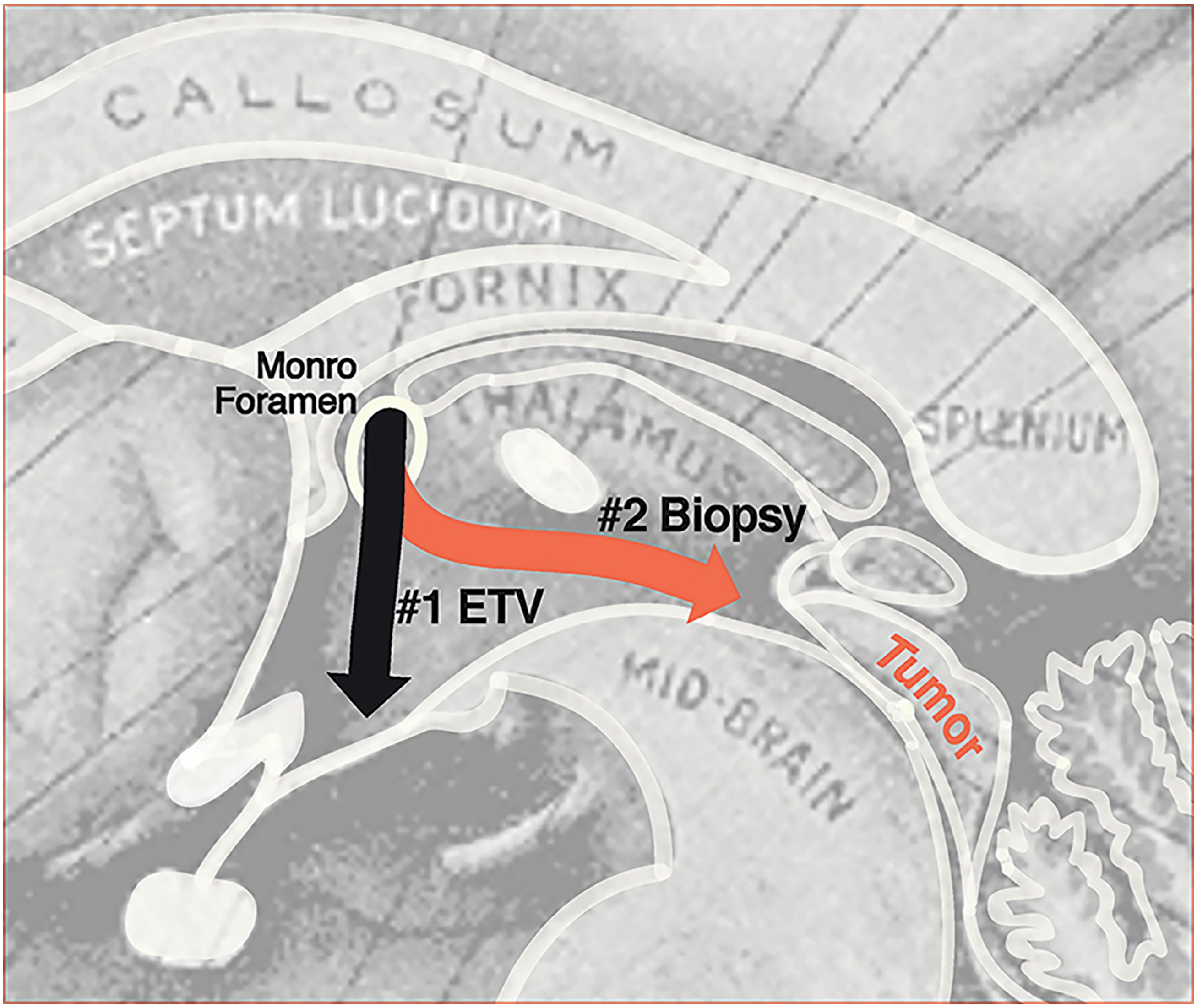

FIGURE 3. *Scheme of an endoscopic procedure tailored for tectal lesions that determine triventricular hydrocephalus; a thirdventriculostomy is performed with a flexible endoscope (#1), then the tip of the instrument is directed towards the aqueduct to attempt a safe tumor's biopsy (#2). Image modified from Gray's Anatomy, http://www.bartleby.com/107/illus715.html, Public Domain.*

become indicated when the tumor demonstrates growth in size or if a malignant, secondary, or vascular lesion is suspected.[14] Fractioned external beam radiotherapy and SRS were also proposed as therapeutic alternatives (Table), yet with some side effects.[9,18,19]

Although in children, the typical MRI pattern is sufficient to diagnose TPG, in adults, since this tumor is very rare, a histological diagnosis might be important.[11] Obtaining such biopsies is often difficult as the tumor may not be visible at the time of the endoscopy and may be covered by the ependymal lining of the aqueduct; it could also be argued that the biopsy may lead to hemorrhage and therefore added morbidity and mortality.[14] Yet in a large multicentric series, neuroendoscopic biopsies provided meaningful pathological data for over 90% of patients, for a wide range of tumor types, with an overall fairly low—and mostly reversible—complication rate of <13% (in 293 procedures): there was a 5% of severe bleeding, with a single case of death (<0.5%).[20]

Different endoscopic approaches were proposed (see Table): since this approach to the third ventricle is limited by the foramen

TABLE. Literature Review of TPG and How They are Treated

Author	Patient #	Sex	Age (yr)	Surgical treatment first-line	Surgical treatment second-line	Postoperative complications	Flexible/rigid endoscope	Histopathology
Yeh et al[10]	1	–	23	None	–	–	–	–
	2	–	56	None	–	–	–	–
	3	–	64	VP shunt	–	–	–	–
	4	–	46	ETV + biopsy	–	–	–	Pilocytic astrocytoma
	5	–	69	VP shunt	–	–	–	–
Javadpour et al[13]	6	F	9	ETV	Second ETV VP shunt	None	Flexible	–
	7	F	9	ETV + biopsy	None	None	Flexible	Low-grade
	8	F	11	ETV + biopsy	Second ETV VP shunt	Wound CSF leak (resutured)	Flexible	Nonspecific
	9	M	12	ETV	Second ETV	None	Flexible	–
	10	M	16	ETV	None	None	Flexible	–
	11	M	19	ETV	Second ETV	None	Flexible	–
	12	F	24	ETV + biopsy	None	None	Flexible	Probably low-grade
	13	M	27	ETV	None	None	Flexible	–
	14	F	42	ETV + biopsy	None	None	Flexible	Low-grade
	15	M	56	ETV	None	None	Flexible	–
	16	M	59	ETV	None	None	Flexible	–
Mizoguchi et al[26]	17	M	24	ETV + biopsy	None	None	–	Low-grade glioma
Stark et al[14]	18	M	6	Resection, shunt, radiotherapy	ETV	–	Rigid	Ependymoma
	19	F	16	Shunt	–	–	–	–
	20	F	12	Shunt	ETV	–	Rigid	–
	21	F	8	Shunt	Resection	–	–	Low-grade astrocytoma
	22	F	3	Resection, shunt	ETV + biopsy	–	Rigid	Low-grade astrocytoma
	23	M	11	ETV + biopsy	–	–	Rigid	Astrocytoma (WHO I)
	24	M	6 wk	Shunt	–	–	–	–
	25	F	4 wk	Shunt	ETV	–	–	–
	26	F	11 mo	ETV + biopsy	ETV revision	–	Rigid	Low-grade astrocytoma
	27	M	3	ETV	ETV revision	–	Rigid	
	28	F	6 mo	ETV	–	–	Rigid	
	29	F	6 wk	ETV, EAP	Shunt	–	Rigid	
Oka et al[11]	30	F	8	ETV + biopsy	–	None	Flexible	Glial tissue
	31	M	11	ETV + biopsy	–	None	Flexible	Low-grade Glioma
	32	M	14	ETV + biopsy	–	None	Flexible	Low-grade glioma
	33	F	21	ETV + biopsy	–	None	Flexible	Low-grade glioma
	34	F	22	ETV + biopsy	–	None	Flexible	Low-grade glioma
	35	F	23	ETV + biopsy	–	None	Flexible	Low-grade glioma
	36	M	24	ETV + biopsy	–	None	Flexible	Low-grade glioma
	37	F	48	ETV + biopsy	–	None	Flexible	Low-grade glioma
	38	F	24	ETV + biopsy	Radiotherapy	None	Flexible	Anaplastic astrocytoma
	39	F	50	ETV + biopsy	Radiotherapy, ultrasonic endoscopic aspiration	None	Flexible	Anaplastic astrocytoma
	40	F	73	ETV + biopsy	–	None	Flexible	Anaplastic astrocytoma
Constantini et al[20]	41-79	–	–	Biopsy (±ETV)	–	Overall complication ratio 13%	–	–

of Monro, working at the aqueductal region requires modifications to the standard ETV procedure, namely: working with 2 entry burr holes for 2 separate trajectories;[21] working with a single "compromised" burr hole for both the ETV and the biopsy;[22] or using a flexible endoscope.[11,23,24] Flexible endoscopy has already proven to be very effective in diverting CSF (by means of ETV) and obtaining a tumor sample in the context of pineal region tumor surgery; moreover, when feasible, such tumors can also be partially suctioned through the endoscope's operative channel, allowing for a sufficient tumor debulking.[25]

Nowadays, the use of flexible endoscopes is not popular in neurosurgery, since rigid instruments present many advantages (ie, better visualization, multiple operative channels).[26] On the other hand, we believe flexible endoscope still represents the best tool to approach this kind of lesion as explained with these reported cases; like an eel slithering through seaweeds and corals, the flexible scope is a harmless tool that allows the surgeon to reach virtually any site within the ventricles, perform a range of procedures, and allow a judicious call about the safety of other—more risky—maneuvers. In particular, an ETV can easily be performed and—when possible and safe—a tumor biopsy can be done, through a single burr hole. The flexible scope does not cause damage to the brain parenchyma (small diameter), no damage to the columns of the fornix, or other delicate inner brain structures; even in an expert's hands, a rigid tool inserted through the brain and inside the ventricles cannot be bent or moved along the 2 orthogonal space directions unless damaging those very delicate structures. On the other hand, a flexible and bendable tool is more fit to safely enter and navigate the cerebral ventricles. It is important to underlie that resolving the hydrocephalus (followed by radiological follow-up) represents the pillars of TPG management. In the first case, we safely performed a biopsy; in the second case we did not biopsy the tumor because the lesion was mainly expanding into the aqueduct and no abnormal tissue was visible on the intraventricular surface of the tectum. In both cases EVT was performed with success and no complications, relieving hydrocephalus' symptoms; patients are still doing fine and TPG are stable in both cases (follow-up: ∼24 mo).

CONCLUSION

TPG represent a subset of brainstem gliomas, which are typically indolent; their management is not yet standardized. The main goal of the neurosurgeon should be to timely treat hydrocephalus by establishing an alternative CSF flow (eg, ETV), then to histologically define the nature of the lesion, which is a key step to manage such patients. We presented a reappraisal of flexible neuroendoscopy as a safe, minimally invasive approach to this neurosurgical condition. We would like to remind neurosurgeons that flexible endoscopy could be an option to safely perform an ETV, with the advantage of doing a tumor biopsy—when feasible—through a single burr hole. This can reduce both the physician's and the patient's anxiety, by adding an important piece of information to take care of those brain tumor patients where surgical removal is never an option.

Disclosure

The authors have no personal, financial, or institutional interest in any of the drugs, materials, or devices described in this article.

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Supplemental Digital Content. Video. Endoscopic thirdventriculostomy and biopsy of a lesion originating from the lamina quadrigemina; a flexible endoscope allows the surgeon to perform both steps of the procedure with a single burr-hole and a single trajectory through the brain cortex.

COMMENT

This paper illustrates the utility of flexible endoscope in neurosurgery, which appears to be less appreciated at present. The essence of the authors' strategy is potentially applicable to many other disease conditions related to the ventricles. The caveat, though, is how to maintain its minimal invasiveness when unexpected bleeding occurs during procedure.

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