Tectal Tumors of Childhood: Clinical and Imaging Follow-up

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PURPOSE: This study was done to determine which clinical and imaging findings best correlate with outcome in children with tectal tumors.

METHODS: A retrospective review was done of the medical records and imaging studies of 32 children (16 boys and 16 girls; mean age, 8 years) with tectal tumors. Eight children had CT, 11 had MR imaging, and 13 had both CT and MR studies. Findings from surgical and pathologic reports as well as from follow-up examinations (mean follow-up period, 5 years; range, 3.6 months to 17 years) were included in the review.

RESULTS: All patients had hydrocephalus and all but one required CSF diversion. The tectum was the center of the tumor in all cases and the majority of the tumors appeared isodense on CT scans, isointense on T1-weighted MR images, and hyperintense on T2-weighted images. Twenty patients required no further treatment. In this group, the mean maximum tumor diameter was 1.8 cm and enhancement occurred in two cases. At follow-up, 18 patients had stable tumor size, one had an increase in tumor size with cyst formation but no worsening of symptoms, and one had a decrease in tumor size. Twelve patients required further treatment (excision and/or radiotherapy) because of progression as indicated by either increased tumor size or worsening of symptoms. In this group, the mean maximum tumor diameter was 2.5 cm and contrast enhancement occurred in nine cases. Further follow-up in this group showed decreased tumor size in eight and stable residual tumor in three.

CONCLUSION: Tectal tumors in childhood have variable behavior. MR imaging assists in the clinical determination of which children need treatment beyond CSF diversion. Larger tumor size and enhancement are radiologic predictors of the need for further treatment.

Tectal gliomas are a unique subset of brain stem tumors of childhood (1–3). This article evaluates the usefulness of imaging in relation to management and outcome in a large series of children with tectal tumors.

Methods

Through a retrospective analysis of the medical records and imaging studies garnered over a 7-year period (January 1990 through January 1997), we identified 33 children who had tectal tumors and hydrocephalus. One patient was excluded because the tumor was a metastatic focus from a primary renal neoplasm. Initial imaging studies were available in 32 patients and consisted of CT in eight, MR imaging in 11, and CT plus MR imaging in 13. Axial 5- or 10-mm CT sections were obtained before and after intravenous administration of contrast material in three patients. CT examinations were obtained only without contrast enhancement in 14 patients and only with contrast enhancement in four patients. MR examinations were performed on a 1.5-T system in 23 patients and on a 1.0-T system in one patient. Imaging parameters included a section thickness of 5 mm with an intersection gap of 1 mm, a 256 × 128 matrix, and a 24-cm field of view (FOV) for sagittal T1-weighted conventional spin-echo images (600/11/2 [TR/TE/excitations]). Fast spin-echo axial proton density–weighted (2000/17/1) and fast spin-echo axial T2-weighted (3200/85/1) images were obtained with a section thickness of 5 mm and a gap of 2.5 mm, a 256 × 192 matrix, and a 24-cm FOV. Contrast material was administered intravenously in 17 patients at a dosage of 0.1 mmol/kg. Axial, coronal, and sagittal T1-weighted images of the brain were subsequently obtained. All patients had follow-up MR examinations. Imaging features evaluated included tumor location, size, extent, density, intensity, and enhancement. Only patients with primary tumors centered in the tectum were included. The patient population was divided into two groups: one group of 20 patients who required no further treatment after shunting of hydrocephalus and a second group of 12 patients who did require further treatment because of worsening symptoms or increased tumor size.
Tumor size at presentation was compared between the treated and untreated groups using a two-sample Student’s t-test. Proportional data were compared using Fisher’s exact test. Data are expressed in terms of the mean ± standard deviation. Logistical regression analysis was performed to assess whether maximum tumor diameter and the presence or absence of lesion enhancement were radiologic predictors of the need for further treatment. The odds ratio was used as a measure of association, and 95% asymptotic confidence intervals were constructed (4). Stepwise regression was then performed to generate an equation to determine the probability of treatment based on maximum diameter of tumors with and without enhancement. The computational form of the logistic equation for estimating the probability of further treatment from maximum tumor diameter in centimeters and the presence or absence of tumor enhancement has the following relationship:

$$
\text{Probability} = \frac{\exp(\beta_0 + \beta_1 \times \text{tumor size} + \beta_2 \times \text{enhancement})}{1 + \exp(\beta_0 + \beta_1 \times \text{tumor size} + \beta_2 \times \text{enhancement})}
$$

where $\beta_0$ is the intercept term, $\beta_1$ and $\beta_2$ are regression coefficients derived by using the maximum likelihood principle, and $\exp$ is the base of the natural logarithm. Enhancement equals 1 if present and 0 if absent. The likelihood of further treatment (surgery and/or radiotherapy) was calculated by determining the probability using Equation 1 and multiplying by 100%. Data analysis was conducted using version 6.11 of SAS for Windows (SAS Institute Inc, Cary, NC). A two-tailed value of $P < .05$ was considered statistically significant for all analyses.

Biopsies were done of the tumors in eight patients. In five patients, pathologic analysis by light microscopy was obtained at our hospital with the use of standard staining techniques.

Results

Initial Clinical and Imaging Findings

Thirty-two patients (16 girls and 16 boys) had tectal tumors. The mean age at diagnosis was 8 years (range, 2 months to 17 years). The clinical presentations included headache ($n = 19$); papilledema ($n = 11$); visual problems ($n = 10$) such as decreased vision ($n = 3$), diplopia ($n = 6$), sunsetting ($n = 1$), and ptosis ($n = 1$); ataxia ($n = 8$); vomiting ($n = 7$); nausea ($n = 5$); clumsiness/falling ($n = 5$); Parinaud syndrome ($n = 5$); nystagmus ($n = 4$); sixth nerve palsy ($n = 5$); dysmetria ($n = 4$); tremors ($n = 3$); lethargy ($n = 2$); memory deficits ($n = 2$); declining school performance ($n = 2$); developmental delay ($n = 2$); motor disturbance ($n = 1$); increasing head circumference ($n = 1$); and seizures ($n = 1$). One patient had neurofibromatosis, one patient presented after head trauma, and one patient had subarachnoid and intraventricular hemorrhage.

The tectum was the center of the tumor in all patients, including bulbous enlargement and extension into the tegmentum in four and extension into the thalamus in six. In one patient, the tumor was exophytic posteriorly into the quadrigeminal plate cistern. On CT scans, the tumor was isodense with gray matter in 11 patients, of low attenuation relative to gray matter in three patients, and exhibited calcification in three patients. Enhancement was observed in four of the seven patients who received a contrast agent for CT. On T1-weighted MR images, the lesions were isointense with gray matter in 16 patients, hypointense in six patients, and of mixed hypointensity and hyperintensity in one patient (Fig 1). On proton density–weighted images, the lesions were hyperintense in 21 patients and of mixed hyperintensity and hypointensity in two patients. On T2-weighted images, the lesions were hyperintense in 19 patients, of mixed hyperintensity with hypointensity in two patients, and isointense in one patient. In one patient, the MR images obtained before contrast administration were not available for review. The hypointensities noted on the proton density– and T2-weighted MR images corresponded to the areas of calcification on the CT scans. Of the 17 patients who received contrast material, lesion enhancement was observed in seven. The mean maximum tumor diameter in 26 of the patients was 1.8 cm. In five patients, the tumors could not be measured accurately on CT scans because of poor visualization of the margins. No patient had metastatic disease at the time of presentation.

Hydrocephalus was characterized by the ratio of the greatest width of the frontal horns to the diameter of the inner skull, with a ratio of 0.28 to 0.30 indicating mild ventricular dilatation and a ratio over 0.30 signifying significant dilatation (5). Hydrocephalus was considered severe when there was marked enlargement of the lateral and third ventricles or when periventricular edema was present. In five patients, the degree of hydrocephalus was unknown because imaging studies were not available before shunt surgery.

![Image](image.png)
placement. In one patient, the initial studies, consisting of skull films, sonograms, and pneumoencephalograms, were not available for analysis of ventricular size.

All patients had hydrocephalus and all but one required CSF diversion, which consisted of ventriculoperitoneal (VP) shunting in 20, third ventriculostomy in seven, ventriculocervical (VC) shunting in three, and cisternal-cervical shunting in one. VP shunt revisions were required in two patients with VC shunts, in one patient with a VP shunt, in the one patient with the cisternal-cervical shunt, and in two patients with third ventriculostomies. One patient who underwent VP shunting initially has had a subsequent third ventriculostomy.

Follow-up Clinical and Imaging Findings

Untreated Group.—Twenty patients required no further treatment after shunting of the hydrocephalus. In this group, the mean maximum tumor diameter was 1.8 cm ± 0.44 and enhancement occurred in only two patients. Enhancement information was unavailable in eight. On follow-up studies (mean, 5 years; range, 3 months to 24 years), 18 patients had stable residual tumor size, one had an increase in tumor size with cyst formation, and one had a decrease in tumor size. In one of the patients with stable residual tumor size, a 3-mm cyst developed 3 years after initial diagnosis; however, it has not changed on subsequent follow-up studies, and the patient is clinically stable. In another patient with stable residual tumor size, the tumor increased in enhancement from being isointense to having increased density, suggesting calcification, 8 months after initial diagnosis. A 3-mm cyst developed in this same patient 2 years after initial diagnosis, but the patient has remained clinically stable. In two patients, tumor enhancement developed 5 and 6 months, respectively, after initial diagnosis, without a significant change in tumor size or clinical status. The one patient with increased tumor size and cyst formation remains clinically stable, and no further treatment has been done. The cyst appeared 2.5 years after diagnosis and has increased in maximum diameter from 5 mm to 9 mm over a 5.5-year period. The tumor has also shown a slight increase in size. This patient has partial complex seizures controlled with medication. The remaining patients in this group are stable.

Treated Group.—Twelve patients required further treatment, consisting of surgical excision in three and radiotherapy in nine. One of these patients had a ventriculoscopic biopsy, which was nondiagnostic. One patient underwent surgical excision of the tumor at the time of diagnosis because of its large size. The remaining 10 patients required further treatment because of disease progression, as indicated by either an increase in tumor size, as measured by maximum tumor diameter, or worsening of symptoms. The time from initial diagnosis to therapeutic intervention ranged from 2 to 40 months (mean, 13 months). The mean maximum tumor diameter in these patients was 2.5 cm ± 1.0 and contrast enhancement occurred in nine patients before treatment (eight at initial diagnosis and one on follow-up imaging) (Fig 2). Among the patients who received radiotherapy, doses ranged from 5250 to 5620 cGy (mean, 5417; median, 5400). One patient received a whole-brain dose of 3600 cGy plus a coned-down dose of 1800 cGy to the midbrain. Four patients received stereotactic radiotherapy with doses of 5250, 5400, 5400, and 5580 cGy (mean, 5408). Five patients underwent radiation therapy after biopsy or subtotal resection. No patient received chemotherapy.

On follow-up studies (mean, 5 years; range, 3 months to 17 years) in this group, the tumor decreased in size in eight patients and there was stable residual tumor in three patients. In one patient, only 3 months of follow-up had elapsed since the time of diagnosis. All patients were alive and without recurrence at the time of the study. Three months after radiation therapy, one patient had a 3-mm cyst along the anterior margin of the tectal tumor and T2 hyperintensity in the adjacent pons, both of which resolved on subsequent follow-up examinations. This patient has mild attention deficits, delayed puberty, and growth hormone deficiency. Another patient had a 9-mm cyst 1 month after stereotactic radiotherapy. There was an increase in the size of the cyst (maximum size, 1.9 cm) and increased enhancement over the next 10 months, although the patient was asymptomatic (Fig 3). During the next 2 years, the enhancement resolved and the cyst decreased in size. In a third patient, the tectal tumor exhibited increased
enhancement 3 months after stereotactic radiotherapy and resolved in another 3 months. This patient has had mild learning disabilities. A fourth patient had a hemorrhage within the pons 9 years after radiation therapy that subsequently resolved (Fig 4). This event most likely represented a radiation vasculopathy, and the patient has residual left-sided motor weakness and mild learning problems. Except for one patient, who has a daily headache and an upward gaze abnormality, the remaining patients in this group are stable and the symptoms have resolved.

Pathologic Findings

Insufficient biopsy material precluded a pathologic diagnosis in one patient. Two patients who underwent biopsies at outside hospitals both had a low-grade astrocytoma. Five patients had satisfactory biopsy material for pathologic review at our hospital. Each specimen showed mild hypercellularity composed of a monomorphic population of cells with oval nuclei and eosinophilic cytoplasm with fibrillary processes. The background showed a coarse fibrillarity that was accentuated with either phosphotungstic acid-hematoxylin (PTAH) or glial fibrillary acidic protein (GFAP) stains. Four tumors had Rosenthal fibers (80%), two had areas with extensive extracellular material (40%), and one had areas with cyst formation. Two tumors also had eosinophilic granular bodies. There was extensive calcification in one tumor and abundant hemosiderin in another case. No high cellularity or mitosis was detected in any of the specimens. Although the biopsy specimens were generally too small to allow identification of all the characteristic features, the histology in these cases was most consistent with pilocytic astrocytoma.

Statistical Analysis

At presentation, the tumors were significantly larger in the treated group (2.5 cm ± 1.0) than in the untreated group (1.8 cm ± 0.44); (P = .02). Also, a greater proportion of the tumors in the treated group (9/12, 75%) showed enhancement as compared with those in the untreated group (2/12, 17%) (P = .01). The treated group had a significantly greater proportion of patients who presented with diplopia (P = .02) and a marginally significantly greater proportion with
cranial nerve palsies ($P = .06$) than did the untreated group. There were no statistical differences between the two groups with respect to the presenting symptoms of Parinaud syndrome or nystagmus.

Logistic regression analysis revealed that maximum tumor diameter was a significant predictor of the need for further treatment ($P = .015$). The odds of treatment for tectal tumors (Table) were almost five times greater for each increase of 1 cm in maximum tumor diameter (odds ratio, 4.9; 95% confidence interval, 1.3 to 19.3). In addition, the odds of further treatment (Table) were 15 times greater for patients in whom the tumor enhanced than for those in whom the tumor did not enhance (odds ratio, 15.0; 95% confidence interval, 2.2 to 106.5) ($P = .003$). The wide confidence intervals reflect the small sample size.

The likelihood of treatment (surgery or radiotherapy) for a child whose tumor had a maximum diameter of 2.5 cm was estimated to be 90% with tumor enhancement and only 11% without enhancement (Fig 5). An estimated 67% of patients with tectal tumors that had a maximum diameter of 1.5 cm and enhancement were estimated to require treatment, as compared with only 3% of tumors of the same diameter without enhancement. The parameter estimates in the final stepwise logistic regression model were $\beta_0 = -5.75$, $\beta_1 = 1.48$, and $\beta_2 = 4.21$. For example, the likelihood of further treatment for patients who had an enhancing tumor with a diameter of 2.5 cm was

\[
\text{Likelihood} = \frac{\exp(-5.75 + 1.48 \times 2.5 + 4.21)}{1 + \exp(-5.75 + 1.48 \times 2.5 + 4.21)} \times 100\% \\
= \frac{8.67/9.67 \times 100\%}{9.67} = 89.7, \\
\text{or approximately } 90\%.
\]
Similarly, the likelihood of further treatment for patients who had a nonenhancing tumor with a diameter of 2.5 cm was

$$
3) \quad \text{Likelihood} = \frac{\exp(-5.75 + 1.48 \times 2.5)}{1 + \exp(-5.75 + 1.48 \times 2.5)} \\
\times 100\% = \frac{0.129}{1.129} \times 100\% \\
= 11.4, \text{ or approximately } 11\%.
$$

**Discussion**

**Clinical Aspects**

Brain stem gliomas in children classically present with multiple cranial nerve palsies, long tract signs, and late onset of hydrocephalus; they often have a poor prognosis (6, 7). Tectal gliomas are a unique subset of brain stem gliomas in children that are reported to have a more benign clinical course (1, 3, 8–11). In this series of tectal tumors there was no gender predilection. In previous reports a male predilection has been described, but it is of uncertain significance (8–11). One of our patients had neurofibromatosis-1, which has also been reported in association with tectal tumors in other series (8, 12). The presenting clinical signs and symptoms in the majority of children were related to hydrocephalus, including headache (59%) and papilledema (34%). No patient presented with multiple cranial nerve palsies. The majority of all these manifestations resolved after ventricular shunting. Clinical presentations related to hydrocephalus in the absence of brain stem signs have also been reported in other series (3, 8–10).

**Imaging Aspects**

The imaging findings in the majority of patients in our series corresponded with those previously reported in the literature. Tectal tumors on CT scans were usually isodense with gray matter. On MR examinations the lesions were usually isointense on T1-weighted images and hyperintense on proton density– and T2-weighted images (3, 10, 13, 14). Calcification in these lesions was detected in two of our cases and also has been described by others (8, 10, 13). The overwhelming majority of tectal tumors are astrocytomas (1, 10, 12–17). Other reported tectal masses in children include ependymoma, ganglioglioma, primitive neuroectodermal tumor, metastasis, hamartoma, abscess, granulomatous disease, periaqueductal gliosis, and vascular malformations (18–20).

**The Untreated Group**

Of the patients who had no further treatment after ventricular shunting, the majority were stable on follow-up examinations. Imaging changes on follow-up studies included the development of small cysts in three patients, a phenomenon that has been described previously in tectal gliomas and that is often associated with focal neurologic signs (1). However, focal neurologic signs were not found in our patients. The development of calcification in these lesions has also been described previously (10). One patient in our series who had an increase in tumor size and cyst formation remains clinically stable. It is unclear at this time whether further treatment will be required.

**The Treated Group**

Among the group of patients requiring further treatment, there was a significantly greater proportion who presented with diplopia, and a marginally significantly greater proportion who had cranial nerve palsies, which may be related to the larger tumor size in this group. The patients who required further treatment after shunting of hydrocephalus had worsening symptoms or increasing tumor size. The tumors were significantly larger on presentation and showed greater enhancement than those in the patients who did not require further treatment. Larger tumor size and contrast enhancement were significant predictors of the need for further treatment (Fig 5, Table). Tumors that had a maximum diameter of 2.5 cm and
showed contrast enhancement had a 90% likelihood of requiring further treatment.

The significance of contrast enhancement in these lesions is controversial according to the literature. Squires et al (11) associated contrast enhancement with tumor growth and disease progression. Of the 12 patients in that series, the one who had an enhancing tumor at presentation also had disease progression. Of the three patients in that series who had disease progression, all subsequently had enhancing tumors, and all patients with disease progression had accompanying enhancement. In another study, by Bognar et al (13), enhancement at presentation was thought to be independent of tumor grading. In that series, calcification, contrast enhancement, cysts, and an exophytic component were observed in both high- and low-grade astrocytomas. The only imaging parameter correlating with biological behavior in that study was tumor size, which was corroborated in our series, in which the tumors were larger in the treated group. In the series reported by Chapman (1) and by May et al (10), a lack of contrast enhancement was thought to be associated with a benign, indolent clinical course, and follow-up studies in these groups showed no disease progression.

As in our series, previous studies have shown a relatively benign and indolent course for these tumors, with good long-term prognoses (1, 3, 8–11). All the biopsy specimens of the tectal tumors in our series revealed low-grade astrocytoma, a finding that is corroborated in our series, in which the tumors were larger in the treated group. However, in one series by Raffel et al (12), the majority of the lesions in which biopsies were performed were anaplastic astrocytomas.

Most of the patients in our series who received treatment had a decrease in tumor size on follow-up examinations. The changes in imaging findings on follow-up studies after radiation therapy in four of our patients were most likely related to treatment effects rather than to disease progression. These new findings included the development of cysts, increased size, increased enhancement, edema, and hemorrhage—all of which resolved. Such findings after radiation therapy are transient and are usually not associated with changing signs or symptoms (21–27).

**Conclusion**

Tectal tumors in childhood have variable behavior but generally a good prognosis. Larger tumor size and enhancement are significant radiologic predictors of the need for further treatment beyond CSF diversion.

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**References**