Case Report

LORAZEPAM FOR THE MANAGEMENT OF POSTERIOR FOSSA MUTISM: A CASE REPORT

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Abstract

Posterior Fossa Syndrome (PFS) is a condition that can develop in children and adults following a cerebellar tumor resection surgery. The syndrome is characterized by multiple symptoms, including mutism, personality changes, and mobility problems. We present a case of a four-year-old boy diagnosed with posterior fossa tumor with secondary hydrocephalus who underwent an uneventful total resection of the tumor and developed posterior fossa syndrome, including mutism, was managed using lorazepam and made a full recovery of almost all of the symptoms he developed. Posterior fossa syndrome has multiple symptoms, most of which are usually transient. The exact pathophysiology and time to recovery are still not fully known. Patients with posterior fossa syndrome are managed with a multi-disciplinary approach. Mutism resulting from this syndrome is proven to be successfully managed by lorazepam. ***ASEAN Journal of Psychiatry, Vol. 25 (5) May, 2024; 1-5.***

Keywords: Posterior Fossa Syndrome; Mutism; Cerebellar Mutism; Cerebellar Mutism Syndrome; Lorazepam; Ependymoma; Pediatrics

Introduction

Posterior Fossa Syndrome (PFS) is a syndrome that affects children and adults following a cerebellar tumor resection surgery. It is a rare syndrome that mostly affects children but can also occur in adults. It is also known as posterior fossa mutism syndrome and cerebellar mutism syndrome. The syndrome is characterized by mutism, apraxia as well as behavioral and personality changes. Other reported complications commonly associated with cerebellar surgery are cranial nerve palsies and other neuropsychiatric symptom [1]. Patient that experience symptoms of this syndrome usually return to their preoperative state of health with the proper management. The duration and severity of symptoms varies form a patient to another but it has been reported to be in the range of weeks, months or even years [2].

The treatment of posterior fossa mutism syndrome consists of both behavioral and biological modalities. Few studies have been done on the rare complication of posterior fossa syndrome. A study by Catsman-Berrevoets and Aarsen was done in 2010. They reported 148 patients with cerebellar tumor that underwent resection surgery. They found that out of the 148 patients, 41 has developed posterior fossa syndrome [1].

A case report was published in 2016 by Gadgil et al., about a two year old girl with medulloblastoma who underwent a resection of the tumor with a posterior fossa craniotomy [3]. The patient showed mutism but she showed improvement in her mutism by a month following the surgery. She was followed 45 months later and was found to still have dysarthria despite having speech therapy. Another case was also reported that shows a two year old boy who was diagnosed with an ependymoma. The patient underwent a total resection by a posterior fossa craniotomy as well. The patient developed mutism that resolved completely 6 months following the surgery [3].

A more recent study was also done in 2021 which included 178 patients who were diagnosed with medulloblastoma and underwent surgical resection. It was found that 60 of them had developed posterior fossa syndrome. Complete mutism was found in forty of these sixty patients affected while twenty patients had reduced speech only. For those who experienced complete mutism, the median time for full resolution of mutism was 2.3 months. There were no pharmacological protocols for management for post operation complications including mutism. All those affected would undergo behavioral and speech therapy [4].

Very few numbers of studies has been done on the management of posterior fossa syndrome with lorazepam. A case report that presents a 10 year old boy who developed posterior fossa mutism and was managed with lorazepam on an as needed bases. His symptoms improved and he gained speech back soon after [5].

In this case report we present a patient who was diagnosed with posterior fossa mutism and was treated successfully with lorazepam.

Case Presentation

A four year old presented to the emergency department with a history of persistent vomiting for twenty days, associated with headache, photophobia, irritability, bilateral lower limb weakness and significant weight loss.

The patient was admitted in the Pediatric Intensive Care Unit (PICU) and Computed Tomography (CT) brain was done which revealed posterior fossa mass with hydrocephalus, and emergency External Ventricular Drain (EVD) was inserted. Magnetic Resonance Imaging (MRI) with contrast was later performed, which showed large midline posterior fossa mass that seems to be filling the 4th ventricle and extending through the foramen of Luschka, with significant mass effect on Brainstem and cerebellum. MRI whole spine showed no drop metastasis.

He then underwent a posterior fossa craniotomy and gross total resection of the tumor, which was successful and uneventful. The patient was extubated after MRI brain with contrast was done which showed complete resection and no residual, post-operative examination patient found to have right sixth and seventh nerve palsies (facial collicular syndrome), he was also found to be mute, with personality changes, aggression and decreased mobility. Histopathology report was anaplastic ependymoma World Health Organization (WHO) grade III.

Regarding behavior changes, aggression and mutism; psychiatry were involved in the multidisciplinary team managing the patient. Patient was treated with low dose lorazepam and was followed up both during admission and as in outpatients setting. He was initially started on 0.25 mg of lorazepam twice daily but showed almost no improvement. His dose was then increased to 0.5 mg twice daily and that's when he started to show improvement. He did not show any side effects which made the increase in dose possible. His dose was finally increased to 1 mg twice daily and that's when he showed rapid and major improvements. His behavior improved as he became calmer, he started smiling and was able to play with his toys. Patient was able to start gain back his speech abilities soon after the operation and by his two months follow up he was able to speak in words clearly. His behavioral symptoms, aggression and mobility started to improve as well. His medications were finally stopped almost a year later after gradual tapering down as the patient continued to improve to the point of complete recovery.

Discussion

Posterior fossa mutism syndrome is a rare complication of brain tumor excision surgery. Few case reports and case studies on this subject have been published. An even fewer number of studies have been published on the use of lorazepam as a treatment modality for it. Risk factors to developing this syndrome were identified to include pediatric age group, midline tumor and midline incision [6].

We present a patient who was unfortunate to developed posterior fossa syndrome. He is a toddler boy who was diagnosed and treated for ependymoma. He was later presented with a post-operative complication which was identified to be posterior fossa mutism syndrome.

The precise pathophysiology of posterior fossa mutism syndrome is not yet fully understood. There are multiple proposed theories for the pathophysiology of this syndrome and it is believed that the pathogenesis is most likely to be multifactorial.

Its pathophysiology is believed to result from the injury of the Dentato-Thalamo-Cortical (DTC) pathway, leading to Cerebro-Cerebellar Diaschisis (CCD) [7].

Like this presented case, most cases that were displayed in the literature were from a pediatric age group. It was also mentioned earlier in this paper that most patients that end up with the complication of posterior fossa mutism can have spontaneous recovery. However, studies show that recovery is usually partial and incomplete [8]. Studies show that even though mutism is transient, most patients are usually left with dysarthria [9]. This was not the case with the patient shown in our case. He showed subtle progressive improvement shortly after the operation and ended in full resolution of his mutism without the commonly occurring dysarthria.

The management is usually supportive but can include speech and occupational therapy. Most patients with posterior fossa mutism are expected to have spontaneous and complete recovery of most symptoms. However, most patients can be left with slowed speech and ataxic dysarthria [10]. This was definitely not the case with our patient who was treated with lorazepam and showed full recovery of his mutism. His speech was back to normal and matches speech of children his age.

Few pharmacological treatments had been studied including Bromocriptine, fluoxetine, haloperidol, risperidone, delorazepam, and zolpidem have been tested as potential treatments for a certain condition. However, the studies conducted on these drugs involved small sample sizes, making it difficult to draw any meaningful conclusions. The beneficial effects of these drugs usually appear at least 24 hours after the first administration, and complete recovery can take several months, which further complicates the assessment of their contribution to healing.

On the other hand, a study reported that midazolam was able to resolve post-operative Centers for Medicare and Medicaid Services (CMS) within a few minutes after intravenous administration in a 17-year-old boy who had undergone a fourth-ventricle choroid plexus papilloma resection [11].

The mechanism of action of Lorazepam and other benzodiazepine medications like Midazolam work by increasing ionotropic Gamma-Aminobutyric Acid (GABA)-ergic stimulation, an inhibitor of neural activity. An inhibitory effect of this drug on the direct and indirect basal ganglia pathways that regulate the excitatory output of the thalamus. The final result is a reduction of inhibition of the thalamus *via* the striatum, an outcome similar to what is obtained with bromocriptine administration [12].

The management of patients with posterior fossa syndrome is multi-disciplinary team is indeed needed for such patients. In this case we present, the patient was managed by a multi-disciplinary team that included a neurologist, neurosurgeon, oncologist, psychiatrist and a physiotherapist (Figures 1-3).

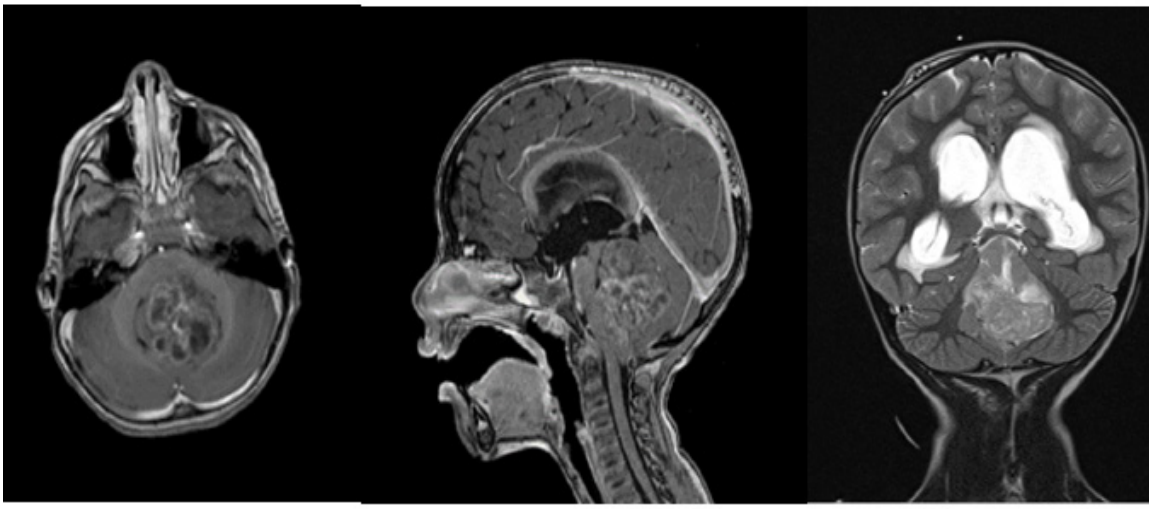

Figure 1. MRI brain post EVD insertion: Showed large midline 4th ventricular mass, causing significant mass effect on adjacent structures including brainstem, and causing hydrocephalus.

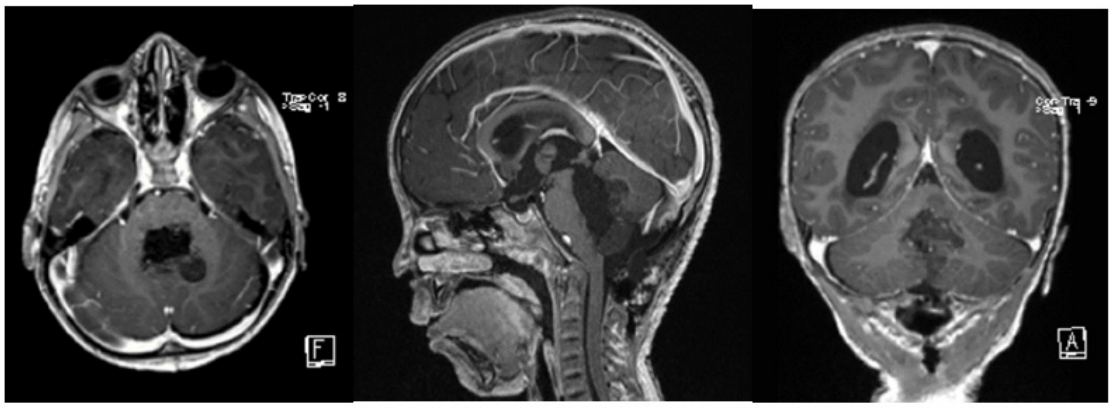

Figure 2. Post-operative MRI Day 1: No residual.

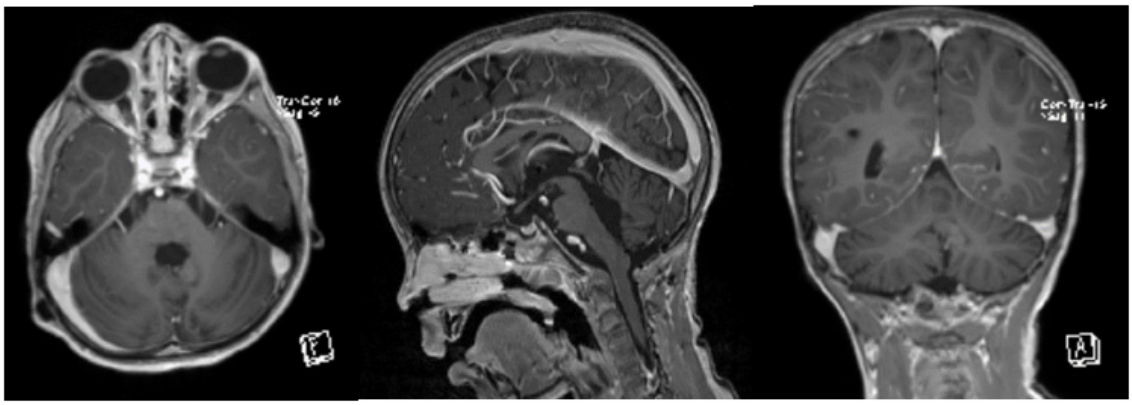

Figure 3. MRI 1 year post-operative: No residual.

Conclusion

Posterior fossa mutism syndrome is a complication of posterior fossa surgery. It is usually transient but the exact duration to witch symptoms improve differs from one patient to another. The exact pathophysiology for how it develops is still not fully understood. A multi-disciplinary team is needed for the management of such patients. This includes, neurosurgeons, neurologists, oncologists, psychiatrist and speech and occupational therapists. No pharmacological management protocol has been established for Posterior fossa mutism yet. Lorazepam shows promising results. However, further studies are definitely needed to confirm its effects.

Posterior fossa mutism represents a complex and challenging phenomenon observed predominantly in pediatric patients following surgical interventions in the posterior fossa region. While its exact mechanisms remain elusive, current research suggests multifactorial involvement, including disruption of cerebellar pathways, direct surgical trauma, and neuroinflammatory responses.

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