

## Case report

# Cerebellar mutism in adult after posterior fossa surgery:

## A rare presentation

<sup>1</sup>Dr. S. V. Rege, <sup>2</sup>Dr. Harshad Patil, <sup>3</sup>Dr. Sharadendu Narayan

<sup>1</sup>Professor & HOD , Department of Neurosurgery, Sri Aurobindo Medical College and PG Institute, Indore, Madhya Pradesh, India. 452010,

<sup>2,3</sup>Senior Resident , Department of Neurosurgery, Sri Aurobindo Medical College and PG Institute, Indore, Madhya Pradesh, India. 452010,

**Corresponding author:** Dr. Harshad Patil

---

### Abstract

Cerebellar mutism is commonly seen after involvement of midline cerebellar structures secondary to degenerative disease, tumors, hemorrhage, or surgery. Typically, cerebellar mutism syndrome (CMS) has been seen in children and rarely described in adults after surgery of the posterior fossa. This syndrome typically arises 48 hours after the initiating event and resolves approximately 7 to 8 weeks later. Herein author has described a rare case of cerebellar mutism after posterior fossa surgery in adult.

**Keywords:** Cerebellar mutism, Adult, Posterior fossa surgery

---

### Introduction:

Cerebellar mutism is characterized by complete absence of speech without impaired consciousness, other cranial nerve deficits, or long tract signs.<sup>1</sup> Cerebellar mutism was first reported in 1979 by Hirsh after a posterior fossa tumor resection.<sup>1</sup> Cerebellar mutism syndrome (CMS) mainly appears after posterior fossa tumor resection in children and there are few case reports of CMS in adults. Cerebellar mutism is commonly seen after involvement of midline cerebellar structures secondary to degenerative disease, tumors, hemorrhage, or surgery. The incidence of CMS in children who underwent surgery of the cerebellum is estimated to be between 8 and 31%.<sup>2</sup> The exact reason for cerebellar mutism is not known but recent reports believe that there is decreased blood flow to the dentate-thalamic-cortical pathway.<sup>2</sup>

Herein authors have described a rare case of cerebellar mutism after posterior fossa surgery in adult.

### Case report:

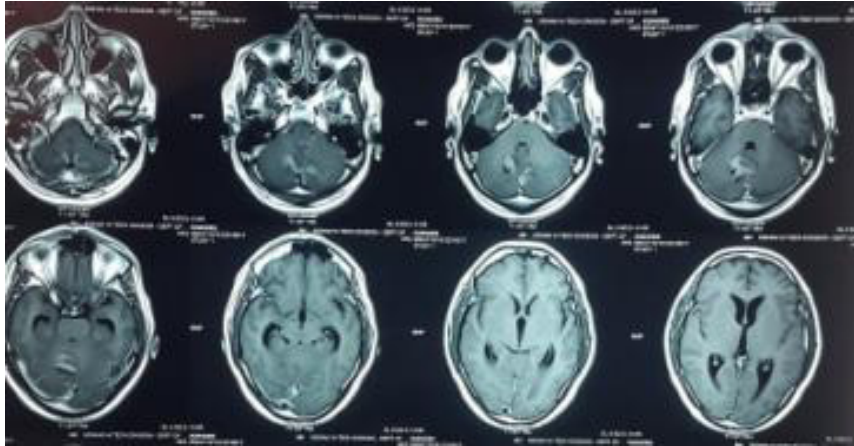
A 45 years old male presented in neurosurgery department with headache and gait disturbance for last three months. Neurological examination revealed cerebellar signs with ataxic gait. Magnetic resonance imaging (MRI) of brain showed heterogeneous intensity mass lesion in posterior fossa mainly involving midline ( Image 1,2). In view of symptoms patient was taken up for surgery. Gross total excision of the tumour was done via midline suboccipital craniectomy.

The histopathological examination revealed medulloblastoma. Immediately after the surgery, the patient was alert, his neurological examination showed only mild truncal ataxia. However on the

second postoperative day he was not able to speak, apart from answering 'yes' and 'no'. He could not form words or sentences, although he was clearly able to understand simple commands. There were no lower cranial nerve deficits. No obvious mass lesion was present in postoperative imaging of brain (Image

3). In view of symptoms and clinical examination diagnosis of cerebellar mutism was made and patient was subjected to speech rehabilitation. About three weeks after surgery, he began to speak few words but with severe dysarthria. At the two month follow-up examination he was able to speak normally.

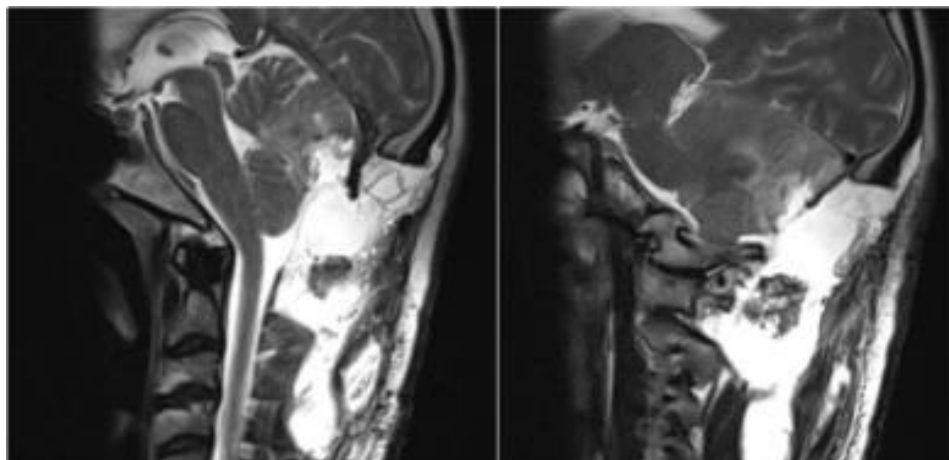
**Image 1:** MRI Brain contrast axial section showing midline cerebellar tumour



**Image 2:** MRI brain contrast sagittal section showing cerebellar



**Image 3:** Post operative MRI scan showing no obvious mass lesion



### Discussion

Cerebellar mutism was first reported in 1979 by Hirsh after a posterior fossa tumor resection and since then over 400 cases has been reported in the literature. Cerebellar mutism is most commonly seen after resection of posterior fossa tumors in children but rarely reported in adults.<sup>1</sup> Several recent prospective studies have reported the incidence of cerebellar mutism after posterior fossa surgery in children to be 11–29%<sup>2,3</sup> The mean age of presentation in pediatric population is 9.1 years and there is no sex prevalence. In adults it is more common in young adults.<sup>4</sup> but in our case it was found in middle age.

The clinical characteristic of mutism after posterior fossa surgery is as follows:

It has delayed onset (1–6 days) and limited duration (1 day–4 months) followed by a recovery period where speech is marked by dysarthria. Mean time of onset is 1.7 days and average duration is 7–8 weeks. Recovery is spontaneous and can occasionally be rapid (a few days) and complete, but the majority of patients are left with long-term speech and language dysfunction such as ataxic dysarthria, dysfluency and slowed speech rate.<sup>4</sup> these characteristics are same in

both pediatric and adult patients. In our case mutism developed after second postoperative day and persisted for three weeks.

Children tend to have a predilection for cerebellar mutism following posterior fossa surgery; however the exact reason is unknown. A simple reason for this may be a higher incidence of posterior fossa and midline tumors in children. However pathophysiological mechanisms also contribute to the high incidence of CMS in children. The incomplete development of motor speech control and language is the first factor. Additionally, the immaturity and incomplete myelination of the reciprocal links in childhood, connecting the cerebellum to thalamus, sensory areas, motor, and supplementary motor area, also make the children more vulnerable.<sup>5</sup>

The pathophysiology of CMS has not been entirely established. There are many theories considering the responsible factors, but none of them provides an entirely satisfactory explanation. The functional hypothesis supports that CMS is a kind of negativism on the part of the child who feels betrayed by his parents and doctors (form of “elective mutism”). The fact that many patients recover as soon as they get home.<sup>6</sup> Damage to a specific anatomical substrate has

been proposed as a pathogenetic factor. The dentate nucleus was the first region that was hypothesized to be involved. Many authors proposed that bilateral damage of the dentate nuclei is a critical factor for CMS while some proposed postoperative edema led to disturbance of the venous circulation, which was responsible for the dysfunction of the dentate nuclei.<sup>7</sup> The majority of case reports and studies support that CMS can occur after the resection of a midline tumor and, as a rule, if vermis is involved.<sup>8</sup>

One of the most famous theories about the pathophysiology of CMS is involvement of the dentato-thalamo-cortical (DTC) tract. This pathway projects to and from the dentate nucleus of the cerebellum on either site, travels through the superior cerebellar peduncle, crosses the midline to the opposite side in the decussation of the superior cerebellar peduncle, continues through the brainstem to the contralateral ventrolateral nucleus of thalamus and then to the contralateral motor cerebral cortex (premotor and supplementary motor cortices). Mutism can occur after lesion anywhere along this tract.<sup>8</sup>

It is difficult to predict the prognosis for the individual patient, and the course of recovery is extremely variable. The duration of symptoms after

surgery seems to correlate with functional prognosis. Patients who's symptoms persist for more than four weeks are at a greater risk of still suffering from speech and language dysfunction at one year postoperatively than those who's symptoms last less than four weeks.<sup>9</sup> In our patient mutism last for three weeks and he recovered completely after two months. There is no established treatment that facilitates recovery from cerebellar mutism. The effects of pharmacological intervention such as dopamine agonist drugs and/or speech therapy are sporadically reported in the literature but randomized controlled trials are still lacking to support this treatment modality.<sup>10</sup> In our patient speech therapy was given and he recovered after two months.

### **Conclusion**

Cerebellar mutism is a rare complication of posterior fossa surgery in children and is still rarer in adults. The exact mechanism of injury, the pathophysiology of delayed onset and its resolution are still not clearly understood. Pharmacological therapy in form of dopamine agonists has been proposed, however this needs validation by randomized control trials. At present speech therapy remains the most effective method of treatment in these patients.

### **References**

1. Pitsika M, Tsitouras V. Cerebellar mutism. *J Neurosurg Pediatr.* 2013 Dec;12(6):604-14. doi: 10.3171/2013.8.PEDS13168. Epub 2013 Sep 27. Review.
2. Pollack IF, Polinko P, Albright AL, Towbin R, Fitz C. Mutism and pseudobulbar symptoms after resection of posterior fossa tumors in children: incidence and pathophysiology. *Neurosurg.* 1995; 37:885-93.
3. Robertson PL, Muraszko KM, Holmes EJ, Spoto R, Packer RJ, Gajjar A, Dias MS, Allen JC; Children's Oncology Group. Incidence and severity of postoperative cerebellar mutism syndrome in children with medulloblastoma: a prospective study by the Children's Oncology Group. *J Neurosurg.* 2006 Dec;105(6 Suppl):444-51.

4. Kai Y, Kuratsu J, Suginozawa K, Marubayashi T, Ushio Y. Cerebellar mutism after posterior fossa surgery--two case reports. *Neurol Med Chir (Tokyo)*. 1997 Dec;37(12):929-33.
5. Catsman-Berrevoets CE, Aarsen FK. The spectrum of neurobehavioural deficits in the Posterior Fossa Syndrome in children after cerebellar tumour surgery. *Cortex*. 2010 Jul-Aug;46(7):933-46. doi:10.1016/j.cortex.2009.10.007. Epub 2009 Oct 29.
6. Tasdemiroğlu E, Kaya M, Yildirim CH, Firat L. Postoperative cerebellar mutism and autistic spectrum disorder. *Childs Nerv Syst*. 2011 Jun;27(6):869-78. doi:10.1007/s00381-010-1316-6. Epub 2010 Oct 30. Review.
7. Aguiar PH, Plese JP, Ciquini O, Marino R. Transient mutism following a posterior fossa approach to cerebellar tumors in children: a critical review of the literature. *Childs Nerv Syst*. 1995 May;11(5):306-10. Review.
8. Clerico A, Sordi A, Ragni G, Festa A, Cappelli C, Maini CL. Brief report:transient mutism following posterior fossa surgery studied by single photon emission computed tomography (SPECT). *Med Pediatr Oncol*. 2002 Jun;38(6):445-8.
9. Grill J, Viguier D, Kieffer V, Bulteau C, Sainte-Rose C, Hartmann O, Kalifa C, Dellatolas G. Critical risk factors for intellectual impairment in children with posterior fossa tumors: the role of cerebellar damage. *J Neurosurg*. 2004 Nov;101(2 Suppl):152-8.
10. Gudrunardottir T, Sehested A, Juhler M, Schmiegelow K. Cerebellar mutism:review of the literature. *Childs Nerv Syst*. 2011 Mar;27(3):355-63. doi:10.1007/s00381-010-1328-2. Review.