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Case Report

Postoperative Cerebellar Mutism Syndrome in an Adult Patient

Todor P. Shamov[1], Ioanna Tivcheva[2], Tihomir Eftimov[3]

[1]*Department of Neurology and Neurosurgery, Military Medical Academy, Sofia, Bulgaria*
[2]*Medical University, Sofia, Bulgaria*
[3]*Department of Neurosurgery, Military Medical Academy, Sofia, Bulgaria*

Corresponding author: Todor P. Shamov, Department of Neurology and Neurosurgery, Military Medical Academy, 3 Georgy Sofiisky St., 1606 Sofia, Bulgaria; E-mail: shamov@abv.bg; Tel.: +359898667552

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Postoperative cerebellar mutism syndrome is a common complication occurring after surgical treatment of medulloblastoma in children. The clinical course of this syndrome is characterized by delayed onset after the surgical intervention, loss of speech, emotional lability, hypotonia, and oropharyngeal dysfunction. This syndrome is rarely seen in adults. We report here a case of postoperative cerebellar mutism in a 25-year-old female patient, who underwent a surgical intervention due to medulloblastoma of the forth ventricle. There are very few reported cases in literature of this syndrome. In the discussion we discuss the likely reasons for the syndrome to develop in adult patients.

Keywords

adult patients, hyperthermia, medulloblastoma, postoperative cerebellar mutism, risk factors

INTRODUCTION

Postoperative cerebellar mutism syndrome (pCMS) is a common complication following surgical interventions for cerebellar tumors in children. The incidence varies from 7% to 50%.[1] According to the expert consensus of Posterior Fossa Society the common symptoms of pCMS are late onset of the mutism, reduction to loss of speech, emotional lability and some additional symptoms such as hypotonia and oropharyngeal dysfunction or dysphagia.[2] This syndrome is frequently accompanied by cerebellar motor symptoms, cerebellar cognitive affective syndrome, dysfunction of the brainstem and cranial neuropathies.[3] The duration of the cerebellar mutism varies from a few days to six months.

Though most authors consider the cerebellar mutism a transient syndrome, some studies regarding the late results of surgical treatment of medulloblastomas demonstrate the possibility of persistent residual symptoms such as dysarthria, cognitive deficit and poor school performance.[5]

Cerebellar mutism is a syndrome common predominantly in childhood and most frequently subsequent to surgical interventions for medulloblastomas. There are a few cases of cerebellar mutism in adult patients, developed after surgical procedures of the cerebellum reported in literature. In the present publication we present a case of pCMS in an adult female patient.

CASE REPORT

The patient, a 25-year-old woman, was admitted to the clinic with headache, episodes of vomiting and swaying in no particular direction. The symptoms started about one month prior to admission. The clinical examinations showed large-amplitude horizontal pendular nystagmus at glance in both directions, muscle hypotonia in all four

limbs, and marked truncal ataxia.

MRI of the patient revealed tumor formation, localized in the fourth ventricle, causing complete occlusion of the ladder and obstructive hydrocephaly. The tumor is hyperintense on T2 weighted echo sequences and hypointense on T1.

Due to the clinically manifest hydrocephaly, it was decided that a third ventriculostomy should be performed as a first stage of treatment, prior to the surgical excision of the tumor. Perforation of the floor of the third ventricle was made with neuroendoscope. The patient received dexamethasone as antiedemic therapy for a few days. The clinical condition of the patient improved and five days later she underwent surgery.

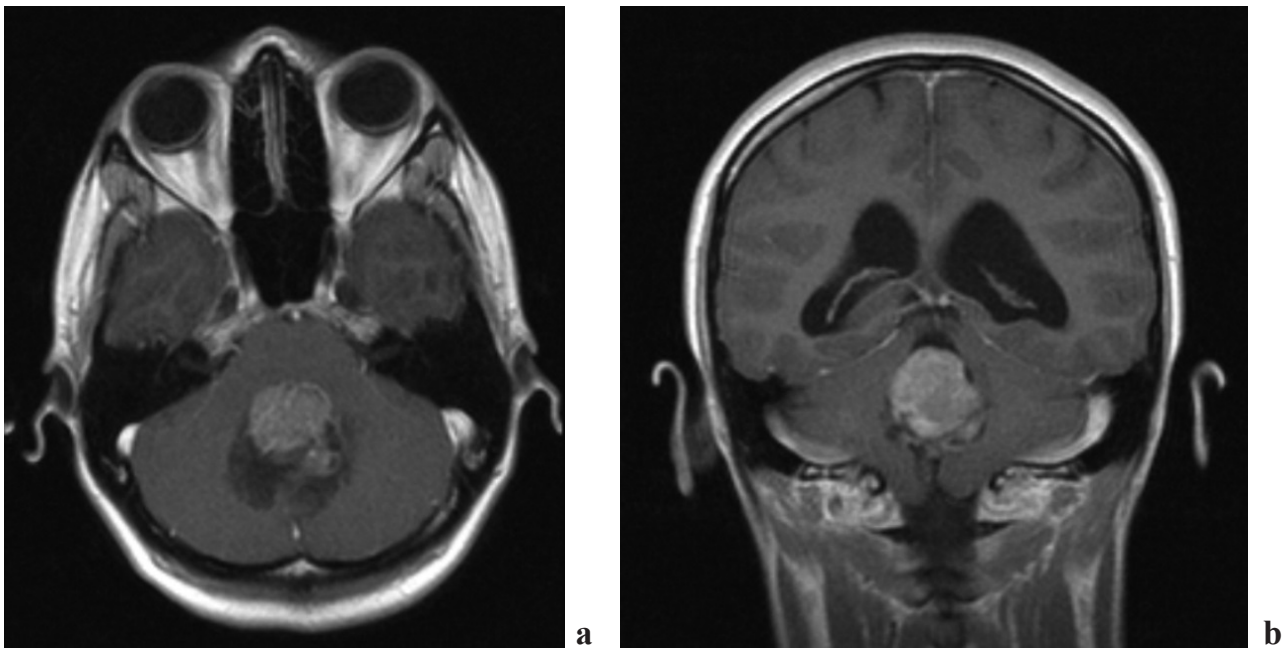

Figure 1. Preoperative MRI (T1 + contrast) of the tumor from axial (**a**) and coronary (**b**) plane.

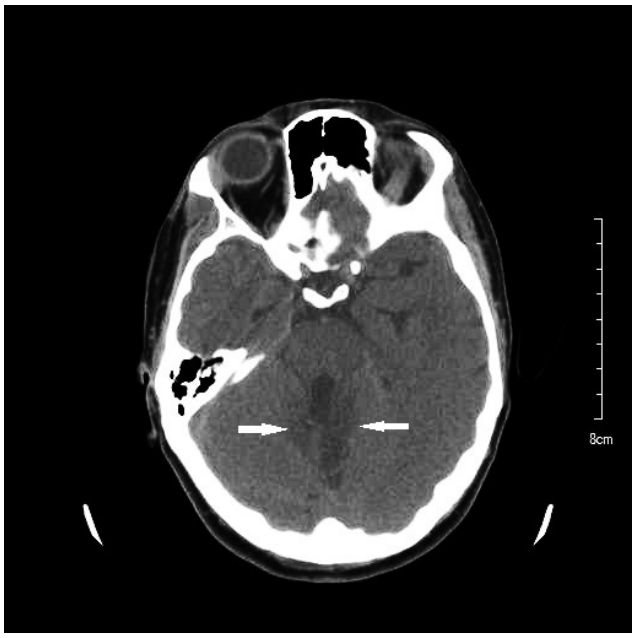

Figure 2. Postoperative CT on third postoperative day showed edema in the area of the cerebellar peduncles (white arrows).

Surgical procedure

The patient was placed on the operating table in lateral position, inverted left, with thorax elevated at 30°, the head was flexed and in an indifferent position. Suboccipital midline craniotomy with removal of the medial third of the atlantic arch. A Y-type incision of dura mater was made. The tumor was reached via telovelar-medullar approach. It was partially dissected and its surface was coagulated. Using an ultrasonic aspirator, internal decompression of the tumor was achieved and with consecutive dissection, the tumor was completely removed. The tumor had partially grown into the brainstem, in the right stria acustici area, but with microdissection a visually total excision was achieved. Hermetic closure of dura mater was performed with uninterrupted suture and fibrin glue. Osteoplastic closure of the posterior cranial fossa.

Histological findings: *desmoplastic medulloblastoma*

The early postoperative period was uneventful. On the third postoperative day the patient was in agitated state with suppressed speech initiative to complete loss of speech and was only capable of making incomprehensible sounds. Due to the development of such clinical symptoms, a control CT was performed. It showed no signs of intra- or extra-axial

blood-equivalent mass lesions, but edema in the area of the cerebellar peduncles was seen.

The patient was diagnosed with cerebellar mutism and discharged 10 days postoperatively. The clinical observation continued in ambulatory manner. Two months after the procedure there was gradual regaining of speech and regress of the emotional lability. On the third postoperative month a MRI was performed which showed no signs of residual tumor. Meanwhile, the patient was referred for review to oncology committee, which recommended that radio- and chemotherapy should be initiated.

DISCUSSION

The term cerebellar mutism in neurology signifies the presence of acquired loss of speech that could result from damage at each level of the nervous system - from the cortex to the peripheral organs of speech.[6] During World War I, Holmes described a speech disorder resulting from gunshot injury to the cerebellum.[7] The first well documented case of cerebellar mutism was reported by Baily in 1939 in a 4-year-old child who had undergone surgery because of ependymoma of the forth ventricle.[8] The postoperative cerebellar mutism syndrome was defined in 1978 by Stein who described mutism resultant of surgical procedure for removal of cystic astrocytoma of the cerebellum, causing significant clinical symptoms.

Some elements of the pathogenesis were revealed in the eighties by Fraioli, because of bilateral stereotactic lesion of nn. dentati during an attempt to treat dystonias.[10] Later it became clear that the pathogenetic basis for the development of this syndrome was lesion of the proximal segments of the efferent cerebellar pathways, passing through the superior cerebellar peduncle. Most of the fibers of these pathways originate from n. dentatus, but also include projection fibers from the other cerebellar nuclei (n. fastigii, n. globosus et n. emboliformis). These fibers cross in the pontomesencephalic tegmentum, in the Guillain-Mollaret decussation, project to the thalamic nuclei and from the thalamus to different cortical areas. In 1914, von Monacoff introduced the term diaschisis and postulated that the lesion of certain zones in the brain can cause suppression of the function of other cortical areas to which there are projections from the injured areas. Ergo the suppressed function of the speech cortical areas, according to some authors, had the characteristics of speech apraxia. Microstructural alterations in the white matter of the superior cerebellar peduncle and the thalamic projections to the cortex have been demonstrated using diffusion tensor MRI images.[11] The same authors have proven that the edema and degeneration of the fibers of the superior cerebellar peduncle could lead to edema of the contralateral oliva in the medulla oblongata and degeneration of the fibers originating in the inferior olivary nucleus. Such changes can be explained by the topical distribution of the fibers in the Guillain-Mollaret decussation.

The mechanisms of the injury of the efferent cerebellar structure are still unclear. The hypothesis for direct surgical trauma is unacceptable explanation, because of the typical delayed onset of symptoms. Some authors consider the possibility of thermal injury of the conductive systems due to the use of ultrasonic aspirator. This hypothesis was renounced, because the incidence of the syndrome has not been lower in the era before the usage of CUSA was a primary method.[12]

One cohort study, comparing children who had undergone surgery for medulloblastoma tumors, concluded that the incidence of the syndrome is significantly higher in the children who had higher temperature on average by 0.5°C in the first few postoperative days. Higher temperature increases the metabolic rate of nervous tissue, making it significantly more vulnerable. Of immense importance regarding the prevention of the syndrome is maintaining normothermia in the postoperative period.[13]

Significant blood loss is another important factor that could explain the development of the syndrome in children. During childhood patients have notably smaller intravascular volume. Increased blood loss leads to reduced blood supply and oxygen deficit, hence the ischemia in the perioperative field.[14]

It has been found that the incidence of pCMS is significantly higher in patients with medulloblastoma compared to ones with other histological types of tumors of the forth ventricle. In childhood the relative volume of the forth ventricle is lower than that in adults. The tumor size is singled out as an independent factor for development of pCMS. The incidence is higher in patients with medulloblastoma tumors larger than 5 cm in diameter.[15]

Postoperative cerebellar mutism is rare in adult patients. That is partially attributed to the overall lower incidence of medulloblastoma tumors in adults. In the literature we were able to access the reported cases are seven.[16-20]

The development of the syndrome in adults could be a result of excessive use of ultrasonic aspirator and the bipolar coagulation in close proximity of important structures, as well as the untimely compensation of blood loss or hyperthermia in the postoperative period.

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Синдром послеоперационного мозжечкового мутизма у взрослого пациента

Тодор Шамов[1], Йоанна Тивчева[2], Тихомир Евтимов[3]

[1]*Кафедра „Неврология и нейрохирургия", Военно-медицинская академия, София, Болгария*

[2]*Медицинский университет - София*

[3]*Кафедра нейрохирургии, Военно-медицинская академия, София, Болгария*

Адрес для корреспонденции: Тодор Шамов, Кафедра „Неврология и нейрохирургия", Военно-медицинская академия, ул. „Георги Софийски"№3, 1606 София, Болгария E-mail: shamov@abv.bg; Tel.: +359898667552

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Абстракт

Послеоперационный синдром мозжечкового мутизма является частым осложнением после хирургического лечения медулобластомы у детей. Клиническое течение этого синдрома характеризуется поздним началом после операции, отсутствием речи, эмоциональной лабильностью, гипотензией, дисфункцией ротоглотки. Этот синдром редко наблюдается у взрослых. Мы сообщаем о послеоперационном мозжечковом мутизме у 25-летней пациентки, перенёсшей операцию по поводу медуллобластомы четвёртого желудочка. В литературе описано очень мало случаев данного синдрома. В этой статье мы обсудим вероятные причины возникновения симптомов у взрослых пациентов.

Ключевые слова

послеоперационный мозжечковый мутизм, медуллобластома, взрослые пациенты, факторы риска, гипертермия