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CASE REPORT

Endoscopic third ventriculostomy and biopsy of a low-grade glioma

Terceiroventriculostomia e biópsia endoscópica em glioma de baixo grau: relato de caso

Maina Tavares Zanoni[1], Vírgina Resende Silva Weffort[1], Roberto Alexandre Dezena[2], Daniel Fonseca Oliveira[2], Raphael Guerra David Reis[2], Rafaela D'Angelo dos Reis[2]

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Abstract

This paper reports the case of a 5-year-old girl with clinical signs consistent with intracranial hypertension caused by hydrocephalus secondary to a posterior fossa tumor, which included headaches, difficulty walking, and vomiting. Head computed tomography (CT) and brain magnetic resonance imaging (MRI) scans confirmed the clinical diagnosis. The patient underwent an endoscopic third ventriculostomy to manage the hydrocephalus and have the tumor biopsied. She improved clinically and the hydrocephalus resolved. The patient was diagnosed with a low-grade glioma.

[1] Universidade Federal do Triângulo Mineiro, Pediatria - Uberaba - Minas Gerais - Brazil.
[2] Universidade Federal do Triângulo Mineiro, Neurocirurgia - Uberaba - Minas Gerais - Brazil.

Correspondence to:
Maina Tavares Zanoni
Universidade Federal do Triângulo Mineiro (Departamento de Pediatria). Avenida Getúlio Guarita, nº 130, Nossa Senhora da Abadia, 2° Andar, Hospital de Clínicas. Uberaba - MG. Brazil. CEP: 38025-440. E-mail: E-mail: mainazanoni@gmail.com

INTRODUCTION

Hydrocephalus is a condition characterized by a mismatch in the production and absorption of cerebral spinal fluid (CSF) resulting in increases in the size of the brain ventricles. It may be divided into communicating – when there is communication between the ventricles and the subarachnoid space – or obstructive hydrocephalus – when the flow of CSF is impeded in the ventricles or subarachnoid space.[1] Neuroendoscopy has had many uses in surgery, with endoscopic third ventriculostomy (ETV) ranking as the most commonly performed neuroendoscopic procedure in the treatment of children with obstructive hydrocephalus.[2] Gliomas are tumors of the glial cells that preferentially affect the posterior fossa in children and cause obstructive hydrocephalus.[3]

Case Presentation

A five-year-old girl was brought to a care center after presenting episodes of vomiting for two weeks. She had also experienced adynamia, difficulty walking, imbalance, and vertigo during a trip with her family. The clinical signs prompted the attending physician to order a computed tomography (TC) scan of the head, which revealed she probably had obstructive hydrocephalus. The patient was discharged after a prescription of topiramate to reduce the production of CSF. Her clinical signs worsened when she arrived at her hometown, and she was taken to a medical service where a contrast-enhanced CT of the head was ordered. The patient was diagnosed with hypertensive obstructive hydrocephalus caused by a tumor located in the posterior portion of the third ventricle (Figure 1). She was referred to a pediatric intensive care unit (PICU), where magnetic resonance imaging (MRI) of the brain was ordered the following day so that she could undergo surgery as soon as possible. The brain MRI scans showed a mass in the region close to the pineal gland and the inlet of the cerebral aqueduct with a tri-axial component consistent with a low-grade tumor – possibly a germ cell or glial tumor – in addition to significant obstructive hydrocephalus with signs of hypertension (Figure1). Neuroendoscopic surgery was chosen based on these findings. The patient was placed under general anesthesia while on a supine position. Trephination was performed on the right Kocher's point to allow access to the right lateral ventricle and the third ventricle. ETV was performed inside the site communicating the ventricular cavity and the interpeduncular cistern (Figure 2). With the neuroendoscope at a posterior angle, a tumor was seen protruding through the inlet of the cerebral aqueduct. Electrocoagulation was performed and fragments of the tumor were removed (Figure 2). The patient was sent back to the PICU after surgery, where she was extubated without complications. The patient was hemodynamically stable. A CT scan of the head performed one day after surgery revealed a small reduction on the size of the ventricles and minor bleeding on the biopsy site without additional significant findings. The patient improved two days after surgery and was referred to the pediatric ward, from which she was discharged three days later. She was asymptomatic, without fever, her gait improved significantly, and her surgical wound was in good shape,

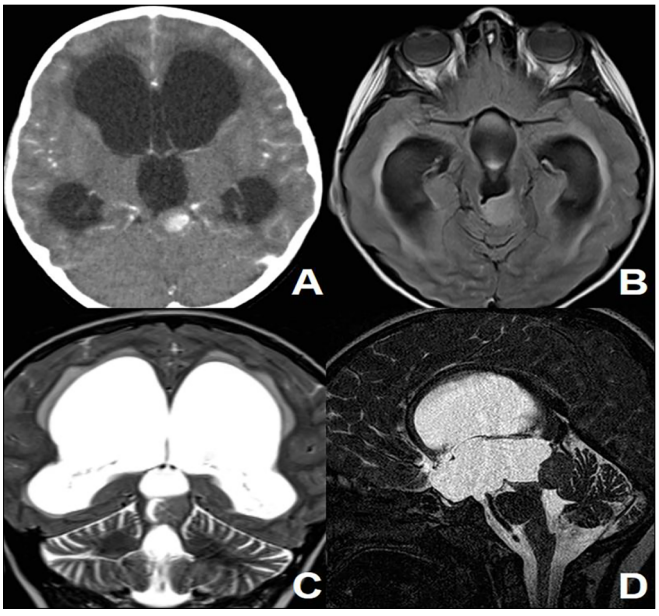


Figure 1. Preoperative imaging. Contrast-enhanced computed tomography scan of the head showing a mass on the right lateral portion of the midbrain **(A).** Axial magnetic resonance imaging scan, FLAIR **(B),** coronal T2 **(C)** and sagittal T2 **(D)** views, showing the intra-axial nature of the mass.

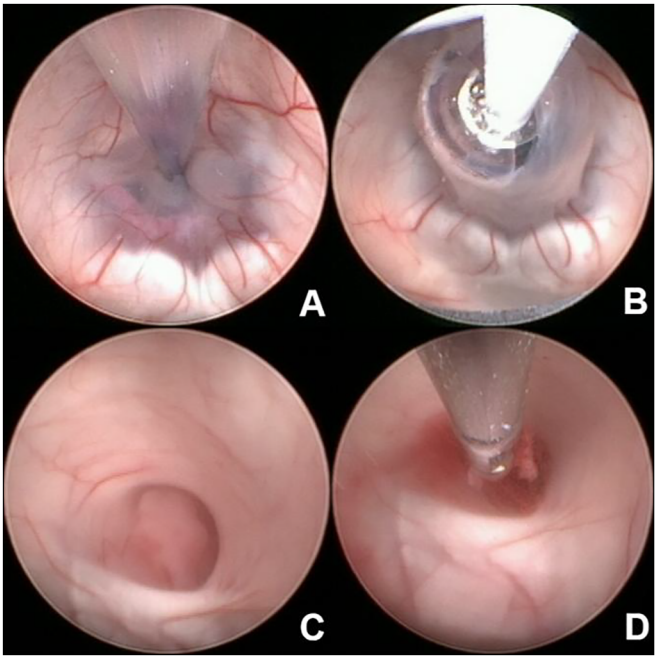


Figure 2. Intraoperative imaging. Third ventriculostomy showing electrocoagulation of the tuber cinereum **(A)** and dilatation of the stoma using a Fogarty catheter **(B).** Tumor biopsy showing a mass protruding through the inlet of the cerebral aqueduct **(C)** and removal of tumor fragments after electrocoagulation **(D).**

requiring only analgesia. Ten days after discharge, the patient came back for a follow-up consult. She was asymptomatic, off medication, and her gait improved further. The pathologist report indicated she had a tumor consistent with a low-grade glioma; immunohistochemistry agreed with the pathologist report and showed positive results for GFAP and OLIGO2, expression of protein H3.3 K27M and partial loss of expression of protein H3.3 K27me, and negative results for IDH1 (R132H).

DISCUSSION

CSF shunts – particularly into the peritoneal cavity – are often used to treat children with hydrocephalus with a setup including an outflow catheter and a valve. Unfortunately, this procedure may fail by obstruction and infection in the shunt system.[1] Endoscopic approaches have gained relevance in the management of pediatric hydrocephalus for not requiring grafts.[2]

The main endoscopic approaches are ETV for obstructions leading to third ventricle dilatation and neuroendoscopic septostomy for communicating lateral ventricles and isolated lateral ventricle in cases of complex hydrocephalus.[4]

ETV is the most commonly performed neuroendoscopic procedure worldwide.[2] The procedure involves opening the floor of the third ventricle to allow CSF to flow away from the ventricular system into the subarachnoid space.[4,5] Indications include primarily the treatment of obstructive hydrocephalus affecting areas ranging from the posterior portion of the third ventricle to the median and lateral apertures of the fourth ventricle.[6] Some conditions must be present to ensure the success of the procedure, such as having a patent subarachnoid space and intact CSF absorption in the arachnoid granulations.[4]

ETV is the main intervention prescribed to tumors in the posterior region of the third ventricle and in areas close to the pineal gland, as in the case in question, since it can be used not only to treat hydrocephalus, but also to provide a window for biopsy, CSF analysis, and reveal tumor implants.[2,4,7] Recent studies listed the following as predictors of ETV success: age greater than six months with stenosis of the aqueduct or tectal tumor, and lack of an anterior shunt. Outcomes are better in patients undergoing ETV as a primary treatment for obstructive hydrocephalus without evidence of previous infection or hemorrhage.[2,4,7,8] As other surgical procedures, ETV is not free of complications.[2] They include ventricular wall bleeding, large vessel bleeding (including veins and branches of the basilar artery), hypothalamo-pituitary disconnection, and pseudoaneurysm of the basilar artery.[9,10] In terms of outcome, clinical improvement is more evident than the improvement seen from imaging findings, since most of the individuals with hydrocephalus present with largely non-compliant ventricles that often do not return to their original sizes.[2,8]

Tumors of the central nervous system account for 20% of the malignant diseases affecting children, 30% of which are gliomas associated with most of the deaths caused by primary brain tumors.[3] Clinical manifestations commonly associated with this tumor type include sudden-onset clinical events without a source, headache, vomiting, visual impairment, gait disorders, ataxia, and seizures. These symptoms are commonly associated with involvement of the posterior fossa in children and increased intracranial pressure derived from obstruction to CSF flow.[7]

Our patient had a diffuse midline glioma harboring the H3 K27M (Histone 3-Lysine-27-Methionine) mutation. This type of tumor is predominantly found in children (aged 5-11 years; both sexes equally affected), although individuals in other age ranges may develop it. It is more commonly found in the brainstem, thalamus, and spinal cord. Tumors located in the brainstem with tumor cells diffusely involving local and distal structures are assigned the status of Grade IV by the WHO.[11]

Nearly all tumors express NCAM1 (CD56), S100, and OLIGO2 in immunohistochemistry. Positive findings for GFAP and MAP2 vary.[3] Although patients with tumors harboring the H3 K27M mutation have a poor prognosis, traits such as low histological grade and having a circumscribed tumor may indicate the presence of less aggressive disease.[11] Despite the sensitive location, surgical resection is the basis of treatment in most cases, with chemotherapy and radiation therapy prescribed depending on patient findings, particularly when atypia is observed.[3,11] Steroids to decrease brain edema and anticonvulsants for patients with seizures may be useful in pre- and postoperative care.[3]

CONCLUSION

Endoscopic third ventriculostomy has been used with good results to treat individuals with hydrocephalus associated with central nervous system tumors. The effectiveness of ETV is closely tied to judicious indication of the procedure. In addition to its use in treating hydrocephalus, ETV also allows the harvesting of biopsy specimens of intraventricular tumors.

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