Case report

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Pediatric tectal glioma presented with acute hydrocephalus and ventriculomegaly. Two case reports

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ABSTRACT

Tectal gliomas represent a subset of low-grade tumors that arise in the tectal region at the roof of the brainstem. 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. In the treatment, ETV (endoscopic third ventriculostomy) or VP-shunt (ventriculoperitoneal) can be applied to treat hydrocephalus. 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. We present two cases of pediatric patients with mesencephalic tectal plate tumors. An 11-year-old boy and a 15-year-old girl applied to the Emergency Department with different complaints. The 11 year-old-boy was treated with VP-shunt due to acute hydrocephalus.

Keywords: *tectum mesencephali; glioma; seizures; hydrocephalus.*

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INTRODUCTION

Tectal gliomas occur in the tectal region at the roof of the brainstem. Occasionally tectal plate is a rare location for a tumor. They typically represent a unique subset of low-grade tumors. Brainstem gliomas in childhood account for 20% or more of primary brain tumors.[1] Brainstem gliomas, depending on their location, are often grouped as intrinsic pontine gliomas, tectal plate gliomas, focal brainstem tumors, dorsal exophytic tumors and cervicomedullary tumors. These tumors tend to develop spontaneously, without an identifiable cause or genetic link, and they are generally assumed to follow a benign course.[2] Symptoms of tectal glioma typically include those caused by increased intracranial pressure due to obstructive hydrocephalus. Symptoms may also include abnormal eye movements and/or Parinaud's phenomenon.[3] Parinaud phenomenon is classically described by the triad of impaired upward gaze, convergence retraction nystagmus, and pupillary hyporeflexia. The tectal glioma can be detected incidentally when imaging is performed for another indication. Tectal plate gliomas tend to progress slowly and they present with few neurological symptoms unless they produce obstructive hydrocephalus with intracranial hypertension.[4] Obstructive hydrocephalus can be treated with ventriculoperitoneal (VP) shunt, and a wait-and-see approach followed by imaging may be the treatment of choice for silent tectal plate gliomas.

Anatomical localizations of tectal tumors

The midbrain is the most superior portion of the brainstem. There are three parts of the midbrain that include: The *tectum*, the *tegmentum* and the cerebral peduncles. The *tectum* is the region of the midbrain posterior to the cerebral *aqueductus* of Sylvius. It contains the nuclei of the superior and inferior *colliculi*. These *colliculi* are involved in preliminary processing of the vision (superior *colliculi*) or audition.[5.]

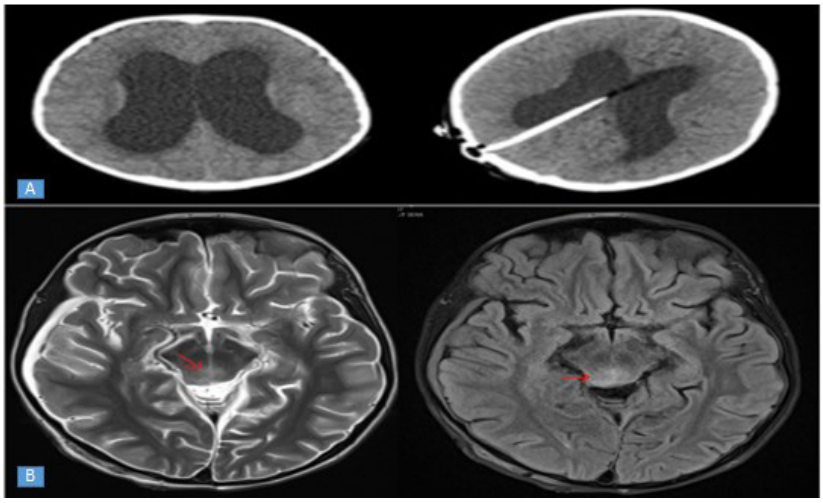
Between the cerebral aqueduct and the pars compacta of the *substantia nigra*, is the larger portion of the midbrain called the *tegmentum***.** The *tegmentum* in the broader sense extends through the whole brainstem, but its superior portion forms a part of the midbrain.[5]

We present two cases of pediatric patients with mesencephalic tectal plate tumors treated in our institution in 2023. The first case was an 11-year-old boy and the second case was a 15-year-old girl.

Case 1

A 11-year-old boy presented to the Emergency Department of our hospital with headache, nausea, double vision and visual blurring. *Pseudotumor cerebri* was the first preliminary diagnosis considered. The computed tomography (CT) scan revealed a noncommunicating triventricular hydrocephalus and a hypodense lesion on the tectal region *(Figure 1)*. Magnetic resonance imaging (MRI) demonstrated a tectal

Figure 1. Case 1



A: *CT axial section. Triventricular moderate to severe hydrocephalus is notable; preoperatively (left) and after VP-shunt surgery (right).*
B: *MRI T1 and T2 axial imaging. 17 x 13 mm mass lesion was observed in the mesencephalon, located in the tectum (mainly involving the right superior colliculus) and extending slightly exophytically towards the ambient cistern (red arrows). It caused obstruction at the aqueduct level.*

plate lesion hypointense on T1-weighted images and hyperintense on T2-weighted images with lack of contrast enhancement ***(Figure 1)***. It was a 17 x 13 mm tumor located in the *tectum* and slightly extending into the *ambient* cistern.

Since we do not have an endoscope at our institution, the VP-shunt was the first option in our cases. He initially underwent VP shunt placement achieving adequate control and improvement of the symptoms. He was discharged a week later. The patient was monitored with imaging studies on an outpatient basis and received radiotherapy. In the patient's evaluation at 6 months, no neurological or clinical worsening was detected, and no tumor growth was observed radiologically. The VP shunt is functional.

Case 2

A 15-year-old girl was admitted with headache. The patient had a history of trauma following a fall 6 months ago. CT and MRI showed mild ventriculomegaly, a T2 FLAIR hyperintense 18 x 28 x 24 mm lesion that thickened the tectum, surrounded the edges of the 3rd ventricle, significantly narrowed the superior *aqueductus* of Sylvius and extended towards the lower cerebellar peduncle on the left. In the single voxel MR spectroscopy examination performed for the T2 FLAIR the hyperintense lesion, that significantly thickens the tectum, extends towards both *thalamus*, surrounds the edges of the 3rd ventricle, narrows the *aqueductus* of Sylvius superiorly, and extends towards the inferior cerebellar peduncle on the left. The cholin peak was increased and the tumor was evaluated primarily in favor of midline glioma *(Figure 2)*. Considering the absence of deterioration or progression of symptoms, no additional studies were performed to evaluate intracranial hypertension. It was decided that the patient did not have surgical indication and was followed up clinically and radiologically on an outpatient basis. The patient applied to the radiation oncology clinic and received radiotherapy.

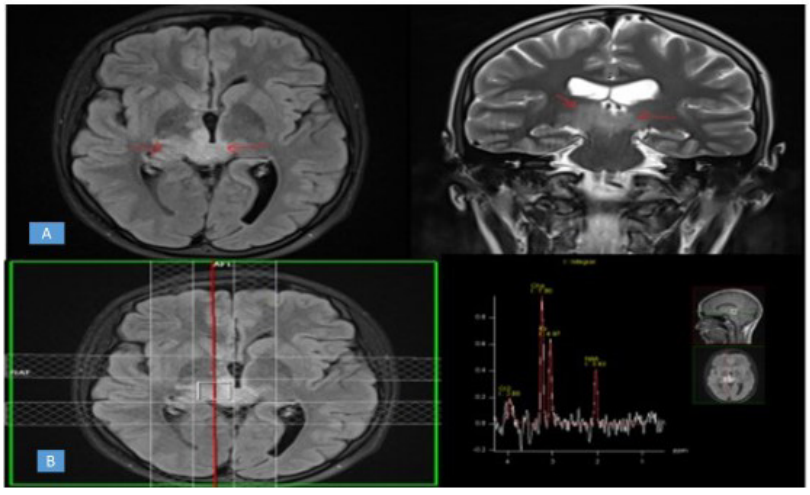
DISCUSSION

Brainstem gliomas without pontine involvement are almost always low-grade tumors.[6-8] The progression rates of brainstem gliomas vary depending on the histology of the tumor, its location within the brainstem, clinical symptoms, and response to treatment.

Tectal plate gliomas are a distinct subgroup of brainstem gliomas with slow clinical progression. They are usually diagnosed in childhood and often occur in adults. The average age at diagnosis is 7-10 years.[1]

Dabscheck et al. evaluated at their institution, clinical records of subjects under 21 years of age at the time of diagnosis of tectal glioma.

Figure 2. Case 2



A: *MRI T1 axial and T2 coronal images: lesion that significantly thickens the tectum, extends towards both thalamus, surrounds the edges of the 3rd ventricle, narrows the aqueductus of Sylvius superiorly, and extends towards the inferior cerebellar peduncle on the left (red arrows). Ventriculomegaly.*
B: *Single voxel MR spectroscopy examination performed for the T2 FLAIR: hyperintense lesion. Cholin peak was increased and the tumor was evaluated primarily in favor of midline glioma.*

They compared the demographic characteristics of 66 patients with tectal glioma. 56 patients underwent VP shunt placement.[3] Our cases were diagnosed at the ages of 11 and 15. Since tectal plate lesions are located close to the cerebral *aqueduct* of Sylvius, they slowly obstruct them causing hydrocephalus. In both of our cases, hydrocephalus was observed and a VP-shunt operation was required in the 11-year-old male patient.

In most patients, symptoms occur due to hydrocephalus. Bauman et al. reported a systematic review for tectal gliomas. Their review included 14 studies with 355 patients. They found that abnormal ocular findings (gaze palsies, papilledema, diplopia, and visual field changes) were most common at presentation as in our first case. Cerebrospinal fluid diversion was the most performed procedure (in the 89.3% of the patients).[9]

When hydrocephalus is diagnosed, patients often undergo surgical intervention such as endoscopic third ventriculostomy (ETV) or VP-shunt operation. It is known that auriculoventricular derivation is also a valid option in specific cases. ETV is the option of choice whenever indicated.

Annual radiological follow-up may be sufficient in monitoring patients with tectal plate gliomas. If the tumor is small the contrast enhancement may not be seen.[10] The increase in tumor size and contrast uptake should be assumed as tumor progression.

Lázar et al. presented in their study, seven patients with mesencephalic tectal plate tumors treated in their institution, between 1994–2005. Their ages were betwen17 and 70 years old.[11]

If the tumor is larger than 2 cm in diameter, is an enhancing lesion and has spread to nearby structures including the pons, this may indicate a malignant tumor. Depending on the location, 91% of tumors seen in the pontine and only 2% of those that are not pontine are considered malignant. In both of our cases, cholin peak was observed in MRI spectroscopy. If the tumor progresses, surgery, radiotherapy or chemotherapy may be necessary. Radiological tumor progression occurs in 15-25% of cases.[12]

In summary tectal gliomas are often benign, slowly progressing lesions. They are mostly diagnosed incidentally; if hydrocephalus develops, they may cause symptoms due to increased intracranial pressure. After the VP-shunt operation if the patient does not have hydrocephalus or neurological symptoms, annual radiological follow-up may be sufficient. ■

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