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REVIEW

Postoperative cerebellar mutism in adult patients with Lhermitte-Duclos disease

Ali Afshar-Oromieh · Heinz Linhart · Dino Podlesek · Wiebke Schrempf · Gabriele Schackert · Dietmar Krex

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Abstract Cerebellar mutism (CM) is a rare and severe form of speech and language impairment, mostly diagnosed in children and adolescents and rarely reported in adults. We here review the literature and summarize all anatomical structures related to the pathogenesis of this rare syndrome. We also report two illustrative cases of CM following surgical treatment of Lhermitte-Duclos disease (LDD; dysplastic gangliocytoma) in two adult patients. LDD is a rare benign cerebellar tumor. Surgical excision appears to be the only effective treatment. However, surgery is hampered by the difficulty to distinguish between tumor and healthy cerebellar tissue, which may result in extensive resection and cause neurological deficits such as CM. A review of the literature and our two cases suggest that lesions or functional impairment of paravermian structures including dentate nuclei, vermis, lateral hemispheres, and cerebellocortical pathways contribute to the development of CM. However, there is no single anatomical structure identified to be associated with CM. It is unknown whether some diseases such as LDD carry a higher risk of postoperative CM than others. As illustrated by our two cases, although there are no special means, optimal preoperative diagnosis might contribute to the prevention of this syndrome. Despite the severity, CM carries a favorable prognosis and generally resolves within a few months.

To conclude, we review the clinical signs and particularly the pathophysiological observations and anatomical structures affected in the development of postoperative CM and contribute two cases illustrating the pathogenesis, prognosis, and possible prevention of this syndrome, to focus that CM might also occur in adults even in association with rare tumors.

Keywords Cerebellar mutism · Posterior fossa · Lhermitte-Duclos disease · Dysplastic cerebellar gangliocytoma

A. Afshar-Oromieh · D. Podlesek · G. Schackert · D. Krex (✉)
Department of Neurosurgery,
University Hospital Carl Gustav Carus,
Technical University of Dresden,
Fetscherstrasse 74,
01307 Dresden, Germany
e-mail: Dietmar.Krex@uniklinikum-dresden.de

H. Linhart
German Cancer Research Center (DKFZ)
and National Center for Tumor Diseases (NCT),
Heidelberg, Germany

W. Schrempf
Department of Neurology,
Carl-Gustav-Carus University Hospital of Dresden,
Dresden, Germany

Introduction

Mutism is defined as a state consisting of decreased impetus with lack of verbal communication, although alertness and perceptual capacity are sustained. It has primarily been observed in patients suffering from bilateral anterior cerebral infarction but is also caused by lesions of thalamic and diencephalic structures. Cerebellar mutism (CM) was first reported in children after surgery of the posterior cranial fossa in 1979 [28]. In the adult, CM was first reported by Salvati et al. [50] in 1991 in a case that occurred after resection of a medulloblastoma in the cerebellar vermis. CM is defined as an impairment of speech and language caused by lesions of the cerebellum without concomitant pyramidal tract signs, superior hemispheric injury, or cranial nerve palsies. It is assumed that a loss of coordination of orofacial movements is primarily

responsible for the development of CM. Some authors consider CM to be the most extreme form of ataxic dysarthria [9, 37, 46, 59, 62]. Patients with CM may also present with a combination of symptoms such as reduced spontaneous activity, ataxia, oropharyngeal apraxia, lack of response to visual stimuli, and emotional disturbances such as depression or emotional instability, apathy, and irritability [13, 44, 52, 55, 60].

CM in adults is a very rare entity and has never been reported in the context of Lhermitte-Duclos disease (LDD) before.

LDD, a rare and benign lesion of the cerebellum also referred to as dysplastic gangliocytoma, is known to be associated with Cowden disease (CD) and with germline mutations of the *PTEN* gene. Due to its rarity, the diagnosis is frequently missed by surgeons, radiologists, and pathologists. Surgical excision appears to be the only effective treatment to improve the symptoms [22, 39]. However, it is difficult to distinguish the tumor parenchyma from the healthy cerebellar tissue, thus hindering effective surgical treatment [8, 40].

We here review the literature on postoperative CM in adults, summarize the anatomical structures affected in this syndrome, and present two illustrative cases of CM after surgery of LDD, which has not been reported before. This might help to avoid similar postoperative complications and aid in effective preoperative workup of patients with LDD.

Review of the literature on CM

Adult patients with postoperative CM—clinical characteristics

Since the first report of postoperative CM in adults by Salvati et al. in 1991, another 16 patients in 11 articles have been published in greater detail including the two cases with LDD presented here (Table 1). There were 10 male and 7 female patients; the median age was 33 years (range, 18–74 years). In addition to our two patients with gangliocytoma, there were 5 patients with medulloblastoma, 3 metastases, 2 astrocytomas, 2 hemangioblastomas, and 1 arterio-venous malformation, pineal gland dermoid, and choroid plexus papilloma each. The median time of duration of CM was 4 weeks (range, 4 days to 1 year). Dysarthria was the most common symptom occurring in 15 of the reported 17 patients.

It has to be mentioned that there are several additional case reports of CM in adults and children involving other disease entities and/or interventions, such as evacuation of an acute subdural posterior fossa hematoma [25], posterior circulation infarction [38], and acute cerebellitis [43]. However, the causal relationship between disease/intervention and CM in these cases is frequently debated in the literature and not universally accepted.

Regardless of its cause, CM is a very rare syndrome. Dubey et al. [17] analyzed the medical records of 500 patients (age ranging from 12 to 78 years) who had undergone posterior fossa surgery. Six of these patients presented with CM postoperatively, suggesting a CM incidence of 1.2%. Although the authors of this report do not specify age and disease of these CM patients, it is generally accepted that the incidence of CM in children is higher than in adults and varies between 2% and 40% [63].

Pathophysiological and anatomical observations

CM mainly affects pediatric patients after posterior fossa surgery; adult cases are rare for unknown reasons. Unfortunately, the pathophysiological mechanisms and anatomical correlates of CM are still not fully understood. Several types of injury and locations in the posterior fossa were reported as possible causes of CM: bilateral lesions of paravermian structures including the dentate nuclei, lesions of the vermis, lateral cerebellar hemispheres, and lesions of cerebellocortical pathways.

The pivotal role of the cerebellum in speech includes the cognitive components of speech generation, the internal generation of words, choice of words, rehearsal of speech, timing of speech, vocal intonation during speech production, the coordination of orofacial muscles during the internal generation of words, the verbal production of words, as well as the timing and intonation of speech [2, 6, 15, 30, 34, 54]. The cerebellum also plays a role in the organization of syllabic timing and sequencing, even at the level of internal speech [1]. Dailey et al [13] and Ryding et al [48] suggested that the cerebellum integrates auditory and sensory information to coordinate the motor production of speech. In agreement with an important role of the cerebellum during speech, the paravermal regions of the cerebellum show enhanced signal intensity and activation in functional magnetic resonance imaging (MRI), positron emission tomography, and single-photon emission computed tomography studies [48, 58, 64]. Neuroimaging studies have also shown activation of the cerebellum when speech is imagined by individuals [15, 34].

Originally, lesions to the paravermian structures including the dentate nuclei were thought to be responsible for CM [3, 5, 11, 16, 21, 27]. Meanwhile, the syndrome has not only been anatomically associated with lesions of the dentate nuclei but additionally with lesions of the vermis, lateral cerebellar hemispheres, and the cerebellocortical pathways [13, 23, 26, 41]. McMillan et al. [35] found indications that the presence of preoperative brainstem compression in association with posterior fossa tumors may increase the risk of postoperative CM. However, larger

Table 1 Reports about adult patients with postoperative CM

Author	Publication year	No. of patients	Sex	Age (y)	Pathology
Salvati et al. [50]	1991	1	M	20	Medulloblastoma
D'Avanzo et al. [12]	1993	2	M	45	Medulloblastoma
			F	20	Medulloblastoma
Cakir et al. [9]	1994	1	M	61	Metastasis
Dailey et al. [13]	1995	1	F	20	Astrocytoma
Bhatoe [7]	1997	1	M	28	Hemangioblastoma
Dunwoody et al. [18]	1997	1	M	54	Arteriovenous malformation
Kai et al. [31]	1997	2	M	71	Hemangioblastoma
			F	74	Metastasis
Caner et al. [10]	1999	1	F	18	Choroid plexus papilloma
Ildan et al. [29]	2002	2	M	32	Medulloblastoma
			M	44	Astrocytoma
Sherman et al. [51]	2005	2	F	33	Pineal gland dermoid
			F	56	Metastasis
Akhaddar et al. [4]	2008	1	M	22	Medulloblastoma
Afshar-Oromieh et al.	Present study	2	M	36	Gangliocytoma
			F	31	Gangliocytoma
Total		17			

M male; *F* female

studies could not support a correlation between tumor size and risk of CM [47, 63].

Isolated lesions of the cerebellar hemispheres are thought to be insufficient to produce CM [60], and indeed, such patients very frequently show vermal lesions in addition to the primary cerebellar defect [58].

Rekate et al. [45] reported on six patients with transient mutism following removal of cerebellar tumors. From these cases, the authors concluded that acute bilateral damage of both cerebellar hemispheres including the dentate nuclei can cause CM.

Supporting the concept that bilateral damage is required, Ozimek et al. [42] found that only bilateral and not unilateral lesions of the dentate nuclei were correlated with CM. In contrast, Dailey et al. [13] suggested that bilateral impairment of the dentate nuclei is not a prerequisite for the development of CM: in nine patients with CM, none had bilateral edema or infarction of the dentate nuclei after surgical treatment; however, it is notable that splitting of the vermis was detectable in all of these subjects. They also reported that 25% of children undergoing procedures associated with the splitting of the vermis develop CM. Similarly, other authors suggested that both superior and inferior vermian incisions are associated with this syndrome [16, 44, 53].

However, it appears that damage to the median and paramedian structures including the vermis alone is insufficient to cause CM [19, 20]. It has been suggested that bilateral damage of pathways between cerebellar structures and the cerebrum (so called dentatothalamocortical pathway) connecting the dentate nuclei with the supplementary motor area through the contralateral and ventrolateral thalamical nuclei are involved [11, 24, 27, 60, 61]. For example, there are a few case reports of mutism following excision of bilateral parasagittal meningioma [11] or a brainstem cavernous angioma [24, 33]. In agreement with the hypothesis that concomitant impairment of cerebral and cerebellar function is required for CM, single-photon emission computed tomography analysis of patients with mutism has shown decreased blood perfusion in the thalami, medial frontal lobes, temporal lobe, as well as cerebellar vermis and both cerebellar hemispheres [49]. In addition, blood perfusion was shown to increase in these structures upon recovery from mutism [13, 27, 49].

Illustrative cases

Case 1

A 36-year-old male patient reported recurrent headaches, vertigo, and absence-like episodes for a period of more than 2 years. At the initial presentation, he showed minor cognitive deficits (due to neonatal hypoxia) and an insecure, broad-based gait. There were no further neurological deficits. The cranial MRI scan revealed a weak-enhancing space-occupying lesion with a perifocal edema in the left hemisphere of the cerebellum with consecutive hydrocephalus and beginning vermal herniation into the foramen magnum. The mass presented with only weak

gadolinium enhancement consistent with gangliocytoma (Fig. 1a).

For tumor excision, the patient was placed in a prone position with the head inclined and fixed in a Sugita head-holder system. A suboccipital median craniectomy approach was performed. After cortex incision above the left cerebellar hemisphere, a blunt preparation through the cerebellar cortex was performed using one spatula to reach the tumor. The tumor was of grayish color and of relatively soft consistency. The histological examination revealed a dysplastic cerebellar gangliocytoma Lhermitte-Duclos, which was confirmed by two independent neuropathological evaluations.

On the first postoperative day, the patient developed significant neurological complaints including dysphagia, disability to cough, and ataxia. In addition, he presented with symptoms that were unexpected after cerebellar resection: reduced spontaneous and dysarthric speech, depressive mood and facial expression, reduced voluntary activity, and concentration deficits. The presenting symptoms were in agreement with the diagnosis of CM. A postoperative cerebral computed tomography (CT) scan showed a defect in the left lateral hemisphere and inferior vermal region after resection of the mass but without additional postoperative complications such as bleeding or infarction (Fig. 1b). Due to the rapid occurrence of postoperative respiratory complications, it was not possible to perform a cerebral MRI scan: during the second postoperative day, the patient had developed dyspnea after aspiration and was intubated. Due to prolonged ventilation, a tracheotomy was performed 9 days after intubation, and respirator assistance was continued for more than a month.

Periodic neurological examinations revealed no signs for isolated cranial nerve dysfunction. The postoperative aspiration and dyspnea were most likely triggered by the reduced voluntary activity and the reduced level of alertness of the patient.

Three weeks after surgery, the patient was discharged for rehabilitation with the continuous presence of mutism. However, in the following weeks, mutism slowly resolved and disappeared completely about 4 months after surgical treatment, although some typical cerebellar symptoms such as dysarthria and ataxia persisted in a mild form.

Case 2

A 31-year-old healthy female patient was referred for cranial MRI scanning in order to exclude a pituitary adenoma for further diagnosis of infertility. The images revealed a slightly contrast-enhancing bilateral lesion predominantly involving nodulus and uvula vermis of the cerebellum and infiltrating the fourth ventricle without any additional pathological signs (Fig. 2a).

The initial examination revealed no neurological deficits, and the patient was admitted for surgery of the cerebellar lesion. The operative procedure including positioning and

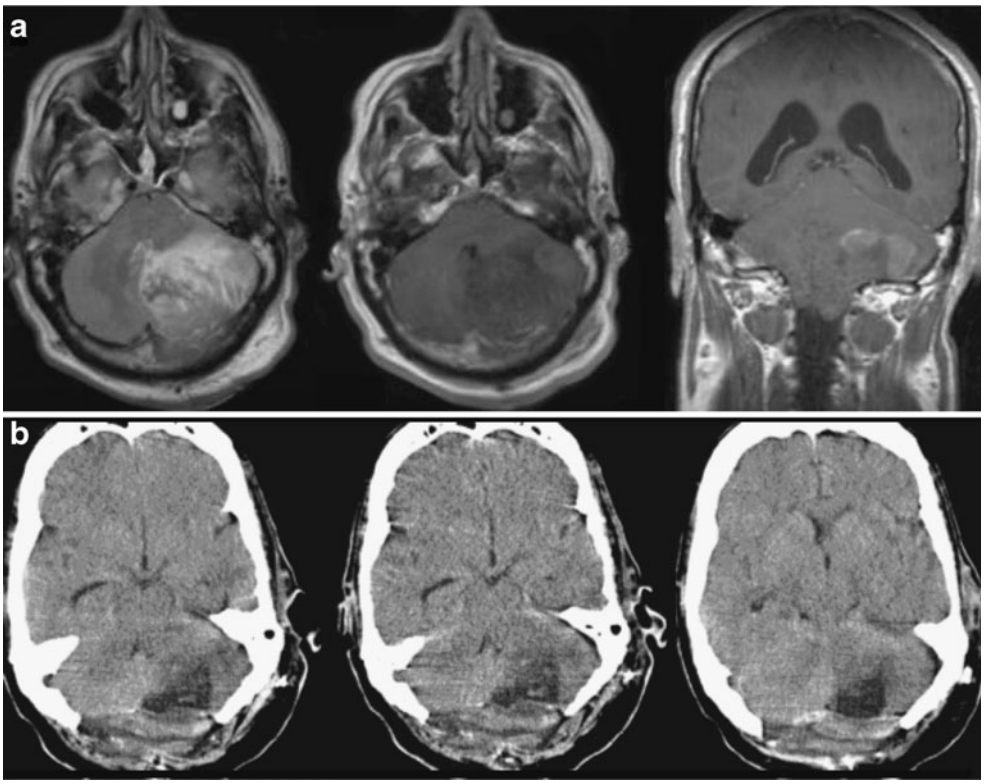

Fig. 1 **a** MRI scan, from left to right: axial FLAIR sequence showing perifocal edema and shifting, T1 postcontrast axial sequence presenting weak enhancement compatible with gangliocytoma; T1 postcontrast axial sequence: vermal herniation into foramen magnum due to expansive lesion. **b.** Computed tomographic scan on postoperative day 1. Isodense defect with lesion in lateral hemisphere and inferior vermal region

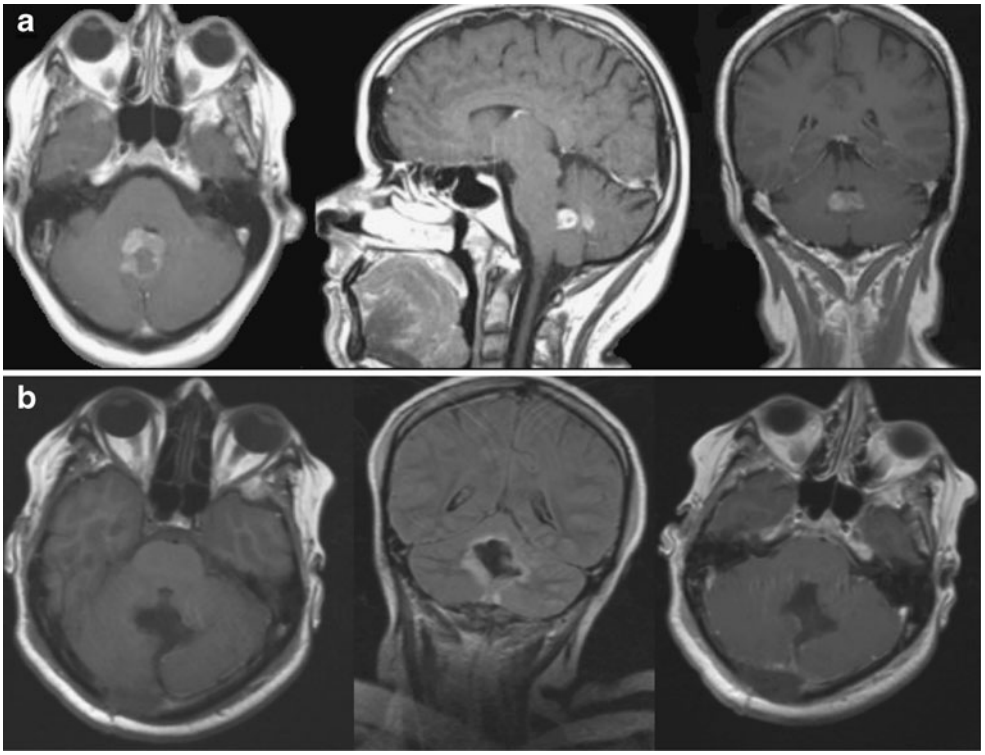

Fig. 2 **a** Preoperative MRI scan (*from left to right*): T1 postcontrast medium, axial sequence: bilateral lesion accentuated on nodulus and uvula vermis; T1 postcontrast medium, sagittal sequence showing the medial and vermal zone infiltration; T1 postcontrast medium, coronar sequence: bilateral nuclei fastigii and dentatus lesion. **b** Postoperative MRI scan of the second patient (*from left to right*): T1 sequence showing the iatrogenic lesion of vermis, FLAIR coronar sequence: possible temporal affection of bilateral nuclei dentatus due to postoperative edema; T1 postcontrast medium, axial sequence: absence of enhancement

approach was similar to the one described in the first case; however, it was conducted by a different surgeon. A suboccipital median craniectomy was performed. After dura opening, two spatulas were used to retract cerebellar hemispheres, thus exposing the inferior part of the tumor. The vermis was split to the smallest possible length to gain access to remove the tumor completely. As we had no negative experience with that transvermian approach, so far we did not consider the alternative telovelar approach [56]. The histological examination revealed a dysplastic cerebellar gangliocytoma Lhermitte-Duclos, similar to the first case.

The patient was asymptomatic on the first postoperative day. However, 1 day later, she presented not only with signs of cerebellar dysfunction such as dysphagia, dysarthria, and disability to cough and severe ataxia but also with the classical symptoms of mutism: severe emotional lability, depression, reduced spontaneous speech and voluntary activity, as well as deficits in memory and concentration. In addition, involuntary orofacial movements were observed. As in the first case, the symptoms were diagnosed as mutism in collaboration with our neurological specialists. The postoperative MRI scan revealed no residual tumor, but a resection cavity affecting the vermis and a possible impairment of both nuclei dentati due to edema (Fig. 2b). The signs of mutism slowly resolved similar to the first case. Four months after surgery, a follow-up examination still revealed slight ataxia and concentration deficits. The patient was able to speak complete sentences, although she still had anomia as well as deficits in phonation and velocity of speech. Taken together, the symptoms of mutism resolved and only typical symptoms of cerebellar dysfunction remained.

The postoperative CT scan of our first patient (Fig. 1b) revealed an isodense defect with lesions in the lateral hemisphere and the inferior vermal region. The images are also compatible with functional impairment of the left nucleus dentatus and further vermal structures due to edema surrounding the resection cavity.

In the second patient, the postoperative MRI scan (Fig. 2b) revealed an iatrogenic lesion of the vermis and is compatible with functional impairment of both nuclei dentati and the cerebellar pars intermedia due to postoperative edema.

Based on the aforementioned reports, lesions to any of these structures may have contributed to the development of CM in both of our patients. Our two cases therefore support a possible role of vermal splitting, bilateral damage to nuclei dentati, impairment of the cerebellar hemisphere, and pars intermedia, respectively, in the development of CM.

To summarize, CM is known to be caused by damage or functional impairment of the following structures: paravermian structures including the dentate nuclei, vermis, lateral cerebellar hemispheres, and cerebellocortical pathways (connections between superior cerebellar hemisphere, dentate nucleus, red nucleus, thalamus, and cerebral cortex).

Prevention and prognosis

Clearly, any pathology and any type of surgery in the cerebellum carry the risk of developing CM. However, it is unclear why CM is so exceedingly rare, even though damage to the abovementioned anatomical regions due to haemorrhage, infarction, surgical treatment, and mass effects is relatively frequent. In addition, the question is still unsolved whether surgical treatment of certain diseases such as LDD might carry a higher risk to develop CM. More investigations and reports may show some tendencies.

Although CM is a severe handicap, it has a self-limiting course and a favorable prognosis for complete resolution. This was also observed in the two cases reported here, who had almost completely recovered 4 months after surgery. Speech therapy may accelerate improvement of symptoms.

In addition to the pathophysiology of CM, our two cases also illustrate current difficulties in the management of LDD that can contribute to such postoperative complications. Even though our second patient presented with no neurological deficits, it was decided to proceed with surgery. The dignity of the tumor was unclear prior to surgery; the lack of symptoms was ascribed to the small size of the tumor. It was anticipated that the risk of developing cerebellar dysfunction after surgery would be minor, whereas no surgical intervention would probably have resulted in tumor outgrowth, secondary cerebellar dysfunction, and aggravation of surgical conditions at later stages. As illustrated by this case, surgery of LDD is hampered by the difficulty to distinguish between tumor and healthy cerebellar tissue. However, LDD can be detected by careful neuroradiological analysis prior to surgery [32, 36, 57]. Typical radiological signs are a striated ("tiger-striped") pattern of hyperintensity on T2-weighted images and a corresponding hypointensity on T1-weighted images, as well as the typical absence of enhancement following gadolinium-diethylenetriaminepentaacetic acid (Gd-DTPA) administration [32, 65]. With such preoperative information, the surgeon will be aware that tumor tissue and healthy tissue cannot be distinguished visually and will leave narrower margins of resection due to the benign nature of the tumor. In addition, instantaneous section should help guide the procedure. These provisions can help prevent severe postoperative complications such as CM and are likely applicable to other benign cerebellar lesions. Since the pathophysiology of CM is not fully understood, the above recommendations might help reduce the incidence of CM; however, this postoperative complication cannot be fully avoided. It is therefore important to inform the patient accordingly.

We agree with de Ribaupierre that until recently, neurosurgeons eagerly removed cerebellar lesions without consideration of future cognitive impairment that might be caused by the resection [14].

Due to our experiences and the aforementioned risks of developing CM, we concretely suggest the following strategy in the management of LDD: First of all, careful neuroradiological analysis should lead to a suspicion of this entity. In case neurological deficits are absent, preoperative stereotactic biopsy should be considered to confirm the diagnosis. Following confirmation of the diagnosis, we recommend a "wait-and-scan-strategy" (MRI initially every 6–12 months). Depending on the further rate of growth and the development of neurological deficits, a complete resection leaving narrower margins should be performed.

In case of neurological deficits at the time of presentation, we suggest an instantaneous intraoperative tumor biopsy for pathological confirmation of the diagnosis followed by complete resection leaving narrower margins.

Conclusions

CM is a rare syndrome more frequently observed in children and adolescents and rarely observed in adults. The leading symptom is impairment of speech and language. There is emerging consensus that lesions of paravermian structures including dentate nuclei, vermis, lateral hemispheres, and cerebellocortical pathways contribute to this syndrome, but there is no single particular site associated with CM**.** The two illustrative cases included in our report support involvement of these structures in the development of CM. It is important to be aware that this syndrome might also occur in adults. Besides an optimal preoperative diagnostics and thorough and careful preparation, there are no particular means that might help prevent such complications. Despite the severity of symptoms, CM generally carries a favorable prognosis and usually resolves within a few months.

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Comments

Ludwig Benes, Marburg, Germany

Cerebellar mutism (CM) is a rare postoperative complication of mainly pediatric posterior fossa surgery and reserved for cases with onset of mutism without corresponding long tract signs or cranial nerve palsies.

The authors present a well written paper analyzing two own cases of CM in young adults after posterior fossa surgery for Lhermitte-Duclos disease (LDD). In this context CM has never been mentioned in the pertinent English literature before and therefore it is worth publishing. The lessons we should learn from this paper is, that surgery close to paravermian structures bears the risk of severe cerebellar mutism especially in pathologies which are difficult to distinguish from normal cerebellar structures. Therefore, it is essential to have a meticulous preoperative planning to avoid this complication.

Mario Ammirati, Columbus, Ohio, USA

Oromieh et al present a nice review of postoperative cerebellar mutism after intracerebellar surgery in adults. This is a rare conditions that points to the involvement of the dentate nuclei in the majority of cases in which neuroradiological imaging was obtained. The relevance of the dentate nuclei in the genesis of this syndrome is highlighted by the work of Guidetti and Fraioli who observed total inability to speak for up to 3 months in 2 patients in whom simultaneous and bilateral lesions of the dentate nuclei were stereotactically created to treat spasticity (Guidetti B, Fraioli B: Neurosurgical treatment of spasticity and dyskinesias. Acta Neurochir [Suppl] 24:27-39, 1977). Cerebellar mutism is a rare condition that fortunately has, as pointed out by the authors, a positive resolution in the overwhelming majority of patients. While no clear reccomendation may be made to decrease its occurrence, we feel that meticulous attention to microneurosurgical techniques and limited use of bilateral cerebellar retraction is certainly warranted when operating in the cerebellar midline. Also we have been using, whenever feasible, the telo-velo-medullary approach to posterior fossa lesions, thus decreasing the cases in which splitting of the vermis is necessary.

Again the authors are to be commended for a well written review of the literature and for the addition of their own two interesting cases.

Engelbert Knosp, Vienna, Austria

The authors described two cases of Lhermitte Duclos disease, who had cerebellar mutism after extensive removal. Dysplastic gangliocytomas are very rare in neurosurgical practice and cerebellar mutism is a rare complication after posterior fossa surgery in adults too.

The authors are congratulated to provide the readers this excellent case report and a review of the current literature. They discussed meticularly the pathology of dysplastic gangliocytoma, the indication for surgery and the rare neurological complication of cerebellar mutism. Although cerebellar mutism is still not fully understood pathophysiologically, I strongly believe, that its development is related to severe–maybe bilateral–damage of the cerebellum and its connection to the brainstem. Therefore approaches like the telovelar approach (1), which avoids splitting of the vermis should be preferred whenever possible. With this approach the upper fourth ventricle and even the aqueducal area are attainable without splitting the vermis.

If the position of the lesion necessitates a supracerebellar approach, utmost care should be taken not or to damage or tear the superior cerebellar peduncle(s). Sometimes it would be preferable to use a supratentional, transtentional route to reach these lesions.

The authors presented very interesting cases and provided us with a excellent overview of a rare complication.