Original Article

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Cerebellar Mutism after posterior fossa tumor surgery: is it predictable, preventable, treatable?

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Introduction: Posterior fossa tumors pose challenges in pediatric neurosurgery and can lead to complications such as cerebellar mutism, affecting speech and motor abilities. This study aims to describe the evolution of cerebellar mutism and identify relevant factors in its pathophysiology and prognosis.
Methods: A review was conducted on three cases of children who underwent surgery for posterior fossa tumors by the same surgical team and developed postoperative cerebellar mutism. The study analyzed preoperative clinical presentation, mutism manifestation, and recovery. Additionally, a literature review was performed on the emergence, evolution, and prognosis.
Results: Case 1 involved a 3-year-old girl with medulloblastoma who experienced mutism, behavioral changes, and self-mutilation after surgery. Symptoms resolved after one month but recurred with a subsequent operation, resolving again after a month. Case 2 featured a 15-year-old girl who developed severe posterior fossa syndrome, including mutism, dysarthria, aphasia, and motor impairments. With support care, she gradually improved over a year. Case 3 involved a 4-year-old boy with cerebral palsy and a posterior fossa tumor. He experienced akinetic mutism, resolved over time with rehabilitation therapies.
Discussion: Cerebellar mutism following posterior fossa surgery has implications for patient recovery. It primarily involves impairment of cerebellar outflow pathways rather than direct brainstem or cortical lesions. Identifying at-risk patients remains challenging, though younger age and specific tumor characteristics may increase the likelihood of mutism. Rehabilitation, including physical and speech therapies, plays a crucial role in promoting recovery. Effective communication and support for patients and their families are essential during the recovery process.

Keywords: Posterior fossa tumors, Cerebellar mutism, Rehabilitation

INTRODUCTION

Posterior fossa tumors are the most frequent brain neoplasms in children [1] and, in the field of neurooncology, represent the most relevant topic in Pediatric Neurosurgery. Medulloblastomas, pilocytic astrocytomas and ependymomas, the most common histological entities, are located in close relationship with the cerebellum and the fourth ventricle, requiring manipulation and wide access to the region for the most complete excision possible, which influences the prognosis in terms of recurrence and disease-free survival [1].

Although the surgical technique has evolved considerably in recent decades, there are important complications to be considered in surgery for posterior fossa tumors in children, such as hemorrhages, lesions of nuclei and long tracts of the brainstem and cerebellar mutism, also called akinetic mutism due to the characteristic inhibition of movement that appears in some cases [2][3][4][5].

This condition affects approximately 25 to 30% of children operated on to remove tumors from the posterior fossa and presents a widely variable picture of clinical manifestations, involving difficulties in speech, behavior, alertness, and motor function. The severity of the condition is also quite variable, as well as the extent and delay of recovery[3][6].

Since cerebellar mutism became better recognized and systematically studied, several theories have been developed to explain its pathophysiology. Injury to the deep cerebellar nuclei has been predominantly involved in this discussion. The most up-to-date understanding points to the impairment of the cerebellar outflow pathways, especially those that travel through the superior cerebellar peduncles, playing a fundamental role [7][8].

In fact, the study of this condition has even contributed to a deeper understanding of the non-motor functions of the cerebellum, advancing in relation to the classic view that attributed a restricted role to the cerebellum in motor

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control [9][10]. The clinical status of children with cerebellar mutism confirms the existence of these functions.

There is also an extensive discussion about risk factors for the occurrence of cerebellar mutism, whether related to individual characteristics of the patient or aspects of the surgical technique. Although some studies point to the size of the lesion, the location in the midline, the age of the child, the diagnosis of medulloblastoma and the transvermian access as elements of increased risk, others indicate the difficulty of predicting which patients are more likely to present postoperative mutism [11][12].

In this context, the aim of this study is to describe the evolution of cerebellar mutism and identify factors involved in the pathophysiology and prognosis of the phenomenon.

METHODS

Three cases of children who underwent surgery to remove tumors from the posterior fossa and who developed postoperative cerebellar mutism were reviewed, with emphasis on the preoperative clinical presentation, the manifestation of cerebellar mutism and the evolution of recovery.

All 3 patients were operated on in the last 3 years by the same team in two different referral hospitals for Pediatric Neurosurgery in Fortaleza, Brazil. Case #1 had a first operation in other city and the neuroimaging exams related to the original diagnosis were not available.

Case reports complied with national ethical regulations and the project was approved by the hospital research ethics committee.

An extensive literature review was also carried out regarding the emergence, evolution, and prognosis of cerebellar mutism.

RESULTS

Case 1

Female patient, 3 years old, presented, 2 months before admission, paresis of the lower limbs, progressively impaired gait, frequent falls, recurrent fever, vomiting, lethargy, irritability and hyporexia. A brain MRI revealed a voluminous tumor filling the fourth ventricle, resulting in supratentorial hydrocephalus and transependymal edema. The patient was referred to a tertiary hospital, in a service other than ours, for tumor resection. In less than twenty-four hours after surgery, the patient developed mutism and behavioral changes, including irritability, emotional lability, aggressiveness and episodes of self-mutilation, which ceased after one month. One year after the onset of the initial symptoms, the patient again presented gait ataxia with associated drowsiness, being diagnosed with tumor recurrence (4,2cm AP x 2,0 width x 3,8 height), requiring a new surgical approach and beig this time referred to our reference center. She evolved again, in the immediate postoperative period, with behavioral disturbances, speech suppression, hypokinesia and hypotonia and could not remain seated without support. These symptoms resolved after one month post-operatively (Fig. 1). Histological analysis confirmed a medulloblastoma.

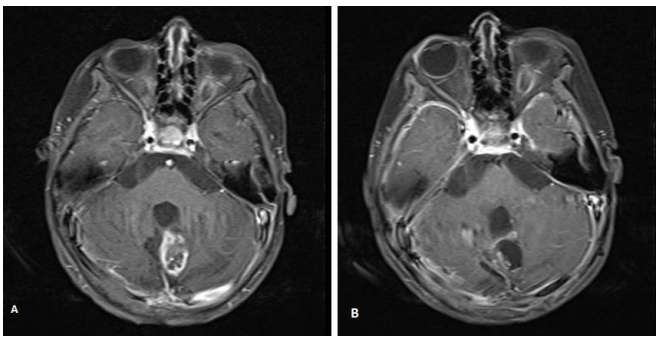

Figure 1 - A) Axial post-contrast T1 pre-operative MRI image of tumor before second operation (case 1). B) Axial post-contrast T1 post-operative image showing resection of the lesion.

Case 2

A 15-year-old girl was admitted due to severe headaches and vomiting. Neuroimaging revealed a fourth ventricle tumor with an extensive mass effect (4,8cm AP x 5,4 width x 5,2 height). She was operated and had near total tumor removal via a telovelar approach, and was diagnosed with classic medulloblastoma, as confirmed by the biopsy. Although surgery had no noticeable complications, she developed a severe posterior fossa syndrome, with immediate prominent mutism, initially presenting a marked degree of dysarthria, progressing to complete aphasia, associated with changes in mood and irritability. She had also an important impairment of motor functions and was restricted to the bed. Followed up in support care due to the inability to self-care and spontaneous swallowing, she was treated with radiotherapy and chemotherapy, staying long periods in hospital care. After one year, she started progressively improving motricity and communication, and became able to be transferred to a wheelchair and to talk with a minor degree of dysarthria.

Case 3

A 18 months old boy, born after a complicated twin pregnancy, required admission to the Neonatal Intensive Care Unit after birth. At 1 year of age, it was noticed that the patient had not reached the expected neurodevelopmental milestones, was not able to walk and had poor interaction. At age 1 year and 5 months, he was diagnosed with cerebral palsy, and put under physical therapy and speech therapy. One month later he started to present ataxia, hypotonia and

progressive neurological involution. This clinical picture was still attributed to prematurity and cerebral palsy but, as it kept worsening a brain MRI was performed and revealed a great posterior fossa mass inside the fourth ventricle (5,8cm AP x 4,6 width x 5,2 height) (Figure 2). Two days after receiving a venticuloperitoneal shunt to treat hydrocephalus, he was submitted to a telovelar approach and total removal of the tumor, posteriorly diagnosed as a pylocitic astrocytoma in biopsy. After surgery, he was hospitalized at the ICU for 30 days and, after 15 days, the patient woke up, but remained with poor interaction besides opening his eyes, characterizing akinetic mutism. After his discharge, the patient continued under physical therapy and speech therapy, and started progressive neurological improvement. At 4 years and 4 months old, he takes his first steps with a walker and is able to communicate.

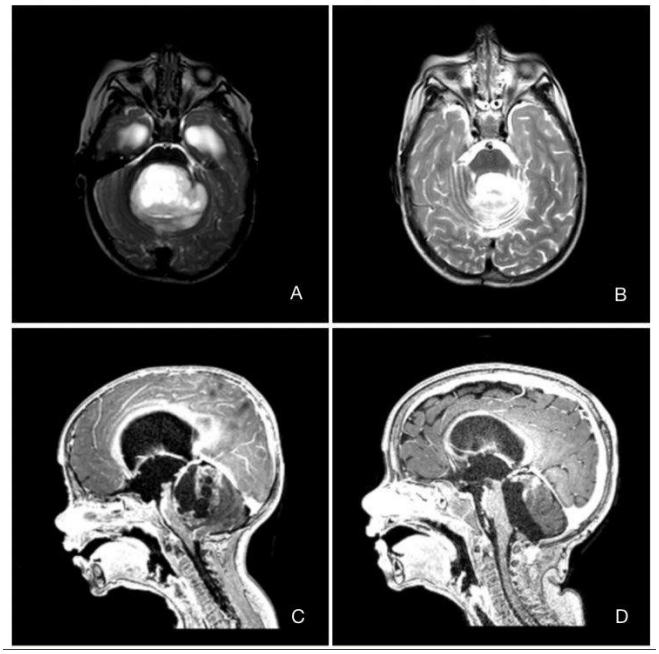

Figure 2 - A) Axial T2 pre-operative image of posterior fossa tumor (case 3). B) Axial T2 post-operative image of the same patient showing enlargement of pre-pontine cistern and brainstem decompression. C) Sagittal post-constrast T1 pre-operative image presenting irregular enhancement of lesion. D) Sagittal post-constrast T1 post-operative image.

DISCUSSION

Although surgical technique has evolved significantly in recent decades, access to the fourth ventricle can still have adverse effects with very harmful consequences for patients, such as cerebellar mutism. Bearing in mind that tumors of the posterior fossa are, by far, the most frequent brain neoplasms in the pediatric age group, this topic becomes very relevant [1]. The three illustrative cases described in this study occurred over a period of 3 years in a large reference service in pediatric neurosurgery and were operated on by the same team. At our pediatric neurosurgery reference center, we operate on approximately 70 pediatric brain tumors each year, most of which are located in the posterior fossa, as is the case in most services around the world. These reports do not reflect the complete incidence of cerebellar mutism in our service, and their main purpose is didactic and demonstrative.

Although most studies indicate the occurrence of cerebellar mutism in 25% to 30% of operated cases, it is difficult to estimate the actual incidence because mild cases of postoperative verbal, cognitive or motor impairment may go unnoticed [3]. Although it is a consensus in the literature and in the experience of the services that cerebellar mutism occurs in a minority of cases, the individual repercussions are very impactful and imply prolonged hospital stays [13].

The matter raises questions already from the nomenclature. The most consolidated designation in practice continues to be cerebellar mutism, an expression originating from the intense speech difficulty observed in most cases. The denomination akinetic mutism seeks to contemplate the motor difficulties that include hypotonia and hypoactivity. More recently, taking into account the wide variability of clinical manifestations, the term posterior fossa syndrome has gained ground [14] (Fig. 3).

CEREBELAR MUTISM SYNDROME:
TRANSIENT MUTISM/REDUCED SPEECH
EMOTIONAL LABILITY
HYPOTONIA
ATAXIA
OROPHARYNGEAL DYSFUNCTION/DYSPHAGIA

Figure 3 – Clinical presentation of Posterior Fossa Syndrome.

Over time, the cause of cerebellar mutism has been the subject of intense debate. The concept itself is counterintuitive when considering the classical and ancient view of cerebellar neurophysiology, which used to focus only on motor control functions. In fact, the study of neuroanatomy has established clear attributions of the cerebellar output pathways in the modulation of behavior, verbal expression and phonation. The very systematization of research on cerebellar mutism contributed to the most modern, complete and complex view on the functioning of the cerebellum. Additionally, the clinical examination that demonstrates hypotonia and hyporeflexia, as observed in the cases described, sustains that there is no lesion to superior cortical areas or long tracts that travel through the brainstem. The most recent studies reinforce the importance of the cerebellar output pathways, especially those that travel through the superior cerebellar peduncles, including activation of the periaqueductal gray matter, with

consequent repercussions on the thalamus and cerebral cortex [7][9][10][15].

It is important to note that cerebellar mutism can occur regardless of clear complications during operation, even with anatomical preservation of the structures and in the absence of hemorrhages or ischemia [5][16]. None of the cases described in this report had intraoperative complications and postoperative CT scans did not raise any additional concerns. The patient in case 2 had an episode of intracranial hypertension caused by dysfunction of the ventriculoperitoneal shunt used to treat hydrocephalus. This may have delayed the recovery of her neurological functions, but mutism was already established in the immediate postoperative period. The patient in case 3 probably had the influence of prematurity and cerebral palsy on his evolution, but the characteristic neurological impairment after tumor removal and recovery over time point to the unequivocal role of cerebellar mutism. Interestingly, the patient in case 1 underwent two operations with access to the fourth ventricle due to tumor recurrence and manifested mutism after both procedures, presenting later progressive recovery on the two occasions she was operated on.

In this context, there is great interest in trying to identify who are the patients with the highest risk of developing cerebellar mutism after surgery to remove a posterior fossa tumor and establish whether there are modifiable factors in the surgical technique that can be used to reduce its incidence. A view reinforced by practice and by studies in the literature has indicated that the main risk factors are younger patients, larger lesions, location inside the fourth ventricle and transvermian approach [11]. However, there are studies that indicate great difficulty in identifying these risk factors and predicting with an acceptable degree of accuracy who are the patients who will develop mutism [17]. Some efforts have also been made to identify risk factors inherent to the specific cases of patients, such as motor laterality [18] and the presence of language disorders in the preoperative period, with insightful findings [19]. Among the patients described in this study, there were very different ages (from less than two years old to 15 years old). In all reported cases, the tumor masses were large and exerted a significant compressive effect, and they were operated on through a telovelar access to the interior of the fourth ventricle. Although a current concept states that the transition from the transvermian approach to the telovelar approach could reduce cerebellar mutism incidence, it is not clearly verified in literature. Perhaps the fact that telovelar approach imposes cerebellar retraction can explain it. We used unilateral cerebellar retraction in the 3/4 prone (park bench) position, but the other positionings used to perform the surgery (seated or ventral) also implicated in cerebellar retraction that can injure cerebellar pathways involved in the genesis of cerebellar mutism. We did not use electrophysiological monitoring in these 3 cases, although we are convinced of the advantage of the method is securing nuclei and long pathways integrity. In fact, we have been using it in more recent cases.

In this context of evaluation, the role of magnetic resonance imaging (MRI) analysis has gained prominence. Toescu et al., in a study with 12 children, were unable to identify pre-operative imaging factors that indicated an increased risk of cerebellar mutism in the post-operative period. However, there was a positive association with T2 hyperintensity located in the superior cerebellar peduncles and the dentate nucleus of the cerebellum, reinforcing the role of Guillain-Mollaret triangle in the pathophysiological process [20]. This seems to be consistent with concerns about cerebellar retraction during surgery.

Faced with these difficulties in anticipation, it is important to know the variability in the evolution of cerebellar mutism and the strategies that can facilitate recovery [11]. It is widely known that most cases actually present neurological recovery after varying intervals of time in the postoperative period, and in different degrees of improvement. Although the studies do not have a high level of evidence for the establishment of these strategies, it is necessary to take measures to maintain homeostasis and cerebral protection in order to offer the best conditions for the reestablishment of temporarily impaired neural functions. A fundamental element in this process is rehabilitation involving therapies with a multidisciplinary team, including physical therapy and speech therapy, which must be maintained in the long term to enable the restoration of functionalities [8][13].

CONCLUSION

In summary, cerebellar mutism continues to be a challenge for pediatric neurosurgery and an entity with a high impact on the evolution of patients and with difficult, if possible, predictability. Therefore, a high degree of technical rigor is recommended in the execution of operations and postoperative surveillance so that the best recovery conditions are offered, including the support of the multidisciplinary team. It is fundamental that there is adequate interaction between the surgical team and the patients' families, so that the appearance of cerebellar mutism is considered as a possible adverse effect of the treatment and so that the proper support is provided during the recovery period.

ACKNOWLEDGMENTS

DISCLOSURES

Ethical approval

This study was performed in line with the principles of the Declaration of Helsinki. Approval was granted by the local Ethics Committee (Hospital Infantil Albert Sabin), number: CAAE 96905018.0.0000.5042

Consent to participate

The patients gave consent to use their information and images for research purposes. *Consent for publication*

The patient gave consent to use his information and images for publication.

Conflict of interest

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

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CONTRIBUTIONS

-**Carlos Eduardo Barros Jucá**: Conceptualization, Data curation, Formal Analysis, Project administration, Supervision, Writing – review & editing
-**Gabrielle Benevides Lima**: Investigation, Writing – original draft
-**Pâmella Christine de Souza Munhoz**: Formal Analysis, Investigation, Writing – original draft
-Patrícia Pereira de Andrade: Formal Analysis, Investigation, Writing – original draft
-Francisco Gleison Albuquerque Ribeiro: Data curation, Investigation, Writing – original draft
-Júlia Praciano Lopes: Formal Analysis, Investigation, Writing – original draftData curation, Investigation, Writing – original draft

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