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The long-term outcome in children with late-onset aqueductal stenosis resulting from benign intrinsic tectal tumors

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Benign intrinsic tumors arising in the dorsal midbrain have long been recognized as a potential cause of late-onset aqueductal stenosis. Where histopathological studies of such lesions have been performed, the majority have been reported to be low-grade gliomas. Because these tumors often present with a paucity of neurological findings and a characteristic radiographic appearance and because there has been substantial uncertainty regarding their potential for long-term progression, the authors have routinely deferred biopsy and/or radiotherapy for these lesions until there has been clear-cut evidence of disease progression. Herein, the authors report their experience with 16 children manifesting this syndrome who were treated between 1979 and 1992. The patients ranged in age from 6 months to 14 years at presentation (median 9.75 years). In general, symptoms of increased intracranial pressure developed insidiously; three of the older children had exhibited profound macrocephaly since infancy, which predated the onset of other symptoms of hydrocephalus by several years. Only one of the 16 children showed evidence of brain-stem dysfunction at presentation, a partial Parinaud's syndrome that resolved following placement of a ventriculoperitoneal shunt. In 12 patients, the tumor was detected by magnetic resonance (MR) imaging at initial evaluation as a bulbous enlargement of the tectal plate. In four patients who presented before the advent of MR imaging, initial computerized tomography (CT) scans failed to delineate the tectal lesion convincingly; however, subsequent MR studies clearly demonstrated the presence of an intrinsic tectal mass. All 16 patients underwent cerebrospinal fluid diversion initially, with conservative management of the tectal lesion and close long-term follow-up monitoring. Four children ultimately demonstrated clinical signs of progressive tumor growth with the insidious onset of partial or complete Parinaud's syndrome, despite the presence of a functioning shunt.

The median interval to symptom progression was 7.8 years from the time of shunt insertion and 11.5 years from the onset of initial symptoms and signs of hydrocephalus. Follow-up CT and MR studies demonstrated obvious tumor enlargement in three of the four patients who then underwent stereotactic or open biopsy. The histological diagnosis in these three was benign mixed glioma, anaplastic astrocytoma, and low-grade astrocytoma. All four patients with clinical evidence of disease progression were treated with conventional radiotherapy; the patient with an anaplastic astrocytoma also received focal stereotactic radiosurgery. These patients subsequently remained clinically stable, with three showing tumor regression and one showing stable disease on serial MR studies (median follow-up period from tumor progression, 4.25 years). One other child was noted to have progressive tumor enlargement during the 2 years after shunt insertion; she remains asymptomatic and has not yet undergone biopsy or radiotherapy.

It is concluded that benign intrinsic tectal tumors, although generally indolent, merit conscientious long-term follow-up monitoring since these lesions may ultimately show evidence of progressive growth and require therapeutic intervention to maintain disease control. These tumors are best visualized on MR imaging, which should be included in the workup of all patients with late-onset aqueductal stenosis.

KEY WORDS • **aqueductal stenosis** • **astrocytoma** • **glioma** • **hydrocephalus** • **tectum** • **children**

BRAIN-STEM gliomas account for 10% to 20% of pediatric brain tumors.[2,17,23] In recent years, it has become apparent that this broad category of neoplasms encompasses several distinct subgroups of tumors that differ significantly with regard to their growth characteristics, histology, and response to therapy.[1,2,4,10,14,16,39] Recognition of this diversity has fostered attempts to better define the long-term biological behavior of the individual tumor subgroups and to refine their management. The vast majority of brain-stem

neoplasms are infiltrative, biologically aggressive intrinsic tumors that typically are not amenable to surgical resection; although radiotherapy often leads to temporary disease control, the long-term prognosis for patients harboring such lesions remains poor with current therapy.[1,2,6,10,13,24,28,35,39] Other less common subgroups such as focal intrinsic lesions of the midbrain[16,39,41] and cervicomedullary junction[14–16,39] and dorsally exophytic tumors arising from the floor of the fourth ventricle[21,30,40] are generally slow-growing, histologically benign lesions. In some instances, radical surgery alone is effective in producing long-term disease control.

In comparison to the above categories of brain-stem tumors, which generally demonstrate relentless progression unless adequately treated, gliomas of the tectal plate have been reported to be particularly indolent, often remaining stable in size for several years;[7,8,26,32] the majority of the lesions that were examined histopathologically have been described as low-grade glial neoplasms. These intrinsic tectal tumors characteristically present with late-onset aqueductal stenosis, often without associated brain-stem signs. Because many of these lesions manifest with a paucity of neurological findings, exhibit a characteristic appearance on neuroimaging studies, and demonstrate indolent growth characteristics, biopsy of the tectal mass is often deferred and the diagnosis of "benign tectal glioma" or "benign intrinsic tectal tumor"[26] is made presumptively. These patients are then managed expectantly with cerebrospinal fluid (CSF) shunting and close follow-up monitoring. However, the natural history of these tumors in terms of their long-term potential for clinical and radiographic progression remains uncertain.

In order to address this issue, we reviewed our experience with a series of intrinsic tectal tumors that presented with symptoms of obstructive hydrocephalus from aqueductal stenosis, but without localizing brain-stem signs. These patients were all initially treated with CSF diversionary procedures; specific surgical and radiotherapeutic interventions for the tectal lesion were deferred until disease progression had been documented clinically or radiographically. The long-term follow-up results of these patients and our recommendations for their management are the focus of the current report.

Clinical Material and Methods

Between 1979 and 1992, 16 children (nine boys and seven girls) with late-onset aqueductal stenosis resulting from a benign intrinsic tumor of the dorsal midbrain were treated at the Children's Hospital of Pittsburgh (Table 1). Two patients had neurofibromatosis type I. Fifteen children presented with severe hydrocephalus and had obvious papilledema. Thirteen had headache, nausea, and vomiting of several months' duration, six manifested mild to moderate ataxia, and seven exhibited deterioration in school performance associated with memory loss and/or subtle personality changes. Four children had an acute exacerbation of their previously subtle symptoms following a minor head injury, thus prompting urgent neurosurgical referral. One

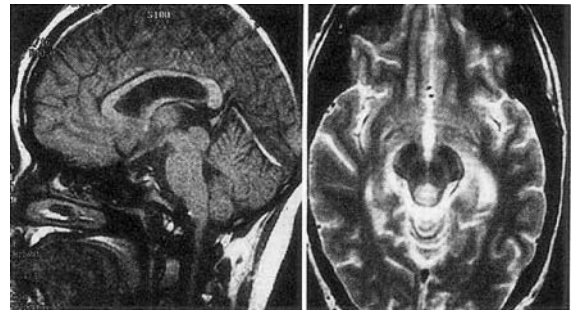

FIG. 1. The typical magnetic resonance imaging appearance for a benign tectal glioma (Case 13). *Left:* Sagittal T_1-weighted image showing a globular tectal mass that is isointense with the surrounding brain and is occluding the aqueduct of Sylvius. The lesion showed no enhancement following administration of gadolinium. *Right:* On T_2-weighted imaging, the tumor is hyperintense in comparison to the surrounding brain.

6-month-old child presented with progressive macrocephaly. Three older children also had obvious macrocephaly that had been noted since infancy, which predated the onset of other symptoms and signs of hydrocephalus by several years. In one child with neurofibromatosis, the tectal lesion was detected on a screening magnetic resonance (MR) image obtained at 8 years of age; this patient subsequently developed obstructive hydrocephalus at the age of 12 years. Only one patient showed evidence of brain-stem signs at presentation; this child had a partial Parinaud's syndrome that resolved following placement of a ventriculoperitoneal shunt.

All patients underwent computerized tomography (CT) initially. This characteristically was either normal or showed abnormal fullness and/or subtle hyperdensity of the tectal region without evidence of enhancement. Focal tectal calcification was detected in three children. Twelve patients who presented after 1985 also underwent MR imaging at or shortly following presentation; this disclosed a bulbous tectal mass that was hypointense or isointense with the surrounding brain on T_1-weighted images, hyperintense on T_2-weighted images, and without enhancement following administration of gadolinium (Fig. 1).

All 16 patients were treated with CSF diversion: 14 children received a ventriculoperitoneal shunt initially, one underwent shunt placement after an unsuccessful attempt at third ventriculostomy, and one was successfully treated initially with third ventriculostomy but later required shunt placement because of recurrent ventricular dilatation. None of the children underwent biopsy or radiotherapy initially. Subsequently, each child was closely followed with serial imaging studies and clinical examinations.

Results

Four children, three of whom were noted to have significant macrocephaly at presentation, had multiple

TABLE 1

*Clinical and radiographic findings in 16 patients with benign intrinsic tectal tumors**

Case No.	Age at Initial Presentation	Presenting Symptoms & Duration	Findings on Initial Examination	CT Appearance at Presentation	MRI Appearance at Presentation‖	Evidence of Disease Progression	Progression-Free Survival	
							From Onset of Symptoms**	From Presentation
1	8.5 yrs	memory loss, headache 3 mos; macrocephaly > 7.5 yrs	papilledema, ataxia, macrocephaly	normal	not done	yes	16.5 yrs	9 yrs
2	5.5 yrs	headache 6 mos; personality changes 12 mos‡	papilledema	tectal fullness§	not done	yes	10 yrs	9 yrs
3	11 yrs	headache, vomiting, ataxia, decreased school performance 3 mos; macrocephaly > 10 yrs	papilledema, ataxia, macrocephaly	normal	globular tectal mass	yes	13 yrs	3 yrs
4	6 mos	progressive macrocephaly	papilledema, tense fontanel, macrocephaly	normal	not done	yes	6.5 yrs	6.5 yrs
5	13 yrs	headache, ataxia 1 yr; macrocephaly > 12 yrs	papilledema, ataxia, macrocephaly	tectal fullness§	globular tectal mass	no	12.5 yrs	6 mos
6	8 yrs	headache 1 mo‡	papilledema	normal	not done	no	12 yrs	12 yrs
7	8 yrs	headache 6 mos; decreased school performance, lethargy 2 mos	papilledema, ataxia	tectal fullness with focal calcification§	globular tectal mass	no	7.5 yrs	7 yrs
8	11 yrs	headache 2 mos	papilledema	tectal fullness§	globular tectal mass	no	6.25 yrs	6 yrs
9†	8 yrs	tectal lesion found on screening MRI	stigmata of NF	tectal fullness§	globular tectal mass	equivocal	1.5 yrs	5.5 yrs
10	13.5 yrs	headache, ataxia 2 mos	papilledema, Parinaud's syndrome	tectal fullness§	globular tectal mass	no	3.25 yrs	3 yrs
11	8.5 yrs	headache, decreased school performance, listlessness 6 mos; decreased visual acuity 3 mos‡	papilledema, ataxia	tectal fullness with focal calcification§	globular tectal mass	yes	2.5 yrs	2 yrs
12	11.5 yrs	headache, worsening visual acuity 6 mos	papilledema	tectal fullness§	globular tectal mass	no	2.5 yrs	2 yrs
13	11.5 yrs	decreased school performance, decreased vision 3 mos	papilledema	tectal fullness§	globular tectal mass	no	2.25 yrs	2 yrs
14	14 yrs	decreased school performance 1 yr; headaches, memory problems 6 mos; monocular blindness 1 wk	papilledema	tectal fullness with focal calcification§	globular tectal mass	no	2.25 yrs	15 mos
15†	9.5 yrs	headache, vomiting 1 wk	papilledema	tectal fullness§	globular tectal mass	no	18 mos	18 mos
16	10 yrs	headache, vomiting 2 wks‡	papilledema	tectal fullness§	globular tectal mass	no	9 mos	9 mos

* CT = computerized tomography; MRI = magnetic resonance imaging; NF = neurofibromatosis.

† Patients with NF type 1.

‡ Symptoms worsened dramatically following a minor head injury precipitating neurosurgical referral.

§ On CT scans these lesions were all isodense or slightly hyperdense and nonenhancing.

‖ In cases with MRI, the lesions were isodense on T_1-weighted images, hyperintense on T_2-weighted images, and nonenhancing.

** From the presumed onset of initial symptoms and signs of hydrocephalus.

problems related to CSF overdrainage following shunt insertion. One such patient suffered chronic low-pressure headaches and required several shunt revisions. Three other children (one of whom had a flow-regulated valve placed initially) developed large subdural hematomas. These were treated by external drainage in two and placement of a subdural peritoneal shunt in the other; in two patients, an antisiphon device was added to the shunt system. During resolution of the subdural collections, all three patients developed a peculiar syndrome of gross ventriculomegaly and symptoms suggestive of elevated intracranial pressure such as headache, nausea, lethargy, and upgaze palsy, but in the setting of extremely low intracranial pressures documented by shunt tapping. Each child required a period of external ventricular drainage at negative pressures to normalize ventricular size and relieve symptoms. Following gradual elevation of the external drain over several weeks, CSF shunts (including an antisiphon device in two cases) were reinserted without further sequelae.

Ten children (Cases 5, 6, 7, 8, 10, 12, 13, 14, 15, and 16) have shown no clinical or radiographic progression of disease 0.5, 12, 7, 6, 3, 2, 2, 1.25, 1.5, and

TABLE 2

*Clinical features, treatment, and outcome in five patients with disease progression**

Case No.	Symptoms of Tumor Progression	CT Changes at Tumor Progression	MRI Appearance at Tumor Progression	Surgical Intervention	Histo-pathology	Adjunctive Treatment	Survival From Tumor Progression, Disease Status	Survival From Onset of Hydro-cephalus
1	Parinaud's syndrome	enlargement, calcification, mixed-density enhancement	exophytic growth with extension into diencephalon & midbrain tegmentum	open biopsy, cyst drainage	low-grade oligoastro-cytoma	5580 cGy external radiotherapy	4 yrs, partial regression	21 yrs
2	Parinaud's syndrome	enlargement, mixed-density enhancement	exophytic growth with extension into midbrain tegmentum	stereotactic biopsy	anaplastic astrocytoma	4300 cGY external radio-therapy, 1800 cGy stereo-tactic irradiation	4.5 yrs, no evidence of disease	14.5 yrs
3	Parinaud's syndrome	no change	no change in appearance of globular tectal mass	none	none	5400 cGy external radio-therapy	1 yr, stable disease	14 yrs
4	Parinaud's syndrome, precocious puberty	enlargement, mixed-density	cystic exophytic growth with extension into diencephalon	stereotactic biopsy	low-grade astrocytoma	5500 cGy external radiotherapy	6 yrs, partial regression	12.5 yrs
11	none	enlargement	exophytic growth into quadri-geminal cistern	none	none	none	6 mos, awaiting repeat MRI	2.5 yrs

* CT = computerized tomography; MRI = magnetic resonance imaging.

0.75 years, respectively, after CSF diversion, and 12.5, 12, 7.5, 6.25, 3.25, 2.5, 2.25, 2.25, 1.5, and 0.75 years after the onset of initial symptoms and signs of hydrocephalus, such as macrocephaly, headache, vomiting, and decreased school performance (Table 1). As noted above, one child with neurofibromatosis (Case 9) developed hydrocephalus from aqueductal stenosis 4 years after the tectal lesion was initially detected on MR imaging, but the lesion had shown no discernible enlargement and has remained stable radiographically 1.5 years after the onset of hydrocephalus.

Four children (Cases 1, 2, 3, and 4) developed symptoms of progressive brain-stem dysfunction 9, 9, 3, and 6.5 years after initial presentation (16.5, 10, 13, and 6.5 years, respectively, after the onset of hydrocephalic symptoms) (Tables 1 and 2). All four patients manifested insidiously progressive Parinaud's syndrome despite the presence of a functioning shunt. One child also developed precocious puberty. In three of these children (Cases 1, 2, and 4), follow-up CT scans disclosed enlargement of the dorsal midbrain lesion associated with a change in the appearance of the tumor from an isodense or slightly hyperdense lesion to one of mixed density (Fig. 2A and B). Irregular enhancement was detected in two of these tumors; calcification also became apparent in one. Magnetic resonance imaging demonstrated a tectal mass with mixed signal intensity on T_1-weighted and hyperintensity on T_2-weighted images, with exophytic growth into the quadrigeminal cistern and infiltration of the posterior diencephalon and midbrain tegmentum (Fig. 2C). Irregular enhancement with gadolinium administration was present in two of the tumors. One other child (Case 3) developed progressive brain-stem signs without conclusive evidence of tumor enlargement on serial CT scans and MR images. Each of the three patients with radiographic progression of disease underwent biopsy of the tectal mass: in one patient, a limited tumor debulking with cyst drainage was obtained via an infratentorial/supracerebellar approach; transfrontal stereotactic biopsy[9] was performed in the other two children. Histopathological examination of specimens from these three children demonstrated low-grade mixed glioma, anaplastic glioma, and low-grade astrocytoma.

All four patients with clinical evidence of disease progression were treated with conventional radiotherapy (5580, 4300, 5400, and 5500 cGy); the patient with an anaplastic astrocytoma also received a boost dose of 1800 cGy (50% isodose at the tumor margin) to the tumor using stereotactic radiosurgery techniques. Subsequently, these patients have remained clinically stable 4, 4.5, 1, and 6 years after tumor progression (21, 14.5, 14, and 12.5 years, respectively, after the onset of hydrocephalus), with improvement or resolution of their Parinaud's syndrome (Table 2). Serial MR studies have demonstrated tumor regression in three children (Fig. 2D) and stabilization of tumor size in one child.

One child (Case 11) demonstrated slowly progressive tumor enlargement on serial images while remaining clinically stable (Table 2). The lesion enlarged approximately 50% in volume during the 1st year of follow-up monitoring and changed in appearance from

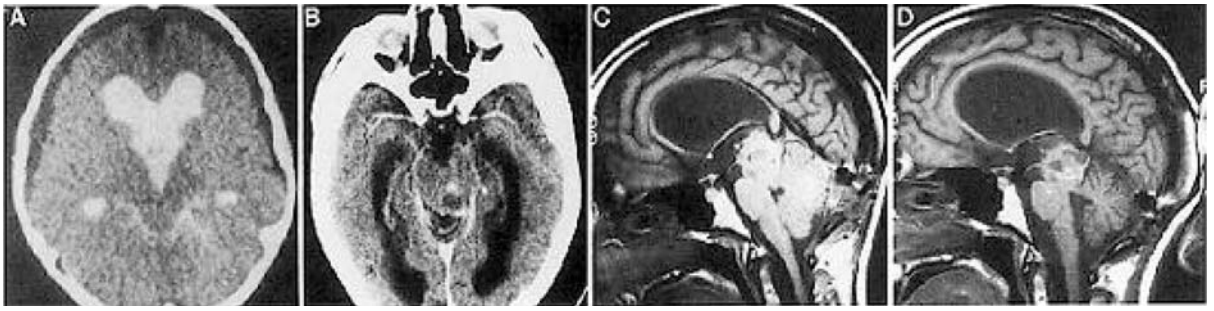

FIG. 2. Neuroimaging in Case 1, a 22-year-old woman who initially presented at 8 years of age with long-standing macrocephaly as well as headaches and memory loss. Tumor progression was documented both clinically and radiographically in this patient. A: Metrizamide computerized tomography (CT)-cisternogram obtained in 1979, several days after performance of a third ventriculostomy, showing profound ventriculomegaly without convincing evidence of a tectal tumor. A ventriculoperitoneal shunt was required several months later because of worsening ventriculomegaly. B: A CT scan obtained in 1985 after the onset of Parinaud's syndrome showing a mixed-density tectal lesion. C: Sagittal T_1-weighted magnetic resonance image showing an exophytic tectal mass with extension into the midbrain tegmentum. D: Magnetic resonance image obtained 4 years after biopsy, cyst drainage, and external radiotherapy, showing a decrease in lesion size; the patient's Parinaud's syndrome had resolved.

a globular to a pedunculated exophytic mass (Fig. 3). During the ensuing year, the tumor has shown comparatively minor additional enlargement. Because the patient is completely intact neurologically she has not yet undergone biopsy or radiotherapy although these are planned if there is any further tumor enlargement or if neurological signs develop.

Discussion

Low-grade gliomas of the dorsal midbrain have long been recognized as a cause of late-onset aqueductal stenosis.[3,11,12,18–20,22,27,29,31,34,36–38] Until recently, these lesions were often diagnosed only on postmortem examination, supporting the statement of Kernohan and Sayre[22] that these are "in all probability the smallest tumors in the human body that lead to the death of the patient." In reality, many of the early deaths resulted from uncontrolled hydrocephalus or complications of operative management rather than from actual tumor progression. With improvements in neuroimaging techniques, intrinsic tectal tumors are now being diagnosed at an early stage, allowing a more realistic appraisal of their long-term biological behavior.

This and other reports have provided new insights into the mode of presentation of these lesions and their characteristic neuroimaging features.[7,8,26,32] In contrast to the majority of brain-stem gliomas, which are inherently invasive and produce progressive cranial nerve deficits and long-tract signs in the absence of hydrocephalus, intrinsic tectal tumors almost invariably present with increased intracranial pressure from severe hydrocephalus, often without associated brain-stem signs. In many cases, the clinical history suggests a lesion that enlarges slowly, if at all, over time. Some patients, like the three in this series, have impressive macrocephaly,[8] which dates their hydrocephalus back to early childhood. Others manifest gradual deterioration in school performance or subtle personality changes over a period of several months. Eventually, more flagrant signs of increased intracranial pressure such as headache, vomiting, and ataxia lead to the diagnosis of hydrocephalus. Despite the anatomical location of the tumor within the dorsal midbrain, Parinaud's syndrome is surprisingly uncommon at presentation.[7,8,26,32]

As noted by several authors,[7,8,32,34,37] these lesions often elude detection by CT since they are usually small and isodense with the surrounding brain, and typically enhance poorly or not at all with intravenous contrast material. Although calcification may occur with time,[26] it is infrequently present at initial evaluation. High-resolution CT with its improved visualization of the tectal region has increased the chances of detecting these lesions by CT,[26] but MR imaging is vastly superior and is recommended for the evaluation of all cases of late-onset aqueductal stenosis. As noted in our study and elsewhere,[5,7,8,26,37] the typical appearance of these lesions on MR images is a globular tectal mass that is isointense or hypointense in comparison to the surrounding brain on T_1-weighted images, hyperin-

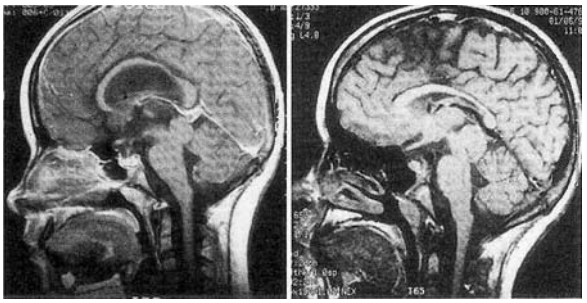

FIG. 3. Serial magnetic resonance (MR) images in Case 11 showing slowly progressive tumor enlargement, although the patient remains asymptomatic. *Left:* Sagittal MR image at presentation in January, 1991. *Right:* Two-year follow-up MR image showing tumor growth with exophytic extension into the quadrigeminal cistern.

tense on T_2-weighted images, and devoid of gadolinium enhancement. This appearance is clearly distinguishable from congenital aqueductal stenosis, where the tectum is never bulbous and rarely shows increased signal intensity on T_2-weighted images.[5] Because of the characteristic features of these tumors on MR imaging and the typical finding of "low-grade glioma" in those lesions that have been examined histopathologically,[7,8,37] biopsy confirmation of the true identity of the tectal tumor is often not obtained. However, it has been our presumption that these lesions represent low-grade gliomas.

The management of the hydrocephalus in association with benign intrinsic tectal tumors is frequently more challenging than for the usual hydrocephalic patient. Since many patients with tectal tumors have gross macrocephaly and craniocerebral disproportion, the risk of shunt-induced complications is substantially higher. Three of the 16 patients in our series and two of seven patients with CSF shunts described by Chapman[8] had large, shunt-induced subdural hematomas and impairment in neurological function. In three of our patients, a curious syndrome of neurological depression and gross ventriculomegaly in the setting of ultra-low intraventricular pressures developed following resolution of the subdural collections. This syndrome may result from transient alterations in the viscoelastic properties of the brain in patients with long-standing hydrocephalus who have undergone CSF drainage. These patients generally require a period of external ventricular drainage at negative pressures to reduce ventricular size and re-establish baseline brain compliance; the ventricles can then be gradually readjusted to a more normal pressure-volume relationship to sustain a low or medium pressure shunt. A detailed biomechanical study of this special type of "ultra-low pressure hydrocephalus" is the subject of a separate report. Several strategies have been used to avoid the initial period of overdrainage in these patients, such as incorporation of an antisiphon device or a flow-regulated valve into the shunt system and slow, gradual assumption of the upright position in the postshunt period, but these have only partially reduced the complication rate in this subgroup of hydrocephalic patients. Alternatively, attempts have been made to avoid a shunt entirely by performing a third ventriculostomy, but the long-term effectiveness of this procedure for these patients remains uncertain.

The management of the tectal tumor itself varies in the different reported series, reflecting uncertainty regarding the potential of these neoplasms for late progression. The combined experience suggests that tectal gliomas follow several distinct growth patterns that require individualized treatment approaches. Vandertop, *et al.*,[41] described a comparatively more aggressive group of tectal tumors that differ clinically and radiographically from those considered in the present study. These tumors characteristically present with focal neurological deficits in addition to hydrocephalus, show intense enhancement on CT and MR images with intravenous infusion of contrast material, and frequently extend into the thalamus or pons; CSF dissemination has also been noted.[33] If untreated, these patients typically suffer progressive neurological deterioration due to local tumor extension. Such lesions therefore require early therapeutic intervention; because they are often well circumscribed, surgical debulking followed by radiotherapy has been successful in achieving long-term disease control.

In contrast, the tectal tumors considered here and in the reports by May, *et al.*,[26] Boydston, *et al.*,[7] and Chapman[8] seem to have a distinctly more indolent course in that they rarely present with brain-stem signs or show contrast enhancement, and the majority remain clinically stable for several years without treatment. However, even within this group, there is variability in the propensity for late progression. For example, May, *et al.*, reported six children who had globular noncystic tectal masses, presumed to be low-grade tectal gliomas, that appeared to follow a particularly benign course. None of these lesions showed clinical progression in spite of minor changes on CT in two, and none required specific treatment during a mean follow-up period of 6.4 years. In contrast, Raffel, *et al.*,[32] described five patients with late-onset aqueductal stenosis and tectal gliomas who had a somewhat less favorable course. These patients all showed tumor enlargement on serial imaging studies, and one patient had CSF dissemination at diagnosis. Three of the four lesions biopsied were anaplastic astrocytomas and all four patients received external radiotherapy; the patient with CSF dissemination was also treated with chemotherapy. One patient died 9 years after the initial diagnosis, but the remaining three were alive $2\frac{1}{2}$ to 9 years after the onset of symptoms. The patient who did not undergo biopsy or receive specific treatment for the tectal tumor remained stable 9 years after the onset of symptoms.

In the present series, four of 16 tumors ultimately demonstrated clinical progression, with the onset of Parinaud's syndrome in all four children and precocious puberty in one. The interval until clinical disease progression averaged 6.9 years from the time of shunt insertion and 11.5 years from the onset of hydrocephalic symptoms. Three of these tumors had clearly enlarged and developed enhancement and/or cavitary changes on serial CT, and MR imaging evidence of exophytic growth into the quadrigeminal cistern and infiltration into the posterior diencephalon or midbrain tegmentum. In spite of these changes, only one lesion was anaplastic. One other lesion showed radiographic evidence of tumor enlargement during the 2 years after shunt insertion, although the patient has remained stable clinically. Since we did not perform a biopsy at presentation in our patients, it remains uncertain whether the tumors that progressed differed histologically at the outset from those that remained quiescent. Chapman[8] also reported the late development of focal signs in four of eight patients with tectal gliomas, 5, 8, 9.5, and 26 years after the presumed onset of hydrocephalus. As in the present series, these tumors all showed exophytic growth by the time brain-stem signs appeared. All four lesions were grade 1 astrocytomas,

and all were treated with open exploration and cyst drainage; only one patient received postoperative radiotherapy. Similarly, Boydston, *et al.*,[7] noted clinical progression and exophytic enlargement in one of six tectal gliomas; this lesion was also treated with cyst drainage and radiotherapy.

Thus, a certain percentage of patients with benign intrinsic tectal tumors will manifest disease progression if followed long enough, while the majority will enjoy long periods of good-quality survival following CSF shunting alone. Since the overall course tends to be one of extremely slow growth and minimal functional compromise, the management of patients with intrinsic tectal tumors must correspondingly be conservative and individualized for the patient. We currently recommend serial MR imaging and clinical examinations every 6 months for the 1st year after diagnosis, and then yearly for 4 years in patients who are asymptomatic. Following that, yearly clinical evaluation is sufficient and imaging studies are obtained every 2 to 3 years or in the interim if new symptoms develop. In view of the potential risks of surgery and radiotherapy, we favor deferring biopsy of these lesions and radiotherapy until disease progression is conclusively documented clinically and/or radiographically, as was suggested by Chapman,[8] May, *et al.*,[26] and Boydston, *et al.*[7] Because these lesions typically remain focal even at the time of progression, highly coned-down radiation fields should be employed, or part of the dose can be given by stereotactic radiosurgery[25] to minimize exposure to the surrounding brain.

Conclusions

Benign intrinsic tectal tumors presenting with late-onset aqueductal stenosis constitute a distinctive group of brain-stem tumors that generally carry a favorable long-term prognosis. Although these lesions may be visualized on CT scans, they are most effectively delineated by MR imaging, which is strongly recommended in all patients older than 6 months of age who present with aqueductal stenosis. Although lesions that are densely enhancing and present with brain-stem signs may require surgical and perhaps radiotherapeutic intervention at the outset, the intrinsic tectal tumors described in the present series can be managed conservatively with CSF diversion and conscientious follow-up monitoring. Further intervention is best deferred until there is clinical and/or radiographic evidence of tumor growth. Disease progression is generally heralded by the insidious development of Parinaud's syndrome. Histopathologically, the majority of these tumors are benign at the time of progression; however, a percentage will show anaplastic features. Following radiotherapy, many of these patients will continue to enjoy good-quality long-term survival.

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