

PALLIATIVE TREATMENT IN A PEDIATRIC PATIENT WITH SEVERE MACROCEPHALY CAUSED BY A BRAIN CYST: CASE REPORT

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Abstract: This is a clinical case of a pediatric patient with severe macrocephaly caused by a brain cyst, who had unsuccessful surgical interventions and underwent palliative care. The importance of a multidisciplinary approach and palliative care is highlighted to alleviate suffering and improve the quality of life of patients and their families, in situations of therapeutic limitation. The report also highlights the need for more research into serious neurological conditions in pediatric patients to establish an early approach and avoid serious neurological complications.

INTRODUCTION

The concept of “cerebral neoplasia”, which involves abnormal cell proliferation in the brain, either locally or through metastasis (1). The World Health Organization has categorized more than 100 types of brain neoplasms into 13 distinct categories, each with specific symptoms (2). Furthermore, non-neoplastic brain cysts can mimic tumor symptoms (3). This case study describes a patient with a brain cyst that resulted in severe macrocephaly. Macrocephaly in pediatric patients, when associated with brain cysts, has implications for neurological and physical development (4). This article presents a clinical case of palliative treatment in a pediatric patient with severe macrocephaly due to a brain cyst, focusing on improving quality of life and relieving symptoms. Although a cure for macrocephaly is not possible, treatment seeks to provide comfort to the patient and support for the family (5).

CASE PRESENTATION

We report the case of a patient who was born vaginally, the third child of a mother who had two children with another father. The patient was born without vital signs and was resuscitated. He weighed 4100 g and had a head circumference of 38 cm at birth, but

did not cry or breathe spontaneously. He remained hospitalized for three days in the shared accommodation, without the need for a neonatal ICU. Due to macrocephaly, a computed tomography of the skull was requested, which revealed moderate ventriculomegaly, with mild asymmetry of the lateral ventricles and an area of focal left frontal corticosubcortical hypodensity, suggestive of encephalomalacia. He was referred to a more complex service for evaluation and follow-up by neurosurgery, which indicated the performance of a ventriculoperitoneal shunt (VPD). However, the family did not have the financial means to purchase the catheter and only managed to acquire it three months after surgery was indicated. During this period, the patient showed progressive worsening of his head circumference (reaching up to 87 cm) and neuropsychomotor development. On 06/03/2022, he underwent the first PVD, without complications. The following day, he developed a cerebrospinal fluid leak and was taken to the general surgical center (CCG), being admitted to the pediatric ICU after the procedure.

Mother reports that she came to Brazil brought by her mother to seek dignified and adequate care, as health conditions in the city where her mother lived were not good. After 3 days of arriving in Brazil, he presented new episodes of recurrent seizures. Taken by his mother for evaluation at an emergency room, where he received a dose of Diazepam and was sent for evaluation at a pediatric reference center.

Patient using valproic acid and L-carnitine, and feeding on infant formula. Incomplete vaccination schedule and allergic to dipyrone and paracetamol. He needed neurological, nutritional, infectious and hematological evaluation, as well as care for skin lesions and control of seizures.

Upon admission to the pediatric emergency room, the patient was in regular general condition, presenting with a generalized clinical tonic seizure and fever, weight of 9300 g, head circumference of 83 cm, signs of intracranial hypertension, such as the setting sun look, enlarged and tense fontanelle, and ulcer pressure with exposure of the catheter in the occipital region. Immediately after admission, an evaluation was requested from the Pediatric Intensive Care Unit (PICU) and the neurosurgery team. An urgent ventriculo-external shunt (LVD) was indicated and admission to the pediatric ICU due to the diagnosis of hydrocephalus, bacterial meningitis and possible brain abscess. forwarded to the surgical center to perform an external ventricular shunt (EVD) and remove the old PVD, showing yellowish liquor with debris when the old PVD was removed.

Post-surgical control computed tomography (CT) of the skull showed that the EVD catheter was outside the lesion, and that the lesion could be a brain abscess. A new approach was planned for the following day. The patient remained stable, awake and slightly irritated, without fever or hemodynamic changes. He received broad-spectrum antibiotics covering the central nervous system (CNS) due to suspected bacterial meningitis and an enteral diet. After the procedure, the patient remained in a regular general drowsy state, looking like the setting sun, slightly tense enlarged fontanelle, with occasional seizures.

The following day, the patient underwent a new EVD exchange. two catheters were passed: in Keen's point on the left and Kocher's point on the left. in Keen's point, normotensive crystalline liquor is released. in Kocher point, with output of turbid yellowish material with macroscopic crystals, slightly hypertensive, not fetid. Uneventful procedure. After a neurosurgical approach, magnetic resonance

imaging (MRI) was requested to evaluate the possibility of resection of the intracranial cyst. CSF analysis of the intracranial cyst content: high protein content. Assessed by neuropediatrics post-operatively, who advised starting baclofen 1.25 mg every 6 hours, and maintaining valproic acid.

Skull MRI showed a wide area of diffuse cerebral encephalomalacia (sequelae of old brain injury) without intracranial solid neoplastic lesions and with septate ventricles.

In the reevaluation by neurosurgery with MRI results, the patient was considered to have severe neurological impairment. Laboratory evaluation indicates high protein content that makes the peritoneal diversion procedure unfeasible and with MRI a neoplastic lesion was excluded. Patient with poor prognosis. Patient with no indication or conditions for a neurosurgical procedure for resection of an intracranial lesion due to a high risk of death, due to the size and location of the lesion.

A multidisciplinary meeting was held (neuropediatrics, neurosurgery, pediatric palliative care, PICU, general pediatrics) together with the child's mother and stepfather; opted to proceed with an exclusive palliative care protocol. After ruling out a surgical procedure to remove the lesion, the neurosurgery team removed the EVDs: one of them is now in the ventricle and the other, which is in the cyst, currently has low output; Neuroimaging shows that the cyst is drained and there is a risk of infection if the EVD persists for a long time.

After finishing antibiotic treatment for neuroinfection, anticonvulsant therapy was adjusted by neuropediatrics and a gastrostomy was requested to ensure a route of nutrition to improve the severe malnutrition that the patient had at admission and due to the severe neurological sequelae and difficulty in sucking evidenced in the evaluation by speech therapy.

Patient who was discharged from hospital with guidance for outpatient return for pediatric palliative care, neurosurgery, pediatric neurology, nutrition and high-risk childcare. Upon outpatient return 4 months after the removal of the 2 EVDs, the patient had a head circumference of 56 cm, a decrease of 27 cm, and a weight of 12,450 g. The objective of palliative care was achieved, because the living conditions of the patient and family were optimized since they arrived from Venezuela to live in Brazil.

DISCUSSION

Palliative care focuses on alleviating suffering and improving the quality of life of patients with serious illnesses, including those with neurological involvement (1). This case highlights the complexity of managing patients with large brain cysts, especially when they present with a serious and irreversible neurological condition. Despite several intervention attempts, the patient's condition remained difficult to manage, emphasizing the need for a multidisciplinary approach to care (6). However, the decision to offer palliative care was based on several factors, including the patient's prognosis and the potential risks associated with additional interventions (7).

According to Bogetz and Lemmon, pediatric patients with severe neurological sequelae are generally affected by congenital or chromosomal disorders and face greater vulnerability and morbidity. Despite representing less than 1% of the pediatric population, they contribute significantly to length of hospitalization and pediatric intensive care unit (PICU) admissions. The complex nature of these symptoms, including discomfort, gastrointestinal complications, seizures and sleep disorders, requires a specialized and multidisciplinary assessment with pharmacological and non-pharmacological interventions (8).

Although the decision to provide palliative care can be challenging for patients and caregivers, it can offer significant benefits, such as pain and symptom control, emotional support and better communication between patients, caregivers and healthcare professionals (7). In a study focused on the feasibility of randomized clinical trials (RCTs) in pediatric palliative care in patients with childhood cancer, the implementation of an electronic reporting system on disease progression was evaluated, these reports were reported by the patient in a multicenter environment. The feasibility of the RCT of pediatric palliative care in advanced cancer cases was demonstrated, with satisfactory retention and altruistic motivations for participation. The main objective of this study was to evaluate the patient's insight into their own disease (9).

Management of patients with large brain cysts and severe neurological involvement presents significant challenges. A multidisciplinary approach is necessary to provide comprehensive care, make informed decisions, and help the family and patient understand and understand the illness they are experiencing. Wolfe, et al. addressed the lack of understanding of the experiences of children with advanced cancer. The need for intensive symptom management strategies is highlighted, especially in situations of progression or intense treatments in children with advanced cancer (3).

Adequate monitoring is essential, but coordination for subspecialty and home nursing assessments is everything. Families of these children face considerable caregiving responsibilities, financial hardship, and ongoing emotional distress as they navigate their child's medical journey. These parents demonstrate remarkable resilience, although they also go through grief and adapting to new routines (8).

Families seeking appropriate settings for their child's final moments and discussing goals of care often turn to palliative care specialists. However, obstacles such as poor collaboration between neurology and palliative care teams, prejudices among health professionals and misunderstandings prevent the full integration of palliative services (10).

In 2018, Eklund published a study that focused on the lack of support programs for families with seriously ill minor children and in the context of palliative care. It is one of the few studies on interventions in terminally ill patients and focused on the importance of family involvement in measures to increase the understanding and morale of sick children (5). The prospects of starting and maintaining early palliative care are viable, involving a diverse team that collaborates in different care environments (8;10).

The involvement of family members, the treating medical team and the nursing team in palliative care improves discussions about the end of life and treatment approaches for young patients in serious general condition. This type of care is effectively integrated into clinics that focus on neuromuscular diseases, providing relief, pain control and comprehensive end-of-life support (10).

CONCLUSION

Early diagnosis and intervention are crucial to prevent serious neurological complications and improve patient outcomes. In this case, unfortunately the neurosurgical approach to resection of the lesion had a very high risk of death but the patient benefited after the measures taken by pediatric palliative care. More research is needed to improve the management of similar cases and advance the field of pediatric neurology.

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