Sudden intraaqueductal dislocation of a third ventricle ependymoma causing acute decompensation of hydrocephalus

Case report

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The authors report an unusual sudden intraaqueductal dislocation of a third ventricle ependymoma causing acute loss of consciousness, and they detail its neuroendoscopic treatment. This case is unique and has never been described in the literature before.

The patient suffered from headache and recurrent nausea. Admission MR images revealed a contrast-enhancing lesion within the posterior part of the third ventricle. During the course of the day of admission, the patient suddenly became comatose. Emergency MR imaging demonstrated a deeply intraaqueductal dislocation of the pedicled tumor, with complete obstruction of the aqueduct and ventricular dilation. Immediate surgical intervention with endoscopic third ventriculostomy and complete tumor removal was performed. After surgery, the patient made a rapid recovery.

In this case, the authors presume a sudden intraaqueductal dislocation of the ependymoma caused by aspiration of the tumor as a result of the caudal CSF flow. (DOI: 10.3171/2011.5.PEDS10414)

Key Words • ependymoma • third ventricle • neuroendoscopy • intraventricular tumor • endoscopic tumor removal • hydrocephalus

Ependymomas are neuroepithelial tumors that arise from the ependyma of the ventricles. They are rare, and account for only approximately 3%–9% of all neuroepithelial tumors in the CNS. Usually they occur in the first decade of life. Most ependymomas originate within the fourth ventricle, rarely in the lateral, and even less frequently within the third ventricle. Because of their intraventricular location, ependymomas usually become symptomatic because of an obstruction of the CSF pathway, with a resulting occlusive hydrocephalus. We report for the first time an unusual sudden intraaqueductal dislocation of a third ventricle ependymoma causing acute loss of consciousness, and we detail its neuroendoscopic treatment.

Case Report

History and Examination. This 12-year-old boy suffered from intermittent headache and recurrent nausea for approximately 2 days. One day prior to admission he started vomiting. The family doctor presumed that the boy had a viral infection and treated him with analgesics and antiemetics. On the following day, the patient was admitted to the hospital because of deterioration of the symptoms. Admission MR images revealed a contrast-enhancing lesion within the posterior part of the third ventricle arising from the tectum and causing an occlusive hydrocephalus due to aqueductal obstruction (Fig. 1). Therefore, the patient was transferred to our institution and endoscopic third ventriculostomy and tumor biopsy were planned for the next day. However, during the course of the day of admission, the patient suddenly became comatose. Emergency MR imaging demonstrated a deeply intraaqueductal dislocation of the pedicled tumor, with complete obstruction of the aqueduct (Fig. 2).

Operation. Immediate surgical intervention with endoscopic third ventriculostomy and complete tumor removal was performed. The procedure was done using the Gaab universal neuroendoscopic system (Karl Storz GmbH and Co.) after general anesthesia had been induced. After a right precoronal bur hole had been placed, the operating sheath was inserted freehand into the right
lateral ventricle. The endoscope was navigated through the foramen of Monro into the third ventricle. An endoscopic third ventriculostomy was made halfway between the mammillary bodies and infundibular recess. Afterward, the tumor was inspected at the entry of the aqueduct, and was blocking it like a cork (Fig. 3). The tumor was barely vascularized and attached only with a small pedicle to the tectal plate. In accordance with the preoperative MR images, there was no endoscopic evidence for tumor dissemination. The lesion could easily be dissected and totally removed. After hemostasis had been achieved, the endoscope was removed and the skin closed in layers. The insertion of an externalized ventricular drain was not necessary.

Postoperative Course. After surgery, the clinical symptoms improved rapidly. At the time of discharge, the boy had no neurological deficits. The histological examination of tissue samples revealed an ependymoma (WHO Grade III). An MR imaging study obtained 5 days after surgery demonstrated complete removal of the tumor (Fig. 4). We concluded that the contrast enhancement in the rostral aspect of the aqueduct was a postoperative dysfunction of the blood-brain barrier at the resection site, because preoperative MR images demonstrated no enhancement of this area.

Further imaging of the central neuraxis yielded no evidence of metastatic disease. The patient was treated with adjuvant radiation and chemotherapy according to the protocol HIT 2000, including 60 Gy of radiation. To date, after 5 years of follow-up, no tumor recurrence has been observed.

Discussion

To the best of our knowledge, we present for the first time a case of a sudden occlusion of the aqueduct caused by a deeply intraaqueductal dislocation of a pedicled ependymoma arising from the tectum. Whether tectal tumors are small or large, they may cause an occlusion of the cerebral aqueduct, and a subsequent hydrocephalus may occur. According to Kernohan and Sayre,4 tectal tumors are supposed to be the smallest lesions in the body that may lead to the death of patients. However, usually, the symptoms of increased intracranial pressure (ICP) develop more slowly. Only a few reports of patients who presented with acute neurological deterioration and sudden death caused by lesions of the pineal region, the tectum, or the aqueduct have been published in the literature. Six cases of pineal cysts5,7,8,10,11 have been described
as responsible for a sudden death because of an acute obstruction of third ventricular CSF outflow. This is in contrast to colloid cysts, which usually block third ventricular inflow. Intralesional hemorrhage might be the cause in some cases. Hemorrhage may lead to rapid expansion of the lesion, resulting in compression of the aqueduct and a sudden increase in ICP.

In our patient, we saw a sudden intraaqueductal dislocation of the ependymoma caused by the caudal CSF flow, which aspirated the tumor into the aqueduct. The sudden neurological deterioration was caused by the acute elevation of ICP in a patient with preexisting hydrocephalus. We think that in the situation shown in Fig. 1, the CSF flow is still not completely blocked. Only after tumor dislocation was the CSF flow completely stopped and, in our opinion this explains the sudden deterioration of the patient. Of course, the increased ICP may have contributed to the movement of the tumor into the aqueduct.

Our case required only 1 bur hole located approximately 2 cm in front of the coronal suture, to access both the floor of the third ventricle and the posterior part of the third ventricle. On the MR images obtained prior to the surgery, we saw a large foramen of Monro, which allowed a tilting of the endoscope within the foramen to visualize the ventricle floor and entry of the aqueduct. Of course there is a risk of damaging sensitive structures such as ependymal veins or the fornix during this maneuver. However, with careful insertion and slow movements of the scope, the risk seems to be low. It is of utmost importance to analyze the preoperative MR images very accurately. Of course, a small foramen of Monro requires a 2-bur-hole approach when a rigid scope is used, because injury of veins and/or fornix is very likely.

Conclusions

In our opinion, endoscopic management with third ventriculostomy and gross-total tumor resection is the procedure of choice in these cases, even in this emergency situation, when an experienced neuroendoscopist is available.

Disclosure

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Author contributions to the study and manuscript preparation include the following. Conception and design: Rosenstengel, Schroeder. Acquisition of data: Müller. Drafting the article: Rosenstengel. Critically revising the article: all authors. Reviewed submitted version of manuscript: all authors. Approved the final version of the manuscript on behalf of all authors: Rosenstengel.

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