Tectal glioma

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Tectal gliomas fall under the grouping of childhood midbrain gliomas and unlike the other tumours in that group they are typically low grade astrocytomas with good prognosis.

Epidemiology

Tectal plate gliomas are encountered in children and adolescents.

A male predilection has sometimes been reported although this is by no means certain.

An association with neurofibromatosis type I (NF1) has been reported [1].

Classification

For tectal gliomas, the classification would depend on the specific features observed in the tumor tissue. Here is a general overview:

Low-grade Tectal Gliomas (Grade I and II): These tumors are typically slow-growing and less

aggressive. They may be classified as pilocytic astrocytomas, which are characterized by well-differentiated cells and a relatively favorable prognosis.

High-grade Tectal Gliomas (Grade III and IV): These tumors are more aggressive and may be classified as anaplastic gliomas (Grade III) or glioblastomas (Grade IV). High-grade tumors are associated with a poorer prognosis and may require more aggressive treatment approaches.

Molecular markers, such as mutations in specific genes like IDH (isocitrate dehydrogenase) and 1p/19q co-deletion, are increasingly considered in glioma classification as they provide additional information about the tumor's biology and can influence treatment decisions.

Pediatric tectal glioma

Clinical features

Symptoms of tectal glioma include those caused by increased intracranial pressure due to obstructive hydrocephalus. Headache, blurred vision, double vision, nausea, and vomiting are common symptoms.

Their expansion within the brainstem causes narrowing the aqueduct of Sylvius and causing obstructive hydrocephalus with presentation usually secondary to headache.

Additional symptoms may include gaze palsy, due to compression of the medial longitudinal fasciculus leading to an upgaze palsy, diplopia or Parinaud syndrome, although these are uncommon.

Pathology

The vast majority of lesions are low grade astrocytoma, although occasionally other glial series tumours are encountered in the tectal region including ependymoma, ganglioglioma and primitive neuroectodermal tumours (PNET).

see Tectal ganglioglioma.

Since many of these are not biopsied, meaningful statistical analysis is not possible.

Diagnosis

Diagnosis is based on initial suspicion fostered by the presentation of an obstructive hydrocephalus followed by physical exam which may potentially reveal indications of pyramidal tract dysfunction or cranial nerve palsies.

Radiographic features

The superb sensitivity of MR and its multiplanar imaging capability permit unparalleled diagnostic

accuracy in this region. The sagittal and axial planes are ideal for evaluating the tectum. CT remains important in the detection of acute hemorrhage and calcification. Grouping of abnormalities on the basis of anatomic boundaries (tectum, aqueduct, and quadrigeminal plate cistern) is useful in establishing the correct diagnosis [2)].

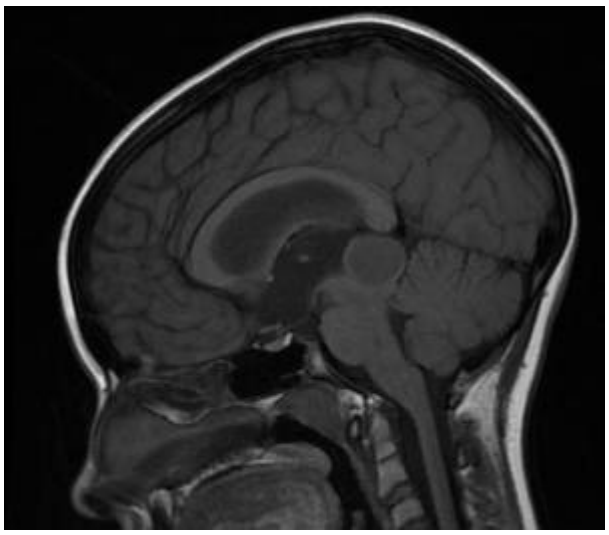
CT

Typical CT finding is homogeneous expansion of tectal plate, isodense to grey matter with minimal enhancement on postcontrast images.

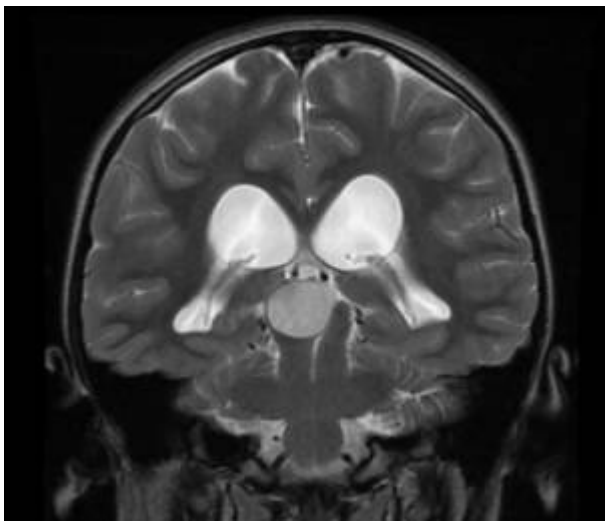
On CT it is not uncommon to find a central tectal calcification.

MRI studies reveal a characteristic well-circumscribed, isodense or hypodense mass on T1-weighted images, with hyperdensity on T2 imaging. Yet current radiological methods insufficiently distinguish tectal plate gliomas from brainstem tumors or gliomas in the neighboring structures, and a definitive diagnosis requires biopsy and histopathological analysis.

Typically the tumours demonstrate expansion of the tectal plate by a solid nodule of tissue.



T1: iso to slightly hypointense to grey matter



T2: hyperintense to grey matter

T1 C+ (Gd): usually no enhancement

With time the mass can develop small cystic spaces (sometimes associated with neurological deficits) or calcification.

Higher grade tumours tend to be larger and tend to enhance more vividly.

Differential diagnosis

Tumors leading to occlusion of the aqueduct of Sylvius include those of pineal, thalamic, and tectal origins.

see Aqueductal tumor.

The differential diagnosis from germ cell tumor or pineal cyst is essential for treatment.

When the tectum is near-normal then the differential is largely limited to:

aqueductal stenosis

no mass lesion

a focal stenosis or web may be visible

With larger lesions, where the mass is not definitely arising from the tectal plate then the differential is essentially that of a pineal region mass and therefore includes:

pineal parenchymal tumours and germ cell tumours

pineal cyst

meningioma

cerebral metastases

cavernous malformation

In patients with NF1 a hamartoma should also be considered. They tend to have some T1 hyperintensity.

Treatment

Tectal gliomas are usually diagnosed in childhood and often occur in adults. They are often benign, slowly progressing lesions; outpatient clinical and radiological follow-up is sufficient.

In the treatment, ETV (endoscopic third ventriculostomy) or VP-shunt (ventriculoperitoneal) can be applied to treat hydrocephalus.

Management is planned according to the degree of associated signs and symptoms, and may range from diligent observation and periodic screening for advancing tumor development, to cerebrospinal fluid shunting in an effort to resolve obstructive hydrocephalus, to radio- and chemotherapy. A wide range of minimally invasive approaches using endoscopy is available for the neurosurgeon, including endoscopic third ventriculostomy and endoscopic aqueductoplasty [3)]

Tumors leading to occlusion of the sylvian aqueduct include those of pineal, thalamic, and tectal origins. These tumors cause obstructive hydrocephalus and thus necessitate a CSF diversion procedure such as an endoscopic third ventriculostomy (ETV), often coupled with an endoscopic biopsy (EBX).

As tectal plate gliomas are low grade and often very slow growing, shunting is often the only required intervention for long term survival. As surgical biopsy can have significant morbidity in this area, usually the diagnosis is made on imaging findings alone.

Imaging predictors of patients who will need further treatment include a size greater than 2.5 cm and presence of contrast enhancement.

Outcome

In the minority of patients who progress, radiotherapy often leads to local control or even tumour regression.

Surgical excision is sometimes necessary.

Tectal plate gliomas showed indolent clinical courses, even after radiologic tumor progression. After the treatment of obstructive hydrocephalus, clinical and radiologic follow-up can be recommended for indolent tectal plate gliomas [4)].

Case series

2015

Between October 2002 and May 2011, 11 patients with tectal gliomas were treated with Gamma Knife radiosurgery. Five patients had pilocytic astrocytomas and six nonpilocytic astrocytomas. Ten patients presented with hydrocephalus and underwent a CSF diversion procedure [7 V-P shunt and 3 endoscopic third ventriculostomy (ETV)]. The tumor volume ranged between 1.2-14.7 cc (median 4.5 cc). The prescription dose was 11-14 Gy (median 12 Gy).

Patients were followed for a median of 40 months (13-114 months). Tumor control after radiosurgery was seen in all cases. In 6/11 cases, the tumors eventually disappeared after treatment. Peritumoral edema developed in 5/11 cases at an onset of 3-6 months after treatment. Transient tumor swelling was observed in four cases. Four patients developed cysts after treatment. One of these cases required aspiration and eventually disappeared, one became smaller spontaneously, and two remained stable.

Gamma Knife radiosurgery is an effective and safe technique for treatment of tectal gliomas. Tumor

shrinkage or disappearance after Gamma Knife radiosurgery may preclude the need for a shunt later on [5].

Dabscheck et al., conducted a retrospective review of all patients with tectal gliomas over a 22-year period at a single institution. Data extraction included sex, age at presentation of tectal glioma and age of presentation with seizures, magnetic resonance imaging (MRI) findings, seizure frequency and semiology, and EEG findings. They identified 79 patients, 66 of whom had adequate imaging and clinical data for further analysis. Eight patients (12.1%) had a history of seizures. Three patients had a clear symptomatic cause of seizures. Three patients were diagnosed with a tectal glioma as an incidental finding after a first seizure. One patient had a history of febrile convulsions. One patient had a generalized seizure 5 years after presenting with macrocephaly. Although the risk of seizure in children with known tectal glioma was relatively high, we did not identify specific clinical, radiologic, EEG, or MRI features that are predictive of increased risk. Thus, in children with tectal gliomas who have seizures, alternative causes for the seizures must be sought [6].

Among 26 patients, 19 presented with signs or symptoms of increased intracranial pressure (73 %) versus an incidental finding in 7 (27 %). Median follow-up was 46 months (range 8-143 months). Six of 26 (23 %) experienced progressive disease after diagnosis. Five of 26 (19 %) required more than one surgical procedure due to failure of initial endoscopic third ventriculostomy. Seven of 26 had history of endocrine dysfunction, of which, five presented with endocrine dysfunction (precocious puberty or short stature), 1 developed menstrual irregularities after surgical intervention and 1 had preexisting pan hypopituitarism. Of 12 patients with available neuropsychological testing, eleven had at least one indicator of executive functioning in the low-average to impaired range. While tectal plate gliomas have been considered indolent tumors that are rarely progressive, 23 % of patients in this cohort experienced disease progression and required further therapy. Neurocognitive deficits may occur, while endocrine deficiency is uncommon. Regular multidisciplinary oncology follow-up, routine monitoring with MRI and formal neurocognitive evaluation are imperative to provide early recognition of disease progression or recurrent hydrocephalus and to improve school functioning in this population [7].

2014

Forty-four patients with a mean age of 10.2 years who harbored tectal plate gliomas were included in the study. The mean clinical and radiological follow-up was 7.6 ± 3.3 years (median 7.9 years, range 1.5-14.7 years) and 6.5 ± 3.1 years (median 6.5 years, range 1.1-14.7 years), respectively. The most frequent intervention was CSF diversion (81.8% of patients) followed by biopsy (11.4%), radiotherapy (4.5%), chemotherapy (4.5%), and resection (2.3%). On MR imaging tectal plate gliomas most commonly showed T1-weighted isointensity (71.4%), T2-weighted hyperintensity (88.1%), and rarely enhanced (19%). The initial mean volume was 1.6 ± 2.2 cm(3) and it increased to 2.0 ± 4.4 cm(3) ($p = 0.628$) at the last follow-up. Frontal and occipital horn ratio (FOHR) and third ventricular width statistically decreased over time ($p < 0.001$ and $p < 0.05$, respectively).

The authors' results support existing evidence that tectal plate gliomas frequently follow a benign clinical and radiographic course and rarely require any intervention beyond management of associated hydrocephalus [8].

2013

Twenty-two children were identified from a 15-year retrospective database of neuroendoscopic procedures performed at the authors' institution, Children's Hospital of Alabama, in patients with a minimum of 1 year of follow-up. Clinical outcomes, including the need for further CSF diversion and symptom resolution, were recorded. The frontal and occipital horn ratio (FOR) was measured on pre- and postoperative, 1-year, and last follow-up imaging studies.

In 17 (77%) of 22 children no additional procedure for CSF diversion was required. Of those in whom CSF diversion failed, 4 underwent successful repeat ETV and 1 required shunt replacement. Therefore, in 21 (96%) of 22 patients, CSF diversion was accomplished with ETV. Preoperative and postoperative imaging was available for 18 (82%) of 22 patients. The FOR decreased in 89% of children who underwent ETV. The FOR progressively decreased 1.7%, 11.2%, and 12.7% on the initial postoperative, 1-year, and last follow-up images, respectively. The mean radiological follow-up duration for 18 patients was 5.4 years. When ETV failed, the FOR increased at the time of failure in all patients. Failure occurred 1.6 years after initial ETV on average. The mean clinical follow-up period for all 22 patients was 5.3 years. In all cases clinical improvement was demonstrated at the last follow-up.

Endoscopic third ventriculostomy successfully treated hydrocephalus in the extended follow-up period of patients with TPGs. The most significant reduction in ventricular size was observed at the the 1-year followup, with only modest reduction thereafter [9)].

2012

A 5-year-old boy who had tectal plate Low-grade glioma with obstructive hydrocephalus was managed with Codman programmable ventriculoperitoneal shunt. There was a spontaneous change in the opening pressure of the shunt valve leading to shunt malfunction. Routinely used household appliances produce a magnetic field strong enough to cause change in the setting of shunt valve pressure and may lead to valve malfunction [10)].

2003

Tumors involving the tectal region constitute a distinct subgroup of brain stem gliomas with an indolent clinical course. Here, we present the clinical and neuroradiologic features of 9 children with tectal tumors. All patients presented with signs and symptoms of hydrocephalus and were managed with ventriculoperitoneal shunt insertion. MRI studies revealed focal hyperintense lesions on T2-weighted images without any contrast enhancement, and no evidence of progression was demonstrated in any patient. We also reviewed the published series of tectal gliomas in the literature to compare with our results. Based on these and other published series, it was concluded that intrinsic tectal gliomas of childhood with sizes less than 2 cm in diameter and without any tumor extension or contrast enhancement constitute a specific subgroup of tectal masses which rarely display invasive clinical behavior and should be managed conservatively. CSF diversion procedures and long-term yearly follow-up examinations with MRI scans are sufficient in these patients [11)].

1999

A 10-year retrospective review has identified 11 consecutive children with tectal plate lesions. Headache, vomiting, a decline in school performance, tremor, and complex partial seizures were common presenting symptoms. All patients presented with signs and symptoms of hydrocephalus. Magnetic resonance (MR) imaging delineated an intra-axial mass lesion of the midbrain primarily localized to the tectal plate which uniformly was hyperintense on T2-weighted imaging and had a more variable appearance on T1-weighted imaging and rare enhancement with gadolinium. No patient underwent surgical resection, chemotherapy, or radiotherapy. Three of 11 patients (27%) showed evidence of progression in size or a new focus of enhancement on MR imaging, which was clinically asymptomatic. In this series, no patient with a tectal plate lesion less than 1.5 cm in maximal diameter and without gadolinium enhancement showed any evidence of clinical or radiological progression. Although intrinsic tectal lesions in children are clinically indolent and the initial management consists of CSF diversion, these lesions may eventually progress and still warrant long-term follow-up with serial MR imaging [12)].

1998

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.

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.

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 [13)].

1994

A series of 12 patients with tectal plate gliomas, is presented treated by direct surgery. Mean age was 19 years. All patients presented with signs of raised intracranial pressure and supratentorial hydrocephalus on CT scan. Diplopia was the most common local sign. CT scan and MR imaging showed 4 intrinsic, 6 exophytic, and 2 ventrally infiltrating tectal tumours. The histological diagnosis was low-grade astrocytoma in 7, high-grade astrocytoma in 2, oligodendroglioma in one, oligo-astrocytoma in one, and ependymoma in one case. The suboccipital supra- and transtentorial approach was used in every cases. Tumour resection was generous at the level of the superior

colliculi, but on the contrary, resection was limited at the level of inferior colliculi due to the auditory risk. Tumour removal was total (macroscopically) in 9 cases and partial in 3 cases. There were 4 surgical complications and one death related to surgery. Parinaud's syndrome was the most-common postoperative sequelae. Auditory hallucinations and the acoustic neglect syndrome were seen once. In three cases additional radiotherapy and chemotherapy were given once with severe sequelae. The treatment of tectal plate gliomas is controversial. The role of different therapeutic options remains open. We consider the tectal plate as a relatively safer territory for surgery than the ventral part of the midbrain. The brain stem auditory evoked potentials (BAEPs) and middle latency potentials (MLPs) monitoring can help to determine the appropriate limit of surgery [14].

Case reports

see Tectal glioma case reports

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