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ORIGINAL PAPER

Endoscopic third ventriculostomy for hydrocephalus in brainstem glioma: a case series

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Abstract
Object A brainstem glioma is an incurable brain tumor that can be complicated by hydrocephalus. A ventriculoperitoneal (VP) shunt is generally performed for the control of hydrocephalus. This study aimed to reveal the safety and efficacy of an endoscopic third ventriculostomy (ETV) for hydrocephalus in brainstem gliomas.
Methods Six patients who had pontine glioma with hydrocephalus underwent an ETV between May 2010 and November 2015. In all the cases, there were one or more symptoms of hydrocephalus (headache, nausea, vomiting, or lethargy). Retrospective review of these patients was performed using the medical records and neuroimagings.
Result The ETV was performed safely and there were no intraoperative complications in all patients. The mean follow-up period was 12.3 months. An immediate symptomatic relief of hydrocephalus and an adequate control of symptoms were achieved without a VP shunt in all patients.
Conclusions The ETV is considered to be an effective and safe procedure for the treatment of hydrocephalus in brainstem gliomas. Determining the ventriculostomy site according to the preoperative MRI in each case is considered to be important for the safe procedure.

Keywords Endoscopic third ventriculostomy · Brainstem glioma · Hydrocephalus · ETV

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Introduction

A brainstem glioma is an incurable brain tumor that can be complicated by hydrocephalus. Generally, the brain stem is pushed anteriorly against the clivus by the tumor and the prepontine cistern is narrowed, which can pose a greater risk for an endoscopic third ventriculostomy (ETV) [1]. Therefore, a ventriculoperitoneal (VP) shunt is usually performed for the control of hydrocephalus. Here, we report six cases of brainstem glioma with hydrocephalus treated by endoscopic third ventriculostomy (ETV) without any complications and try to elucidate the efficacy of the procedure.

Methods

We retrospectively reviewed the hospital charts, surgical reports, and imaging tests of six patients having pontine glioma with hydrocephalus. They underwent ETV at the Department of Neurosurgery, National Center for Child Health and Development, between May 2010 and November 2015. We reviewed patient characteristics, including age, sex, symptoms, symptom resolution, dissemination, reoperation (including additional shunts), and follow-up periods.

Regarding the surgery, a frontal approach was selected for all patients. In five cases, nondominant side was selected. In case 3, the ETV procedure was performed through the left burr hole, because we planned to fenestrate just adjacent to the right side of the floor of the third ventricle. All the procedures were performed using a flexible endoscope. The ventriculocisternostomy was made using the forceps in all cases then the stoma was enlarged by inflating a French #3 Fogarty balloon catheter. No or little ventricular bleeding is expected, and it is easily controlled by gentle irrigation with normal saline solution at body temperature. The catheter,

which was connected to the subgaleal Ommaya reservoir, was inserted the lateral ventricle in all cases.

All patients received additional chemotherapy and/or radiotherapy after the ETV. Magnetic resonance imaging (MRI) was performed about 1 week after the operation to check the function of ETV. At the time of the end of additional therapy and worsening of the neurological symptom, CT or MRI was performed to assess the efficiency of the therapy and the size of the ventricles.

Result

The study patients were treated at the National Center for Child Health and Development. The clinical details of this cohort are provided in Table 1. Five patients were diagnosed with diffuse intrinsic pontine glioma while one patient was diagnosed with secondary glioma after undergoing radiotherapy for an ependymoma in the fourth ventricle. The average age at surgery was 6.1 years. The mean and median follow-up periods were 12.3 and 8 months, respectively. In all cases, there were one or more features of symptomatic hydrocephalus (headaches, nausea, vomiting, or lethargy), and hydrocephalus was documented in the preoperative imaging studies. All obstructive hydrocephalus patients with pontine glioma were treated by ETV.

In five cases, the area of the prepontine cistern was narrowed, but there was a subarachnoid fluid space in the upper part of the cistern (Fig. 1a). In these cases, the ventriculostomy was performed with the forceps at the midline between the mammillary bodies and the infundibulum. The forceps was advanced toward the clivus in order to avoid the injury to the brainstem or the basilar artery.

In one case, the prepontine cistern was occupied by the tumor and the subarachnoid space was present only in the right side of the cistern. In this case, we made ventriculostomy at the right side of the third ventricle floor (Figs. 2 and 3).

In all patients, the ETV was performed without any intraoperative complications, and an immediate symptomatic relief of hydrocephalus was achieved. Immediate postoperative imaging of all the cases showed a decrease in ventricular size. The mean follow-up period was 9 months (range 1–36 months). A VP shunt was not needed in all patients at the last follow-up. In five cases, we could show the flow void in the third ventricle in the follow-up MRI. Five patients were still alive at the last follow-up.

Illustrative case

Case 1

A 2-year-old boy presented with lethargy. Head CT scan revealed a low-density area in the pons and dilated ventricles. MRI showed a T2-hyperintense lesion in the pons and dilated ventricles. An emergency ETV was performed, and the patient condition immediately improved. Temozolomide and local radiotherapy (total 54 Gy) were given. The follow-up period was 39 months; he had no symptoms of hydrocephalus. Although the residual tumor increased, the MRI, which was taken 36 months after the surgery, showed the flow void and the opening of the ventriculostomy site (Fig. 1).

Case 3

A 4-year-old girl was referred to our hospital with a history of fatigue and diplopia for 1 month. Brain MRI demonstrated an expansive, T2-hyperintense pontine mass. It was the typical history and imaging of a DIPG, and thus, radiotherapy without biopsy was planned. However, before starting the radiotherapy, vomiting and lethargy were observed and a CT scan revealed progressively dilated ventricles. The tumor had expanded to the left prepontine cistern, and the subarachnoid fluid space was only found at the right side of the cistern. We planned to fenestrate just adjacent to the right side of the floor of the third ventricle. An emergency ETV was performed, and the tumor was observed through the opening site of the ETV. Simultaneously, the biopsy was performed without profuse bleeding or injury to the pyramidal tract. After the surgery, vomiting and lethargy resolved immediately. The biopsy revealed anaplastic astrocytoma. She received

Table 1 Patients' characteristics on operation

Case no.	Age	Sex	Symptom	Diagnoses	Metastasis	Treatment	Follow-up (month)
1	2	M	Lethargy	Low-grade glioma	N	R + C	31
2	8	M	Headache, nausea	Diffuse astrocytoma	N	R + C	6
3	6	F	Lethargy	High-grade glioma	N	R + C	5
4	4	F	Vomiting, nausea	Low-grade glioma	N	R	7
5	8	M	Headache, nausea	Fibrillary astrocytoma	N	R	6
6	9	F	Lethargy	N	N	R	1

F female, *M* male, *N* none, *R* radiation therapy, *C* chemotherapy

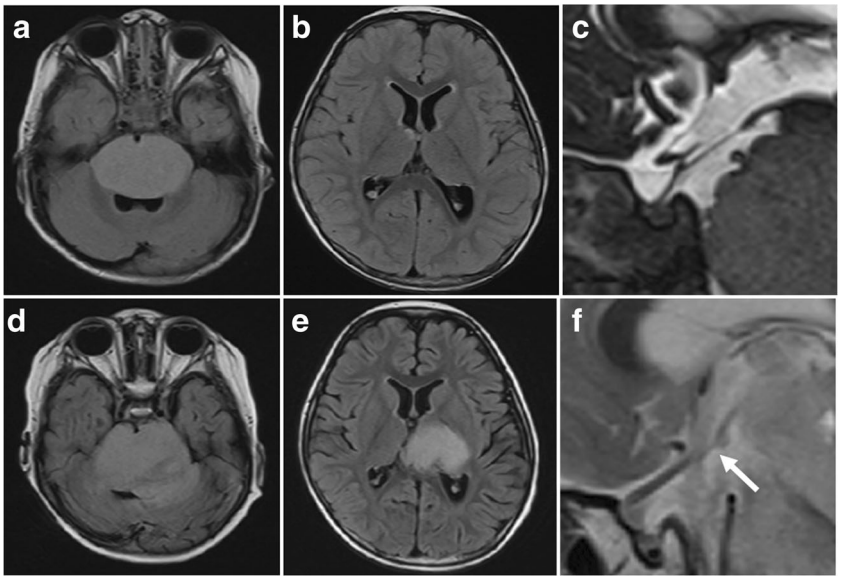

Fig. 1 MRI at presentation showed the hydrocephalus and brain stem glioma in case 1. **a**–**c** MRI taken 36 months after the ETV showed no worsening of ventriculomegaly (**d**–**f**). The flow void at the fenestration site was demonstrated (*arrow*)

chemotherapy (cyclophosphamide and vincristine) and local radiotherapy (total dose of 54 Gy). The follow-up period was 5 months. She had been without worsening of the hydrocephalus (Fig. 2).

Discussion

Brainstem glioma is a primary glial neoplasm that accounts for 10 to 15 % of all the childhood brain tumors [2]. Tumor progression or dissemination may lead to cerebrospinal fluid (CSF) pathway obstruction and development of hydrocephalus. Therefore, many patients will develop hydrocephalus during the course of the disease [3–5].

The ETV has become the alternative method for the treatment of obstructive hydrocephalus [6, 7]. Potential complications of the ETV include injury to the basilar artery, the cranial nerves, or the brainstem [8]. The prepontine cistern of patients with a brainstem glioma is usually very narrow due to the tumor. So far, it is considered that there can be a greater risk of injury.

In our series, all patients underwent the ETV safely. Preoperative imaging is considered to be important for determining the ventriculostomy site. In five cases, the area of the prepontine cistern was narrowed, but there was a

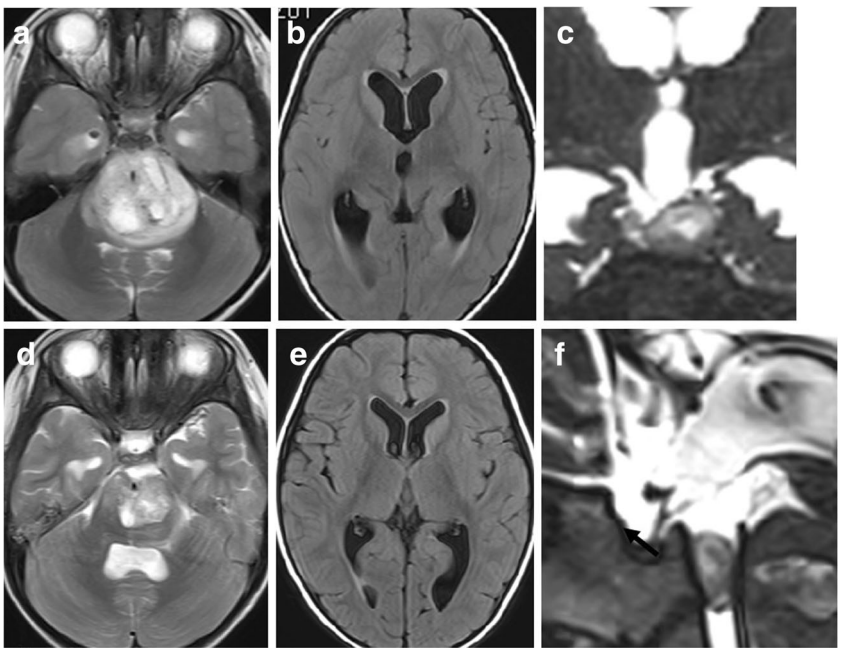

Fig. 2 MRI at presentation showed the hydrocephalus and brain stem glioma in case 3 (**a**–**c**). The subarachnoid space of the prepontine cistern was only seen at the left side. MRI taken 3 months after the ETV showed improvement of hydrocephalus (**d**–**f**)

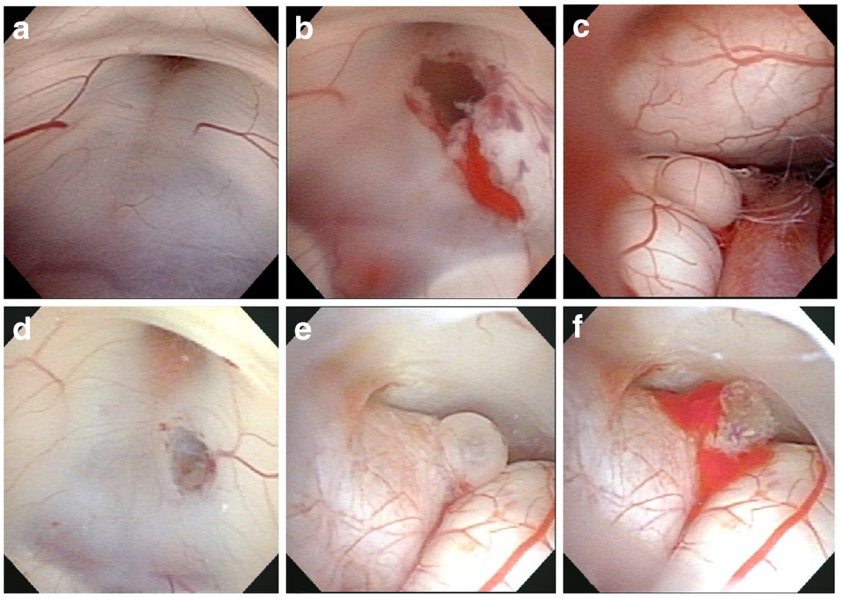

Fig. 3 Intraoperative imaging in case 1 (**a**–**c**) and case 3 (**d**–**f**). In both cases, ETV was performed safely. In case 3, we opened just the right side of the third ventricle floor. Then, the tumor was demonstrated in the ventral side of the pons through the fenestration site. The biopsy of the tumor was also performed safely

subarachnoid fluid space in the upper part of the cistern. In these cases, the ventriculostomy was performed with the forceps at the midline between the mammillary bodies and the infundibulum. The forceps was advanced toward the clivus in order to avoid the injury to the brainstem or the basilar artery. In one case, the tumor expanded anteriorly with left predominance. The prepontine cistern was occupied by the tumor, and the subarachnoid space was present only in the right side of the cistern. In this case, we made ventriculostomy at the right side of the third ventricle floor in order to avoid penetration of the tumor or injury to the basilar artery. Thus, the ETV can be an effective and safe procedure also for these patients.

A VP shunt is another method for the treatment of hydrocephalus [9]. However, infection and obstruction of the shunt are well-known and relatively frequent complications of this method. In addition, hydrocephalus with posterior fossa tumor may entail other complications such as upward herniation, hematomas in the tumor, and metastasis through the VP shunt [10–14]. El-Ghandour et al. compared ETV and VP shunt as a treatment for hydrocephalus in posterior fossa tumors: the patients treated by VP shunt had a greater risk of upward herniation and intratumoral hemorrhage [15]. In addition, four cases of a brainstem glioma were reported to have metastasis through the VP shunt [10, 16–18]. In some cases, additional treatment for metastasis to the peritoneal cavity was needed.

At the time of this review, all but one patient had died of the disease. None of the patients had a closure of the ETV site and required the placement of a VP shunt. All the patients had an immediate relief of the symptoms of increased intracranial pressure. The ETV is considered to be an effective and safe procedure for the treatment of hydrocephalus in these tumors.

Conclusions

The ETV is considered to be an effective and safe procedure for the treatment of hydrocephalus in brainstem gliomas. Determining the ventriculostomy site according to the preoperative MRI in each case is considered to be important for the safe procedure.

Acknowledgments We would like you to review our paper " Endoscopic third ventriculostomy for hydrocephalus in brainstem gliomas." This paper mentioned our experience of the endoscopic third ventriculostomy for hydrocephalus in brainstem glioma and some insight for the management of hydrocephalus in these patients.

Compliance with ethical standards We declare that the manuscript has not been previously published in whole or in part or submitted elsewhere for review.

Conflict of interest There are no conflicts of interest.

Disclosure There is no financial disclosure.

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