A Novel Treatment and its Complications in Pediatric Neuro-Oncology: Encephaloclastic Cyst Induced by Intraventricular Topotecan in a Pediatric Patient with an Atypical Rhabdoid Teratoid Tumor

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Abstract

Advances in cancer treatment and multidisciplinary working teams are essential to provide the best chance of survival and a good quality of life for patients with brain tumors. With the advent of new treatments, we recorded isolated cases of rare complications. Here we report a treatment-associated complication, which has not been previously described in the literature, in a pediatric patient with an atypical rhabdoid teratoid tumor. Within the therapeutic scheme for this entity, intraventricular topotecan is administered through an intraventricular Ommaya reservoir. The formation of a symptomatic encephaloclastic cyst and its surgical resolution with complete clinical recovery is described.

Keywords: Encephaloclastic Cyst; Ommaya Reservoir; Topotecan; Oncological Treatment; Atypical Rhabdoid Teratoid Tumor

Abbreviations

ATRT: Atypical Rhabdoid Teratoid Tumor; CSF: Cerebral Spinal Fluid

Introduction

The atypical rhabdoid teratoid tumor (ATRT) of childhood is a malignant tumor (Grade IV of the WHO classification) distributed in a similar manner between the supra and infratentorial space, with an average age of presentation between 2 and 4 years. After surgical treatment, an early oncological management is mandatory.

Currently, one of the therapies options consists of the administration of intrathecal topotecan through an Ommaya reservoir.

The Ommaya reservoir is a quick and safe tool for intrathecal administration of chemotherapy.

Case Report

We present the case of a 2-year-old female patient with no relevant history of disease who debuted with a left-sided brachio-crural hemiparesis, which motivated the consultation. The diagnosis of a right supratentorial tumor was made. Surgery was performed twice because of residual tumor (Figure 1). Histology revealed an atypical rhabdoid teratoid tumor.

One month after tumor removal, an Ommaya reservoir was placed in the left frontal horn for twice-weekly administration of intrathecal chemotherapy (Figure 2) [1]. Nine months after treatment initiation, the patient developed right-sided crural hemiparesis with gait impairment and generalized deterioration. CT scan and MRI of the brain were performed evidencing pseudocystic formation associated with the catheter path (Figure 3 and Figure 4). Chemotherapy was discontinued. A sample of cerebrospinal fluid (CSF) was obtained from

the reservoir [2]. The cell count and glucose and protein levels were normal. CSF culture was negative. The brain CT scan was repeated with iodinated contrast (iopamidol) through the reservoir, ruling out leakage into the parenchyma (Figure 5). Administration of intraventricular topotecan was discontinued and the intraventricular reservoir was removed. After one month, complete clinical recovery and resorption of the encephaloclastic cyst was observed on serial images (Figure 6 and Figure 7). Once the patient recovered, systemic oncological treatment was reinstated. Currently, after 20 months of follow-up, the patient shows no tumor recurrence and has recovered independent gait with left hemiparetic gait sequelae associated with the tumor, but no complication-related sequelae.

**Figure 1:** Enhanced T1-weighted MRI with residual tumor after the first surgery.

**Figure 2:** CT scan showing the catheter in the left frontal horn, without residual tumor

**Figure 3:** CT scan showing the encephaloclastic cyst associated with the catheter path.
Discussion

The placement of an Ommaya reservoir facilitates intrathecal drug delivery. Topotecan rapidly clears from the CSF after the administration of an intraventricular dose which makes it a safe drug with low neurotoxicity [3]. The most common side effects of topotecan are fatigue, fever, nausea, and CSF pleocytosis for 24 to 120 hours after the drug is administered [3-7]. The formation of a catheter-associated

encephaloclastic cyst after intrathecal administration of topotecan is an atypical complication. Only two cases have been described in the literature in adult patients with breast cancer and central nervous system (CNS) metastasis [8]. To the best of our knowledge, ours is the first report of this complication in a pediatric patient. The pathophysiology possibly underlying the formation of the encephaloclastic cyst are changes in CSF pulsations with retrograde flow of chemotherapy to the cerebral parenchyma and the subsequent development of a chemical encephalitis rather than leakage of the substance during its instillation, as this latter mechanism could be ruled out in our patient by infusing iopamidol [8].

**Conclusion**

In conclusion, an encephaloclastic cyst is a rare complication of intraventricular treatment in patients with brain tumors. Intraventricular treatment discontinuation and the removal of the intraventricular reservoir resolved the problem resulting in clinical recovery.

**Conflict of Interest**

Authors report no conflict of interest.

**Bibliography**


