Involution of enhancing intrinsic tectal tumors after endoscopic third ventriculostomy

Report of two cases

AHMED M. ALKHANI, M.D., FREDERICK A. BOOP, M.D., AND JAMES T. RUTKA, M.D., PH.D., F.R.C.S.(C)

Division of Neurosurgery, The Hospital for Sick Children, University of Toronto, Toronto, Ontario, Canada; and Division of Pediatric Neurosurgery, Arkansas Children’s Hospital, University of Arkansas, Little Rock, Arkansas

For benign intrinsic tectal tumors causing triventricular obstructive hydrocephalus, cerebrospinal fluid diversion followed by neuroimaging is a widely accepted treatment plan. In this report, the authors describe two children with focal enhancing tectal lesions that caused acute, symptomatic hydrocephalus. One child had neurofibromatosis Type 1 (NF1). In both children the hydrocephalus was effectively treated by endoscopic third ventriculostomy. Following this procedure, serial imaging studies revealed not only that the ventriculomegaly had resolved, but also that the enhancing tectal tumors had regressed and disappeared over time. The time to complete involution of these tumors was 18 months for the child with NF1 and 12 months for the other child. To the authors’ knowledge, this is the first report of the involution of enhancing tectal tumors after endoscopic third ventriculostomy. The possible mechanisms for this unexpected result are discussed.

KEY WORDS • astrocytoma • tectal tumor • endoscopic third ventriculostomy • endoscopy • involution • children

BRAINSTEM TUMORS are a heterogeneous group of lesions with respect to their clinical and histopathological features and biological behavior. Astrocytomas are the most common form of intrinsic brainstem lesion and account for some 10 to 20% of pediatric brain tumors. The classification of brainstem astrocytomas has been revised since the advent of magnetic resonance (MR) imaging. Use of MR imaging has enabled neurosurgeons to recognize several different brainstem astrocytomas in children, including focal midbrain, diffuse and focal pontine, dorsally exophytic brainstem, and cervico-medullary and benign intrinsic tectal astrocytoma subtypes. With the exception of the diffuse intrinsic and some focal pontine astrocytomas, brainstem astrocytomas in children are usually benign low-grade neoplasms.

Since the description of tectal astrocytomas as “pencil gliomas” by Sanford, et al., benign intrinsic tectal tumors have come to be recognized as indolent, slow-growing lesions that occlude the aqueduct of Sylvius, producing obstructive triventricular hydrocephalus. In the majority of children presenting with obstructive hydrocephalus in whom a small, tectal neoplasm is identified, cerebrospinal fluid (CSF) diversion alone suffices in treating the patient’s symptoms. In recent times, endoscopic third ventriculostomy has become an effective means by which aqueductal stenosis that causes hydrocephalus can be overcome without resorting to the implantation of permanent CSF shunting devices. Success rates as high as 70% have been reported for long-term control of hydrocephalus after endoscopic third ventriculostomy. In this report, we describe two children in whom acute obstructive hydrocephalus was caused by enhancing tectal tumors. Endoscopic third ventriculostomy was performed in both children, leading to the resolution of the acute obstructive hydrocephalus. Interestingly, after endoscopic third ventriculostomy we observed over time involution and disappearance of the enhancing tectal tumors in these patients. Although rare, the involution of low-grade astrocytomas in children has been described previously. However, to our knowledge, the two cases reported here represent the first in which the involution of enhancing tectal tumors was associated temporally with endoscopic third ventriculostomy. The possible mechanisms for this unexpected result after endoscopic third ventriculostomy in these two children are discussed.

Illustrative Cases

Case 1

History. This 9-year-old girl presented to the Neurosurgery Service at The Hospital for Sick Children with a 3-
month history of progressive, intermittent headaches. She had been born at 37 weeks’ gestation following an uncomplicated pregnancy. At 2 years of age, she was found to have neurofibromatosis Type 1 (NF1) after the discovery of multiple café-au-lait spots on her body trunk. A computerized tomography scan performed when she was 2 years old demonstrated a small left-sided intraorbital and intracanalicular optic glioma. No other lesions were noted, and the ventricles were small. When the patient was 7 years old, T2-weighted MR imaging of the brain revealed several small hyperintense lesions in the white matter of the temporal and frontal lobes and within the cerebellum.

At 9 years of age, she presented with increasingly frequent headaches, and examination revealed blurring of disk margins and decreased visual acuity in her left eye (20/60). The acuity in the right eye was normal, and results of the rest of the neurological examination were unremarkable. An MR image revealed a 2 × 1-cm irregularly shaped lesion in the tectum that enhanced on contrast administration (Fig. 1). The aqueduct of Sylvius was obliterated, and acute obstructive hydrocephalus was observed (Fig. 2).

Operation and Postoperative Course. Because of the patient’s symptoms of headaches and decreased visual acuity, endoscopic third ventriculostomy was performed. Using a rigid endoscope (Storz and Co., Tuttingen, Germany) according to previously published techniques,6 the floor of the third ventricle was perforated. After endoscopic third ventriculostomy and before withdrawing the endoscope, the posterior portion of the third ventricle was inspected, and a lesion in the tectal region was visualized but no biopsy sample was obtained. Postoperatively, the patient recovered well, and her headaches disappeared. A 6-month follow-up MR image revealed a marked diminution in ventricular size, indicating a patent and functional ventriculostomy (Fig. 2). In addition, serial MR images obtained at 6-month intervals have shown a progressive decrease in the size of the enhancing tectal tumor such that the lesion could not be identified 18 months after endoscopic third ventriculostomy (Fig. 3). Now, 3 years after the procedure, the patient is well aside from mild weight gain. A complete endocrinological workup revealed that her pituitary hormone levels are all normal. The tectal tumor has not recurred.

Case 2

History. This 11-year-old right-handed girl had a long-standing history of headaches. One month before admission, the headaches had become progressively worse and were associated with photophobia and diplopia. She was referred to the Arkansas Children’s Hospital for further evaluation. Her medical history was unremarkable, and she had been doing well in school.

Examination. Results of the general physical examination of the patient were unremarkable. No café-au-lait spots or cutaneous lesions were found. A neurological examination revealed bilateral papilledema with flame-shaped retinal hemorrhages. The results of the rest of the cranial nerve examination, as well as motor, sensory, and cerebellar testing, were all normal. An MR image of the brain revealed a tectal tumor that enhanced with gadolinium administration (Fig. 4). Acute obstructive hydrocephalus...
Disappearing tectal tumors after endoscopic third ventriculostomy

Fig. 4. Case 2. Magnetic resonance images obtained before endoscopic third ventriculostomy. a: Sagittal T₁-weighted MR image demonstrating enlarged ventricular system and hypointense mass within the tectum (arrow). b: Sagittal T₁-weighted gadolinium-enhanced MR image revealing enhancing lesion in the tectum. c: Axial T₁-weighted gadolinium-enhanced MR image demonstrating small enhancing tumor nodule predominantly in the left tectum with obstructive hydrocephalus involving the third and lateral ventricles. The tumor is causing mass effect and distortion of the collicular plate.

Fig. 5. Case 2. Follow-up MR images obtained 12 months after endoscopic third ventriculostomy. a: Sagittal T₁-weighted MR image with gadolinium enhancement revealing complete regression of enhancing tectal tumor and reduction in size of the ventricular system. b: Axial T₁-weighted MR image revealing restoration of normal tectal anatomy, visualization of the aqueduct of Sylvius, and the opening in the floor of the third ventricle (arrow).

Discussion

We have described two patients in whom enhancing tectal tumors regressed after endoscopic third ventriculostomy for acute obstructive triventricular hydrocephalus. Although the spontaneous involution of low-grade astrocytomas in childhood has previously been the subject of some controversy, many cases with adequate histopathological documentation following biopsy or subtotal resection of the primary tumor have now been reported in the literature. Although focal enhancing lesions are rare in NF1, Morris, et al., described the case of a child with NF1 in whom an enhancing lesion within the basal ganglia regressed spontaneously over the course of 3 years.

The brainstem is a common site of involvement in patients with NF1. Between 5% and 10% of patients with NF1 will show evidence of brainstem involvement on MR imaging. Pollack, et al., described four types of brainstem lesions in patients with NF1: 1) diffuse, poorly circumscribed lesions involving the entire brainstem that are hyperintense on T₂-weighted MR images; 2) focal enhancing brainstem masses; 3) sharply marginated, nonenhancing lesions on T₁-weighted MR images; and 4) intrinsic tectal tumors. Of the five patients with intrinsic tectal tumors in their series, four required only CSF diversion, are high-signal-intensity foci seen on repetition-time MR images. These foci are found in approximately two thirds of patients with NF1 and are found most commonly in the basal ganglia, cerebellum, internal capsule, and brainstem. These lesions have been called “focal areas of signal intensity” or “unidentified bright objects” and correspond to pathological findings of areas of vacuolar or spongiotic change. Although focal contrast-enhancing lesions are rare in NF1, Morris, et al., described the case of a child with NF1 in whom an enhancing lesion within the basal ganglia regressed spontaneously over the course of 3 years.

The brainstem is a common site of involvement in patients with NF1. Between 5% and 10% of patients with NF1 will show evidence of brainstem involvement on MR imaging. Pollack, et al., described four types of brainstem lesions in patients with NF1: 1) diffuse, poorly circumscribed lesions involving the entire brainstem that are hyperintense on T₂-weighted MR images; 2) focal enhancing brainstem masses; 3) sharply marginated, nonenhancing lesions on T₁-weighted MR images; and 4) intrinsic tectal tumors. Of the five patients with intrinsic tectal tumors in their series, four required only CSF diversion,
and there was stabilization of disease at a median follow-up of 4 years. 37 Pollack and colleagues 38 concluded that NF1-associated intrinsic tectal tumors in patients with this syndrome behaved similarly to benign tectal tumors in patients without NF1. In contrast to the child with NF1 whom we present in this report, enhancing tectal tumors were not found in the large series published by Pollack, et al., 37 and Bilaniuk, et al. 37 However, an enhancing pontine lesion observed on MR images obtained in a 12-year-old boy with NF1 spontaneously regressed without treatment; 31 and an enhancing right middle cerebellar peduncle lesion in an 18-month-old boy with NF1 also spontaneously decreased in size and lost contrast enhancement. 1

Although the natural history of brainstem tumors in patients with NF1 is not known with certainty because of the small numbers of patients studied, these lesions have been reported to be associated with a more favorable patient prognosis than those with similar appearances in patients without NF1. 17 Therefore, the presence of NF1 in one of our patients may have impacted favorably on the response of her tectal tumor to endoscopic third ventriculostomy. The mechanism by which NF1 leads to a more favorable prognosis is still unknown. However, the other child with a tectal tumor in this study did not have NF1 and responded to endoscopic third ventriculostomy in a very similar manner. The regression of the tectal tumors reported here may reflect decelerating growth kinetics, induction of spontaneous apoptosis within the tumor cells, 21 the development of host immune reactions, a vascular response, or a hormonal imbalance in response to endoscopic third ventriculostomy. With respect to the latter potential mechanism, one of the known complications of endoscopic third ventriculostomy is injury to the hypothalamopituitary axis following perforation of the floor of the third ventricle. 6 Although our first patient developed mild weight gain after endoscopic third ventriculostomy, a complete endocrinological workup has not uncovered any overt hormonal imbalance.

Conclusions

We report two cases of spontaneously regressing enhancing tectal tumors in children treated with endoscopic third ventriculostomy for acute obstructive hydrocephalus. Because of the delayed onset of aqueductal stenosis, these children were ideally suited for and their hydrocephalus responded well to endoscopic third ventriculostomy. These two cases illustrate the importance of obtaining serial MR images after CSF diversion in patients with enhancing mass lesions in the tectum before a commitment is made to therapies such as radiation or chemotherapy.

References


A. M. Alkhani, F. A. Boop, and J. T. Rutka

Manuscript received March 24, 1999. Accepted in final form June 17, 1999. 
Address reprint requests to: James T. Rutka, M.D., Ph.D., Division of Neurosurgery, Suite 1504, The Hospital for Sick Children, 555 University Avenue, Toronto, Ontario, M5G 1X8 Canada. Email: rutka@sickkids.on.ca.

J. Neurosurg. / Volume 91 / November, 1999